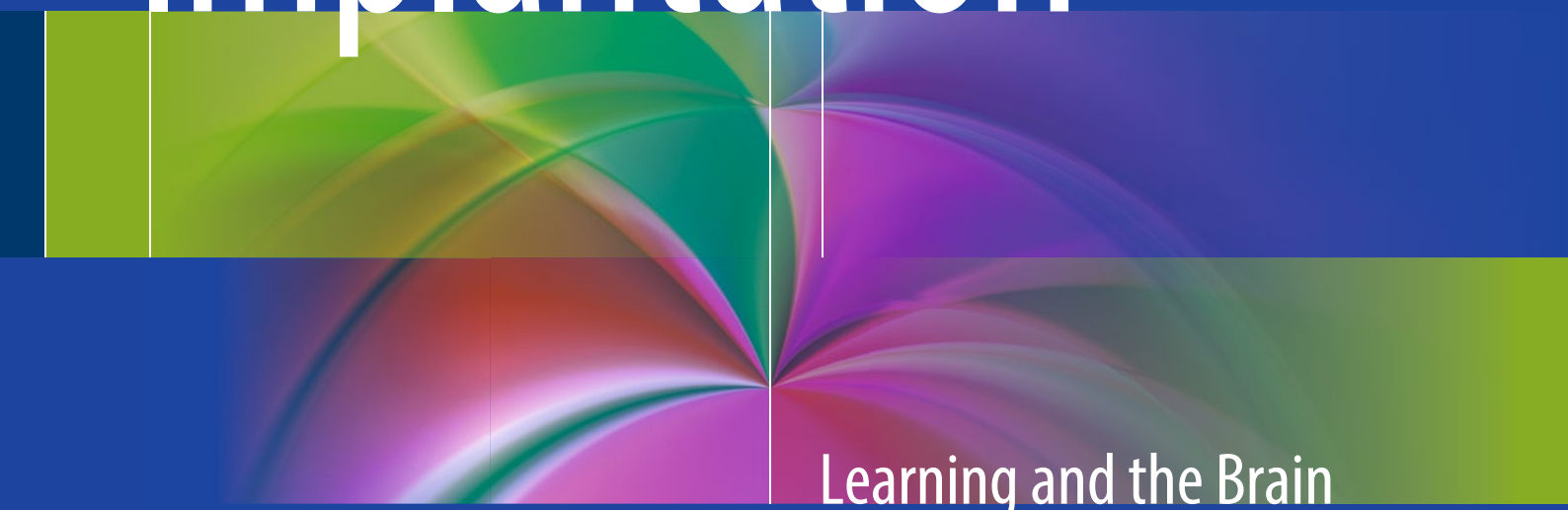
Nancy M. Young
Karen Iler Kirk
Editors

Pediatric Cochlear Implantation



Learning and the Brain

Springer

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In memory of John K. Niparko, M.D., a renowned clinician–scientist and leader in cochlear implantation, who cared deeply about improving the lives of children.

Preface: Cochlear Implants in Children—A Personal Perspective

The cochlear implant is the most successful of all neural prostheses, both in terms of the degree to which it can restore function and in the number of people who have received this device worldwide. Children implanted at a young age can now acquire speech recognition, speech production, language, and reading abilities that previously were not possible for the vast majority of hard-of-hearing or deaf children. The effectiveness of the cochlear implant also has transformed the education of these children. The majority of children with cochlear implants now are educated in oral classrooms that emphasize the use of spoken language and listening skills; inclusion of children with cochlear implants in mainstream classrooms with their typically hearing peers is increasingly common. Classrooms with instruction in Total Communication (i.e., the combined use of manually coded and spoken English) now are integrating listening and spoken language development into the curriculum more than ever before. This change has likely been driven in part by research demonstrating that children using manually coded (signed) English may experience a doubling of the rate of language acquisition after implantation. These expanded educational options have in turn contributed to enhanced language and literacy outcomes. A generation of hard-of-hearing and deaf children is graduating high school, enrolling in postgraduate education, and finding employment to an extent never before seen. To observe these changes unfold over our careers has been most rewarding.

Our goal is for this book to serve as a resource for a broad audience of clinicians and clinical researchers regarding current and emerging best practice in pediatric cochlear implantation. We hope to inspire research in new areas of importance, especially the role of cognitive processing skills in cochlear implant outcomes. We were fortunate to enlist participation from a highly experienced and distinguished group of coauthors. We are very grateful to them for their willingness to share their knowledge and the considerable time they devoted to this project.

Our lead chapter is by Blake Wilson and coauthors Michael F. Dorman, René H. Gifford, and David McAlpine. Wilson's pioneering work in signal processing enabled development of the modern cochlear implant. Over more than 30 years, his work at the Research Triangle Institute in North Carolina has transformed this neuroprosthesis and the lives of many children. In contrast to early implant devices, modern cochlear implant systems utilizing speech processing based on his work have enabled most recipients to understand speech through listening alone and even to use a cell phone. In recognition of his significant contribution to the development of cochlear implants, Wilson was one of several scientists awarded the Lasker-Debakey clinical medical research award in 2013 and the Fritz J. and Delores H. Russ Prize in bioengineering in 2015. The former is commonly referred to as the American Nobel and the latter as the Nobel of engineering. These high honors have brought much recognition to our field. Cochlear implantation is now recognized as the first successful electronic device to replace a human sensory end organ and as one of the major medical advances of the twentieth century. Wilson points out that the brain may be the most important determinant of cochlear implant outcomes. He advocates for the use of a "top down" or "cognitive neuroscience" approach to improve the effectiveness of this and other neural prostheses.

Part II of the book focuses upon clinical management. Rene Gifford gives us a comprehensive description of the interdisciplinary evaluation of pediatric candidates and the significant evolution of candidacy beyond those children with bilateral profound deafness. Tina Tan, an

infectious disease specialist and an expert in pneumococcal disease, provides an overview of the risk of infectious disease in implanted children. She explains the rationale for the Center for Disease Control's current vaccination guidelines that are designed to reduce bacterial meningitis due to pneumococcal disease and Haemophilus influenzae type b. These recommendations now include annual influenza vaccination to reduce the risk of otitis media which may lead to bacterial meningitis. Sharon Cushing, Susan Blaser, and Blake Papsin discuss medical and radiological aspects of unusual cases. They provide us with their medical approach and intraoperative techniques for children with conditions that often precluded cochlear implantation in the past. Pearls they share also include a practical approach to pediatric vestibular assessment and the relationship between poor vestibular function and increased risk of internal device failure. Brandon Isaacson and Peter Roland provide a state-of-the-art overview of surgical considerations including hearing preservation techniques and specialized approaches to address unusual surgical anatomy. Holly Teagle's chapter highlights the important role audiologists play in maximizing cochlear implant outcomes. She describes a sophisticated and logical approach to speech processor programming and postimplant follow-up. Teagle also emphasizes the child, family, and educational factors that come into play when optimizing device programming for individual children. Terry Zwolan and Casey Stach have contributed a chapter on diagnosis and management of device problems that limit use or benefit and may result in reimplantation. They describe the process of recognizing and managing "soft" failures including the important role of imaging, monitoring of auditory progress and electrophysiological measures to identify this challenging problem. The role of electrophysiological testing in clinical management is elucidated by Karen Gordon. Gordon makes a compelling case for more widespread use of electrophysiological measures as part of the test battery to aid in programming and monitoring of progress over time.

Outcomes after implantation are the focus of Part III. Karen Iler Kirk and Michael Hudgins provide insight into the assessment of spoken word recognition abilities in infants and children, review long-term cochlear implant outcomes in children, and highlight factors that have been shown to influence speech and language development in children with cochlear implants. Ruth Litovsky explains the differences between true binaural hearing and the bilateral hearing permitted by current implant technology. She describes measurable gains in speech recognition in noise and in sound localization obtained by bilaterally implanted children, despite receiving degraded binaural cues. Litovsky attributes these findings to the brain's ability to integrate the uncoordinated information received from two implants. Susan Nittrouer and Amanda Caldwell-Tarr review language and literacy of implanted children, including the results of their own ongoing longitudinal investigations. They observed dramatic initial improvements in language and literacy following implantation. However, over time the cochlear implant users in their study developed language and literacy skills that fell within the lower end of the normal range for children with normal hearing. The authors propose that changes in cochlear implant design as well as effective postimplant behavioral intervention may be necessary to ameliorate this situation. Alexandra L. Quittner, Ivette Cejas, Jennifer Barnard, and John Niparko share important research findings regarding psychosocial development gleaned from the Childhood Development after Cochlear Implantation (CDaCI) study, the first longitudinal multicenter national cohort study to systematically evaluate early cochlear implant outcomes in children. Their research demonstrates that, despite the many areas of great improvement, implanted children often face challenges in psychosocial and social-emotional functioning. They suggest a family-centered management approach, as well as proactive screening of young children for delays in cognition, behavioral development, and social-emotional function and health-related quality of life.

In the early days of pediatric cochlear implantation, often only children thought to be "ideal" candidates were eligible to receive an implant. Today cochlear implantation is the accepted treatment for deafness. Therefore it is only natural that a growing number of children with a broad range of co-occurring complicating conditions that may slow progress or reduce expecta-

tions for spoken language development are being evaluated for cochlear implantation. These children are the focus of Part IV. An overview of many of these conditions, the range of benefits obtained, and increased need for communication between the implant team and others serving these children is provided by Nancy Young, Elizabeth Tournis, and Constance Weil. The goal of their chapter is to encourage a redefinition of implant candidacy and a better understanding of the potential impact of hearing in the lives of these children and their families beyond what typically is measured in current clinical practice. Children with cochlear nerve deficiency are a very special population who present a management challenge. The literature demonstrates that this problem is more common than previously recognized. An overview of diagnosis and management options is provided by Claire Iseli, Oliver Adunka, and Craig Buchman. The authors describe the incidence and clinical presentation of children with cochlear nerve deficiency and consider audiological and radiological assessments used to diagnosis this condition. They also describe functional electrophysiological assessments that may help to improve preoperative and intraoperative prediction of cochlear implant benefit in these children. Their contribution is followed by a discussion of auditory brainstem implantation (ABI) for congenital deafness by Robert Shannon, Lilliana Colletti, and Vittorrio Colletti, pioneers in this area. The authors report that pediatric ABI users demonstrate a broader range of speech recognition skills, including open-set word recognition, and they consider the role of neuroplasticity in pediatric ABI outcomes. And finally, the new frontier of cochlear implantation to address single-sided deafness is described by David Friedmann, Susan Waltzman, and J. Thomas Roland. Because single-sided deafness can be congenital, acquired, or result from a progressive hearing loss, they highlight the need for careful monitoring in children with unilateral hearing loss. The authors describe the negative impact of single-sided deafness on language and educational outcomes in children. Finally, they consider the relative merits of various treatment options for single-sided deafness, including cochlear implantation.

What factors beyond the implant affect learning and how outcomes may be maximized are addressed by multiple authors in Part V. Motivated by a desire to understand individual differences in cochlear implant outcomes, David Pisoni and collaborators from the Speech Research Laboratory within the Department of Psychological and Brain Sciences at Indiana University have conducted pioneering research into the relationship of cognitive processing and working memory to spoken word recognition, speech perception, and language skills in pediatric implant recipients. This work has led to the development of novel interventions to improve spoken language processing. Angela AuBuchon, David Pisoni, and William Kronenberger explore neurocognitive processes underlying verbal working memory; they suggest that prelingually deaf cochlear implant users appear to be at risk for slow and inefficient phonological recoding and verbal rehearsal processes because of the early atypical auditory and language environments in which their verbal working memory systems develop. Kronenberger and Pisoni describe neurocognitive training procedures that have been used to improve working memory in children with normal hearing and children with cochlear implants. Speech perception training is explored by Patrick Wong and Erin Ingvalson. They recommend an individualized approach which selects training targets based on a child's ability to perceive the acoustic properties of speech. Wong and Ingvalson propose that training outcomes can be enhanced through the use of an adaptive strategy that matches the listener's skill set throughout the training paradigm. Susan Nittrouer provides a chapter on intervention to improve language and literacy skills in children with cochlear implants. Based on empirical evidence, she recommends that children receive intensive support for language learning throughout childhood. She presents a set of organizing principles that underlie an integrated approach to language intervention. Nittrouer also makes recommendations to enhance the effectiveness of intervention such as the use of audiovisual speech signals to strengthen the child's internal representations of speech, and bimodal cochlear implant use (i.e., the use of a hearing aid on the contralateral nonimplanted ear) in unilaterally implanted children. Kate Gfeller considers the importance of music in children's daily lives and describes the limitations of cochlear implant signal processing in conveying the structural properties of music. Gfeller provides a comprehensive summary of

research concerning pediatric cochlear implant users' perception of music and the role of music training paradigms in improving music perception and enjoyment. Finally, she explores music training's potential to impact speech and language development and describes principles for the application of music-based training for children with cochlear implants.

Two chapters on educational management comprise Part VI. Marybeth Lartz and Tracy Meehan provide a comprehensive overview of the therapy needs of children with cochlear implants who are younger than 3 years of age and are enrolled in early intervention programs. They describe a framework for designing and delivering intervention for children across a range of chronological ages and language levels. They also provide strategies to enhance listening, language, and literacy skills that can be used with parents and their children before and after cochlear implantation. Finally Nancy Mellon and her colleagues at the River School in Washington DC, a superb inclusion program integrating children with implants and typically hearing children, provide an excellent overview of building auditory skills, spoken language, academic, and socioemotional skills within the classroom.

This book reflects the remarkable progress that has been made in the interdisciplinary field of cochlear implantation, yet so much still remains to be done. Variability is a hallmark of cochlear implant outcomes, and not every child reaches his or her maximum potential. Current cochlear implant technology provides a crude signal, at best, in comparison to normal hearing. Even cochlear implant recipients with the best results may have difficulty in challenging situations such as when background noise or competing speakers are present. Other reasons for poorer performance include further degradation of the auditory signal due to suboptimal programming or lack of identification of a "soft" failure, inadequate or ineffective therapy, and less than adequate school services to support auditory and spoken language development. In addition, access to cochlear implantation remains a serious problem even in developed countries. Many parents of newly diagnosed deaf children remain unaware of the effectiveness of cochlear implantation. Candidates may not be recognized or referred early in life for candidacy evaluation. Financial barriers may prevent or delay implantation of one or both ears. Children with medical complexity, especially those with conditions associated with cognitive impairment, may not be viewed as viable candidates. For children in less developed countries, access to cochlear implantation is far more limited and may be the exception for deaf children. Much work remains for the next generation of clinicians and investigators.

Numerous studies of speech perception, spoken language, and literacy of early implanted children have yet to reveal the reason for differences in low- and high-performing children on standard measures of performance. Differences in cognitive processing and learning likely underlie the variability in outcomes after implantation. In other words, the brain is as important as the ear. Using this framework, the redesign of cochlear implant systems and implementation of novel therapies may enable more rapid progress and improved outcomes after implantation. Methods of evaluation that provide insight into each child's cognitive strengths and weaknesses may permit development of customized therapy to preemptively address the needs of those children likely to have poorer outcomes. These are just some of the areas of future research that we hope the readers of this book will embrace.

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Acknowledgements

Nancy M. Young, M.D.
Development of a multidisciplinary program capable of meeting the diverse needs of children with hearing loss and their families would not have been possible without the many individuals who deeply believe in our mission of helping children achieve their full potential. I gratefully acknowledge the Buehler Family Foundation, the Lillian S Wells Foundation, and the Foundation for Hearing and Speech Rehabilitation for their crucial support. I also wish to thank our implant team members, past and present, and the community-based professionals with whom we collaborate for all they have taught me. I am profoundly grateful to the children and families I serve. They continue to provide me with an invaluable education in humility and a reminder of the struggles that families endure and often overcome.

An important influence early in my career was Alfred E. Mann. Observing first hand his relentless drive and faith in his own vision to alleviate human suffering by translating scientific discovery into clinical interventions remains a source of inspiration. I am also indebted to the many clinicians and researchers in our multidisciplinary field who have generously shared their knowledge. I am fortunate that many have become good friends. One such colleague is Karen Iler Kirk, Ph.D., to whom I am forever grateful for her wisdom and friendship.

My husband Mitchell Marinello, despite his own demanding career, has been unswerving in his support. I am truly blessed to have found such a wonderful husband and father to our three daughters.

Karen Iler Kirk, Ph.D.
I owe a debt of gratitude to William F. House, M.D., who gave me the opportunity to work with his cochlear implant team in 1981, near the beginning of cochlear implantation in children. My work at the House Ear Institute in Los Angeles inspired me to pursue further graduate study, choose an academic career, and conduct research in cochlear implant outcomes. I have been privileged be a part of this pioneering research field and to see the transformative effect this revolutionary technology has had on children and families. Many families gave generously of their time through participation in research to further our understanding of speech and language development following cochlear implantation. I am most grateful to them.

None of this would have been possible without the love and support of my family. My husband, Gerald Kirk, M.D., has been an equal partner in our home and family life, thus allowing me to pursue my career goals. I also want to acknowledge my wonderful children: Andrew and his wife Shawna, Brian and Sarah. I am so proud that they are compassionate, productive young adults who want to make a difference in the world. Finally, I want to thank my dear friend and collaborator, Nancy M. Young, M.D., for having the vision and drive to bring this book to fruition. It wouldn't have been possible without her.

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Part I

Introduction

Cochlear Implant Design Considerations

1

Blake S. Wilson, Michael F. Dorman, René H. Gifford, and David McAlpine

Abbreviations

AB	Arthur Boothroyd (as in the AB words)
AzBio	Arizona Biomedical Institute (as in the AzBio sentences)
BKB	Bamford-Kowal-Bench (as in the BKB sentences)
BPF	Band-pass filter
CI	Cochlear implant
CIS	Continuous interleaved sampling
CNC	Consonant-nucleus-consonant (as in the CNC words)
CUNY	City University of New York (as in the CUNY sentences)
EAS	Electric and acoustic stimulation (as in combined EAS)
F0	Fundamental frequency
HINT	Hearing in Noise Test (as in the HINT sentences)
HSM	Hochmair-Schulz-Moser (as in the HSM sentences)
LPF	Low-pass filter
PTA	Pure tone average
RF	Radio frequency
RM ANOVA	Repeated-measures analysis of the variance
S/N	Speech-to-noise ratio
SEM	Standard error of the mean

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Introduction

The performance of the present-day cochlear implants (CIs) is made possible with (1) multiple and perceptually separable sites of stimulation in the cochlea; (2) good use of those sites with processing strategies aimed at representing in a clear way most of the information that can be perceived with CIs; and (3) the remarkable ability of the brain to make good use of a sparse and otherwise unnatural input from the periphery. When all of the parts come together, the results can be surprisingly good. For example, the great majority of patients today can understand everyday speech in quiet with the CI alone, in the absence of any visual cues. Indeed, most patients today use their cell and landline phones routinely. This restoration of function is a long trip from total or nearly total deafness. As the esteemed Professor Dr. Jan Helms put it (Helms 2009), "From my perspective, cochlear implants are the most significant medical development in the second half of the twentieth century, as they replace an entire sensory organ."

In this chapter, we describe (1) the designs of the present-day unilateral CIs; (2) the performance of those CIs; (3) stimulation that might be added to a unilateral CI to improve performance; and (4) other possibilities for improving performance. Additional information about the performance of the current CI systems is presented in Chap. 9, and information about the designs and performance of prior systems is presented in Wilson (2004, 2006, 2015), Wilson and Dorman (2008, 2009), and Zeng et al. (2008).

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Design and Performance of the Present-Day Cochlear Implants

Components shared by all CI systems now in widespread use are shown in Fig. 1.1. The components include an external sound processor, an external transmitting coil, an implanted receiver/stimulator, a cable from the receiver/stimulator to the implanted electrodes, and the array of the electrodes that are inserted into the scala tympani at the time of surgery. In the illustrated device, the array has 12 sites of stimulation along its length. Other arrays have 16 or 22 sites, although more sites than 8 may not confer further benefits, at least with the present designs and placements of electrodes, and at least with the present processing strategies (see Fig. 1.7 and the accompanying text in the section on "Performance of Unilateral Cochlear Implants"). The other major component in the systems, the user's brain, is not shown in Fig. 1.1. All components are important and must work together to produce good outcomes.

Electrodes

Multiple sites of stimulation and multiple channels of sound processing are needed to maximize performance with CIs. Prior to the 1990s, some developers of CI systems believed that a single site of stimulation and a single channel of processing could be, or might be, just as effective as multiple sites and multiple channels (e.g., House and Urban 1973; Hochmair-Desoyer et al. 1981). However, results from studies conducted at the University of Iowa (e.g., Gantz et al. 1988), along with results from the first prospective, randomized trial of CI devices (Cohen et al. 1993), demonstrated that the multisite and multichannel systems at the times of the studies were better than the contemporaneous single-site and single-channel systems.

As indicated previously, increasing the number of sites up to about eight can produce significant improvements in speech reception. In addition, monopolar stimulation, with currents passed between each intracochlear electrode and a

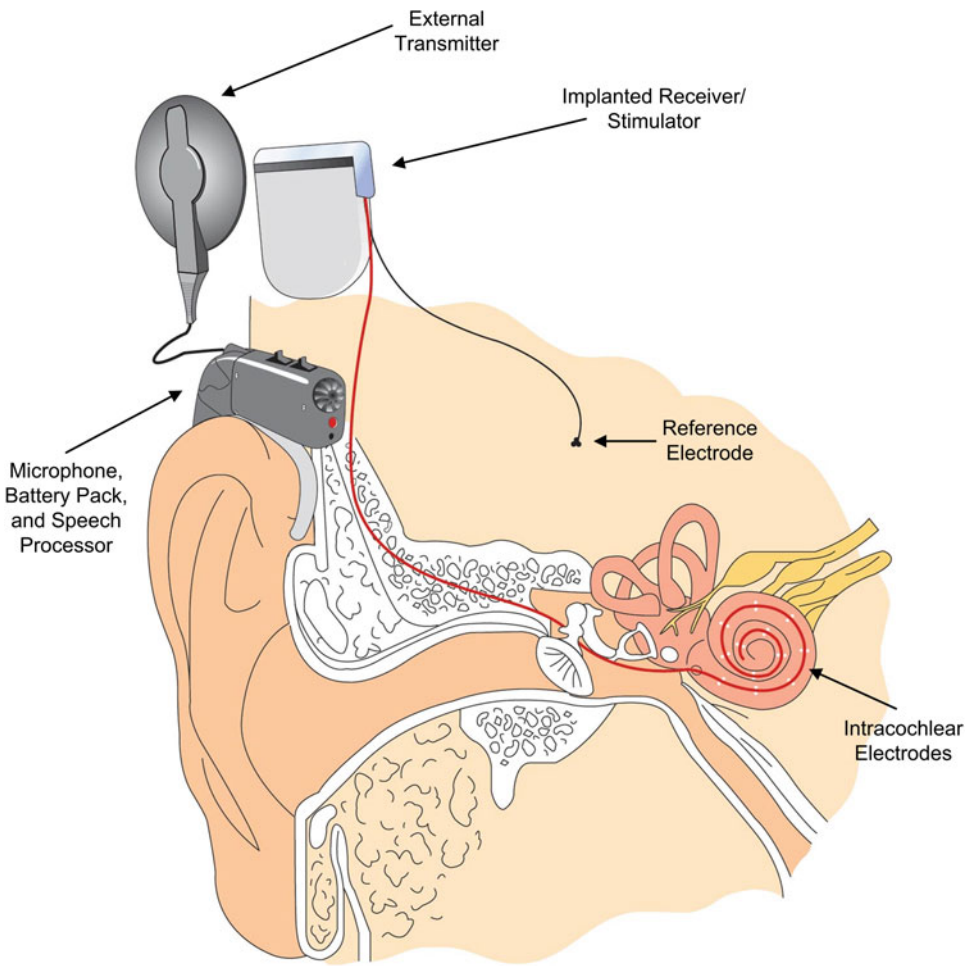

Fig. 1.1 Components of cochlear implant systems (diagram courtesy of MED-EL GmbH of Innsbruck, Austria)

remote reference electrode outside of the cochlea (see Fig. 1.1), is at least as effective as other modes of stimulation, e.g., bipolar stimulation in which currents are passed between intracochlear electrodes, from one electrode to the other in a pair of electrodes. This finding was surprising in that the other modes were thought to produce sharper (more spatially distinct) patterns of electrical stimulation than monopolar stimulation (e.g., Wilson 2004). (Whether the patterns are in fact sharper with the other modes is now an open question; see the two opposing points of view expressed in Kwon and van den Honert 2006, versus Nelson et al. 2011.) However, speech reception scores are at least as good with monopolar stimulation compared to the other modes, and some patients describe monopolar stimulation as sounding more natural than the other modes. Fortunately too, monopolar stimulation requires less battery power than the other modes, and produces much more uniform stimulus magnitudes required for threshold and comfortably loud percepts compared to the other modes. This latter result facilitates the fitting of the device; monopolar stimulation is used in all modern CIs.

Processing Strategies

The external sound processor implements a processing strategy—or a choice among strategies—for transforming the sound input into a set of instructions that are transmitted to the implanted receiver/stimulator via the transcutaneous link that includes the external transmitting coil and the receiving antenna in the implanted receiver/stimulator. Radio-frequency (RF) transmission is used and the instructions are encoded in the RF carrier through some form of modulation. The instructions are decoded upon receipt at the receiver/stimulator using active electronics in that part of the device, and the decoded (recovered) instructions are used to generate the stimuli that are directed to the intracochlear and reference electrodes via cables. (See also the cable for the reference electrode in Fig. 1.1.) Power for the implanted electronics and the stimuli is generated in the receiver/stimulator by rectifying and then smoothing (low-pass filtering) the overall RF signal received by the antenna.

A simple but effective processing strategy for CIs is illustrated in Fig. 1.2. This is the "continuous interleaved sampling" (CIS) strategy that produced a breakthrough in the performance of CIs in the early 1990s (e.g., Wilson et al. 1991; Fayad et al. 2008) and is used to this day as a processing option in all of the devices manufactured by the three largest companies in the field, which together have more than 99 % of the world market. CIS gets its name from the continuous sampling of band energies and presenting that information in an interleaved sequence of stimulation across the utilized intracochlear electrodes in the implant. Multiple other strategies use this same basic approach, with additions to or variations in the processing, as described for example in Wilson and Dorman (2012).

The overarching goal of the design was to represent in the clearest possible way most or all of the information that could be perceived with CIs. That information included the "place" and "temporal" codes for the frequencies of components within sounds, and the amplitude or charge of electrical stimuli for the intensities of the components.

Coding Frequency by Place of Stimulation

For most patients, stimulation of electrodes near the basal end of the cochlea elicits percepts with relatively high pitches and stimulation of electrodes at progressively more apical locations elicits percepts with progressively lower pitches. This representation based on the site of stimulation is the place code for frequencies, a topographic organization that is maintained throughout the ascending auditory pathways all the way up to and including the A1 area of the auditory cortex. Some patients are able to discriminate among a high number of electrodes when the electrodes are stimulated in isolation and at the same pulse rate or sinusoidal frequency. For example, some patients can discriminate among the 22 sites of stimulation provided with the implant devices manufactured by Cochlear Ltd. (Nelson et al. 1995; Zwolan et al. 1997), although results for most patients demonstrate poorer discrimination and no patient tested to date has more than about eight effective sites when the electrodes are stimulated in a speech processor context with rapid sequencing of stimuli among the utilized electrodes. Thus, having more than about eight sites may be "overkill" and we shall return to this point in the sections on "Performance of Unilateral Cochlear Implants" and "Possibilities for Improvements."

Coding Frequency by Rate or Frequency of Stimulation

In addition to the place code for frequencies, stimulation at different rates (for pulses) or frequencies (for sinusoidal or "analog" stimuli) at any one of the multiple sites can produce different pitches up to a maximum rate or frequency beyond which further increases in pitch are not produced. This maximum rate or frequency is called the "pitch saturation limit" for CIs and typically approximates 300 pulses/s or 300 Hz (e.g., Zeng 2002). However, the limit can be higher for exceptional patients, up to or a bit beyond 1000 pulses/s or 1000 Hz for at least one of each patient's electrodes (Hochmair-Desoyer et al. 1983; Townshend et al. 1987; Wilson et al. 1997b). Frequencies in the modulations of pulse trains also have the same limits; for example, the great majority of patients can perceive different frequencies in the modulation waveforms as different pitches up to about 300 Hz but not higher. Discrimination among rates or frequencies below the pitch saturation limit is progressively

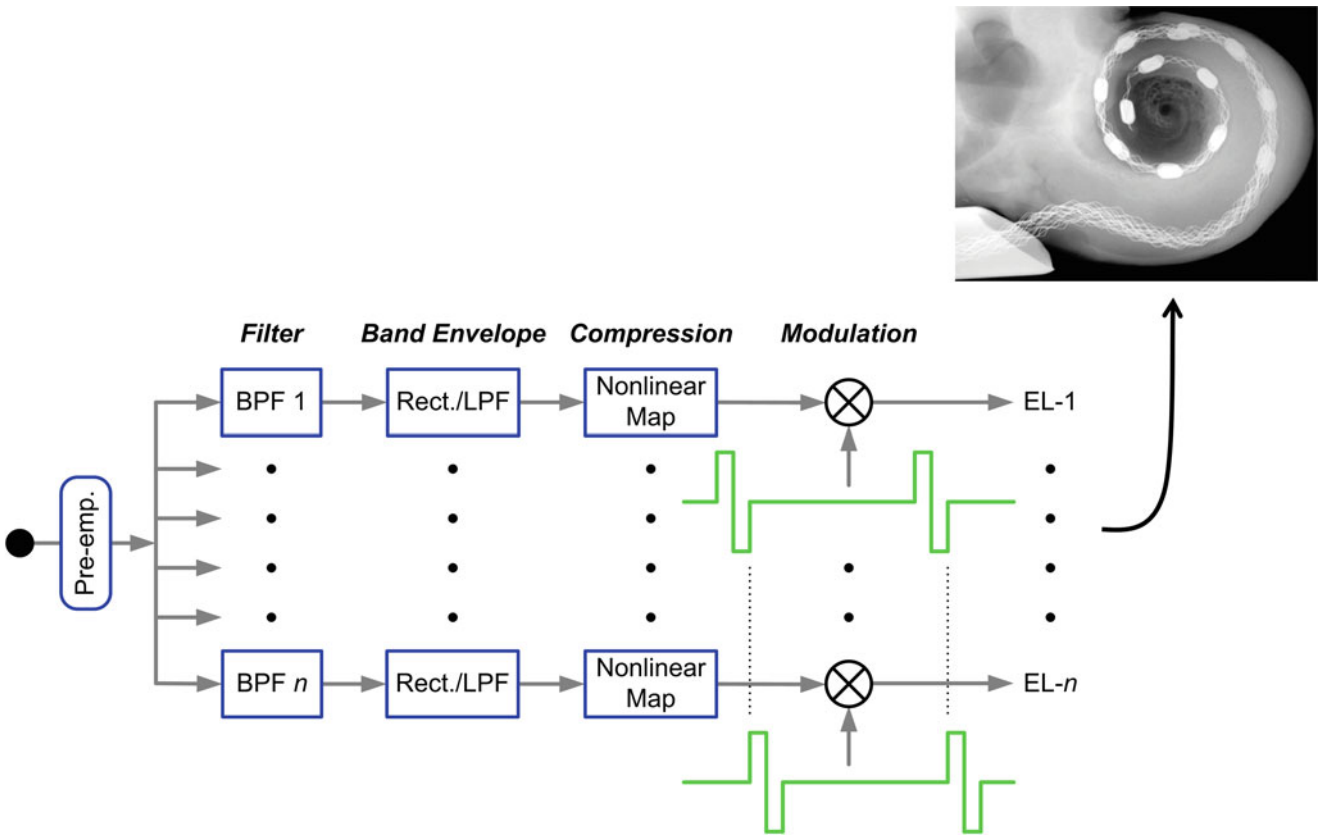

Fig. 1.2 Block diagram of the continuous interleaved sampling (CIS) processing strategy. The diagram is explained in the text. Abbreviations: *Pre-emp.* pre-emphasis filter, *BPF* band-pass filter, *Rect.* rectifier, *LPF* low-pass filter, *EL* electrode. The *circles* with "x" *marks* within them are multiplier blocks, and the *green traces* beneath those blocks are stimulus waveforms (the diagram is adapted from Wilson et al. (1991) and is used here in its modified form with the permission of the Nature Publishing Group; the *inset* is from Hüttenbrink et al. (2002) and is reproduced here with the permission of Lippincott Williams & Wilkins)

worse as the limit is approached and even for low rates or frequencies the discrimination for implant patients is typically ten times worse than the frequency difference limens for subjects with normal hearing (e.g., Zeng 2002). Variation in the rates or frequencies that can produce different pitches for CI users is the temporal code for frequencies; the code is a coarse, but nonetheless useful, representation of frequencies below the pitch saturation limit.

Coding Signal Amplitude

Matching of loudnesses between electric and acoustic stimuli for implant patients who have partial or normal hearing contralateral to the implant has demonstrated the same growths in loudness with (1) equal dB (ratio) increases in intensity on the hearing side or (2) linear increases on the implant side (Eddington et al. 1978; Zeng and Shannon 1992; Dorman et al. 1993). This finding indicates that the growth of loudness in normal hearing can be approximated with a logarithmic mapping (compression) function for electric stimuli, i.e., a logarithmic transformation of acoustic intensities to electric intensities.

The number of discriminable steps in intensity for electrically elicited hearing using balanced biphasic pulses as the stimuli can range from about 7 to about 45 among subjects and electrodes within subjects (Nelson et al. 1996), with an average of about 25 across the subjects and their tested electrodes (Nelson et al. 1996; Chua et al. 2011). These numbers are lower, but not dramatically lower, than the average number of discriminable steps in intensity for listeners with normal hearing, about 83 steps (Nelson et al. 1996).

Design of CIS

In CIS, frequencies of components in the input sound are represented with both the place and temporal codes, and the energies of those components are represented with stimulus intensities. As indicated in Fig. 1.2, multiple channels of processing are used and the output from each channel is directed to a corresponding electrode in the cochlea. Each channel includes a band-pass filter (BPF), an energy (or "envelope") detector, a nonlinear mapping function, and a multiplier, which multiplies (modulates) a train of balanced biphasic pulses with the output of the nonlinear mapping function.

The energy detector illustrated in the figure includes a rectifier (Rect.) followed by a low-pass filter (LPF). Other types of energy detectors can be used, e.g., a Hilbert transform, but the illustrated one is simpler than the alternatives and functions similarly.

The only difference among the channels is the frequency response of the BPFs. The frequency responses range from low to high along a logarithmic scale, which approximates the mapping of frequencies along the cochlear partition in normal hearing. For a six-channel processor, for example, the overall range of frequencies might be from 300 to 6000 Hz and for that range the corner or "break" frequencies of the BPFs would be 300–494, 494–814, 814–1342, 1342–2210, 2210–3642, and 3642–6000 Hz. (The range of frequencies between each pairing of the corner frequencies is the "pass band" for each BPF.) The modulated pulse train for the channel with the lowest center frequency for the bandpass filters is directed to the apicalmost among the utilized electrodes in the implant; the train for the channel with the highest center frequency is directed to the basalmost among the utilized electrodes; and the trains for the channels with intermediate center frequencies are directed to the utilized electrodes at intermediate positions in the implant. This representation approximates the place coding of frequencies in normal hearing, in which high frequencies produce maximal displacements of the basilar membrane at basal positions along its length, and lower frequencies produce maximal displacements at more apical positions.

Within channels, the effective "cutoff" frequency for the energy detectors is set by the frequency response of the LPF. The upper end of the frequency response typically is between 200 and 400 Hz and most commonly at 400 Hz. With that latter setting, frequencies as high as 400 Hz and a bit beyond may be included in the derived energy signal for each channel, and that range of frequencies encompasses and slightly exceeds the pitch saturation limit for most (nearly all) patients. Thus, the temporal information that can be perceived by CI users as a variety of pitches is represented in the energy signal. Representing higher frequencies in this way most likely would be fruitless for all but an exceedingly small fraction of patients, and might well present conflicting cues for the great majority of patients.

The nonlinear mapping function in each channel is used to map the wide dynamic range of the energy signals, which can be as high as about 70 dB, onto the much narrower dynamic range of electrically evoked hearing, which for short-duration pulses usually is between 5 and 20 dB. The mapping is a logarithmic (or sometimes a power law) transformation of the energy variations into pulse amplitude (or pulse charge) variations. This transformation preserves a normal growth of loudness across the dynamic ranges of the energy variations and the derived pulse amplitudes (or charges), as described previously. The mapping also preserves the maximum number of discriminable steps within the dynamic ranges, for each patient and electrodes within patients. (The values of the parameters for the mapping function for each electrode for a patient are derived mathematically from measurements of the currents or charges needed to produce threshold and comfortably loud percepts, and thus the function is customized for each of the utilized electrodes in the implant.)

The modulated pulses for the different channels and corresponding electrodes are interlaced in time so that stimulation at any one electrode is not accompanied by simultaneous or overlapping stimulation at any other electrode in the implant. This interleaving of pulses across electrodes eliminates a principal component of "channel" or electrode interactions due to the direct summation of the electric fields from the different (simultaneously stimulated) electrodes. This direct summation component is much larger than other components of interaction that are produced by neural refractory and temporal summation effects, which are present even with nonsimultaneous stimulation (e.g., Favre and Pelizzone 1993). Without the interleaving, the high levels of interaction (or "cross talk") among electrodes would (1) produce spurious cues that are not related to the signal at the input to the sound processor and (2) greatly degrade the independence of the represented channel outputs.

For an undistorted representation of the temporal variations in the modulation waveforms, the pulse rate for each channel and corresponding electrode must be at least four times higher than the highest frequency in the waveform for the channel (Busby et al. 1993; Wilson et al. 1997a). This fact became known as the "4× oversampling rule" for CIs. Thus, in a typical implementation of CIS the cutoff frequency for the energy detectors might be 400 Hz and the pulse rate for each channel and corresponding electrode might be around 1600/s or higher. (Such high pulse rates cannot be supported by all transcutaneous links and receiver/stimulators, so in those cases at least the pulse rate must be reduced.)

Most fortunately, the typical cutoff frequencies for the energy detectors also include most or all of the range of fundamental frequencies (F0s) in human speech. Thus, the represented modulation waveforms may convey information about the overall energy in a band, F0s for periodic sounds, and the random fluctuations in energy that are characteristic of aperiodic sounds. (Whether other representations of F0s and periodic/aperiodic distinctions might be more salient is not clear at this point and is a topic of current research.) This within-channel information may be especially helpful for perceiving different voices as such; distinguishing interrogative versus declarative intent by a speaker; and discriminating among voiced, unvoiced, and mixed voiced plus unvoiced consonants in speech. In addition, the information could enable perception of F0s and periodic versus aperiodic components in other sounds such as music.

With CIS, frequencies above about 300 Hz are represented with the site(s) of stimulation, and frequencies below about 300 Hz are represented with the temporal variations in the modulation waveforms. Intensities of energies within bands are represented with the modulated pulse amplitudes. The pulses for the different channels are presented from one electrode to the next until all of the utilized electrodes are stimulated. This cycle of stimulation across electrodes is repeated continuously and at a high rate so that energy variations up to the pitch saturation limit for most patients are represented at each of the electrodes. The pulse rate for all electrodes is the same. No assumptions are made in the design about sounds in the environment or in particular how speech is produced or perceived.

Prior processing strategies either (1) extracted specific features from the input and represented those features only in the stimuli directed to the intracochlear electrodes or (2) presented stimuli simultaneously or with substantial overlaps at the electrodes. In addition, prior strategies using nonsimultaneous pulses as the stimuli presented the pulses at relatively low rates. CIS produced a large jump up in performance compared to the prior strategies and is the basis for many of the strategies that were developed subsequently. CIS remains as the principal strategy against which other strategies are compared.

A more detailed description of CIS and how it departed from the past is presented in Wilson (2015). And considerable further information about the prior and subsequent strategies is presented in Wilson (2004, 2006), and in Wilson and Dorman (2008, 2009, 2012).

CIS exemplifies the design principles that are used in all of the current CI systems. Those principles include (1) representing at least most of the information that can be perceived according to place, frequency, and intensity of stimulation; (2) minimizing electrode interactions; and (3) using appropriate mapping functions and other aspects of processing to minimize perceptual distortions.

Performance of Unilateral Cochlear Implants

Snapshots of the performance of unilateral CIs as of the mid-1990s and today are presented in Fig. 1.3. The two left panels show results from a multicenter study in Europe to evaluate the COMBI 40 implant device (Helms et al. 1997) and the two right panels show results for all postlingually deafened adults who were implanted at the Vanderbilt University Medical Center, USA, from the beginning of 2011 through early 2014 using a variety of the devices now in widespread use (data provided by author RHG). The columns in each panel show scores (circles) for different times after the initial fitting of the device for each subject in the Helms et al. study or for each implanted ear in the Vanderbilt dataset. All of the subjects in the Helms et al. study were implanted unilaterally, and most of the subjects in the Vanderbilt cohort were implanted unilaterally as well, but the rest received bilateral CIs, either sequentially or simultaneously. For those latter subjects, each ear was tested separately and thus data are shown for ears rather than subjects in the right panels. The subjects who participated in the Helms et al. study also were postlingually deafened adults. Selection criteria for those subjects included (but were not limited to) a patent scala tympani as demonstrated in preoperative radiologic scans; at least relatively normal gross anatomy of the cochlea as also demonstrated in the scans; no prior CI; no middle- or outer-ear pathologies that could impede cochlear implantation; a duration of profound hearing loss less than or equal to 50 % of a candidate's lifetime; and "general health good enough to allow testing at scheduled intervals." In contrast, the Vanderbilt cohort included all adults with postlingual onsets implanted there from 2011 through early 2014, many of whom would not have met one or more of the selection criteria for the Helms et al. study. On the other hand, the patients in the Vanderbilt cohort had more residual hearing on average than the subjects in the Helms et al. study, as the audiological restrictions for implant candidacy had been relaxed in multiple steps from the mid-1990s onward.

The tests in the Helms et al. study and at Vanderbilt included measures of word and sentence recognition. Scores for the sentence tests are shown in the top panels in Fig. 1.3, and scores for word tests in the bottom panels. The Hochmair-Schulz-Moser (HSM) sentences (Hochmair-Desoyer et al. 1997), or their equivalents in languages other than German, were used in the Helms et al. study, and the Arizona Biomedical Institute (AzBio) sentences (Spahr et al. 2012) were used at Vanderbilt. The AzBio sentences include multiple talkers and fewer contextual cues than the HSM sentences, and thus are far more difficult than those or other "everyday" sentences uttered by a single talker (Gifford et al. 2008; Spahr et al. 2012). For the word tests, the Freiburger monosyllabic words (Hahlbrock 1953, 1970) or their equivalents for languages other than German were used in the Helms et al. study, and the monosyllabic consonant-nucleus-consonant (CNC) words (Peterson and Lehiste 1962) were used at Vanderbilt. All tests at Vanderbilt were conducted in English. The word tests were comparable in difficulty for the different languages in the Helms et al. study, and between the Helms et al. study and the Vanderbilt measures. In particular, none of the tests included contextual cues and all of the tests used the CNC structure and single talkers. All 55 subjects who participated in the Helms et al. study were tested at all intervals after the initial fitting and thus the number of data points in each column in each of the left panels is 55. Most of the presented data are from Helms et al. (1997), and the remaining data were collected after that publication and provided by Jan Helms (the supplemental

data are reported in Wilson 2006). A total of 267 ears were tested at Vanderbilt, and different numbers of ears were tested at the different intervals, depending on patient availability and when the ear was implanted (of course, recent implants could preclude tests at the later intervals). One of the columns in the right panels includes 183 scores, and the other columns include fewer scores. The numbers of scores vary across the columns in each panel and between like columns in the two panels. The mean of the scores in each column in each panel of Fig. 1.3 is shown with the horizontal red line. CIS was used for the COMBI 40 device, and a variety of processing strategies including CIS were used for the various devices implanted at Vanderbilt, which included devices manufactured by Cochlear Ltd., Advanced Bionics LLC, and MED-EL GmbH. All tests were conducted with hearing alone and without feedback as to correct or incorrect responses. The test items were unknown to the subjects prior to testing. Direct-input or live-voice presentations were used in the Helms et al. study for the word and sentence tests, respectively, whereas free-field presentations in audiometric booths were used at Vanderbilt for all tests. The level of the presentations at Vanderbilt was 60 dBA at the subject's

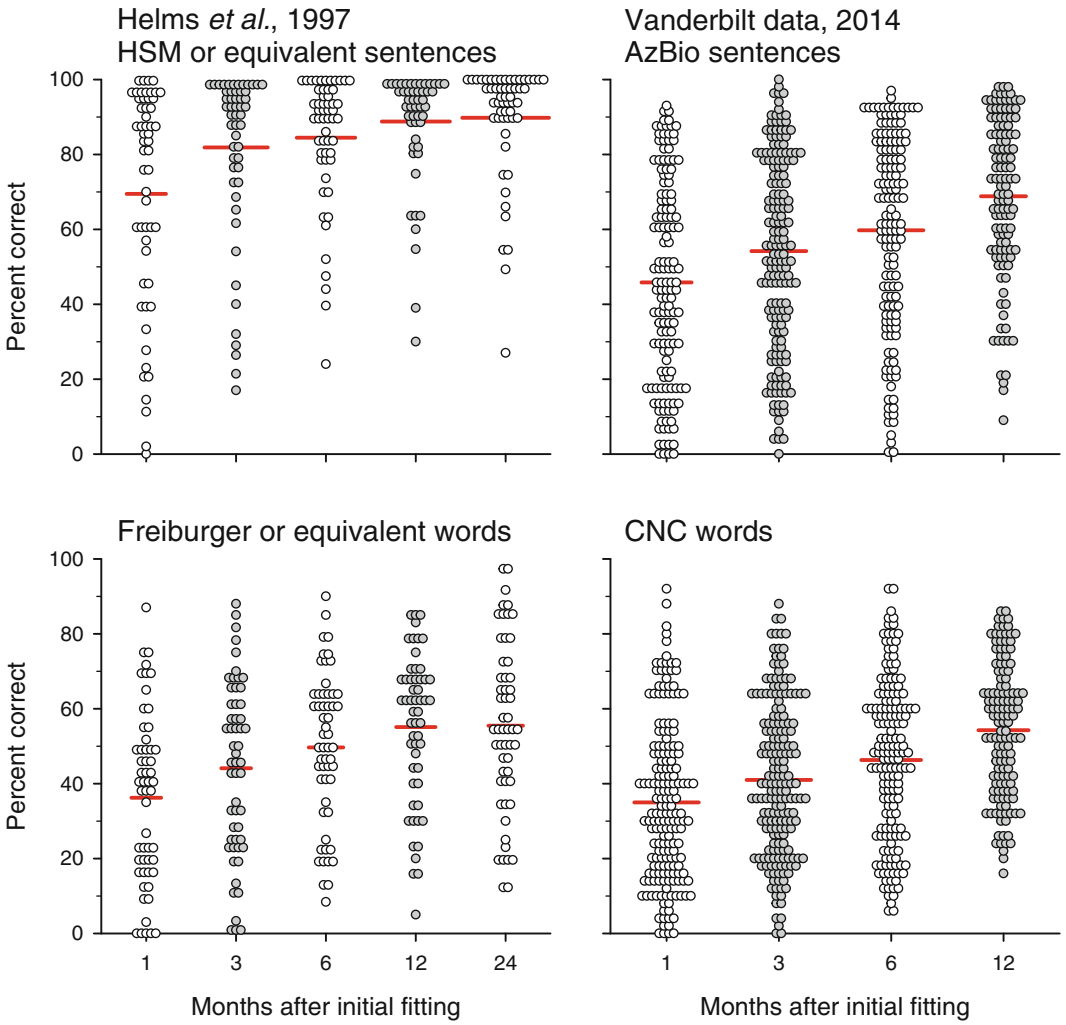

Fig. 1.3 Speech reception scores (*circles*) for cochlear implant (CI) subjects. Data from a multicenter study in Europe by Helms et al. (1997) are presented in the *left panels*, and data collected between 2011 and the first part of 2014 at the Vanderbilt University Medical Center in the USA are presented in the *right panels*. Tests for Helms et al. subjects included recognition of the Hochmair-Schulz-Moser (HSM) sentences or their equivalents in languages other than German, and recognition of the monosyllabic Freiburger words or their equivalents in languages other than German. Tests for the Vanderbilt subjects included recognition of the Arizona Biomedical Institute (AzBio) sentences and recognition of the monosyllabic consonant-nucleus-consonant (CNC) words. The *columns* within the panels show scores for different times after the initial fitting of the device. All tests were conducted with hearing alone and without feedback as to correct or incorrect responses. The Helms et al. subjects used the COMBI 40 CI device and its implementation of the continuous interleaved sampling (CIS) processing strategy, and the Vanderbilt subjects used a variety of the latest devices and strategies. All of the subjects were adults with postlingual onsets of severe or profound hearing loss. The *red horizontal lines* show the means of the scores at each of the test intervals for each of the tests. Many of the scores are displaced slightly along the abscissa and binned into small intervals along the ordinate to aid in the reading of each panel

location, and a "conversational level" was used for the sentence presentations in the Helms et al. study, which typically approximates 60 dBA.

In both sets of data, scores for both the word and sentence tests improve over time, out to 3–12 months after the initial fitting of the device. Scores for the sentence tests are substantially higher than the scores for the word tests; this finding is not surprising given that the word tests do not include the contextual cues available in the sentence tests. Scores for the sentence tests become progressively more clustered near the top with increasing time after the initial fitting out to 1 year for the Helms et al. data and some slight clustering is observed in the Vanderbilt data at the 6- and 12-month intervals only. As might be expected, scores for the AzBio sentences are substantially lower than the scores for the HSM or equivalent sentences. Indeed, the latter scores show clear ceiling effects, whereas ceiling effects are not obviously encountered with the AzBio sentence tests. At the 2-year interval for the HSM or equivalent sentence tests, the mean score is about 90 % correct and the median score is 95 % correct. This high level of performance is completely consistent with high levels of everyday communications. In fact, most CI recipients today have little or no difficulty in using the telephone for communications even with unfamiliar persons or unknown and varying topics. Scores for the other tests are consistent with everyday communications as well; the lower scores for those tests simply reflect their greater difficulty.

An especially interesting aspect of the data in Fig. 1.3 is the time course of improvements for each of the tests. This aspect is easier to see in Fig. 1.4, which shows the means and standard errors of the means (SEMs) for the cases in which both sentence and word scores are available at all of the test intervals. Those cases include the 55 subjects in the Helms et al. study (data in the left panels of Fig. 1.3) and 29 of the ears that were tested at Vanderbilt (subset of the data in the right panels of Fig. 1.3). In addition, means and SEMs are shown in Fig. 1.4 for the additional intervals included in the Helms et al. study for the sentence tests, for the same 55 subjects.

One-way, repeated-measures analyses of the variance (RM ANOVAs) indicate highly significant improvements in the mean scores over time for the recognition of both sentences and words for the Helms et al. subjects and for both sentences and words for the ears tested at all intervals at Vanderbilt ($p < 0.001$ in all cases). Results from *post hoc* pairwise comparisons using the Holm-Sidak method are presented in Table 1.1. In broad terms, significant improvements in the scores for each of the tests are observed out to 3–12 months after the initial fitting of the device for each subject or ear. Asymptotic scores are reached earlier with the HSM or equivalent sentences than for the other tests, most likely due to ceiling effects. (The onset of ceiling effects can be seen in the narrowing of the error bars in Fig. 1.4 starting at the 3-month interval.) Six to 12 months (or perhaps more in the Vanderbilt data) are required to reach asymptotic performance with the more difficult tests. A clear plateau in performance starting at 12 months is seen in the scores from the word tests in the Helms et al. data.

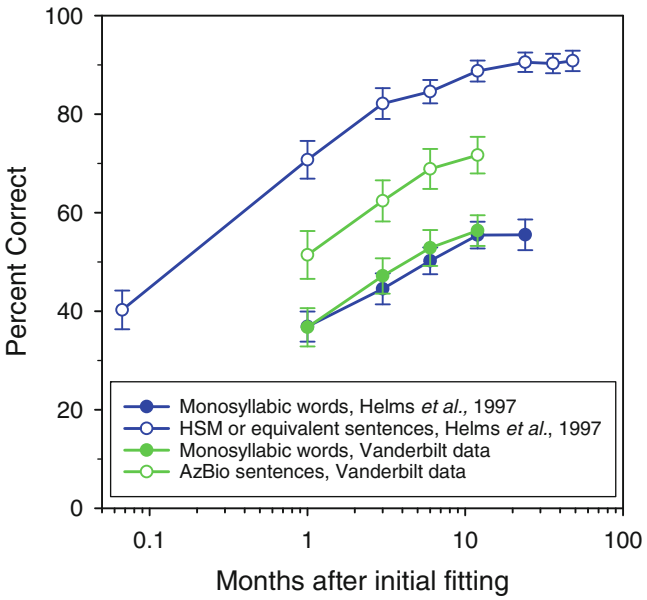

Fig. 1.4 Means and standard errors of the means (SEMs) for the recognition of words (*closed symbols*) and sentences (*open symbols*). The data are from (1) the 55 subjects tested at all of the indicated times after the initial fitting of the cochlear implant in the study by Helms et al. (1997) and (2) the 29 ears tested with both words and sentences at all of the indicated times at Vanderbilt. Details about the tests are provided in the text. Note that the time scale is logarithmic

These long time courses of improvements (on average) are consistent with changes in brain function (e.g., Moore and Shannon 2009; Lazard et al. 2012), but not with changes at the periphery, which are far more rapid. For example, any fibrous encapsulation of the electrode array is usually complete within several weeks of implantation and reductions in electrode impedances typically asymptote within minutes or hours following initial stimulation. Those or other short-term changes are not correlated with the improvements in speech reception.

Scores for the AzBio test are intermediate to the scores for the word tests on the one hand, and the scores for the HSM or equivalent sentence tests on the other hand. In addition, the scores for the AzBio test do not exhibit ceiling effects for the population (although some scores are above 90 % correct and the distribution of scores is slightly skewed toward the top at the 6- and 12-month intervals, as shown in Fig. 1.3). These facts make the AzBio test more useful than easier sentence tests for discriminating possible differences among devices, subjects, processing strategies, and different amounts of experience with a CI. Also, the AzBio test more closely approximates real-world situations than the monosyllabic

Table 1.1 Results from *post hoc* comparisons using the Holm-Sidak method, following significant repeated-measures ANOVAs for each of the tests in Fig. 1.4

Data set	Test	Results
Helms et al.	Sentences	Scores at the intervals between and including 1–48 months are > the score at the 2-day interval; scores at the intervals between and including 3–48 months are > the score at the 1-month interval
	Words	Scores at the intervals between and including 6–24 months are > the score at the 1-month interval; scores at the 12- and 24-month intervals are > the score at the 3-month interval; the score at the 3-month interval is > the score at the 1-month interval
Vanderbilt	Sentences	All pairwise comparisons are significantly different except for the scores at the 12- and 6-month intervals
	Words	All pairwise comparisons are significantly different except for the scores at the 12- and 6-month intervals

The criterion for significance for the *post hoc* comparisons was $p < 0.05$

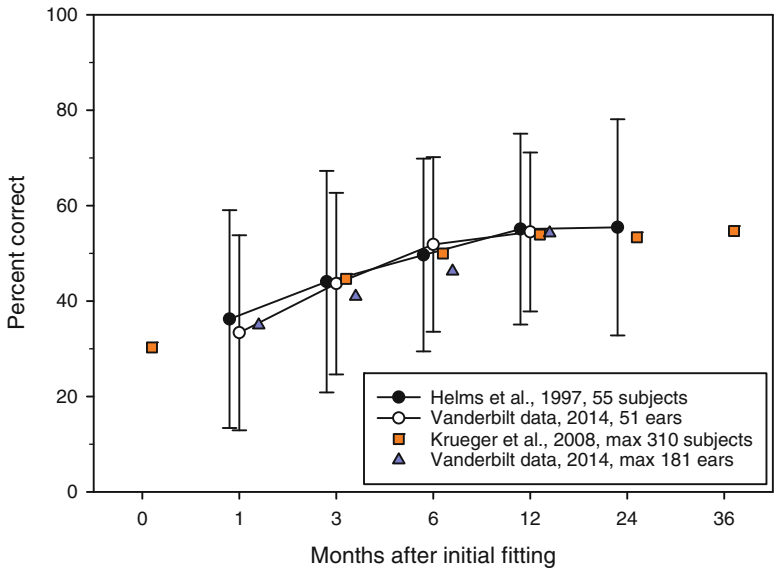

Fig. 1.5 Means of percent correct scores for the recognition of monosyllabic words by cochlear implant subjects. The sources of the data are indicated in the legend. Standard deviations are shown for (1) the 55 subjects in the Helms et al. study who took the test at all five intervals after the initial fitting of the device and (2) the 51 ears tested at all four intervals at Vanderbilt. Some of the symbols are offset slightly along the abscissa to aid in the reading of the figure (the figure is adapted from Wilson (2015), and is used here with the permission of Elsevier BV)

word tests, and thus might be a better measure of everyday performance. Author RHG and colleagues have therefore advocated the use of the AzBio test, instead of easier tests, in standard clinical practice (e.g., Gifford et al. 2008).

A striking finding shown in Fig. 1.4 is the complete overlap in scores for like intervals from the word tests conducted at Vanderbilt and the word tests conducted in the Helms et al. study (two sets of filled symbols in the figure). That is, no improvement in the recognition of monosyllabic words was observed between the mid-1990s, when the Helms et al. data were collected, and 2011–2014, when the Vanderbilt data were collected, despite (1) the introductions of new processing strategies and electrode designs; (2) greater numbers of processing channels and associated sites of stimulation; and (3) substantial relaxations in the audiological restrictions for implant candidacy during the intervening period. This observation is a little sobering and frustrating of course, given the tremendous efforts by our teams and many others to improve performance in the period. The COMBI 40 device, with its eight sites of stimulation in the cochlea and its use of CIS, has yet to be surpassed, at least according to these data and with the caveat that some of the experimental conditions were different between the Helms et al. study and the Vanderbilt measures.

We would like to emphasize here that the data presented in Figs. 1.3 and 1.4 are from large populations of sequentially implanted patients that include all adults with postlingual onsets who received their implants as part of a large clinical trial (the Helms et al. study) or from a large clinical center (Vanderbilt) and who were all treated and tested uniformly. The subjects from such populations represent the broad clinical experience and include CI users with relatively poor outcomes even with the best medical care and the best of the available devices. Higher scores have been reported (e.g., Blamey et al. 2013; Holden et al. 2013; Gifford et al. 2014a), but those scores were obtained with more highly restricted populations and sometimes with different tests and test conditions within the populations. The figures provide an accurate and fully representative picture of where we were as a field in the mid-1990s and recently, from 2011 through early 2014.

The similarities in performance across the years shown in Fig. 1.4 are even more evident in Fig. 1.5, which shows data

for the recognition of monosyllabic words from three sources and at various points in time. The data include the scores for the 55 subjects who participated in the Helms et al. study, and the scores for the 51 ears that were tested at all intervals at Vanderbilt, for the recognition of monosyllabic words. Means and standard deviations are shown. In addition, the means of the scores for all ears tested at Vanderbilt at each of the intervals are shown with the purple triangles, and the means for subsets of the 310 subjects in Group 5 in a retrospective chart study by Krueger et al. (2008) are shown with the orange squares. (Group 5 included the subjects using the latest devices and processing strategies as of 2008; the subjects in Groups 1–4 used earlier devices and processing strategies.) A maximum of 310 subjects were tested at each of the indicated intervals in the Krueger et al. study, and a maximum of 183 ears were tested at any one interval at Vanderbilt. As mentioned previously, the Freiburger or equivalent tests were used in the Helms et al. study and the CNC word test was used at Vanderbilt. The subjects in the Krueger et al. study were all implanted at the Medizinische Hochschule Hannover in Hannover, Germany, and the Freiburger word test was used exclusively for those subjects. All of the subjects from the various studies were adults when they received their implant(s) and had postlingual onsets of severe or profound hearing loss. In addition, malformations of the cochlea or a handicap or handicaps in addition to hearing loss were among the exclusion criteria in the Krueger et al. study. The mode and level of presentations of the test items were not specified in the paper by Krueger et al. The Helms et al. data were collected in the mid-1990s, the Kruger et al. data in the mid-to-late 2000s, and the Vanderbilt data from 2011 to early 2014. With the noted exceptions, the Krueger et al. data also include scores from every single adult patient with a postlingual onset implanted at a large clinic (in fact, the world's largest CI clinic) over a span of years, ending in the year 2008 in this case.

The result from a one-way RM ANOVA for the Helms et al. data was mentioned previously, and a one-way RM ANOVA for the larger set of ears for the monosyllabic word test only in the Vanderbilt data also was highly significant ($p < 0.001$). *Post hoc* comparisons using the Holm-Sidak method for the latter data again showed that the mean score at any one interval is significantly different from the scores at the other intervals, except for the scores at the 6- and 12-month intervals.

The scores for these two sets of data completely overlap, at all of the common intervals. In addition, the standard deviations are the same for the two sets of data.

Further, the mean scores for all of the ears tested at each interval at Vanderbilt, and the mean scores for subsets of subjects in Group 5 tested at each of the intervals in Hannover, closely approximate each other and the mean scores for the 51 ears tested at all intervals at Vanderbilt and for the 55 subjects tested at all intervals in the Helms et al. study. In all sets of data, scores increase out to 6–12 months after the initial fitting of the device, and the score for 12 months and beyond is around 55 % correct. That latter score has become the "gold standard" for present CI devices and processing strategies, at least for unilateral CIs and large groups of unselected subjects.

Of course, experimental conditions varied somewhat among the studies, and one or more of the differences may have affected the results. However, the correspondence in the data across the studies is remarkable and the available results do not indicate any improvements in isolated word recognition performance from the mid-1990s through the beginning of 2014.

Although the mean scores for the recognition of monosyllabic words often approximate 55 % correct for experienced users of unilateral CIs only, the variability in scores is high, as shown by the high standard deviations in Fig. 1.5 and the nearly uniform distributions of the individual scores for those tests in Fig. 1.3. In the Helms et al. data, for instance, the individual scores at the 24-month interval range from about 10 % correct to nearly 100 % correct. One of the principal remaining questions in implant research is why the range is so large, and a related question is how to bring up the lower end of the range.

With that said, performance for some patients is spectacular. Some examples can been seen in Fig. 1.3 and a further example is presented in Fig. 1.6, which shows scores for a CI subject (subject HR4) versus scores for six undergraduate students at Arizona State University with clinically normal hearing (the data are from Wilson and Dorman 2007). The tests included recognition of the CNC words; recognition of the City University of New York (CUNY) sentences; recognition of the Hearing in Noise Test (HINT) sentences (here presented without noise); recognition of the AzBio sentences; identification of 20 English consonants in a /e/-consonant-/e/ context; identification of 13 American English vowels in a /b/-vowel-/t/ context; and recognition of separate lists of the CUNY and AzBio sentences presented in competition with a four-talker babble at the speech-to-babble ratio of +10 dB for both tests and at +5 dB for the AzBio test. The sentences and words were unknown to the subjects prior to the tests and the presentations of the consonants and vowels were randomized. No feedback was given as to correct or incorrect responses and all tests were conducted with hearing alone. The CI subject used an implant device manufactured by Advanced Bionics LLC and its implementation of the CIS strategy. For this subject, 16 channels of processing and corresponding sites of stimulation were used, and the pulse rate at each site was 1449/s.

The scores for the CI subject (HR4) are statistically indistinguishable from the means of the scores for the subjects with normal hearing for all of the tests except for the AzBio

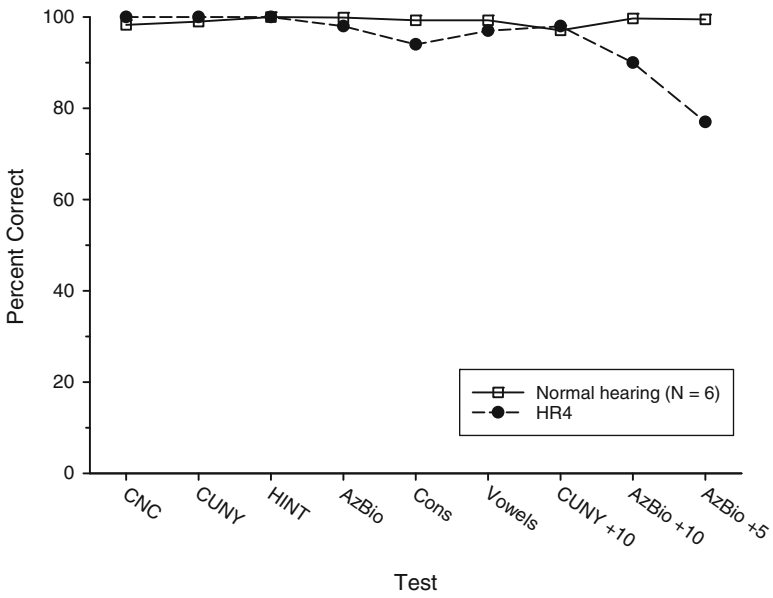

Fig. 1.6 Scores for cochlear implant subject HR4 (*closed circles*) and six subjects with normal hearing (*open squares*) for a battery of speech reception tests. The tests are identified in the text. Means are shown for the subjects with normal hearing; the maximum standard error of the means was 1.1 %. The +10 and +5 labels denote the speech-to-babble ratios of +10 and +5 dB, respectively (the data are from Wilson and Dorman (2007), and the figure is adapted from Wilson (2015), and is used here with the permission of Elsevier BV)

sentences presented in competition with the multitalker babble. The CI subject achieved a perfect score in the monosyllabic word test, and that subject scored at or near the ceiling for most of the other tests, including tests such as the AzBio sentences in quiet that are far more difficult than the sentence tests administered in standard clinical practice.

HR4 and other similarly performing subjects were postlingually deafened and had been profoundly deaf before receiving their CIs. After the CIs and some experience with them, the speech reception scores achieved by these subjects with their restored hearing alone are in the normal ranges according to the standard clinical measures.

This is not to say however that these subjects have normal hearing. As seen for example in Fig. 1.6, the top performers still have difficulties in listening to speech presented in competition with noise. That is a serious problem, because real-world environments such as restaurants and workplaces are notoriously noisy, with typical speech-to-noise ratios (S/Ns) on the order of zero to +5 dB (Pearsons et al. 1977). In addition, and what is not shown in Fig. 1.6, is the great concentration that must be exerted by the CI subjects in achieving their high scores. In contrast, subjects with normal hearing achieve the scores without obvious conscious effort. Furthermore, even the top-performing CI subjects have trouble in fully perceiving sounds that are more complex than speech, such as most music. Full perception of those sounds may require a finer grained representation of frequencies than is possible with the present-day unilateral CIs.

Nonetheless, the high levels of speech reception shown in Fig. 1.6 and elsewhere are impressive. Indeed, they are an existence proof that the peripheral representation is adequate for reception of difficult speech items—and for reception of speech presented in adverse situations—for some patients. That such a sparse representation could be adequate is both surprising and fortunate.

The Number of Electrodes Does Not Equal the Number of Perceptual Channels

The top and average performances of the present-day unilateral CIs are all the more surprising when one appreciates that no more than eight effective channels are available to the users of those CIs, even if the users have a higher number of discriminable electrodes. This fact is illustrated in Fig. 1.7, which shows speech reception scores with different numbers of processing channels and corresponding sites of stimulation in the cochlea. The top panel shows results from one of author BSW's prior laboratories at the Research Triangle Institute in North Carolina, USA, for a subject using a Cochlear Ltd. implant with 22 sites of stimulation in the scala tympani, and the bottom panel shows results from a laboratory within the Manchester Cochlear Implant Programme in the UK, for 11 subjects using MED-EL COMBI 40+ implants with 12 sites of stimulation in the scala tympani (data from Garnham et al. 2002). The tests for the subject in the top panel were identification of 24 English consonants in a /a/-consonant-/a/ context, presented in quiet and in competition with speech-spectrum noise at the S/N of +5 dB (additional details about the tests and results from additional subjects are presented in Lawson et al. 1996; Wilson 1997). The tests for the subjects in the bottom panel were recognition of the Bamford-Kowal-Bench (BKB) sentences; identification of 16 English consonants also in a /a/-

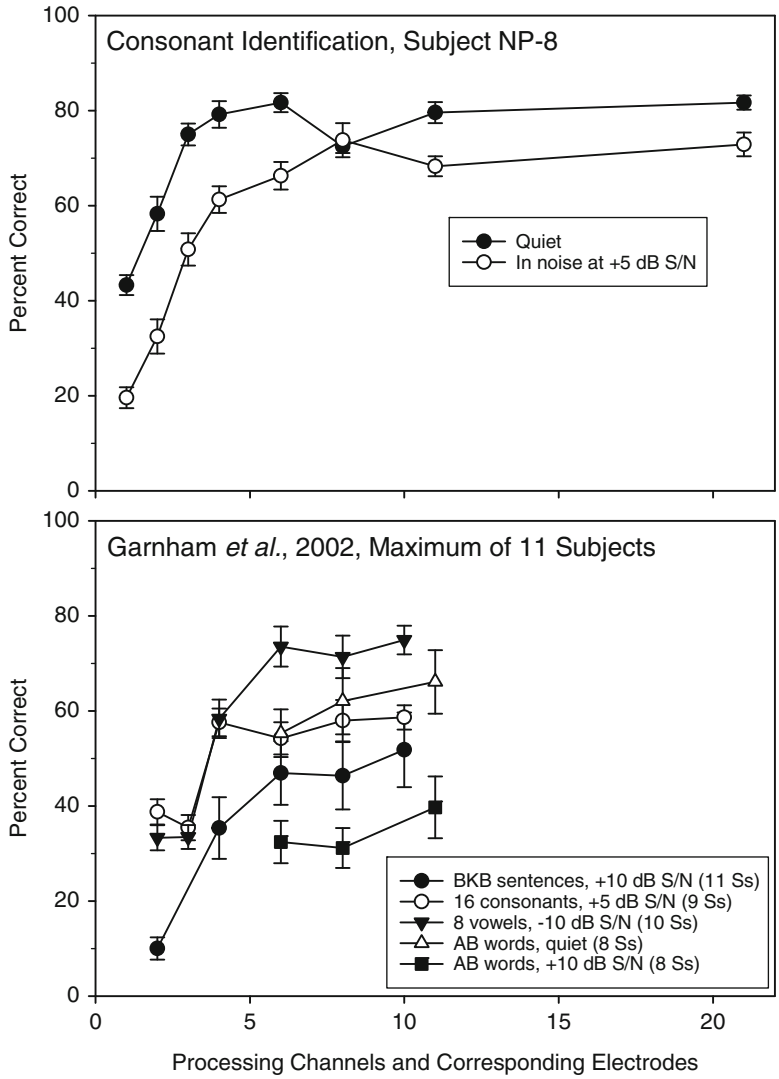

Fig. 1.7 Speech reception scores for different numbers of processing channels and corresponding electrodes. The continuous interleaved sampling (CIS) processing strategy was used. Means and standard errors of the means are shown. The data presented in the *top panel* are from one of author BSW's prior laboratories and the experimental conditions are described briefly in the text and more completely in Lawson et al. (1996) and Wilson (1997). The data presented in the *bottom panel* are from Garnham et al. (2002). The tests are identified in the text, and additional abbreviations in the figure are S/N for speech-to-noise ratio and Ss for subjects (the figure is from Wilson and Dorman (2008), and is reproduced here with the permission of Elsevier BV)

consonant-/a/ context; identification of 8 British vowels in a /b/-vowel-/d/ context; and recognition of the Arthur Boothroyd (AB) monosyllabic words. Many of the speech items were presented in competition with pink noise at the S/Ns indicated in the legend for the figure. A maximum of 11 subjects took each test. All 22 of the intracochlear sites could be discriminated by the subject in the top panel when the sites were stimulated in isolation, one after the other with pauses in between. Users of the MED-EL implants (such as the subjects for the tests in the bottom panel) usually can discriminate a high proportion if not all of their intracochlear sites, also when stimulated in isolation. (The center-to-center distance between adjacent sites in the COMBI 40+ and some other MED-EL implants is 2.4 mm, whereas the distance for the Cochlear Ltd. implants is 0.75 mm; the greater separation in the MED-EL implants generally leads to higher discrimination among the sites.) All tests were conducted with hearing alone and without prior knowledge of the test items or feedback as to correct or incorrect responses. CIS was used for all subjects. Means and SEMs are shown in the figure.

The results from both sets of tests demonstrate asymptotes in performance as the number of processing channels and corresponding electrodes is increased beyond 3, 4, or 6, depending on the test. This limitation is present despite the fact that many patients—including the subjects in the two studies in Fig. 1.7—can discriminate more electrodes when stimulated in isolation, compared with the number of effective channels in a speech processor context, when all of the utilized electrodes are stimulated in rapid sequences. Possibly, temporal interactions among electrodes, such as might be produced with neural refractory or temporal summation effects, impose limits on the maximum number of effective channels with the present-day processing strategies, electrode designs, and electrode placements. An increase in the number of effective channels also could lead to a breakthrough in the design and performance of unilateral CIs.

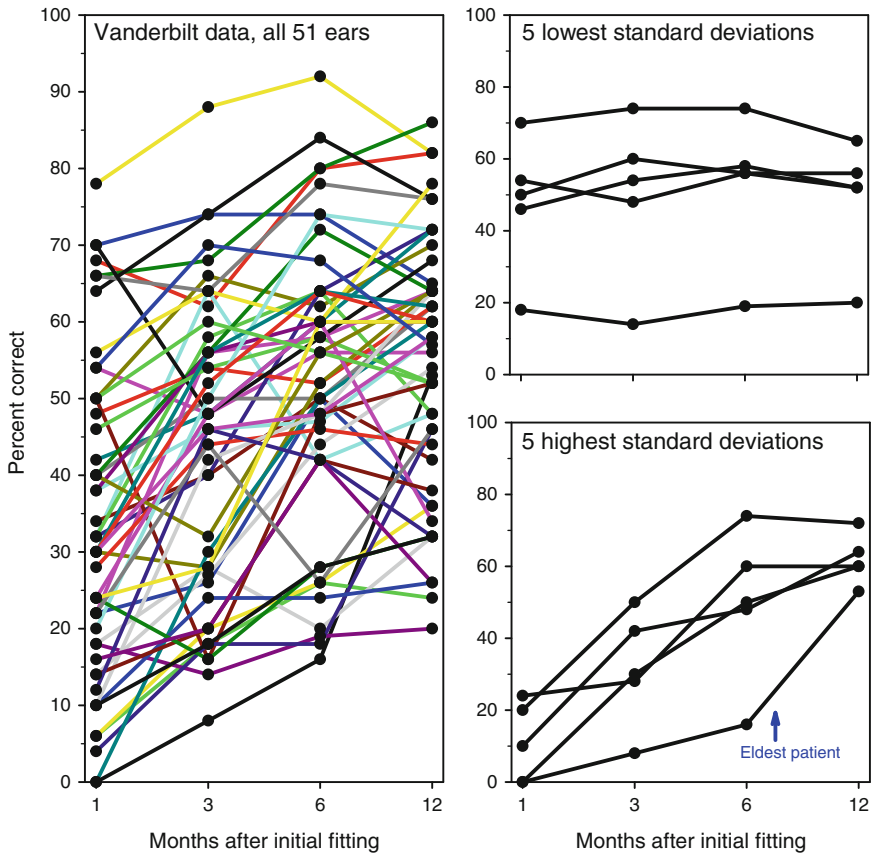

Fig. 1.8 Scores for the recognition of monosyllabic words for (1) the 51 ears tested at all four intervals at Vanderbilt (*left panel*); (2) the 5 among the 51 ears with the lowest standard deviations for the scores across the intervals (*upper right panel*); and (3) the 5 among the 51 ears with the highest standard deviations (*lower right panel*). The data are from 46 patients, 5 of whom were implanted bilaterally. The scores for the ear implanted the latest in life among the patients are identified in the *lower right panel*. That ear was implanted at age 83.6 for the patient, and that was the only ear implanted for the patient. The ear implanted the earliest in life among the 46 patients was implanted at age 27.7 for the patient

Results from the studies in Fig. 1.7 are mirrored by the results from multiple other studies, in which the CIS and other strategies were used, and in which a variety of implant devices were used (Lawson et al. 1996; Fishman et al. 1997; Wilson 1997; Kiefer et al. 2000; Friesen et al. 2001). The findings are always the same; that is, no more than about eight channels are effective for any subject, device, test, or processing strategy. And in many cases, the number is lower than eight.

Thus, the representation provided by the present-day unilateral CIs is even sparser than the number of intracochlear electrodes. In addition, many aspects of the intricate processing in the normal cochlea are not included in the processing or in the neural representations provided by those CIs. (Many of these missing aspects are listed and described in Wilson and Dorman 2007.) The brain can somehow utilize this seemingly impoverished input, and that ability enables the performance of the present-day devices.

Variability in Performance

A final important aspect of the speech reception data for unilateral CIs is the variability among subjects in improvements in scores during the initial 6–12 months of implant use. Although the mean scores for populations of subjects improve in that period (Figs. 1.3, 1.4, and 1.5), not all subjects show improvements. This fact is illustrated in Fig. 1.8, which presents in the left panel recognition of monosyllabic words for all 51 ears that were tested at all intervals at Vanderbilt (the mean scores are shown with the open circles in Fig. 1.5) and in the right panels the sets of scores with the five lowest (upper panel) and five highest (lower panel) standard deviations across the intervals. The high variability in improvements over time is evident in the left panel, and patterns of scores showing no improvements and large improvements are evident in the right panels. The upper right panel shows patterns for the lower tenth of the population in terms of improvements over time, and the lower right panel shows patterns for the upper tenth. A one-way RM ANOVA for the data in the upper right panel is not significant, whereas a one-way RM ANOVA for the data in the lower right panel is highly significant ($p < 0.001$). A further remaining question in implant research is why results for some patients demonstrate large improvements during the first 6–12 months of implant use but results for other patients do not. The age of the patient does not appear to be a limiting factor, as the results for the ear tested for the eldest patient in the group demonstrated some of the largest improvements over time. In addition, the starting point (the scores at 1 month after the initial fitting) does not seem to predict whether scores will stay flat with time, as those points range from about 18 % correct to about 70 % cor-

rect in the upper right panel. However, the largest improvements with time in the present data were associated with relatively low starting points, in the range from 0 to about 23 % correct, as shown in the bottom right panel. The heterogeneity of the patterns merits further investigation. Possibly, the heterogeneity reflects differences in brain adaptability or "plasticity" among CI users.

The hearing brain may well be the most important part of the prosthesis system. It is able to utilize no more than eight broadly overlapping sectors of stimulation in the cochlea, and, for most patients, make progressively better use of that input during the first 6–12 months of implant use. Those eight sites do not compare favorably with the 30,000 neurons in the healthy auditory nerve, the 3500 inner hair cells distributed along the length of the cochlear partition, or even the 20+ "critical bands" spanning the speech range of frequencies in the normally hearing ear. These disparities are huge and yet most CI users can understand everyday speech using their restored hearing alone.

Stimulation in Addition to That Provided by a Unilateral Cochlear Implant

The performance of unilateral CIs has been relatively stable since the introductions of the (then) new processing strategies into widespread clinical use in the early and mid-1990s, as described in the section on "Performance of Unilateral Cochlear Implants." Fortunately, another avenue for improving performance has been found, and that is to present stimuli in addition to the stimuli provided by a unilateral CI. The additional stimuli can be from a second CI on the contralateral side, for electrical stimulation on both sides, or with acoustic stimulation for persons who have residual hearing (usually at low frequencies) in either or both ears. Such adjunctive stimulation can produce large improvements in speech reception for substantial fractions of patients who either have access to bilateral CIs or have residual hearing.

An example of the benefits is presented in Fig. 1.9, which shows CNC word scores for 15 subjects who had a full insertion of a CI on one side and residual hearing at low frequencies on the other side. For reference, the monosyllabic word scores at the 2-year interval in the Helms et al. data are also shown. Those scores for 55 subjects are presented in the left column and the scores for the other 15 subjects, who were studied by author MFD and his coworkers (Dorman et al. 2008), are presented in the middle and right columns. The middle column shows scores for electrical stimulation only (with the unilateral CI) and the right column shows scores for that stimulation plus acoustic stimulation of the contralateral ear. All tests were conducted with hearing alone and without prior knowledge by the subjects of the test items. No feedback was given during the tests as to correct or incorrect responses. The subjects in the Dorman et al. study used the

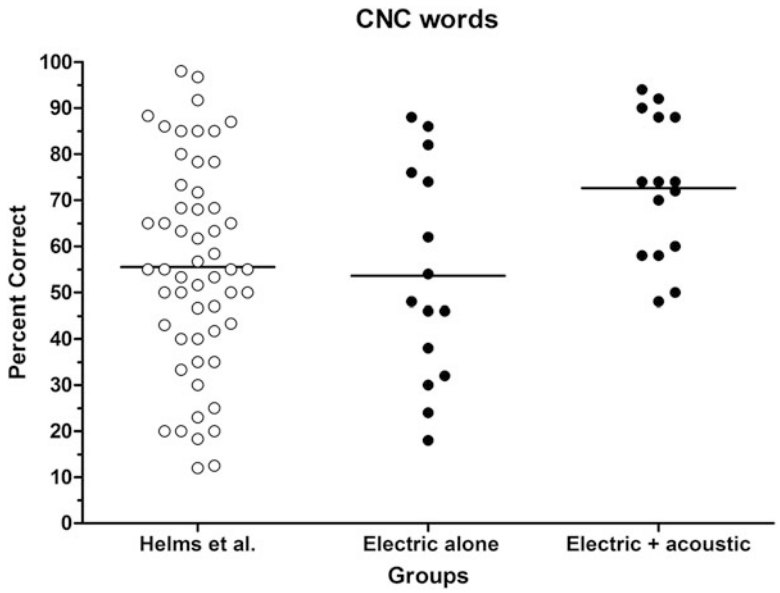

Fig. 1.9 Recognition of monosyllabic words by the 55 subjects who participated in the Helms et al. study (*left column*) and by a separate group of 15 subjects who participated in a study conducted by author MFD and his coworkers in 2007 and 2008 (Dorman et al. 2008). The data for the subjects in the Helms et al. study are from the 2-year test interval (these data also are presented in Fig. 1.3). The subjects in the Dorman et al. study each had a full insertion of a then-current cochlear implant (CI) on one side and residual hearing at low frequencies on the contralateral side. Scores for electric stimulation only, with the CI alone, are presented in the *middle column*, and scores for electric plus acoustic stimulation are presented in the *right column*. The *horizontal line* in each column shows the mean of the scores (the figure is from Dorman et al. (2008), and is reproduced here with the permission of Karger AG)

latest implant devices and processing strategies at the time of the tests, and at that time the subjects had had between 5 months and 7 years of daily experience with their CIs. The means of the scores in each of the columns are shown with the horizontal lines.

Comparison of the scores in the left and middle columns again shows no difference in the performance of unilateral CIs between the mid-1990s and the mid-to-late 2000s. The means and variances in the scores from the two studies are statistically identical.

In contrast, comparison of the scores in the middle and right columns shows a highly significant improvement with the addition of acoustic stimulation on the side contralateral to the CI. The means of the scores for the two columns are 54 % and 73 % correct, respectively, and the variance of the scores in the right column is significantly lower than the variance of the scores in the middle column. In broad terms, the additional stimulation reduced the incidence of scores below 50 % correct; indeed, 8 of the 15 subjects scored below 50 % correct with electric stimulation only (middle column), whereas only 1 of the 15 subjects scored below 50 % correct with combined electric and acoustic stimulation (combined EAS). That is a large jump up in performance, similar to the jump achieved previously with CIS.

Of course, subjects who have relatively high scores with electric stimulation alone don't have much room for improvement and therefore may not gain much if any benefit from combined EAS. In addition, subjects who have relatively low levels of residual hearing may not benefit as much from combined EAS compared to subjects with relatively high levels of residual hearing.

These possibilities were investigated in a subsequent study by Dorman et al., which included 105 subjects with a CI on one side and a wide range of residual hearing at low frequencies on the contralateral side (Dorman et al. 2015). The subjects were grouped according to the level of residual hearing into mild, moderate, and severe loss of hearing categories. The average of hearing thresholds for the frequencies of 125, 250, and 500 Hz was used for the grouping. Subjects assigned to the mild-loss group had pure tone averages (PTAs) for those frequencies of 40 dB HL or better; subjects assigned to the moderate-loss group had PTAs between and including 41 and 60 dB HL; and subjects assigned to the severe-loss group had PTAs of 61 dB HL or worse. The tests included recognition of CNC words in quiet, and recognition of AzBio sentences in quiet and in competition with noise at the S/Ns of +10 and +5 dB. All of the subjects were tested with CNC words and fewer and different numbers of subjects were tested with the other speech items. The mild-loss group included 20 subjects; the moderate-loss group 41 subjects; and the severe-loss group 44 subjects. Tests were conducted with the implant only and with the implant plus acoustic stimulation of the contralateral ear. No visual cues were provided, and no feedback was given during the tests as to correct or incorrect responses. The test items were not known to the subjects prior to testing.

Representative results are presented in Fig. 1.10, which shows the percentage-point gain in the scores for each of the tested subjects when acoustic stimulation was added to the electric stimulation provided by the unilateral CI. The left panel shows the gains for the CNC word test, and the right panel shows the gains for the AzBio test at the S/N of

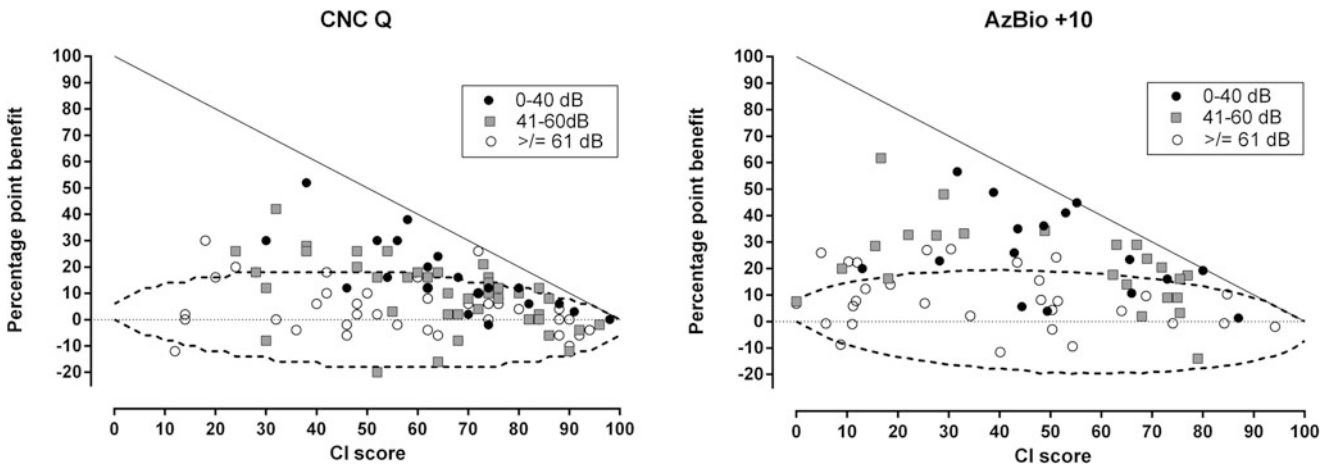

Fig. 1.10 Percentage point improvement in monosyllabic word scores in quiet (CNC Q) and sentence scores in noise (Arizona Biomedical Institute sentences presented in competition with noise at the speech-to-noise ratio of +10 dB, AzBio +10) by users of unilateral cochlear implants (CIs) when allowed to use low-frequency hearing in the ear contralateral to the CI. The abscissa in each panel is the score obtained with the CI alone. The *dashed lines* are the 95 % confidence intervals for the test material. The *solid diagonal line* indicates the maximum score that could be obtained for each point along the abscissa. Each *symbol* represents the performance of one patient. Patients are sorted into three groups based on the averages of each patient's hearing thresholds at the frequencies of 125, 250, and 500 Hz (the figure is adapted from Dorman et al. (2015), and is used here with the permission of Elsevier BV)

+10 dB. Results for all of the subjects are included in the left panel, and results for 69 of the subjects are included in the right panel. The dashed lines in the panels show the 95 % confidence intervals for no gain with combined EAS, and the diagonal lines in the panels show the maximum gain possible according to the recognition scores obtained with the CI alone.

As is evident from the figure, significant gains are produced for many of the subjects with the addition of the acoustic stimulus. The gains are constrained in cases of high scores with the CI alone. Also, few subjects in the severe-loss group have significant gains, whereas many subjects in the moderate- and mild-loss groups have large and highly significant gains. In broad terms, high gains are observed for the subjects with (1) recognition scores of about 60 % correct or lower when using the CI alone and (2) PTAs of 60 dB HL or better. A somewhat greater preponderance of gains is seen for the AzBio sentences presented in competition with noise than for the CNC words, although substantial gains for many subjects are seen for both types of tests.

The acoustic stimulus for combined EAS may be directed to the same ear as the implant if that ear has useful (and preserved) residual hearing (e.g., von Ilberg et al. 1999; Gantz and Turner 2003), or to the ear contralateral to the implant (as in the studies just described), or to both ears. The side of acoustic stimulation does not appear to matter, so long as the residual hearing at low frequencies is at least moderately good (Wilson 2012; Sheffield et al. 2015). The results presented in Fig. 1.10 suggest that the boundary for "useful residual hearing" appears to be around a PTA of 60 dB for the frequencies of 125, 250, and 500 Hz.

On average, the gains obtained with combined EAS are on the order of the gains obtained previously with CIS. Combined EAS is a major step forward.

Additional stimulation with a second CI can also produce large improvements in speech reception, especially for speech presented in competition with an interfering sound (e.g., noise or another talker) at a different location. Indeed, the benefits of bilateral CIs are most evident at highly adverse S/Ns or with especially difficult speech items presented in quiet (e.g., Wilson et al. 2003; Wackym et al. 2007).

Figure 1.11 shows the performance of bilateral CI subjects in a complex, noise-filled environment (Loiselle 2013). In this environment, directionally appropriate noise, recorded originally in a crowded restaurant, was played from eight loudspeakers surrounding the listener. The test signals, sentences from the Pediatric AzBio sentence test corpus (Spahr et al. 2014), were presented, in random order, from the five loudspeakers in the 180° arc in front of the listener. The S/N was adjusted for each subject so that, in the best single CI condition, performance was in a range that would allow improvement to be observed, if present, when the second implant was added.

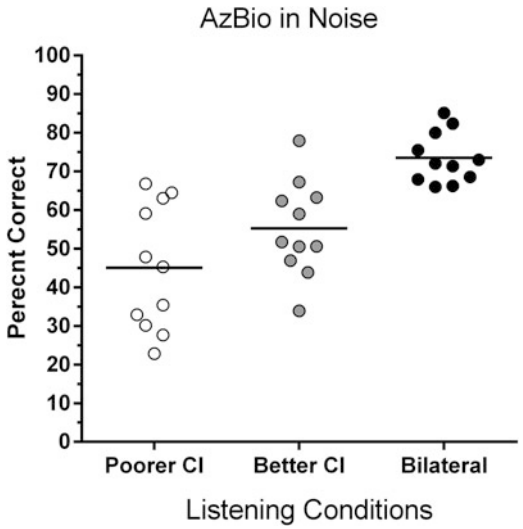

Fig. 1.11 Percent correct scores for recognition of sentences by users of bilateral cochlear implants (CIs). The sentences were presented in competition with restaurant noise from eight loudspeakers surrounding the subject for each test. The sentences were from the pediatric version of the Arizona Biomedical Institute (AzBio) sentences, and presentations of the different sentences in each test session were roved among the five loudspeakers in the frontal 180° plane. The conditions included tests with the poorer of the unilateral CIs, the better of the unilateral CIs, and both CIs. Rankings of the unilateral CIs were made on the basis of scores for the recognition of monosyllabic words. The speech-to-noise ratio was adjusted for each subject to avoid floor or ceiling effects (the data are from Loiselle (2013), and are used here with her permission)

When the best single CI (according to CNC word scores) was tested in this complex listening environment, performance from the 11 subjects averaged 55 % correct. When the subjects were allowed to use both implants, performance increased to 73 % correct; the increase was highly significant. Thus, either a second implant or combined EAS can provide large benefits to speech understanding for CI patients. These outcomes together suggest that a further useful condition for speech understanding in complex noisy environments might be bilateral CIs with at least one ear having residual low-frequency acoustic hearing. Recent results from preliminary studies support this possibility (Dorman et al. 2013).

A further benefit of bilateral CIs, and of acoustic stimulation on both sides for CI users with residual hearing on both sides, is reinstatement of sound lateralization abilities (e.g., Schoen et al. 2005; Grantham et al. 2007; Dunn et al. 2010; Gifford et al. 2014b; Loiselle et al. 2015). Although the accuracy of lateralization is not normal (approximately 20–30° mean root-mean-squared error versus 7–8° for subjects with normal hearing), such abilities provide an alerting function and are likely to be helpful for attending to a primary speech signal in acoustic environments with competing sounds at other locations.

Joachim Müller and his coworkers at the Julius-Maximilians-Universität in Würzburg, Germany, were the first to demonstrate large advantages of bilateral CIs (Müller et al. 2002). Those advantages for speech reception are comparable to the advantages of combined EAS, with some variation across different tests and test conditions. For listening in complex acoustic environments with multiple interferers, bilateral CIs or combined EAS with bilateral acoustic stimulation may be better than combined EAS with acoustic stimulation at one side only (e.g., Gifford et al. 2013, 2014a; Rader et al. 2013). In addition, bilateral CIs or combined EAS with acoustic stimulation on both sides can restore at least some sound lateralization abilities for many of their users.

Possibilities for Improvements

The performance of the present-day CIs is great but not perfect. The top and average scores are high, but some patients do not achieve high scores even with the latest CI devices and processing strategies. In addition, room remains for improvements even for the top performers.

Adjunctive stimulation with bilateral CIs or combined EAS can be helpful for many but not all patients. The patients who are helped the most are those who score below about 50 % correct in the recognition of monosyllabic words using a unilateral CI only. That is a wonderful result, as those "low-performance" patients are in the greatest need of help and should be the patients that we, as a field, should worry about the most.

Possibly, consideration of the brain and its roles in determining outcomes with CIs will be helpful as well. Indeed, accumulating and now compelling evidence indicates that differences in the function of the "hearing brain" among CI users may explain a substantial portion of the remaining variability in outcomes with CIs and the related treatments, including the treatments using adjunctive stimulation. As described elsewhere (e.g., Moore and Shannon 2009; Wilson et al. 2011; Lazard et al. 2012), the function of the hearing brain can be compromised in many ways and those compromised brains may not be able to make the best possible use of the sparse inputs from the periphery provided by the present-day unilateral CIs. Adjunctive stimulation can help, as noted previously, perhaps by providing a richer and more detailed input. At the other end, improving the processing abilities of the compromised brain may produce even greater gains. Ways in which such improvements might be achieved are described for example in Wilson et al. (2011); as noted there, the improvements if realized could be especially helpful for patients presently shackled with the relatively poor outcomes.

Although adjunctive stimulation can boost performance for many patients, not all patients have access to bilateral CIs due to national health policies or to restrictions in insurance coverage, and not all patients have useful residual hearing, which is required for combined EAS. In addition, the unilateral CI and its performance is the foundation for the treatments that include adjunctive stimulation, and thus an improvement in that performance would most likely improve the performance of the adjunctive stimulation treatments as well. For these reasons, continued development of unilateral CIs and the processing strategies used with them is important.

Some among the many promising possibilities for improving the performance of unilateral CIs include:

- Considering the brain in designs and applications of CIs, as mentioned previously and described a bit further below
- A closer mimicking of the intricate processing in the normal cochlea, as described for example in Wilson et al. (2010)
- An increase in the number of effective channels, with more focal stimulation in the cochlea or in the trunk of the auditory nerve, or by minimizing temporal electrode interactions or forward masking effects both peripherally and centrally
- Improving or at least maintaining the biological condition of the implanted cochlea, for better transmission of sound information to the brain, as described for example in Pfingst et al. (2011)
- Prudent pruning of interfering or otherwise detrimental electrodes, as described most recently in Zhou and Pfingst (2012), Garadat et al. (2013), and Noble et al. (2013, 2014)

With respect to the first bulleted possibility, we further note that there are at least two ways to consider the brain as a vital part of the prosthesis system. The first way is to understand that differences in the function of the "hearing brain" may underlie much of the remaining variability in outcomes and to design processing strategies and training procedures with those differences in mind, as mentioned previously. The other way is to ask whether all of the important pathways in the hearing brain are activated and activated appropriately with the present-day processing strategies, electrode designs, and electrode placements (Middlebrooks and Snyder 2010; McAlpine 2011; Shannon 2014). If the answer is no, as has been suggested by the authors just cited, then that could motivate changes to engage the hearing brain more fully. The two approaches are complementary and each may produce large improvements.

With respect to the third bulleted possibility, more focal stimulation might be achieved with optical rather than electrical

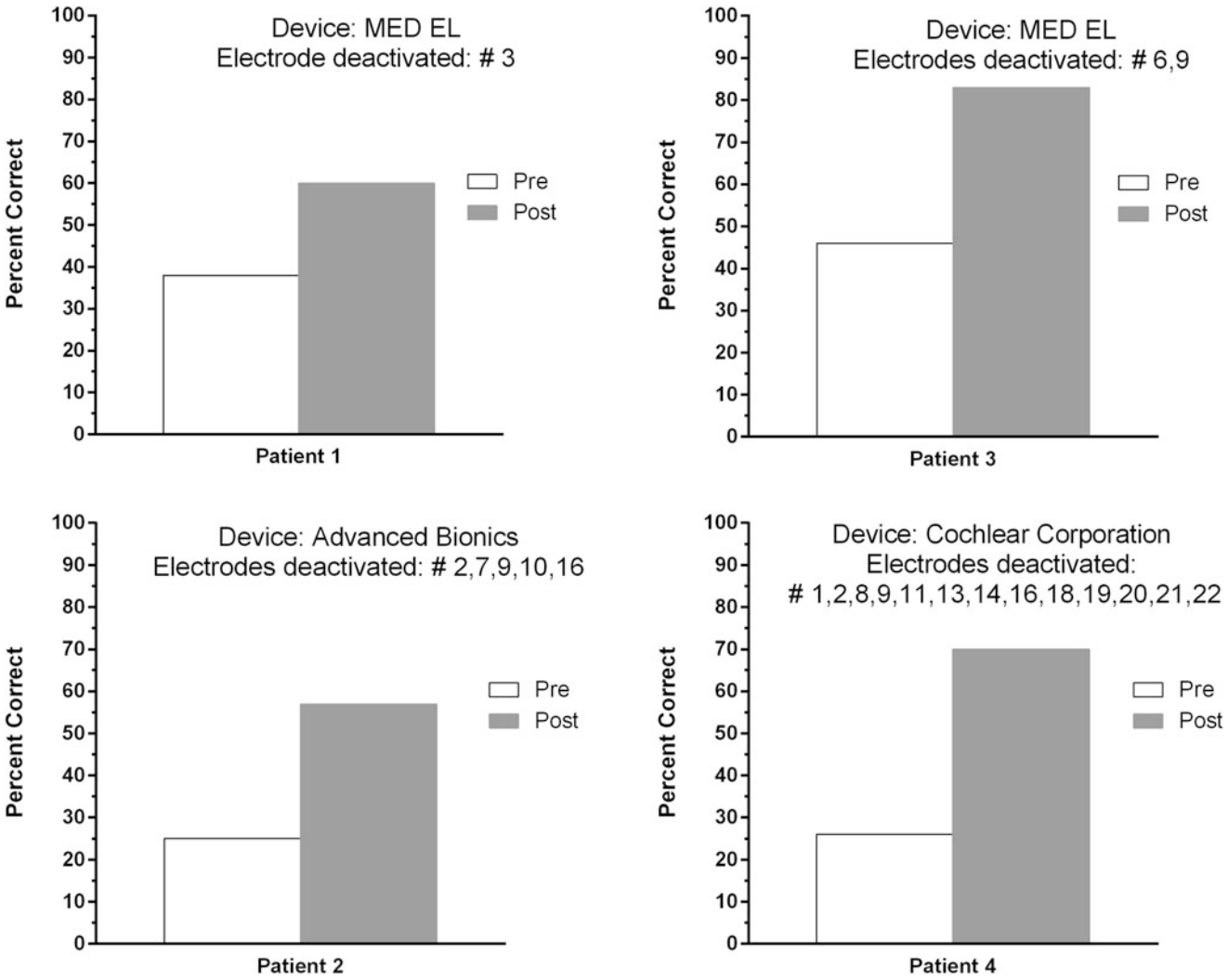

Fig. 1.12 Percent correct scores for recognition of Arizona Biomedical Institute (AzBio) sentences in noise (at the speech-to-noise ratio of +10 dB) by users of unilateral cochlear implants (CIs). The conditions included conventional programming of the CI (Pre) and programming with one or more of the electrodes deactivated (Post). The different panels show results for each of four subjects; the subjects were tested at the Vanderbilt University Medical Center

stimulation (e.g., Matic et al. 2013; Hernandez et al. 2014; Richter and Tan 2014; Jeschke and Moser 2015; Moser 2015) or with electrical stimulation but using new designs or placements of the electrodes, such as placements immediately adjacent to the target neurons in the trunk of the auditory nerve within the modiolus (Middlebrooks and Snyder 2007). More focal stimulation or lower thresholds or both also might be achieved by promoting the growth of neurites from spiral ganglion cells toward the electrodes in the scala tympani using appropriate drug or gene therapies, and safe techniques for drug delivery (Jolly et al. 2010; Clarke et al. 2011; Pfingst et al. 2011; Budenz et al. 2012; Landry et al. 2013; Pinyon et al. 2014; Shepherd and Wise 2014).

And with respect to the final bulleted possibility, optimal results might be achieved by pruning the number of utilized electrodes, inasmuch as the present designs and placements of electrodes for scala tympani implants do not support more than eight effective channels for any patient or processing strategy tested to date (see, e.g., Fig. 1.7). Support for this approach is presented in Fig. 1.12, which shows recognition by CI subjects of the AzBio sentences presented in competition with noise (at the S/N of +10 dB) for two conditions. In the "Pre" condition, the subjects were tested with the conventional programming of their CIs. In the "Post" condition, varying numbers of electrodes were deactivated and the frequency boundaries of the band-pass channels were reassigned appropriately for the smaller number of utilized electrodes, and then the subjects were tested again. Deactivation of the electrode(s) for each subject was based on a CT scan imaging technique that allows identification of

electrodes that would be likely to produce strongly overlapping patterns of neural excitation. Only one among each set of those electrodes is retained for subsequent use in each subject's modified "map," and thus the number of utilized electrodes is reduced and sharply so in some cases (Noble et al. 2013, 2014). As shown in Fig. 1.12, the benefits of such pruning can be large; for the four subjects whose results are included in the figure, the improvements range from 22 to 44 percentage points.

Although not all subjects show such impressive gains, many do, and that latter finding indicates the potential of customizing fittings for CI patients. Additional approaches for customized fittings that include electrode pruning are presented in Zhou and Pfingst (2012) and Garadat et al. (2013). In general, one might want to prune electrodes down to no fewer than eight retained electrodes, as up to eight may be useful for some patients and tests even if some among the eight may still be less than fully desirable. Certainly, no more than eight are needed, at least with the present processing strategies and designs and placements of the stimulating electrodes.

Concluding Remarks

Modern CIs are responsible for the first substantial restoration of a human sense using a medical intervention. They are extraordinary devices. However, not all users have high levels of performance and even the top performers do not experience a complete restoration of hearing and overall auditory function. Most fortunately, further improvements are possible and are being pursued by talented teams worldwide.

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Part II

Clinical Management

Cochlear Implant Candidacy in Children: Audiological Considerations

2

René H. Gifford

Introduction

The Food and Drug Administration (FDA) officially approved cochlear implants for use with children on June 27, 1990 following the successful completion of a multicenter trial with 142 enrolled participants. The initial approval was granted for the Nucleus 22 device for children aged 2–17 years. The current labeled indications for pediatric cochlear implantation have been in effect since 2000 at which time the minimum age was lowered from 18 to 12 months. It is quite likely that in the near future, that the minimum age will be lowered yet again to 9 months or perhaps even younger.

The cochlear implant candidate selection process involves the careful consideration and coordination of audiological assessment, early intervention, and intense habilitation with appropriately fitted amplification. This process does not fall entirely within the scope of the audiologist; rather a multidisciplinary team of professionals is involved ensuring that we are able to consider the needs of the whole child including the wishes and goals of the family. This chapter will discuss pediatric cochlear implantation from the perspective of the audiologist, speech-language pathologist (SLP), deaf educator, social worker, and/or psychologist (see Chap. 4 for medical/surgical considerations in the candidacy process). This chapter will describe implant candidate selection per today's labeled indications as well as discuss those elements that we may want to consider for future assessment.

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Audiological Evaluation

Assessment of Auditory Function: Behavioral Measures

Prior to scheduling the cochlear implant workup evaluation, children are typically seen in the diagnostic Audiology clinic for objective assessment of auditory function. These appointments will typically include otoacoustic emissions, auditory brainstem response (ABR) testing, tympanometry, acoustic reflexes, and if old enough, behavioral hearing assessment. Thus, it is most generally the case that the initial diagnosis of severe-to-profound sensorineural hearing loss does not occur during the implant workup appointment. Rather, families often present to the implant evaluation with an established diagnosis, familiarity with home-based intervention, and at least some hearing aid experience. For audiologists practicing all aspects of pediatric audiology, a patient and his/her family may be followed from initial diagnosis to hearing aid fitting, follow up, assessment of auditory progress, implant candidacy, and postoperative management. Such audiologists, however, are a rarity as cochlear implant audiologists are increasingly finding their schedules full with those needing immediate preimplant and longitudinal postimplant management.

Despite the fact that children will typically have had objective and also perhaps prior behavioral testing, comprehensive audiological assessment is still recommended. Behavioral assessment with a slightly older and more experienced infant or older child may yield greater opportunity for obtaining additional ear specific pure tone and speech understanding information. This may be particularly true for frequencies that prior audiograms may have been lacking, such as 250 and 8000 Hz. Most cochlear implant teams require multiple assessments of behavioral hearing status prior to *finalizing* implant candidacy. This is recommended to ensure that the child does in fact have an implant-qualifying profile. Because identification and then confirmation of behavioral hearing status is essential for determining

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implant candidacy in the U.S., the earliest cochlear implant workups are generally scheduled once an infant is 6+ months of age. The earliest implant workups involve visual reinforcement audiometry (VRA) and/or behavioral observation audiometry (BOA). For infants and children with diagnoses that may delay motoric development such as sitting up and/or independent head and neck control as well as global developmental and/or cognitive abilities, behavioral audiometry is more complicated. Thus for obvious reasons, final determination of implant candidacy is generally delayed for children with comorbidities.

Assessment of Auditory Function: Physiologic Measures

Otoacoustic emissions (OAEs) provide diagnostically valuable information for pediatric implant candidate selection. Though most universal newborn hearing screening programs use OAEs as a first-pass screen (e.g., NIH 1993; White et al. 2005; Gravel et al. 2005; JCIH 2007), not all babies are screened prior to discharge and not all births occur in medical facilities. Further, many experienced pediatric audiologists working with cochlear implants have a story or two about identifying auditory neuropathy spectrum disorder (ANSD) during the course of the implant workup. Such cases do not necessarily implicate prior test or tester error. In fact, early OAE screens and/or diagnostic tests can been confounded due to a number of potential factors including the size of the infant ear canal, placement of the transducer against the canal wall, presence of vernix or other debris in the canal, middle ear effusion, or some combination thereof. ANSD does not preclude a child from implant candidacy (e.g., Shallop et al. 2009; Peterson et al. 2003; Rance and Barker 2008; Teagle et al. 2010; Breneman et al. 2012); however, it is critical that we have an accurate diagnosis in place and that we explore all possible habilitative options prior to recommending cochlear implantation. It is also vital that if ANSD is identified in an infant with history of premature birth, that incomplete auditory maturation is not overlooked as a potential variable.

In addition to OAEs, acoustic reflex testing can help confirm a child's diagnosis and should corroborate the audiometric thresholds or minimal response levels. The obvious exception to this rule occurs in cases of ANSD for which acoustic reflexes are generally absent or elevated (Berlin et al. 2005, 2010). For these children, absent or elevated acoustic reflexes may thus not agree with behavioral thresholds or minimal response levels to pure tone or even narrowband stimuli.

Audiometric Criteria for Implantation

Children <24 Months

There is considerable confusion across clinics and professionals regarding who exactly meets candidacy for cochlear implantation. A primary factor contributing to this confusion is that the audiometric criteria for pediatric FDA-labeled indications differ across implant manufacturers. Adding to the confusion is the fact that for pediatric candidacy, there is an additional element of age-specific criteria with respect to severity of hearing loss. The current audiometric criteria for pediatric cochlear implantation are shown in Table 2.1 for all 3 FDA approved systems. As shown in Table 2.1, the current indications specify bilateral *profound* sensorineural hearing loss for children aged 12–24 months (Cochlear Americas package insert, Advanced Bionics package insert; MED-EL package insert). This does not necessarily imply that children with less severe hearing losses do not benefit from cochlear implantation. More exactly, there have been historical concerns regarding reliable, behavioral assessment of infant hearing status. Thus, the criteria are most stringent for the youngest candidates. This historical concern may not be as relevant in today's clinical environment given the audiologic checks and balances available for validation of behavioral and physiologic assessment. In fact, many professionals have used this as a talking point in the argument for lowering

Table 2.1 Current audiometric criteria for pediatric cochlear implantation for all 3 FDA approved systems; current indications specify bilateral *profound* sensorineural hearing loss for children aged 12 to 24 months

	Audiometric criteria		Speech recognition criteria	
Advanced Bionics	Profound SNHL, ≥ 90 dB HL		<4 years old	4+ years old
			<20 % open-set words (e.g., MLNT) via MLV at 70 dB SPL	<12 % for difficult, recorded open-set words (i.e., PBK) or <30 % open-set sentences (i.e., HINT-C) at 70 dB SPL
Cochlear	<24 months	24+ months	≤30 % on open-set words MLNT or LNT	
	Profound SNHL	Sev-to-profound SNHL		
MED-EL	Profound SNHL, 90+ dB HL at 1 kHz		<20 % for MLNT or LNT words	

the minimum age from 12 months, 9 months, or even younger (e.g., Cosetti and Roland 2010; Kim et al. 2010; Nicholas and Geers 2013; Holman et al. 2013; Leigh et al. 2011). There are more and more studies suggesting that earlier implantation yields increasingly higher levels and more rapid word learning and language development (Bergeson et al. 2010; Houston and Miyamoto 2010; Houston et al. 2012; Niparko et al. 2010; Moog and Geers 1990; Leigh et al. 2013), speech perception (Tajudeen et al. 2010), speech intelligibility (Habib et al. 2010), and vocabulary development (Hayes et al. 2009; Houston and Miyamoto 2010; Houston et al. 2012; Tomblin et al. 2005).

Infants with severe-to-profound hearing loss are missing critical language learning opportunities that occur within the first year of life. This is true even for infants with appropriately fitted hearing aids given that audibility may not be sufficient to allow for consistent auditory access to spoken language at average conversational levels. Developmental changes occurring rapidly in first year of life include word segmentation, auditory memory development, and phonological/lexical/semantic representation. Word segmentation is the process of dividing connected discourse into meaningful units, such as individual words. Researchers have shown that word segmentation develops rapidly between 7.5 and 10.5 months of age (e.g., Jusczyk 2002; Bortfeld et al. 2005). By 8 months of age, infants have the capacity for auditory memory and long-term storage of new words—both being prerequisites for auditory-based language learning (Jusczyk and Hohne 1997; Houston and Jusczyk 2003). In addition, development of phonological, lexical, and semantic representations also rapidly emerges during the first year of life (e.g., Gupta and MacWhinney 1997; Hollich et al. 2002; Soja et al. 1991; Storkel 2009). Given these incredible developmental changes occurring during the first year of life for the typically developing child, an infant with severe-to-profound sensorineural hearing loss and limited aided audibility is missing out on the development of critical auditory-based, language learning opportunities. In fact, these language learning opportunities may even begin before birth! Recent research has shown that neural reorganization in response to speech may occur even before birth. Newborns have been shown to respond differentially to familiar sounds that they were exposed to as fetuses (e.g., DeCasper and Fifer 1980; DeCasper and Spence 1986; Kisilevsky et al. 2003; Moon et al. 2013) with greater brain activity observed in response to familiar sounds (e.g., Partanen et al. 2013).

Indications for Children 2+ Years

For children over 2 years of age, audiometric criteria are slightly more lenient than defined for younger children. Cochlear Americas specifies audiometric candidacy including bilateral *severe*-to-profound sensorineural hearing loss thereby allowing for slightly more residual acoustic hearing for candidacy (Cochlear Americas package insert). Both Advanced Bionics and MED-EL, however, list bilateral profound sensorineural hearing loss in the labeled indications children of all ages. Audiometric criteria for FDA-labeled indications are listed in Table 2.1 for all three manufacturers.

The Role of the FDA in Determining Labeled Indications

FDA-labeled indications (i.e., candidacy criteria) for cochlear implantation are listed in the physician's package insert. The physician's package insert is included in each cochlear implant device packaging and refers to the product information which includes the device characteristics, risks, and indications for use. Despite common misunderstanding regarding package inserts and labeled indications, the FDA is not responsible for defining the criteria or indications for cochlear implantation. Instead, it is the manufacturer that ultimately defines the indications for use. The manufacturer submits an application to the FDA for approval outlining the proposed criteria for their device—which is often tied to a multicenter clinical trial. The role of the FDA is to approve or reject the submitted application and the *manufacturer-defined indications* following the completion of the clinical trial. If approved, the manufacturer-defined "indications for use" are listed on the package insert as the FDA-labeled criteria for cochlear implantation.

The role of the FDA is in the regulation of industry, *not the individual clinician or implant center*. Industry is prohibited from promoting any off-label usage of a device. In other words, even in the presence of evidence supporting expanded criteria for implantation, implant manufacturers are strictly prohibited from recommending device usage for individuals not meeting labeled indications. In contrast, clinicians are able to exercise professional clinical judgment and make clinical recommendations for their patients about the suitability of cochlear implant candidacy. For this reason, the FDA has published a document specifically outlining the "off-label" usage of pharmaceuticals and medical devices called *Off-Label and Investigational Use Of Marketed Drugs, Biologics, and Medical Devices* (FDA 2011). This document authorizes off-label use of biomedical devices when the intent is for clinical practice (i.e., excluding research applications). In this document, the FDA states that should a clinician recommend off-label usage of a device, the clinician has the responsibility to ensure that s/he is (1) well informed about the product, (2) basing its use on firm scientific rationale and on sound medical evidence, and (3) maintaining records of the product's use and effects.

Clinicians on the Cochlear Implant Team hold responsibility to provide professional, clinical determinations regarding implant candidacy. Exercising professional clinical judgment is what separates clinicians from technicians.

Thus, the Cochlear Implant Team may determine implant candidacy for infants with sensorineural hearing loss not making auditory progress with *full-time use* of appropriately fitted hearing aids and compliance with the recommended intervention and therapy, based upon professional clinical judgment—regardless of whether or not the infant has met each of the labeled indications for implant candidacy. Similarly, if an older child has moderate sloping to profound sensorineural hearing loss and is not making auditory progress with appropriately fitted hearing aids and intervention, the cochlear implant team should still consider recommending implantation.

Speech Perception Testing

Recorded Materials at Average Conversational Levels

Pediatric cochlear implant evaluations for older children involve behavioral assessment of auditory-based speech understanding. Children with hearing loss rely heavily on visual cues such as lip reading and nonverbal communication. Speech stimuli are presented without visual cues in an effort to gain an understanding of the child's auditory-based skills.

Presenting speech via monitored live voice (MLV) is known to yield highly variable outcomes (e.g., Roeser and Clark 2008). Thus, in order to assess pediatric implant candidacy in a standardized manner, the use of recorded speech stimuli is critical. Despite the stability of outcomes obtained with recorded stimuli, all pediatric audiologists are aware that MLV testing may be required, at least initially, to elicit reliable responses. This is particularly true for the younger child and for children with developmental delay. Recorded stimuli include an unfamiliar voice that lacks the affect that is typically found in the speaking style of an individual who works with children. However, should a child respond only to MLV stimulus presentation and continue to exhibit poor recognition, it is reasonable to recommend cochlear implantation as speech understanding with recorded stimuli would expectedly be poorer (Roeser and Clark 2008).

Another important considerable for speech perception testing involves appropriate presentation levels. Although audiologists had historically used 70 dB SPL, this level is no longer considered appropriate as it does not represent *average conversational level speech* (Pearsons et al. 1977; Olsen 1998). Despite the lack of a standardized protocol for pediatric candidacy and outcomes assessment, the current adult recommendations—as found in the adult minimum speech test battery (Minimum-Speech-Test-Battery 2011)—include the use of recorded speech materials presented at 60 dBA[1] for assessment of speech recognition performance in both pre- and postimplant testing. Given these recommendations for adult implant candidate selection, it would follow that 60 dBA should be *highest* presentation level used for pediatric candidacy assessment. In fact, data supporting an even lower presentation level demonstrate that average, casual speech levels for pediatric and female talkers range from 50 to 56 dBA (Pearsons et al. 1977; Olsen 1998). Consequently, to accurately evaluate a child's understanding of both female speakers and fellow children, lower presentation levels should also be considered for speech recognition testing, such as 50 dBA.

Specifying presentation levels in terms of SPL can be difficult for busy audiologists who work with audiometers specifying HL as the reference. Unfortunately, there is no uniform conversion value for speech stimuli as there are a number of factors involved. Calibrating speech stimuli for sound field presentation requires that both the input and output of the audiometer be calibrated. For recorded materials, a calibration tone is provided in order to calibrate the *input level* to the audiometer. To calibrate the input level, the audiologist must set the audiometer to External A/B or CD1/2 or Tape 1/2—or whatever setting the individual audiometer specifies for the input port used to connect to the CD player, MP3 player, or computer. Next the transducer is set to *speaker*, though it is not critical at this point as only the input is being calibrated. To avoid presentation of a high-level stimulus through the speaker, it is recommended that the dial be set to a low level. Next the *interrupt* or *stimulus* button is selected to present the *input calibration tone* through the audiometer. Once the calibration tone is played, the sensitivity dials for External A/B (or CD 1/2 or Tape 1/2) are adjusted to ensure that the VU meter is set to 0 or just below in order to avoid distortion and clipping of the input signal. It is important to note that once the *input* is calibrated, the sensitivity dials should *not* be further adjusted. Instead, calibration of the output signal is completed by manipulating the audiometer dial setting.

Calibration of the audiometer *output* requires the use of a sound level meter. A sound level meter microphone can be placed on a stand or suspended from the ceiling via extension cable so that the microphone is placed at the position of a typical child's head when seated in a chair.

A calibration noise accompanies speech stimuli on CD or via wav or mp3 stimuli for sound field calibration. Output calibration involves setting the sound level meter to A

[1]The use of A weighting for sound level meter measurements is recommended given that linear weighting yields noisier recordings due to flat frequency response through the lower frequency region. In contrast, the A weighted frequency response rolls off for lower frequencies reaching 20 dB of attenuation at 100 Hz.

weighting and fast response and then adjusting the audiometer dial in 1-dB increments until the display reads 60 dBA[2]. It is at this audiometer dial setting, and accompanying EXT A/B (CD 1/2 or Tape 1/2) sensitivity dial setting, that the speech stimuli will also be presented at the desired presentation level.

Recorded speech stimuli for sound field presentation would ideally be calibrated prior to each assessment. For busy clinics without a dedicated sound level meter, this is not a realistic option. A second best option would be daily calibration—another unlikely prospect for the typical clinical environment. Thus, it is advised to complete quarterly calibration and to post the dial settings required to achieve the desired presentation levels in the control room.

Speech Recognition Test Materials and Implant Candidate Selection

Children ≤ 3 Years

Development of auditory skills and speech perception for younger children are most generally evaluated with auditory questionnaires, parental report, and speech/language assessment. The labeled indications for children in this age range with respect to "speech understanding" are quite vague and all manufacturers make reference to *limited benefit from appropriately fitted hearing aids and habilitation* as evidenced by lack of progress in the development of simple auditory skills.

Current labeled indications do not reference the use of closed-set measures of speech perception for determining implant candidacy for the toddler age group. However, this does not mean that closed-set measures are not valuable instruments for assessing speech recognition in candidate selection. It is simply the case that these are not listed. Closed-set measures should be used to assess speech recognition for our youngest patients if for no other reason than to provide a baseline measurement against which preoperative progress with hearing aids and/or postoperative progress cochlear implants can be calculated.

Children 3+ Years

Labeled indications for older children are based on either mono- or multisyllabic word recognition—depending on which is more developmentally appropriate for the child being assessed. The speech measures referenced by the implant manufacturers in the package insert are as follows (presented in order of developmentally appropriate progression): Early Speech Perception (ESP) test (Moog and Geers 1990), Multisyllabic Lexical Neighborhood Test (MLNT) (Kirk et al. 1995), Lexical Neighborhood Test (LNT) (Kirk et al. 1995), Phonetically Balanced Kindergarten (PBK) word recognition test (Haskins 1949), and Hearing in Noise Test (HINT) sentences for children (HINT-C, Gelnett et al 1995).

Older children for whom these open-set measures are considered developmentally appropriate are required to exhibit substantially lower performance than even that listed for adult implant criteria (see Table 2.1). Candidacy criteria for older children based on word recognition scores range from 12 to 30 % correct in the best-aided condition (see Table 2.1). Advanced Bionics further lists performance up to 30 % correct for HINT-C sentences—when developmentally appropriate for children over 4 years. This same manufacturer has set its HINT sentence recognition criteria for adult candidacy at 50 % correct. Thus, the pediatric implant criteria are disproportionally restrictive even though children are acquiring auditory language.

The expansion of pediatric cochlear implant criteria has historically been a hot topic. There have been a number of studies reporting highly successful outcomes for nontraditional implant recipients who were either under 12 months of age, had less severe hearing losses, asymmetric hearing loss, and/or above criterion-level performance on measures of speech understanding (e.g., Dettman et al. 2004; Dowell et al. 2004; Leigh et al. 2011, 2013; Sunderhaus et al. 2012; Nicholas and Geers 2013; Holman et al. 2013). In light of data suggesting a relatively narrow critical period for the development of listening and spoken language (Hayes et al. 2009; Moog and Geers 2010; Bergeson et al. 2010; Niparko et al. 2010; Habib et al. 2010; Houston and Miyamoto 2010) as well as auditory pathway maturation (Ponton et al. 2000; Sharma et al. 2002; Eggermont and Ponton 2003; Gordon et al. 2005; Kral et al. 2006; Sharma and Dorman 2006; Gilley et al. 2008; Sharma et al. 2009), it is reasonable that professionals are questioning the restriction implant labeling for the youngest auditory language learners.

A future consideration for pediatric implant candidate selection includes the assessment of speech understanding in noise. Children are rarely in quiet listening environments.[2] In fact, occupied classroom noise ranges from 48 dBA to 69 dBB with mean levels approximating 65 dBA for an early elementary classroom (e.g., Sanders 1965; Nober and Nober 1975; Bess and Tharpe 1984; Finitzo-Hieber 1988). Clark and Govett (1995) reported that children's Leq 24-h levels range from 87.3 dBA for all students and are as high as 95.5 dBA for fifth graders. Given that a child's everyday listening environment is *much noisier* than even that encountered by

[2]Current FDA indications do not suggest the use of speech in noise testing for determining either adult or pediatric implant candidacy.

the typical adult, and that FM is rarely utilized for all listening environments, it would follow that speech recognition in noise should be standard practice for determining implant candidate selection.

Evaluation of Auditory Skills and Progress in Infants and Younger Children

Birth to 3 Years

When it comes to quantifying a child's auditory skills and progress with hearing aids prior to implant candidate selection, many factors will be involved that will provide information beyond the audiogram. The reality is that similar audiograms can yield highly disparate outcomes in speech and language. Because we are unable to assess speech understanding performance for infants and even for some toddlers, it is absolutely critical that the candidacy workup include evaluation of auditory skills, development, and progress with amplification. From the perspective of the audiologist and otologist, these skills are most commonly assessed via parental case history and auditory questionnaires.

According to a recent survey of pediatric cochlear implant programs in the U.S. (Uhler and Gifford 2014), the most frequently used questionnaire for children birth to 3 years is the Infant-Toddler version of the Meaningful Auditory Integration Scale (IT-MAIS, Zimmerman-Phillips et al. 2000). In fact, the package inserts for all three FDA-approved cochlear implant systems list the IT-MAIS for the assessment of auditory progress with amplification. The IT-MAIS was developed to be administered via structured parental interview and thus requires that the clinician interprets open-ended responses and assigns numerical scores ranging from 0 (never) to 4 (always).

Another parental questionnaire designed for use with infants and toddlers is the LittlEARS (Copyright MED-EL, 2004; Weichbold et al. 2005; Coninx et al. 2009). The LittlEARS includes 35 yes/no questions assessing auditory-based responsiveness. The questionnaire is organized in a hierarchical fashion with a progression of difficulty so that after six consecutive 'no' answers, no further answers are necessary. The LittlEARS was designed for children with normal hearing up to 24 months of age. For this reason, administration is typically completed up to 24 months following implant activation; however, typically developing children implanted early who use the implant all waking hours will generally reach ceiling levels prior to that test point. The LittlEARS takes 5–7 min and because it was not designed for parental interview, administration does not need to take up clinical time.

Yet another questionnaire appropriate for use with infants and toddlers is the Auditory Skills Checklist (ASC, Meinzen-Derr et al. 2007). The ASC is a 35-item questionnaire designed to assess detection, discrimination, identification, and comprehension. The ASC was developed for parental interview and/or clinician observation. Scoring includes the parent/administrator assigns a score from 0 to 2 as follows: (0), child does not have the skill; (1), child has emerging skill development; (2), child consistently demonstrates the skill. The ASC can be used along with the IT-MAIS and/or LittlEARS as the ASC may be readministered over a longer period of time (for children implanted up to 3 years of age) and will provide a multidimensional assessment revealing smaller increments in the development of auditory skills.

The Functioning After Pediatric Cochlear Implantation (FAPCI) questionnaire (Lin et al. 2007) is another questionnaire that was designed primarily for use following implantation; however, it may provide value-added information when administered during the candidacy process serving as a baseline measurement. The FAPCI includes 23 items and was designed for children aged 2–5 years. The FAPCI assesses a child's behaviors as related to auditory-based responsiveness and expressive verbal communication. The FAPCI does not require parental interview and completion takes approximately 5–10 min.

There is no one questionnaire or set of questionnaires that is recommended for best practices use in the pediatric cochlear implant clinic. Thus, individual implant programs will determine which questionnaires best meet the needs of its patients and families. What is most important regarding questionnaire administration is that all clinicians within a program be consistent across all patients adhering to the clinic protocol. Protocol adherence affords each implant program the creation of its own normative data for patient outcomes, which is critical for family counseling. Protocol adherence and the development of clinical norms will also provide clinics with the data needed to compare its outcomes to average patient outcomes in the literature. Finally, since most implant programs have multiple pediatric-focused clinicians, protocol adherence allows clinician substitution without sacrificing clinical quality or assessment accuracy.

Preschool and School-Aged Children

The assessment of auditory skills development for older children cannot be well predicted by the audiogram. Despite the fact that behavioral assessment of auditory skills will always be investigated for preschool and school-aged children, a number of factors may limit or even preclude complete behavioral assessment at any given appointment. For this reason, there are several auditory questionnaires designed for supplementary use with preschool and school-aged children.

Uhler and Gifford (2014) showed that the most commonly used auditory questionnaire for children over 3 years is the Meaningful Auditory Integration Scale (MAIS,

Robbins et al. 1991). Like the IT-MAIS, the MAIS is a 10-item parental interview style questionnaire designed to assess auditory skills for 3- to 5-year-old children. As mentioned previously with reference the IT-MAIS, all implant manufacturers reference the MAIS for candidate selection with respect to the assessment of auditory progress—or lack thereof—with appropriately fitted hearing aids.

Another questionnaire designed for use in children aged 3 years and up is the Parents' Evaluation of Aural/oral performance of Children or the PEACH (Ching and Hill 2007). The PEACH is a 13-item questionnaire designed for parental assessment of a 3- to 7-year-old child's aural and oral abilities in everyday life. The PEACH asks parents to consider their child's listening behaviors over the past week and then assigns a numerical value to the answers ranging from 0 (Never or 0 %) to 4 (Always or 75 to 100 %). The PEACH questions assess listening behaviors in both quiet and noisy surroundings.

The FAPCI (Lin et al. 2007) can also be of use in toddlers and preschoolers as it was intended to gauge postoperative progress for children aged 2–5 years. Although the FAPCI was designed to track postoperative progress, administration during the candidacy selection process can also provide clinicians and families with a baseline against which future growth in auditory skills may be measured.

There are other parental questionnaires available for use with children who have severe-to-profound sensorineural hearing loss including the Meaningful Use of Speech Sounds (MUSS; Robbins et al. 1991), Children's Home Inventory for Listening Difficulties (CHILD; Anderson and Smaldino 2000), and Developmental Index of Audition and Listening (DIAL; Palmer and Mormer 1999). As stated previously, individual clinical teams may determine the appropriateness of the different instruments for their patient population and must strive to maintain consistency across clinicians and patients.

Language Development and Hearing Aid Trial

Hearing aid trials generally coincide with a baseline speech/language assessment so that amplification-related gains in development can be accurately measured. The measures chosen by the SLP will be dependent on both the chronologic age and the hearing age of the child. Even infants who would expectedly not be able to engage in a diagnostic speech/language evaluation should be assessed to document communicative and auditory skills. Another reason for scheduling early SLP assessment and therapy, even for infants, is that the clinician can provide the family with tools and strategies necessary for the facilitation of spoken language and auditory skills development (e.g., Cole and Flexer 2007; Estabrooks 2006).

A common misconception is that any auditory progress made with amplification precludes cochlear implant candidacy. Instead, a child should demonstrate month-per-month growth in functional auditory skills as well as in development of speech and language. For example, for a child making full-time use of hearing aids for 3 months should demonstrate *at least* 3 months of progress in auditory skills and speech/language development. Infants and children not making month-for-month growth in functional skills are at greater risk for language delay and later academic failure as they are already behind with respect to early language exposure (e.g., DeCasper and Fifer 1980; DeCasper and Spence 1986; Kisilevsky et al. 2003; Moon et al. 2013; Partanen et al. 2013). In fact, early language competence predicts later communicative competence, reading ability, and overall academic performance (e.g., Catts et al. 2002; National Early Literacy Panel 2009; NICHD Early Child Care Research Network 2005). Thus, it is critical for professionals and families to consider not only the growth in skills but also the appropriate amount of growth. Should month-for-months growth in functional skills not be observed with full-time use of hearing aids and recommended intervention, cochlear implantation should be considered. It is important here to note that although the implant manufacturers specify the need for a 3–6-month trial with amplification prior to determining implant candidacy, that the length of the trial may be compressed in cases of meningitis for which concerns about cochlear ossification and electrode insertion are present.

In order to begin an suitable hearing aid trial, hearing aid settings must first be verified using either probe microphone measurements or test box verification with patient-specific real ear to coupler difference (RECD) measurements (Pediatric Amplification Protocol 2003). Given that audiologists will generally fit both adults and children with nonlinear hearing aid circuitry to ensure audibility at various input levels, a prescriptive fitting formula such as DSL m[i/o] (Seewald et al. 1985; Cornelisse et al. 1995; Scollie et al 2005) is used to verify target audibility at speech levels corresponding to soft, average, and loud (e.g., 50, 60, and 70 dB SPL). Such practice is also helpful in determining implant candidate selection as those children for whom target audibility cannot be achieved at lower input levels run the risk of missing critical speech information in everyday communicative settings.

During the hearing aid trial, it is recommended that a child receives at least two diagnostic speech/language evaluations. Though it may not be possible to administer multiple norm-referenced measures over such a short time period, there are available criterion-referenced measures that can provide vital clinical information about a child's progress with hearing aids. Criterion-referenced measures gauge progress using the child as his or her own control, such as a *within-subjects* comparison.

Cochlear Implant Team: Beyond the Surgeon, Audiologist, and Speech/Language Pathologist

Social workers and psychologists can play a vital role in the process of cochlear implant candidate selection. A psychologist or social worker can assist families with the acceptance of hearing loss and the realistic implications of having a child who will be dependent upon technology for communication. Also within the scope of the psychologist and/or social worker is the evaluation of family dynamics and level of family dedication to the recommended postoperative intervention schedule. Such guidance may also be recommended following cochlear implantation, as well. Another area in which a psychologist could assist is in cases for which concerns arise regarding a child's overall cognitive and global development. In addition to an evaluation by a developmental pediatrician which can take months to years to schedule, an evaluation by a psychologist or developmental psychologist may also be recommended and diagnostically valuable to the team and family.

Social workers specialize in providing counseling and support for families. Many social workers are also trained to provide the necessary information regarding financial requirements for assisted medical insurance. Social workers may be particularly valuable for families of children who will be undergoing cochlear implantation. Most families will be unaware of the financial resources available for insurance coverage of the device as well as the required intervention and assistive listening devices, such as amplified telephones and FM systems. Social workers employed by medical centers not only provide financial counseling but will also help families with the required paperwork to help them navigate through the application process. Some social workers are even able to assist families by coordinating medical and therapy appointments to ensure that all medical specialties and evaluations have been either scheduled or completed.

Deaf educators work in conjunction with otologists, audiologists, speech/language pathologists, and early interventionists to help empower families with the knowledge needed to help meet their communicative and educational goals for their child. Deaf educators are especially knowledgeable regarding the development of an Individualized Family Service Plan (IFSP) or Individualized Education Plan (IEP). The IFSP and IEP—which fall under the Individuals with Disabilities Education Act (IDEA)—will help guide the rehab process for children birth to 3 years (IFSP) and 3–21 years (IEP), respectively. Deaf educators can help families through the process determining whether their child qualifies for services under IDEA and can act as a liaison between the clinical and educational teams to help the family develop the IFSP or IEP.

Preoperative Counseling and Education

Variables Affecting Postoperative Outcomes

Counseling families is a large part of the job description for pediatric audiologists. Indeed the process of cochlear implant candidate selection requires in-depth counseling and family education. Educating a family about how cochlear implants function and what they can do is just as important as focusing on what implants *cannot* do. For example, it is critical to counsel families about the fact that cochlear implants cannot restore "normal" hearing and auditory function nor can implants change the underlying diagnosis of sensorineural deafness.

The most commonly asked question is how well one's child will perform following surgery. To be more explicit, families want to know how well their child will be able to communicate via listening and spoken language. In most cases, it can be impossible to predict how well a child will perform as there are a number of factors that have the potential to impact postoperative outcomes. Some of the most critical variables affecting outcomes include age at implantation, wear time, intervention, integrity of cochlear and neural structures, and etiology.

Age at Implantation

There is a relatively narrow critical period for cochlear implantation for the development of listening and spoken language and auditory pathway maturation (Hayes et al. 2009; Moog and Geers 1990; Dorman et al. 2007; Bergeson et al. 2010; Niparko et al. 2010; Habib et al. 2010; Houston and Miyamoto 2010; Ponton et al. 2000; Sharma et al. 2002; Eggermont and Ponton 2003; Gordon et al. 2005; Kral et al. 2006; Sharma and Dorman 2006; Gilley et al. 2008; Sharma et al. 2009; Tobey et al. 2013). Houston and colleagues have shown significantly higher levels of word learning in children implanted under 13 months of age (Houston et al. 2012; Houston and Miyamoto 2010) as compared to children implanted between 16 and 23 months of age. Other studies have shown that children implanted between 18 and 24 months of age demonstrate significantly greater language and vocabulary development—both expressive and receptive—than children implanted over 2 years of age (Hayes et al. 2009; Niparko et al. 2010; Markman et al. 2011; Boons et al. 2012).

Though the data are clearly in support of "earlier is better" for postoperative outcomes, it is important to recognize that even children implanted over 2 years of age still derive *significant benefit* from cochlear implantation. At issue is

that families are educated and appropriately counseled regarding realistic expectations and that they consider the age at activation as a critical variable in this process. The cochlear implant team must inform families that more aggressive intervention will be needed for children implanted at later ages for whom the family's goal remains auditory-oral language.

Device Wear Time

Though cochlear implants provide access to sound, the implant processor(s) must be worn all waking hours in order to derive maximum benefit. The Alexander Graham Bell Association for the Deaf and Hard of Hearing, Inc. provides powerful examples documenting the need for consistent wear time. Specifically, an infant with normal hearing listens for approximately 10 h each day totaling at least 3650 h of listening over the first year of life. Conversely, the infant wearing hearing aids on a part-time basis during the candidacy process—estimated at just 4 h per day—would require 6 years of hearing aid use to gain as much listening experience as a baby with normal hearing or the baby wearing hearing aids and/or implant sound processors all waking hours (Stovall 1982; Rossi 2003). For the older child, a toddler/preschooler with normal hearing listens approximately 12 h per day equaling 4380 h of listening over the course of a year. For a toddler/preschooler wearing hearing aids or implant sound processors on a part-time basis—such as 2.75 h per day while at preschool—it would take 9 years to gain the listening experience obtained over a single year for normal-hearing child or for the child making full-time use of his/her hearing aids and implant sound processors (Rossi 2003).

Regardless of the age at implantation, if full-time use of the sound processor(s) is not enforced, a child will simply not make progress. The importance of wear time is such a critical counseling tool that it bears repeating at pre- and postimplant visits with the family and all involved caregivers. Family counseling on wear time is supported with the emergence of data logging capabilities in the newest generation of cochlear implant sound processors.

Intervention

Though early implantation and wear time are both critical elements contributing to individual success, the best outcomes will not be achieved without early and consistent intervention (e.g., Geers et al. 2009; Nicholas and Geers 2007; Moeller et al. 2000; Moog and Geers 2010). Research has shown that for those families desiring an auditory-oral approach to language focusing on listening and spoken language, that early enrollment in an intervention incorporating parental and overall family involvement is associated with higher levels of receptive and expressive language (Moeller 2000; Moog and Geers 1990). The best outcomes are achieved with regular and intensive habilitation including speech/language therapy, infant and family services with home-based infant/family specialists, enrollment in parent–infant programs, and later involvement in a preschool program focusing on listening and spoken language. The surgical placement of the device and subsequent activation of the device are just the first steps in the hearing journey with the majority of the "work" to follow.

Though this chapter has focused on an auditory-oral approach to communication, it is important to recognize that this may not be the goal of every family nor may it be a reasonable objective due to the age of identification, age at implantation, family dynamics, and/or other developmental delays associated with the underlying etiology and/or other diagnoses. There are modes of communication that incorporate signing, cued speech, augmentative communication, or any combination thereof that may ultimately serve as a better fit for a given child. The family's goals should be paramount; however, we must always consider child-specific expectations and the fact that expectations may change over time based on a child's progress and evolving diagnoses.

Anatomy of Cochlear and Neural Structures

As mentioned in Chap. 5 (Isaacson, Roland), otologists will generally order imaging studies to determine cochlear patency and to rule out abnormalities of the cochlea or temporal bone that could impact the surgical placement of the device. Though abnormalities of the cochlea and temporal bone do not necessarily contraindicate cochlear implantation, it is still critical that the surgeon be aware of such issues. Abnormalities could influence which device is ordered such as a short or compressed electrode array for common cavity or split array for ossified cochleae. Another reason is that anatomic abnormalities can affect postoperative outcomes and thus are useful for preoperative counseling and expectations management.

Up to 35 % of children with sensorineural hearing loss also have cochleovestibular structural abnormalities (Papsin 2005). Structural abnormalities that can be observed in the pediatric population include Mondini dysplasia, common cavity, enlarged vestibular aqueduct (EVA), and atretic or absent internal auditory canal. Structural abnormalities such as those listed here would be observed via CT imaging and would thus be diagnosed prior to implantation provided that temporal bone CT had been completed.

Another structural anomaly is that of cochlear nerve deficiency (CND). CND is an uncommon neural abnormality that is defined by an absent or hypoplastic auditory nerve.

For children with CND and *absent* auditory nerve, cochlear implantation had been considered contraindicated. The reason is that the primary auditory neurons (i.e., spiral ganglion cells located within the modiolus) are the neural stimulation targets for cochlear implant stimulation. Cochlear implantation for children with CND tends to achieve poorer postoperative outcomes; yet, cochlear implantation can still offer significant benefit (Breneman et al. 2012; Teagle et al. 2010; Buchman et al. 2011; Seymour et al. 2010). High-resolution three-dimensional MRI is the most sensitive modality to diagnose CND, although high-resolution CT of the temporal bones often reveals abnormalities suspicious for its presence. For further discussion on this topic, see Chap. 14.

Etiology

The underlying etiology of hearing loss for the majority of pediatric patients will be unknown. It is estimated that approximately half of all congenital hearing losses have a genetic component and it is likely that this estimate will increase with advances in human genomic research (Rehm 2005). For the vast majority of pediatric implant recipients, there will be no concerns about etiology affecting postoperative outcomes—at least not realized at the time of implantation. There are some etiologies associated with poorer postoperative outcomes for pediatric implant recipients and some requiring specialized counseling for guarded expectations. Some of the more common etiologies and associated concerns are as follows:

Meningitis

Prior to widespread use of the pneumococcal conjugate vaccine, bacterial meningitis was a common cause of postnatal deafness. It accounted for approximately 6% of pediatric cases of acquired sensorineural hearing loss in infancy and childhood (Smith et al. 2005). The pneumococcal conjugate vaccine has significantly reduced the incidence of bacterial meningitis in children; however, bacterial meningitis does continue to occur and approximately 5–10% of cases will result in sensorineural hearing loss (Baraff et al. 1993; Smith et al. 2005). Sensorineural hearing loss as associated with bacterial meningitis results from the development of labyrinthitis, subsequent loss of hair cells, spiral ganglion cell degeneration, and may be followed by cochlear ossification (Lu and Schuknecht 1994; Nadol and Hsu 1991). In fact, cochlear ossification and spiral ganglion degeneration can result in bony obliteration of the cochlea, loss of auditory function, significantly poorer postoperative outcomes, or even contraindication for implantation.

In cases of severe cochlear ossification, it is still possible that the surgeon may be able to either fully insert or insert a portion of the electrodes on the array. In cases of shallow insertion depth and/or spiral ganglion cell degeneration, it is also possible that the implanted electrodes yield little to no auditory stimulation. In cases of incomplete insertion, many recipients are still able to derive auditory benefit. Postoperative outcomes with meningitis and cochlear ossification are highly variable and therefore difficult to predict (Nichani et al. 2011). It is important to note that not all cases of meningitis will result in cochlear ossification; however, for those children exhibiting evidence of preoperative ossification, counseling with respect to realistic expectations is vital.

Syndromic-Related Deafness

There are a number of syndromes for which various degrees of sensorineural hearing loss are common. There are many cases of successful outcomes following cochlear implantation in children with syndromic hearing loss including Branchial-Oto-Renal syndrome, CHARGE syndrome, Pendred syndrome, Refsum disease, Usher syndrome, and Waardeburg syndrome (e.g., Loundon et al. 2003; Raine et al. 2008a; Cullen et al. 2006; Vescan et al. 2002; Arndt et al. 2010; Lina-Granade et al. 2010). Syndromes also affecting visual acuity—such as Usher syndrome and Refsum disease—have the potential to affect outcomes as speech/language therapy and everyday communication makes use of visual cues. In addition, syndromes associated with global developmental delay and cognitive impairment, such as CHARGE, may also impact outcomes and thus require extensive preimplant counseling regarding appropriate expectations and communication-related goals.

Chromosomal-Related Deafness

Trisomy 21, commonly known as Down syndrome, is also linked with sensorineural hearing loss. In fact, sensorineural hearing loss is observed in 5–20% of cases (Roizen et al. 1993; Hans et al. 2010). Cochlear implantation is a viable treatment option for children with Down syndrome and significant sensorineural hearing loss affecting communication (Hans et al. 2010; Cruz et al. 2012). Expectations management will be heavily dependent upon a child's cognitive status as well as the increased risk for middle ear disease. Given that children with Down syndrome have a predisposition for recurrent middle ear disease, it is particularly important that appropriate vaccinations are up to date should a family pursue cochlear implantation (see Chap. 3 (Tan) for greater detail regarding recommended immunizations).

Auditory Neuropathy

Auditory neuropathy is another diagnosis having the potential to affect outcomes. Auditory neuropathy can involve a true neuropathy of the auditory branch of cranial nerve VIII, which is generally accompanied by other peripheral neuropathies including numbness, tingling, reduced sensitivity, and weakness. A diagnosis of auditory neuropathy can also involve auditory neuropathy spectrum disorder (ANSD), which is typically associated with pediatric diagnoses. ANSD typically involves variable audiometric thresholds, poorer than expected speech recognition, absent acoustic reflexes, presence of otoacoustic emissions that are incongruent with the audiogram, and presence of cochlear microphonic with either absent or abnormal auditory brainstem response (ABR) (e.g., Starr et al. 1996; Berlin et al. 1993, 2005, 2010). Postoperative outcomes for children with ANSD—without concomitant peripheral neuropathies or CND—are no different than for children with non-ANSD sensorineural hearing loss (e.g., Teagle et al. 2010; Berlin et al. 2010; Breneman et al. 2012; Ching et al. 2013).

A true auditory neural neuropathy is important to diagnose prior to considering cochlear implantation as it can significantly affect outcomes as "neuropathy" infers neural damage or dysfunction. The prognosis for cochlear implantation is guarded in cases of auditory neuropathy for which the integrity of the auditory nerve is significantly compromised. True auditory nerve neuropathy is rarely diagnosed in childhood and when present, is most frequently a diagnosis secondary to a primary peripheral neuropathy such as associated with Charcot-Marie-Tooth disease.

Other Variables Affecting Postoperative Outcomes

There are a number of variables that can potentially affect outcomes with cochlear implantation including age at implantation, type and frequency of early intervention, educational placement, postoperative degree of audibility with implants, as well as a number of variables of familial and social relevance (e.g., Niparko et al. 2010; Barker et al. 2009; Geers et al. 2003; Szagun and Stumper 2012). Familial- and social-related outcomes most likely to affect postoperative outcomes include family size, intelligence, socioeconomic status, maternal level of education, and the amount of time spent talking to the child at home. Clearly it is not appropriate to discuss the implications for all of these variables during preoperative counseling particularly those including family size, maternal level of education, and socioeconomic status. One variable that demands significant attention during both the pre- and postactivation periods is the amount of time spent talking to the child at home. Engaging a child in auditory-based communication should begin well before the surgery date as it fosters a learned behavior and habit of incorporating language modeling and functional auditory-oral communication. Though this seems to be an obvious recommendation, many parents and family members will often report that they feel reluctant to engage in verbal communication with their child who has severe-to-profound sensorineural hearing loss. This can result from an uncertainty about how to effectively communicate with their child as well as residual emotion regarding the diagnosis. Thus, we as clinicians can help educate families and provide them with the tools necessary to help them successfully navigate the beginning stages of their child's journey to better hearing.

Outcomes

Postoperative outcomes can be highly variable due to a number of hearing- and nonhearing related issues. An increasingly greater number of cochlear implant recipients are able to achieve a normal to near-normal trajectory of auditory-oral speech, language, and vocabulary growth (e.g., Niparko et al. 2010; Geers et al. 2003; Yoshinaga-Itano et al. 2010) and exhibit high levels of academic success within a mainstream educational environment. For children receiving implants at older ages, have additional diagnoses and/or comorbidities affecting cognition, behavior or global development, or who have incorporated manual communication, the rate of auditory-oral growth in speech, language, and vocabulary will expectedly be slower.

The communication goals for each family will be different and should be set individually for every child based on his/her ability, environment, and family goals for communication and quality of life. It is in rare cases that cochlear implantation will yield little to no benefit. Despite this, there are children who ultimately become nonusers due to a myriad of factors that may include repeated device failure[3], behavioral issues, lack of consistent device use, older age at implantation, changing family goals regarding mode of communication, and/or educational placement (Raine et al. 2008a, b).

Conclusion

Pediatric cochlear implant candidate selection is truly a *process* that is accomplished via the collective teamwork of an interdisciplinary group of professionals working together with each child individually and his/her family. Patients and

[3]Device failure in pediatric populations has been estimated to range from 2.9 % (Eskander et al. 2011) up to 10 % over device analysis periods up to 18 years postimplant (Soli and Zheng 2010).

their families are often surprised by the complexity of a cochlear implant evaluation and by the long-term management for the pediatric implant recipient. With the expansion of pediatric cochlear implant criteria and increasing research in this area, the candidate selection process and postoperative follow up will also continue to evolve.

This technology has the potential to change the course of a child's life. In the best cases, cochlear implants and associated intervention can allow for normal development of audition, speech, and language. Even those children not able to achieve an auditory/oral approach to speech and language derive benefit via sound awareness which can significantly improve the quality of life for the recipient and his/her family (e.g., Loy et al. 2010; Clark et al. 2012; Edwards et al. 2012). Pediatric indications have dramatically evolved over the past several decades such that cochlear implants are no longer only for children with bilateral *profound* deafness. The interdisciplinary cochlear implant team will provide patients with the highest level of care and knowledge needed to progress from the preoperative candidacy to a successful postoperative development of auditory-based communication.

For children making full-time use of appropriate amplification, intervention, services, and the family is compliant with recommendations, but are not making progress in speech and language development, the cochlear implant team should consider cochlear implantation. There are firm data from both clinical practice and the peer-reviewed literature demonstrating that children with significant hearing loss who do not meet all items on the eligibility checklist still derive significant benefit from cochlear implantation. Thus, we must remember to look beyond the audiogram and consider the whole child in the candidate selection process.

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Vaccines and the Cochlear Implant Patient

3

Tina Q. Tan

Background

Vaccines are one of the greatest public health achievements of all time and are one of the most important measures for preventing the spread and transmission of many serious infectious diseases. There are routine vaccines that are recommended for persons of all ages in the USA. Each year, the Advisory Committee on Immunization Practices (ACIP) of the Centers for Disease Control and Prevention (CDC) publishes immunization schedules which summarize the recommendations for the routine vaccines recommended at different ages (birth through adulthood) as shown in Figs. 3.1 and 3.2 (CDC 2014a, b). The recommendations for routine use of vaccines in infants, children, and adolescents are harmonized with recommendations made by the American Academy of Pediatrics (AAP), the American Academy of Family Physicians (AAFP), and the American College of Obstetrics and Gynecology (ACOG). Recommendations for routine use of vaccines in adults are harmonized with recommendations of AAFP, ACOG, and the American College of Physicians (ACP).

Infants, children, adolescents, and adults who have a cochlear implant (CI) or will be receiving one, as well as individuals who have either congenital dysplasias of the inner ear or persistent cerebrospinal fluid (CSF) communication with the oropharynx or nasopharynx, should receive all the routinely recommended vaccines appropriate for age based on the CDC annual immunization schedule. None of the vaccines are contraindicated for these underlying conditions. However, these patients are at greater risk for the development of meningitis and other infections with *Streptococcus pneumoniae*, *Haemophilus influenzae* type b, and influenza viruses and AAP and CDC guidelines especially stress the importance of vaccination against these agents in these patients (Rubin et al. 2010; Nuorti and Whitney 2010; CDC 2010, 2012). There is no evidence that children with cochlear implants are more likely to get meningitis due to *Neisseria meningitidis* than children without cochlear implants; therefore, individuals should be vaccinated against meningococcus according to routine recommendations.

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Cochlear Implants and Increased Risk for Certain Infections

During the last several decades, cochlear implantation has emerged as one of the best methods of providing auditory rehabilitation for the profoundly deaf (congenital or acquired). The goal of this surgery in young children is to provide hearing that is adequate to facilitate the development of receptive and expressive language. The most common infectious complications that may occur in implanted children are cellulitis of the overlying skin flap, meningitis, otitis media, and delayed CI infections leading to extrusion of the implant (McJunkin and Jeyakumar 2010; Hansen et al. 2010; Loundon et al. 2010; Hopfenspirger et al. 2007; Hellingman and Dunnebier 2009; Gluth et al. 2011). Rates of infection range from 0.3 to 0.5 % for meningitis, 2–12 % for cellulitis of the skin flap and delayed cochlear implant infections, and up to 36 % for otitis media.

The reported cases of meningitis in CI recipients are thought to have occurred in several ways: (1) in association with leakage of CSF in persons with a malformed cochlea who undergo cochlear implantation, (2) as a consequence of intracranial spread of a middle ear infection along the electrode pathway, or (3) via pneumococcal bacteremia with hematogenous seeding of the cochlea, for example at a site of tissue necrosis related to the electrode or positioner with contiguous spread to the CSF and meninges. In June 2002, the US Food and Drug Administration (FDA) received

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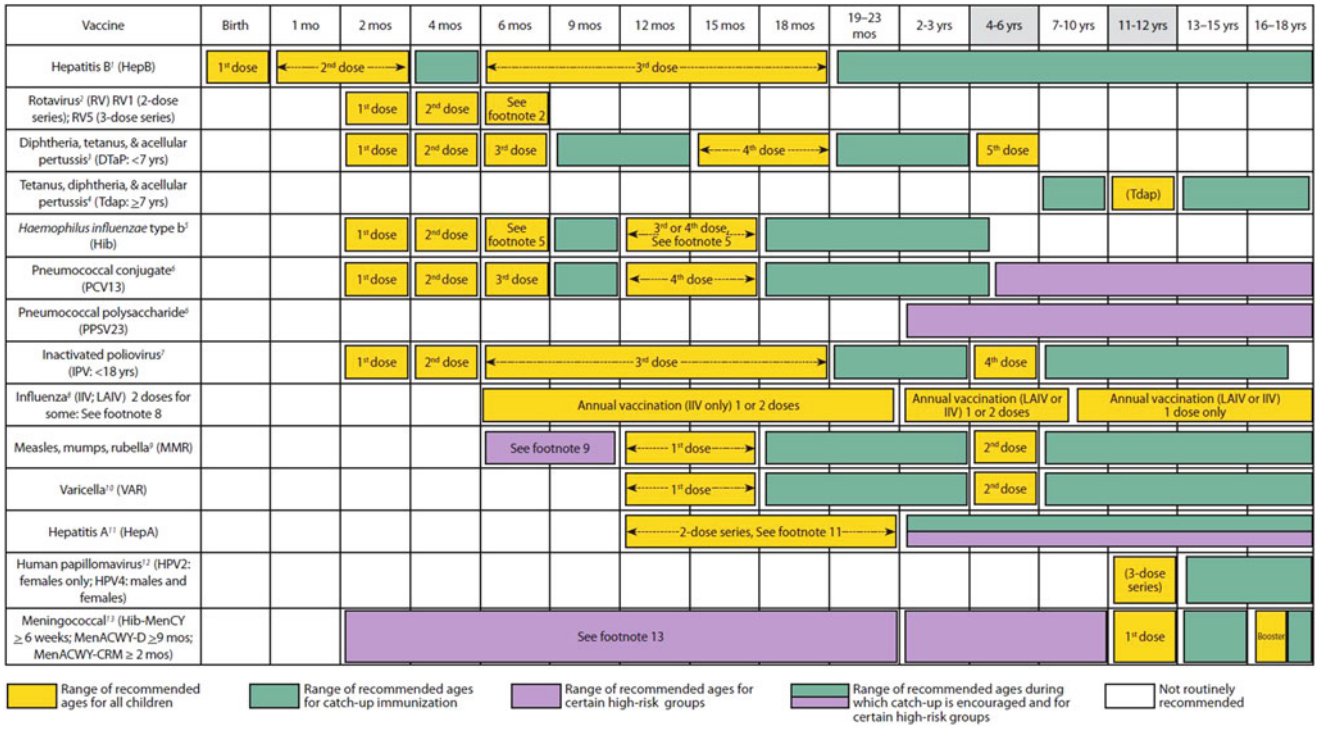

Fig. 3.1 Recommended immunization schedule for persons 0–18 years—USA (2014)

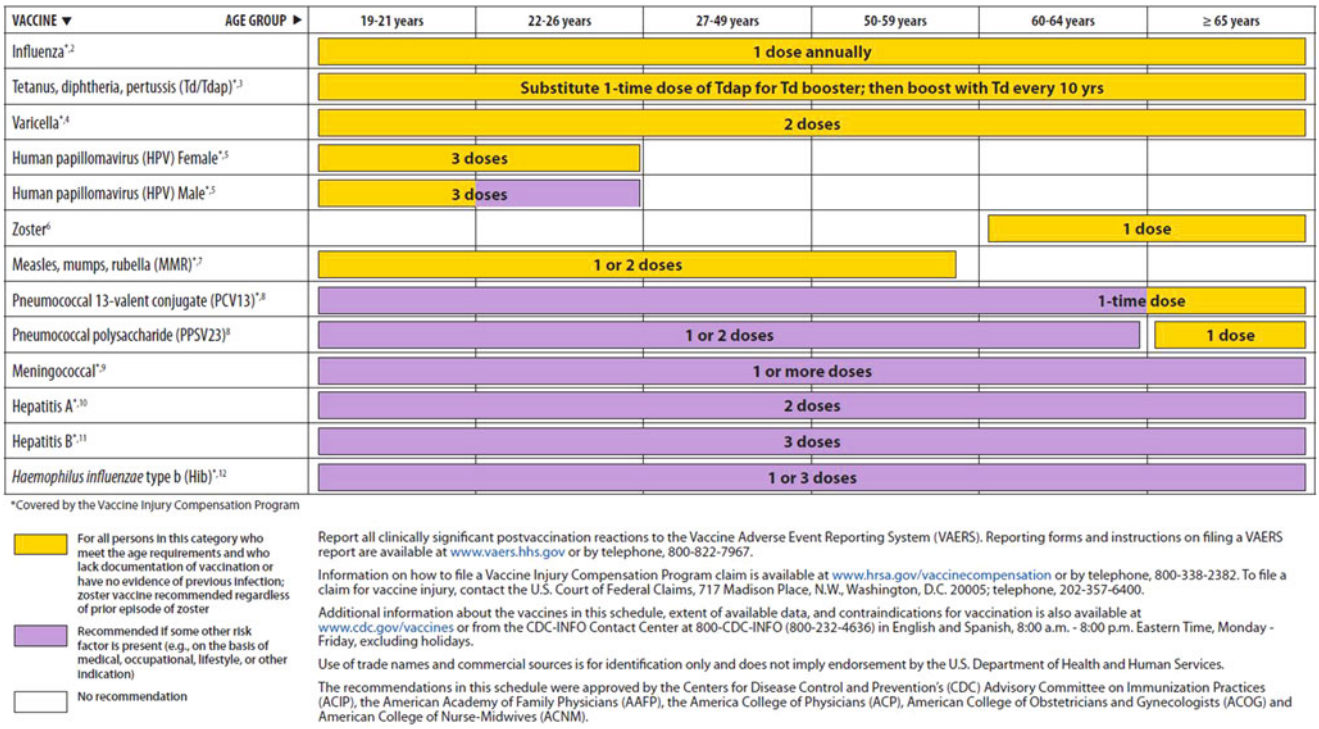

Fig. 3.2 Recommended immunization schedule for persons 19 years and older—USA

Table 3.1 Risk factors for the development of meningitis in cochlear implant patients

Risk factors	Odds ratio (OR), 95 % CI
Presence of positioner	6.0, 1.7–23.1
Cochlear malformation (especially in conjunction with CSF leak)	14.7, 1.7–119.8
Ventriculoperitoneal shunt placement	7.2, 0.6–55.1
Otitis media before implantation	–
CSF leak history	–
Impaired immune system	–
Age less than 5 years	–
Incomplete insertion of the electrode	3.0, 0.5–13.1
Middle ear inflammation at implantation	3.3, 0.7–12.7
Exposure to smoking in the household	2.0, 0.6–6.8
Inner ear trauma due to surgical technique or electrode array design	–
No packing of cochleostomy with soft tissue	–

– no data available to determine risk

numerous reports of bacterial meningitis in implanted children, the majority of whom were younger than age 6 years. The most common causative organism identified was *Streptococcus pneumoniae*, followed by nontypable and type b *H. influenzae*. Other less common organisms that were reported include *Streptococcus pyogenes* (group A beta-hemolytic streptococcus), *Acinetobacter baumannii*, *Escherichia coli*, and *Enterococcus* spp. (Reefhuis et al. 2003). The incidence of pneumococcal meningitis in this group of patients was calculated to be 138.2 cases per 100,000 person-years which was greater than 30 times the incidence in the same-aged cohort in the general population. This increased incidence of meningitis was found to be associated strongly with the use of a CI with a positioner (a wedge-shaped insert available with a Hi Focus I[Advanced Bionics Corporation] that was placed adjacent to the electrode array to position it closer the medial wall of the cochlea) in conjunction with the presence of radiographic evidence of a malformation of the inner ear and leakage of CSF. Cochlear implants with a positioner were voluntarily recalled in the USA in July 2002, although removal of existing implants containing a positioner was not recommended. The risk for bacterial meningitis continues for up to 48 months after implantation, especially among those patients with an implant with a positioner (Reefhuis et al. 2003; Biernath et al. 2006). Table 3.1 shows the risk factors for the development of meningitis in CI patients (Reefhuis et al. 2003; Wei et al. 2010a, b; Cohen et al. 2005).

With the increased risk for meningitis in this population, use of appropriate vaccination against *S. pneumoniae* [7-valent pneumococcal conjugate vaccine (PCV7) and 23 valent-polysaccharide pneumococcal vaccine (PPSV23)] and *H. influenzae* type b is strongly recommended (Rubin et al. 2010; Reefhuis et al. 2003; Biernath et al. 2006; Arnold et al. 2002; Govaerts et al. 2002). On February 24, 2010, a 13-valent pneumococcal conjugate vaccine (PCV13) was licensed by the US FDA. This vaccine contains the seven serotypes included in PCV7 (serotypes 4, 6B, 9V, 14, 18C, 19F, and 23F) and six additional pneumococcal serotypes (1, 3, 5, 6A, 7F, and 19A). This vaccine replaces PCV7 for all scheduled doses of PCV7 in infants (CDC 2010, 2012).

In a study of 50 children who received cochlear implants between 1991 and 1995, researchers found that children prone to the development of otitis media before undergoing implantation were at higher risk for developing postimplantation AOM. The overall prevalence and the severity of AOM, however, were not found to be increased in children with cochlear implants (Luntz et al. 1995). In addition, children's risk of bacterial meningitis may be increased by the presence of a foreign body such as the Positioner that is inserted into the inner ear adjacent to the electrode array. However, even in the absence of a positioner, the electrode lead traverses the middle ear in order for the electrodes to be placed into the inner ear. Therefore there is the potential for spread of middle ear infection into the cochlea along the electrode pathway. Pneumococcal conjugate vaccine and *H. influenzae* type b conjugate vaccine have been shown to decrease nasopharyngeal carriage and the incidence of otitis media caused by these organisms and are a benefit associated with routine use of these vaccines in this population (Nuorti and Whitney 2010; Luntz et al. 1995; Eskola et al. 2001; Cohen et al. 2012; Marom et al. 2014; Agrawal and Murphy 2011).

Vaccination Recommendations for Persons with Cochlear Implants

The following are recommendations for vaccination of patients of various ages and vaccination statuses who have a CI or are scheduled to receive one. These recommendations also apply to children and adults who have congenital malformations of the inner ear, or persistent cerebrospinal fluid (CSF) communication with the oropharynx or nasopharynx who are not CI candidates or recipients (CDC 2012, 2014a, b; Nuorti and Whitney 2010).

1. All infants, children, adolescents, and adults should receive all the routinely recommended vaccines appropriate for age based on the CDC annual immunization schedule.
2. All infants and children under 5 years of age should receive the *Haemophilus influenzae* type b conjugate vaccine according to the routine immunization guidelines.
3. Annual influenza vaccination is strongly recommended for all individuals ≥6 months of age; however, it is of particular importance among CI candidates and recipi-

Table 3.2 Recommended schedule for use of 13-valent pneumococcal conjugate vaccine (PCV13) for *unvaccinated* infants and children

Age at *first* dose (months)	Primary PCV13 series	Recommended regimen	PCV13 booster dose
2–6 months	3 doses	3 doses, 8 weeks apart	Given at 12–15 months of age
7–11 months	2 doses	2 doses, 8 weeks apart	Given at 12–15 months of age
12–23 months	2 doses	2 doses, ≥8 weeks apart	N/A
24–59 months in healthy children	1 dose	1 dose	N/A
24–71 months in children with certain chronic diseases or immunocompromising conditions (includes those with cochlear implants)	2 doses	2 doses, ≥8 weeks apart	N/A

N/A not applicable

Table 3.3 Recommended schedule for use of PCV13 in children <24 months of age who have previously received ≥1 dose of PCV7/PCV13

Age at time of visit (months)	Total number of PCV7/PCV13 received	Recommended PCV13 regimen
2–6 months	1 dose	2 doses, 8 weeks apart; fourth dose at age 12–15 months
	2 doses	1 dose, 8 weeks after most recent dose; fourth dose at age 12–15 months
7–11 months	0 doses	2 doses, 8 weeks apart; third dose at 12–15 months
	1 or 2 doses before age 7 months	1 dose at 7–11 months, with a second dose at 12–15 months or ≥8 weeks after last dose
12–23 months	0 doses	2 doses, ≥8 weeks apart
	1 dose before 12 months of age	2 doses, ≥8 weeks apart
	1 dose at ≥12 months	1 dose, ≥8 weeks after most recent dose[a]
	2 or 3 doses before age 12 months	1 dose, ≥8 weeks after most recent dose[a]
	4 doses of PCV7 or other age-appropriate, complete PCV7 schedule	1 supplemental dose ≥8 weeks after most recent dose

[a]No additional PCV13 doses are indicated for children aged 12–23 months who have received 2 or 3 doses of PCV7 before age 12 months and at least 1 dose of PCV13 at age ≥12 months

ents in order to prevent otitis media, which can lead to the development of bacterial meningitis.

4. All infants and children should be up to date with their *Streptococcus pneumoniae* vaccination according to the routine recommended schedule. Patients who have not received any previous doses of a pneumococcal conjugate vaccine (PCV7 or PCV13) should receive PCV13 as per the routine recommended immunization schedule for infants and children as shown in Table 3.2. Table 3.3 shows the recommended schedule for use of PCV13 in children under 24 months of age who have received prior doses of PCV7 or PCV13. In addition to receiving PCV13, infants and children with CIs should receive one dose of pneumococcal polysaccharide vaccine (PPSV23) at age 2 years or older after completing all recommended doses of PCV13.
5. Children with CIs who have already completed the four-dose PCV7 series and have not received PCV13 or PPSV23 should receive one dose of PCV13. A dose of PPSV23 should be given ≥8 weeks after the dose of PCV13.
6. For children with CIs aged 24 through 71 months who have:
   (a) Not previously received ANY doses of PCV7 or PCV13 should receive two doses of PCV13, ≥8 weeks apart.
   (b) Received an incomplete schedule of <3 doses of PCV (PCV7 or PCV13) before 24 months of age should receive two doses of PCV13, ≥8 weeks apart.
   or
   (c) Received three doses of PCV (PCV7 or PCV13) should receive a single dose of PCV13.

   A dose of PPSV23 should be administered ≥8 weeks after the last dose of PCV13.
7. For children which CIs aged 6 through 18 years who have not previously received a dose of PCV13, a single dose of PCV13 may be administered, regardless of whether they have previously received PCV7 or PPSV23 (Nuorti and Whitney 2010; CDC 2013).
   (a) If the patient has not received PPSV23, the dose of PCV13 should be given first, followed ≥8 weeks later by a dose of PPSV23.

(b) If the patient has been previously vaccinated with ≥1 dose of PPSV23, they should be given a single PCV13 dose ≥8 weeks after the last PPSV23 dose, even if they have received PCV7.

8. CI candidates and recipients should ideally receive all necessary vaccines (PCV13 and PPSV23 doses) at least 2 weeks prior to surgery, if feasible. Vaccination is particularly important in order to prevent otitis media and bacterial meningitis (Rubin et al. 2014). The issue of delaying CI until all doses of both PCV13 and PPSV23 are received is a medical judgment that should be decided on a case-by-case basis. The risk of meningitis for each individual who is partially vaccinated needs to be weighed against the potential impact of delayed intervention.
9. A second dose of PPSV23 is of be consideration 5 years after the first dose. No further additional doses of PPSV23 are recommended.
10. For adults ≥19 years of age who are CI candidates or recipients, the administration of both PCV13 and PPSV23 vaccines is strongly recommended. The dose of PCV13 should be administered first followed by PPSV23 ≥ 8 weeks after the dose of PCV13.

Practical Considerations for Cochlear Implant Programs

Determining which vaccinations a child has actually received can be challenging and time consuming. Confusion may arise as to which pneumococcal vaccinations a child has received. In addition, the marketing of meningiococcal vaccinations as "meningitis" vaccinations has resulted in additional confusion on the part of parents, patients, and primary care physicians (PCPs). For this reason, receiving a copy of the patient's vaccination record, rather than relying upon verbal discussion with medical providers or parental and patient recollection, is advisable.

Certain groups of children require special consideration in order for their vaccinations to be completed. Children implant younger than age 2 years cannot receive PPSV23 prior to implantation. Since PPSV23 is not recommended for widespread use in children or required for school attendance, it typically is not routinely be given by PCPs. Ensuring that these children do receive this vaccination in accordance with CDC recommendations requires effort and vigilance. Hence it is important for the CI program to remind PCPs of the child's need to receive this vaccination and, ideally, to obtain written documentation of its administration. PPSV23 may not be stocked by the PCP office as demand for its use is relatively low. Therefore implant programs may need to find alternative means for families to receive this vaccination. It is also important for CI programs accepting transfer of continued care for already implanted children to review their vaccination status. Adopted children, especially from overseas, also warrant careful scrutiny of their vaccination history. Depending upon their age, these children may need to receive the *Haemophilus influenzae* type b (Hib) vaccination series in addition to pneumococcal vaccinations.

Conclusion

Following the CDC guidelines for vaccination of CI candidates and recipients is important to reduce the risk of bacterial meningitis, a potentially devastating disease. A systematic approach to ensure that children receive these vaccinations is necessary to accomplish this task. Effective communication with PCP about the vaccination status and requirements of these children is essential.

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Medical and Radiologic Issues in Pediatric Cochlear Implantation

4

Sharon L. Cushing, Susan I. Blaser, and Blake C. Papsin

Introduction

This chapter had it been written 20 years ago would have focused on a description of the medical and radiologic factors considered during cochlear implantation that: (1) would have precluded implantation or (2) would risk diminishing the expected outcome following implantation. In contrast, today cochlear implantation is rarely precluded by medical or radiologic factors. The surgical procedure has become so routine in the practiced hands of expert surgeons, that it is possible to overcome most medical and radiologic challenges by employing certain surgical techniques. The major focus of this chapter is demonstrating and outlining these techniques and approaches that can be used to overcome medical and radiological obstacles leading to an optimization of outcome, with a minimization of risk. This chapter focuses on safely approaching the child whose deafness makes them a candidate for cochlear implantation but whose anatomy, physiology or behavior may present other challenges. We focus on the specific part of the candidacy assessment that appraises medical conditions and identifies potential radiologic, and therefore anatomic abnormalities; all issues that have a relatively small impact on candidacy, but much more importantly, need to be addressed with specific and logical techniques in the perioperative period.

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Medical Conditions Affecting Cochlear Implant Candidacy, Surgical Technique, or Outcome

Conditions of the Skin

There are number of skin conditions that impact the decision to implant and might affect the eventual function and longevity of the receiver-stimulator. In our experience, the increased risk of complications in the presence of skin conditions that might interfere with postoperative healing or device use do not preclude the benefits of implantation. We believe careful incision planning, tissue handling and reapproximation are critical to avoiding postoperative wound complications. Fixation of the receiver-stimulator to the bone to prevent micro-movement under a healing wound may in addition improve the chances of success.

One example of a skin disorder that was initially reported to preclude cochlear implantation is keratitis–ichthyosis–deafness syndrome (Hampton et al. 1997). More recent experience has demonstrated that children with this condition may heal and use their external hardware without undue difficulty (Arndt et al. 2010; Barker and Briggs 2009; Cushing et al. 2008a). Children known to form keloids require special consideration. In order to minimize likelihood of keloid formation we infiltrate with depo-steroid at the time of the closure and use non-absorbing sutures. Postoperatively in order to maintain pressure on the wound for a number of months we create a splint made of silicone hearing aid impression material. In some cases additional steroid injections are provided, taking care to avoid injection of the skin overlying the electrodes and

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receiver-stimulator. We have found that avoiding incisions that involve hair baring skin has significantly reduced postoperative keloid formation.

Hematologic Abnormalities

Clotting abnormalities that increase the risk of bleeding (e.g., von Willebrand disease, Thrombocytopenia [e.g., MYH9-related disorders]) provide a challenge to any surgical intervention including cochlear implantation. Successful implantation of these children primarily relies upon collaboration with the hematologist to ensure a medical regimen that minimizes surgical risk. It is not unusual for children with hematologic disorders to require admission prior to surgery to undergo blood work and recommended replacement therapy as well as a longer postoperative stay. Effective co-management between the otologic and hematologic team is central to successful implantation of this special population (Popova and Popov 2009).

Developmental Delay and Multiple Handicaps

Children with developmental delay or multiple handicaps may present challenges to both the cochlear implant (CI) assessment and surgical implantation. The characteristics of the children within these groups are very heterogeneous and span the spectrum from the pervasive developmental disorders to cerebral palsy, and may have any combination of cognitive and physical challenges. Further information on these complicating disorders and the candidacy evaluation of these children may be found in Chap. 13. In regard to our program's approach to the candidacy of the developmentally delayed or multi handicapped child we believe it important for these children to have the capacity to participate to some degree in device programming. An experienced pediatric audiologist's ability to use behavioral techniques to elicit evidence of sound detection and ideally a conditioned response is invaluable in postoperative programming and in recognition of device malfunction. Objective electrophysiological measures can be used when the child's ability to respond sound is limited or in question. We prefer not to rely only on electrophysiologic testing but to use it to augment the data obtained from behavioral assessments. The role of electrophysiology in the difficult to evaluate the child is further addressed in Chap. 8.

A growing number of implant teams, including our own, consider children with developmental delay or multisystem handicap to be good candidates for implantation (Trimble et al. 2008). Any projected outcome in this population must be however considered in the context of their cognitive and motor behaviors (e.g., poor motor function, repetitive movements) that may increase the risk of surgical site complication and ultimately device failure. The following discussion therefore focuses specifically on the techniques used to minimize such complications in response to certain specific behaviors commonly seen in this challenging group of patients.

During the perioperative period to minimize complications there are a number of factors to consider. For example, some children with behavior disorders may pick at their incision resulting in wound healing problems. Despite application of mastoid dressings, soft arm restraints or mittens, it may be very difficult to protect the healing operative site from a determined finger that may carry with it a bacterial inoculate.

For some children with anxiety or behavior disorders, anesthesia consultation prior to the day of surgery may be advantageous for the child and parents.

Children with multiple handicaps that include motoric problems may require use of a headrest when seated. Repeated side to side movements of the head may cause repeated microtrauma to the receiver stimulator that may increase the risk of device failure (Papsin et al. 2011). Poor balance or motor problems may also increase the likelihood of falls resulting in trauma to the implanted device. In addition some children have significant microcephaly (Fig. 4.1) to the degree that placement of the receiver stimulator is best modified. For children who require the support of a head rest or who have significant microcephaly, we position the receiver stimulator in a more superiorly oriented position (Fig. 4.2). Our preferred technique is to create a recessed bed within the cortical bone of the skull and to use tie-down sutures (Fig. 4.3) to secure the device. This approach prevents the tendency for migration of the device more posteriorly. In addition, recessing the receiver-stimulator within a bone bed also lowers its profile thus reducing the risk of trauma and eventual device failure. We have recently described a novel technique for device fixation that involves the additional of linear pedestal to the newer thin bodied models of receiver stimulators (Papsin et al. 2016).

Some children have cervical spine anomalies that limit head positioning thereby making visualization and surgical access to the mastoid challenging. For example, children with the Klippel–Feil have a bone disorder characterized by the abnormal joining (fusion) of two or more spinal bones in the neck (cervical vertebrae) that is often associated with hearing loss and cochlear malformations (Fig. 4.4). These children may be very difficult to position during surgery. Occasionally, restrictions of neck movement may even influence the choice of ear to be implanted. Limitations of neck movement in children with spinal deformities often can successfully be dealt with by strategic positioning of the patient with appropriate cushioning. Elevation and rotation of the surgical table to compensate for inability to optimally position the patient is often necessary. The key is recognition of these issues prior to surgery and thoughtful planning to address these challenges.

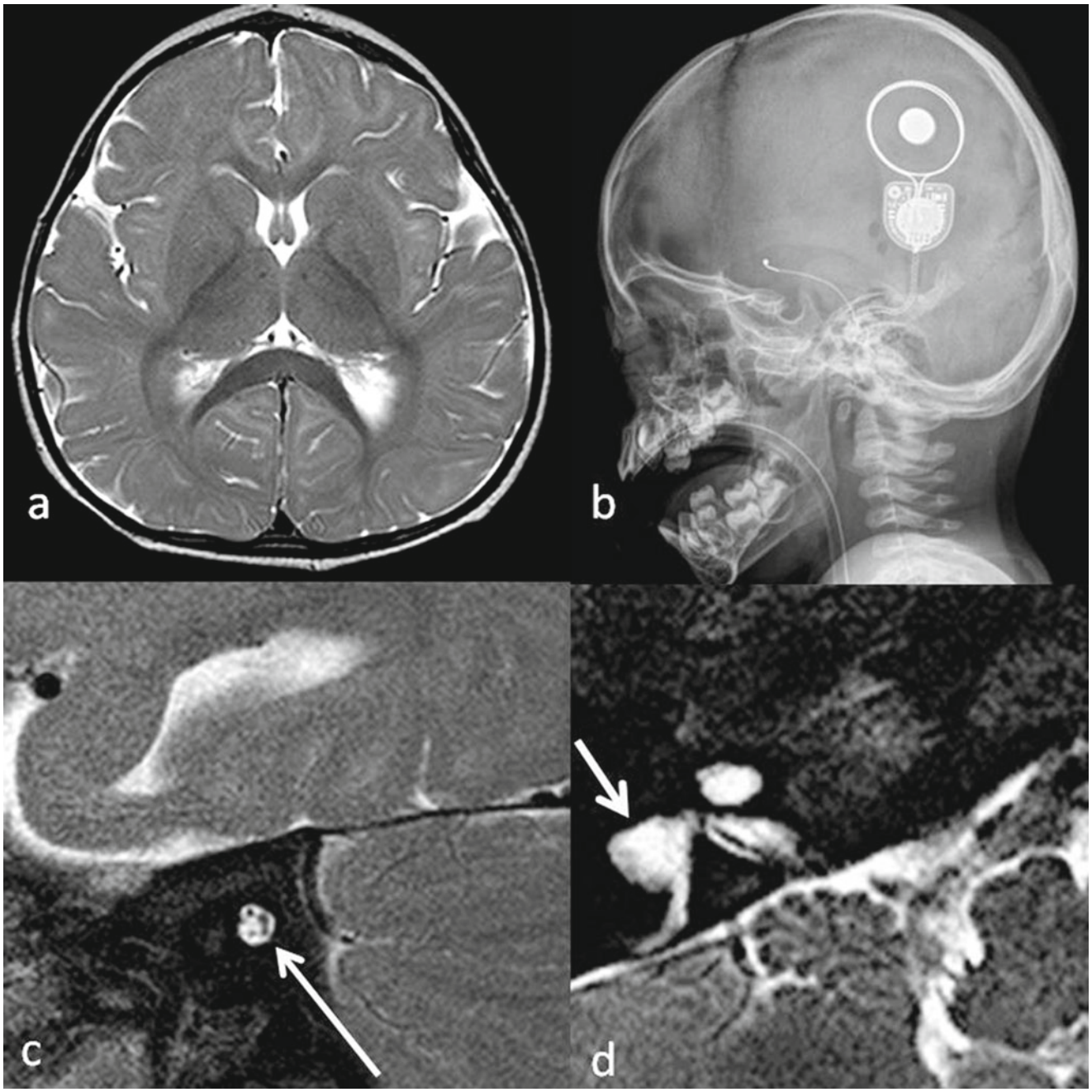

Fig. 4.1 Imaging of child with primary microcephaly. (**a**) Axial T2 MR image with disproportionally small brain and cerebral cortex; (**b**) Lateral radiograph demonstrates more vertical positioning of receiver-stimulator. High resolution 3D MR of temporal bone with (**c**) Sagittal T2 section confirming the presence of all four nerves in the IAC (*arrow*) and (**d**) Axial section demonstrating persistent anlage (*arrow*) of the horizontal semicircular canal and lack of a modiolus (**d**)

Vestibular Abnormalities

There is literature indicating that as many as 70 % of children with profound sensorineural hearing loss (SNHL) have some degree of vestibular dysfunction, with 20–40 % displaying severe to profound vestibular loss which is often bilateral loss (Cushing et al. 2008b, 2009, 2013; Buchman et al. 2004; Licameli et al. 2009). Children with certain etiologies of SNHL are more likely to have vestibular impairment. Etiologies highly associated with vestibular impairment include meningitis, cochleovestibular anomalies, Usher Syndrome (US), and cytomegalovirus. Evidence of vestibular dysfunction does not negatively influence our decision to implant a child at our institution. In fact, our research has shown that restoration of binaural access to sound leads to improvements in balance function for some children (Cushing et al. 2007).

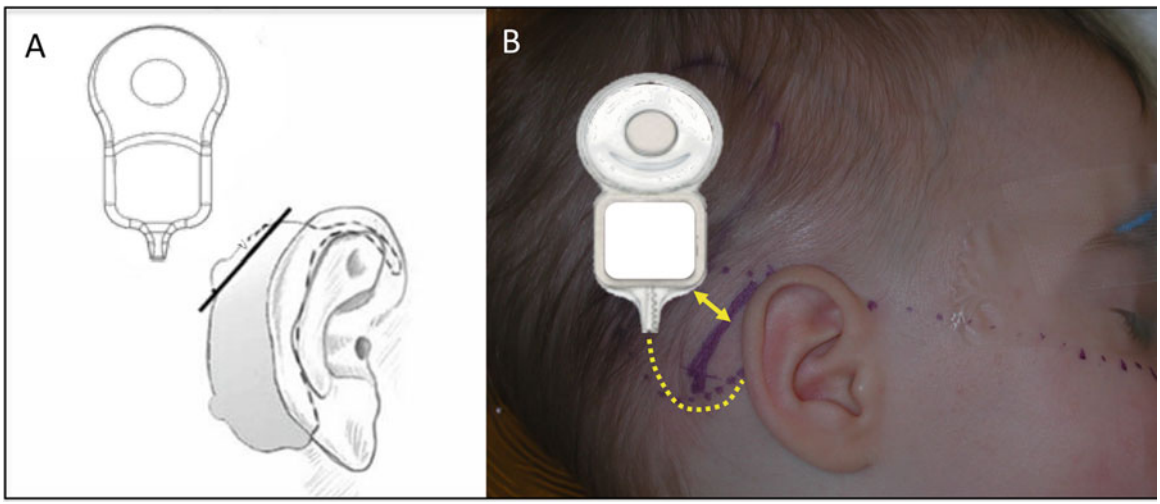

Fig. 4.2 (**a**) Schematic of superior orientation of receiver-stimulator and position relative to incision; (**b**) intraoperative marking of device position and the incision

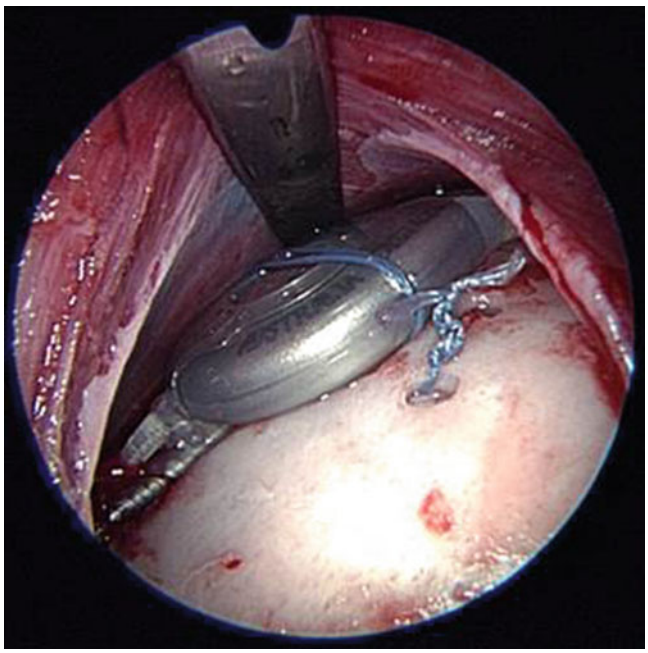

Fig. 4.3 Endoscopic view of tie-down of receiver-stimulator through a small incision approach. Device is secured to skull with suture

From a long-term management standpoint we have found that understanding the vestibular status of pediatric implant recipients to be highly useful. For this reason we recommend that these children be screened for vestibular and balance impairments. Many medical centers have little experience performing formal vestibular testing on young children and may lack a vestibular laboratory that can accommodate this population. However, clinically useful information may easily be obtained in the office. We recommend obtaining a history of motor milestones, a test of static balance (i.e., standing on one foot eyes open and eyes closed) and ideally a clinical test of vestibular end organ function (i.e., head thrust test) to identify children at risk of bilateral vestibular impairment. A particularly useful finding to identify bilateral vestibular impairment is the inability to stand on one foot for more than 4 s (eyes open in younger children and eyes closed in older children). We have found that this test of static balance predicts bilateral vestibular loss with good sensitivity (90 %) and specificity (84 %) (Oyewumi, in press).

The identification of vestibular impairment is relevant to the child with hearing loss and CIs for a number of reasons including identification of US. In our clinic, children with bilateral profound vestibular end-organ dysfunction (areflexia) without cause (i.e., no history of meningitis or cochleovestibular anomaly on imaging) are referred for ophthalmologic assessment which includes an electroretinogram. The electroretinogram measures corneoretinal potentials and is used to diagnose retinitis pigmentosa, which causes progressive visual impairment. The presence of retinitis pigmentosa in a congenitally deaf child with abnormal vestibular function is characteristic of US type 1, the most common clinical form of this disorder. The early identification of US type 1 allows for simple interventions (i.e., minimizing light exposure, vitamin therapy) (Rayapudi et al. 2013; Wang et al. 1997), which may delay the onset and progression of visual impairment. Knowledge of this diagnosis may be of great advantage to the child, as additional therapeutic strategies, experimental or otherwise, become available for the treatment of retinitis pigmentosa. In addition, the identification of US supports proceeding with bilateral implantation and promoting an auditory verbal approach with the goal of listening and spoken language for communication rather than reliance upon sign language and other types of visual communication (Ahmed et al. 2003; Jatana et al. 2013).

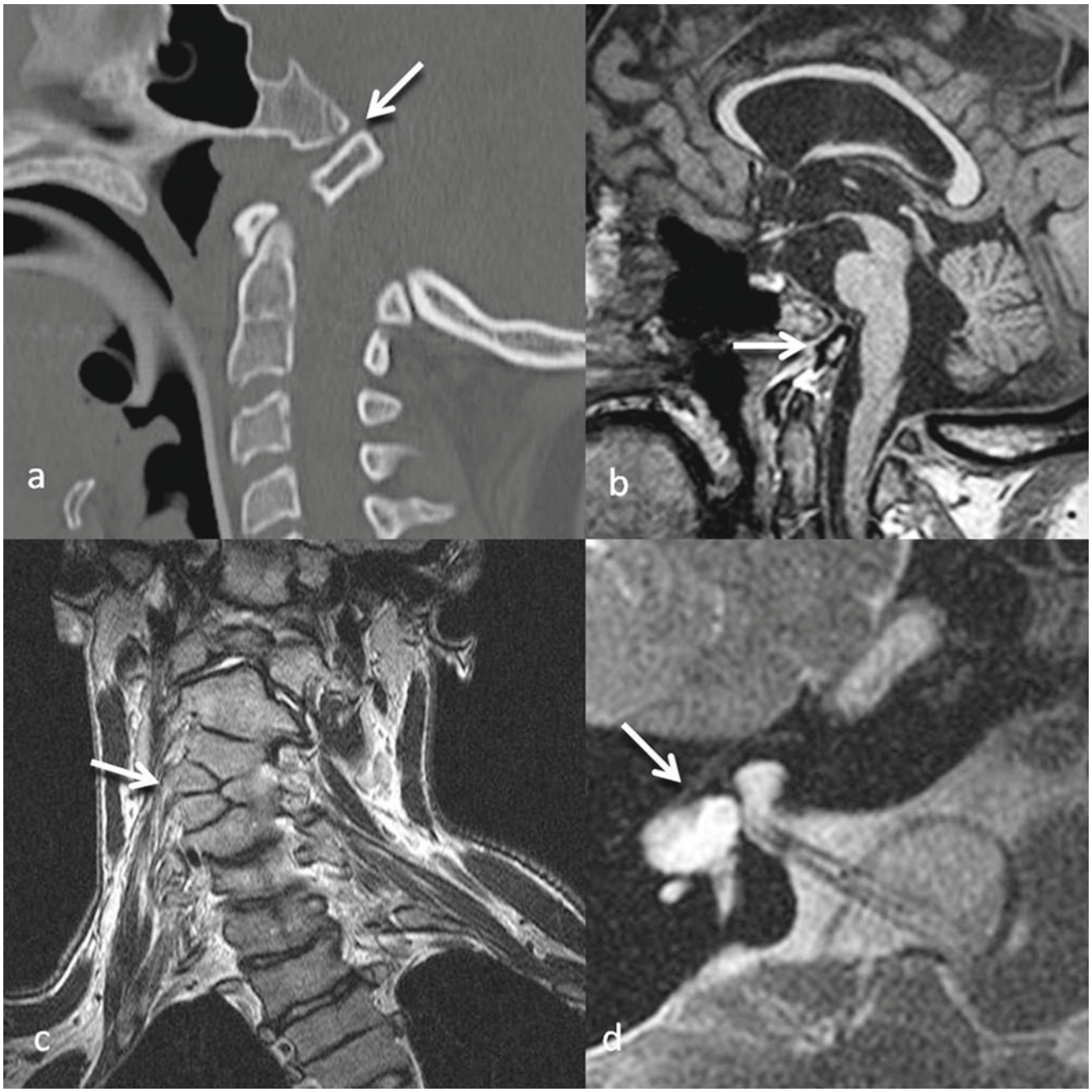

Fig. 4.4 Imaging findings in CHARGE (**a** and **b**) and Klippel–Feil (**c** and **d**). CHARGE: (**a**) Sagittal CT reconstruction demonstrates a rotated basi-occiput fragment separated from remainder of clivus by a transverse basi-occipital fissure (*arrow*); (**b**) MR sagittal T1 image demonstrates sharp angulation at this fragment (*arrow*) indenting the pontomesencephalic junction. Klippel–Feil: (**c**) MR coronal T1 image of spinal malformation in teenager demonstrates complex segmentation and fusion anomalies (*arrow*); (**d**) MR Axial T2 image of the inner ear shows typical IP1 malformation

Identifying vestibular dysfunction in children who are deaf is important beyond etiologic considerations. A recent review of our database revealed that pediatric CI recipients who had an absence of bilateral horizontal canal function (areflexia) had increased odds of mechanical or electrical malfunction of the implanted device by 7.6 times in comparison to children who were not areflexic (Wolter et al. 2015). Likewise poor balance measured on objective tests of function and saccular dysfunction measured by absence of vestibular evoked myogenic potentials, were also significantly more common in children with cochlear implant failure. Multiple previous failure studies, including our own, have noted that children with meningitis are more likely to experience higher implant device failure (Chung

et al. 2010; Eskander et al. 2011). The most likely reason for children with postmeningitic deafness being more at risk for device failure is the fact that this population universally loses their vestibular function, thus increasing their risk of falls resulting in device trauma. Again, many papers report a higher incidence of implant failure in children citing increased risk of fall as the underlying reason; however, none made the connection to poor vestibular end-organ dysfunction (Weise et al. 2005). In summary, poor balance due to vestibular impairment places children who receive an implant at a nearly eightfold increased risk of eventually experiencing device failure. The likely mechanism is an increase in falls leading to device damage. Vestibular dysfunction is therefore the largest patient related factor contributing to CI failure identified to date (Wolter et al. 2015). We believe that magnet displacement from devices containing a removable magnet held within a silastic sleeve are also more likely to occur in children with poor vestibular function as this problem typically occurs subsequent to head trauma (Fig. 4.5).

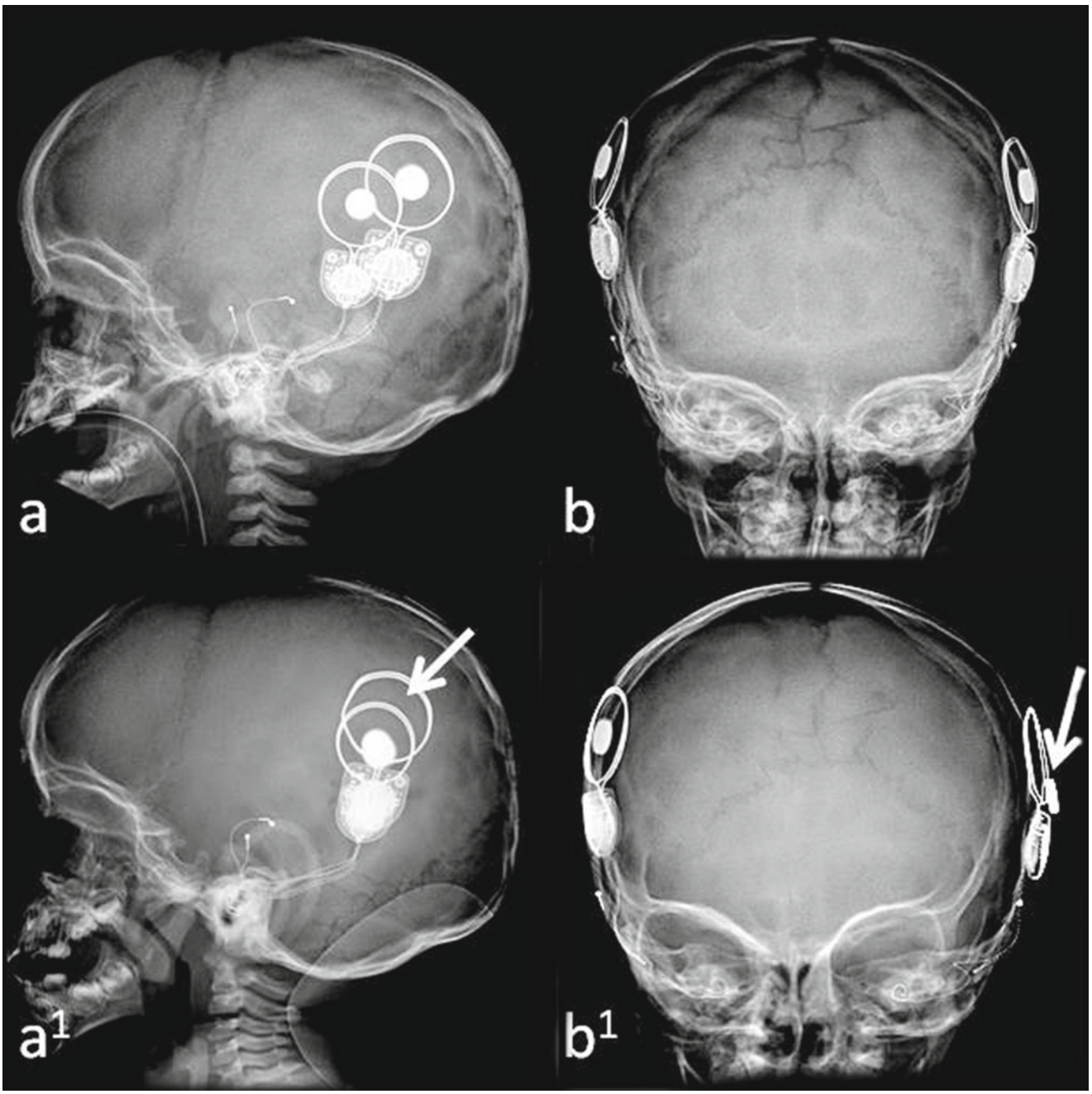

Fig. 4.5 Radiographs demonstrating position of a removable magnet normally contained within a silastic sleeve. Lateral (**a**) and frontal (**b**) radiographs take immediately after surgery demonstrating normal central position of the magnet within the sleeve. Subsequent (**a**) lateral radiograph reveals an "empty" sleeve (*arrow*); (**b**) Inferior displacement of the magnet (*arrow*) is seen on the frontal view

Metabolic, Autoimmune and Inflammatory Conditions

There are number of serious metabolic, autoimmune and inflammatory conditions for which progressive SNHL is a feature. Examples of such conditions include the mucopolysaccharidoses (MPS), neonatal onset multisystem inflammatory disease (NOMID), Muckle-Wells syndrome, primary lymphedema with myelodysplasia (Emberger syndrome) and other primary immunodeficiencies such as MonoMAC syndrome due to GATA-2 mutations. There are published reports of successful cochlear implantation in children affected by each of these disorders (Saeed et al. 2013; Neven et al. 2010; Hall et al. 2013). This fascinating group of children presents additional challenges during the assessment and surgical phase. After implantation these children may experience unpredictable variation in auditory thresholds and speech perception requiring frequent device reprogramming. This situation may be due to their systemic disease causing changes in the electrical milieu surrounding the electrode array.

Intraoperative challenges due to anesthetic considerations arise more often than difficulties related to surgery itself in many children with complex medical problems. From the standpoint of electrophysiological testing, it is important to keep in mind that anesthetic technique may interfere with measurement of the electrically elicited stapedial reflex. This evoked reflex is suppressed in a dose dependent fashion by volatile (inhaled) anesthetics such as nitrous oxide and halothane (Crawford et al. 2009). Total intravenous anesthetic techniques using propofol and remifentanil usually are successful in providing anesthesia while permitting this type of intraoperative testing.

Children with mitochondrial disease such as Kearns–Sayre, Leigh's disease, and mitochondrial encephalomyopathy lactic acidosis and seizure like episodes (MELAS) have a disorder of cellular energy management. They are prone to lactic acidosis and may not tolerate fasting without intravenous dextrose supplementation. Propofol, a commonly used intravenously administered hypnotic/amnestic agent, inhibits the mitochondrial respiratory chain at multiple sites including complexes I and II of the Krebs cycle (Lerman 2011). While propofol has been used safely in single bolus doses and short infusions in a variety of children with mitochondrial myopathies (Driessen et al. 2007), in large cumulative doses it can cause life threatening propofol infusion related syndrome (PRIS) (Allison 2007; Vasile et al. 2003). Dexmedetomidine is a sedative that may be used as part of a total intravenous anesthetic technique and is preferred by many anesthesiologists for this patient population (Rafique et al. 2013). There are no reported deleterious effects of this medication in mitochondrial disorders. At our institution we successfully used dexmedetomidine and remifentanil as anesthetic agents during implantation of a patient with Kearns-Sayre.

Children with mucopolysaccharidoses or congenital dysmorphism undergoing cochlear implantation may have difficult airways and require special management intraoperatively and postoperatively because of greater risk of respiratory complications (Frawley et al. 2012). In addition, these children may also have temporal bone abnormalities such as a thickened calvarium and dilated perivascular spaces due to deposition of mucopolysaccharides (Fig. 4.6).

As a result of the concerns outlined above, successful implantation in this group of children is often best performed in a facility in which there is a wealth of experience with pediatric anesthesia and pediatric airway disorders as well as the availability of a pediatric intensive care unit. From a surgical perspective, minimizing the duration of the surgery and thus the anesthesia exposure is of prime importance. Depending on the fragility of the child it may therefore be in the best interests of some children to undergo implantation of only one ear despite bilateral candidacy.

A number of these conditions have associated deficiencies in immunity that may increase the risk of postoperative wound complications. These children also typically have middle ear disease before and after implantation which can complicate surgery (Fig. 4.7). The latter topic is more thoroughly discussed in Section "Otitis Media" and Section "The Sclerotic Mastoid". Despite these factors, we continue to implant children with metabolic, autoimmune and inflammatory conditions, even if these disorders are expected to be fatal. Our philosophy is that if these children are expected to have a life span of 5 years we will proceed with implantation.

Malignant Disease

In children with underlying malignancy, especially those with intracranial tumors, a number of considerations have to be entertained. For some children, the cause of their profound deafness may be the treatments necessary to achieve remission from their oncologic disease (Lafay-Cousin et al. 2013; Roland et al. 2010; Torkos et al. 2002) (Fig. 4.8). The major consideration in this population is often the need for repeated magnetic resonance imaging (MRI) to monitor the status of their disease. The need for repeated MRI is particularly important for children with intracranial tumors. The presence of the CI, and in particular the internal magnet, may prohibit imaging of adequate diagnostic use, particularly of the head and neck. The options in this instance include: (1) implantation of a magnet-less device, (2) removal of the magnet prior to each MRI, (3) shielding of the magnet during MRI (Walton et al. 2014; Gubbels and McMenomey 2006; Azadarmaki et al. 2014) or (4) use of an alternative imaging modality (e.g., computed tomography). Our experience has

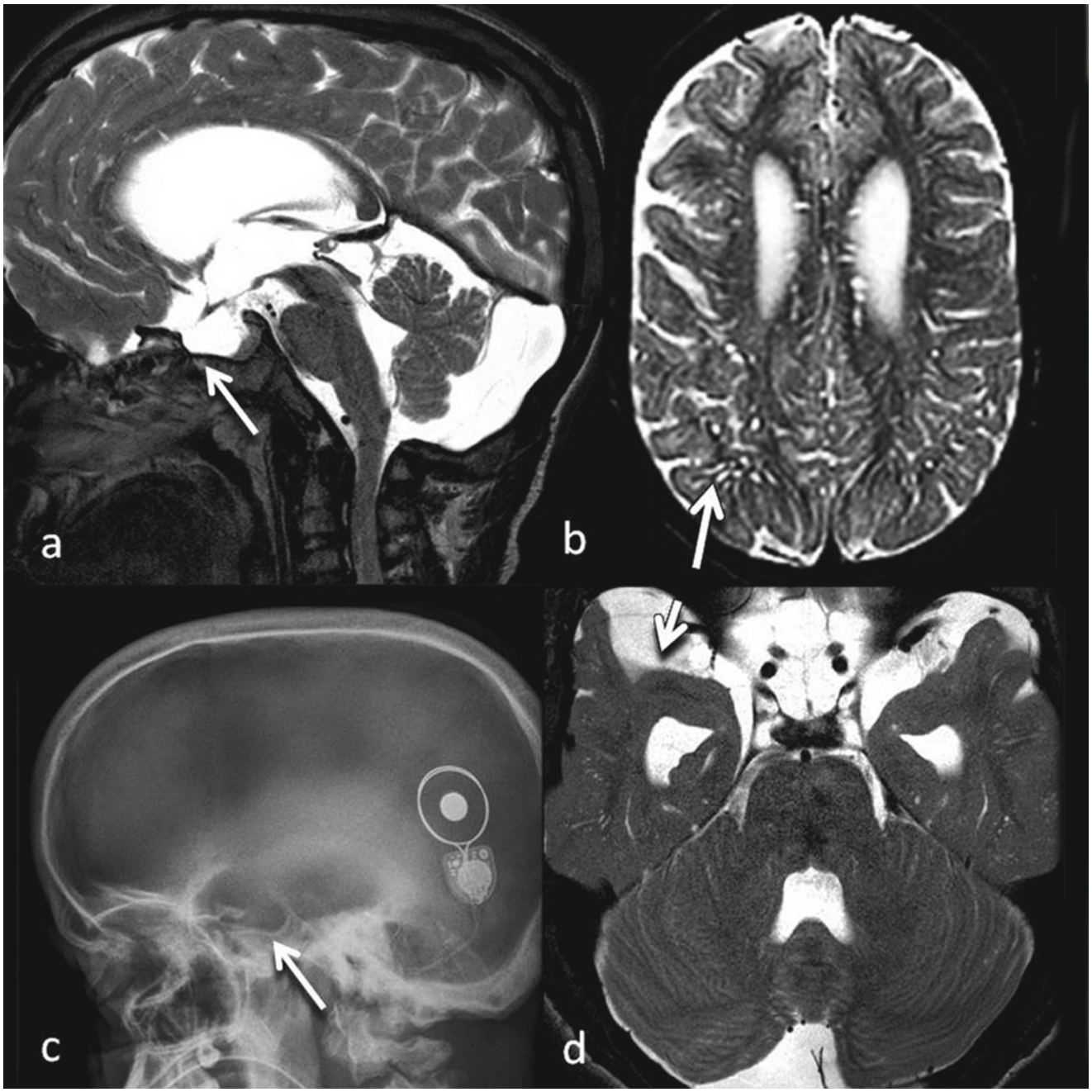

Fig. 4.6 Examples of abnormal imaging findings that occur in mucopolysaccharidosis type 2. (**a**) MR sagittal T2 image and (**c**) lateral radiograph show thickened calvarium and an enlarged sella turcica (*arrows*). (**b**) MR axial T2 image reveals typical dilated perivascular spaces (*arrow*) and (**d**) loculated basal cisterns (*arrow*) due to mucopolysaccharide deposition

primarily been to use a magnet-less device to reduce MRI artifact or to avoid MRI altogether, when the latter approach will not compromise patient care. Surgical removal of the magnet is technically feasible for those devices that have a removable magnet (Migirov and Wolf 2013). However, children who are followed, for example, for brain tumors often require long term repeated imaging at 6–12 month intervals. The removable magnets are not designed for repeated removal and replacement; doing so will likely result in problems with retention of the magnet within its silastic sleeve. In addition the need for repeated surgery with risk of infection is not optimal. Surgical removal with replacement is best reserved for instances in which repeated MRIs are not expected. There is significant literature on the use of shielding of the magnet in lower-Tesla MR scanners (Walton et al. 2014; Gubbels and McMenomey 2006; Azadarmaki et al. 2014), particularly when areas distant to the implant need to be imaged. While this is routinely done in adults, it may not

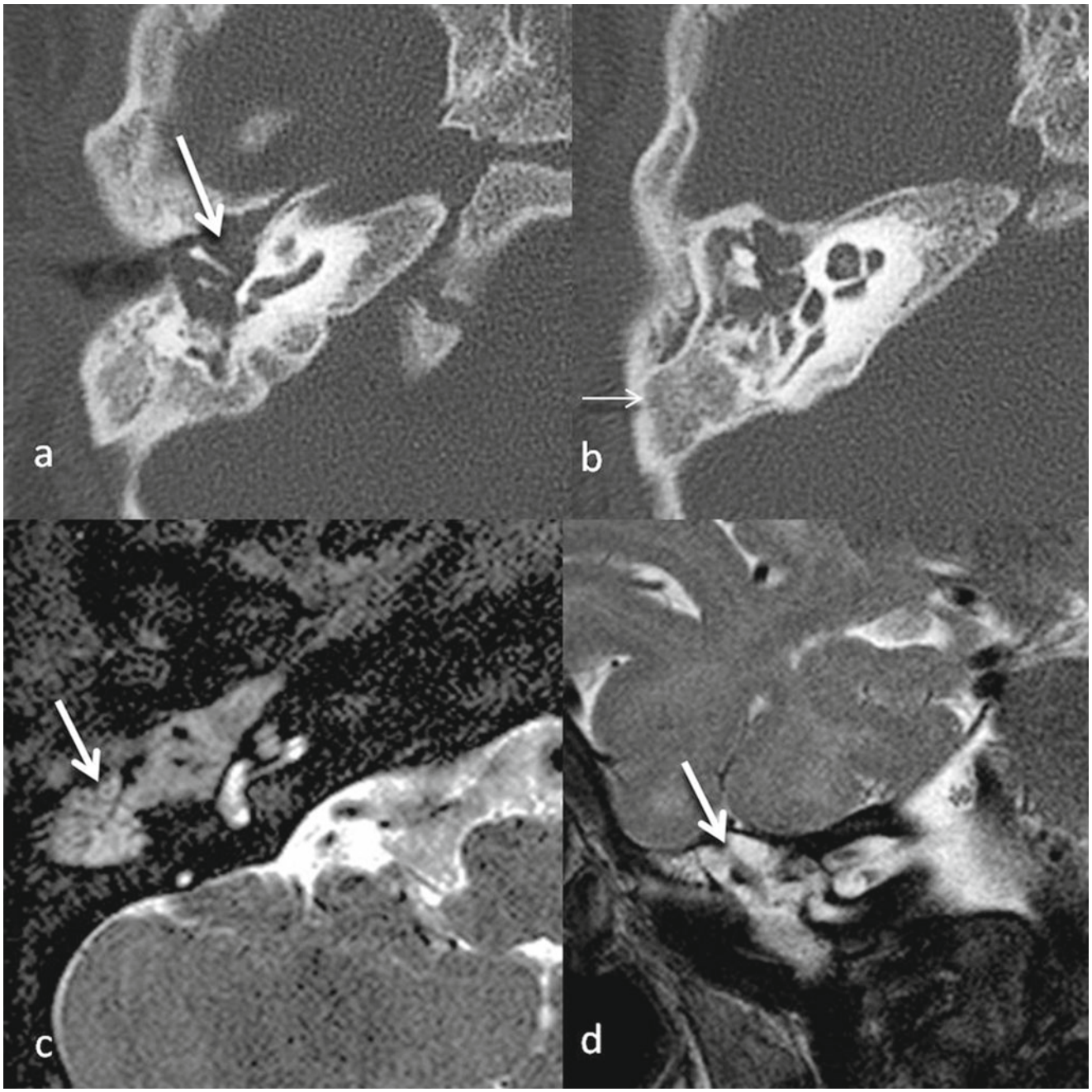

Fig. 4.7 Temporal bone imaging of an 8 month old with deafness secondary to Emberger syndrome demonstrating evidence of middle ear disease with involvement of the mastoid. Axial CT (**a** and **b**) reveals opacification of the middle ear (*arrow*) and the presence of marrow (*small arrow*) within the mastoid. (**c**) Axial and (**d**) coronal MRI also demonstrate opacification (*arrows*)

always be an ideal option in the pediatric setting given that children often require sedation for imaging and therefore are not able to report heat or pain that maybe associated with an impending complication (Hassepass et al. 2014). For children implanted with a magnet-less device, retention of the externally worn headpiece is achieved using double-sided wig tape, a headband or scarf to maintain the externally worn headpiece in alignment and proximity of the implanted internal receiver. A non-magnetic spacer is substituted for the magnet within the silastic sleeve to maintain space for reinsertion of the magnet should the disease no longer require repeated MRI.

Children who receive chemotherapy may develop vestibular impairment secondary to ototoxicity. They therefore maybe at increased risk as discussed above for eventual internal device malfunction due to repeated CI trauma from falls.

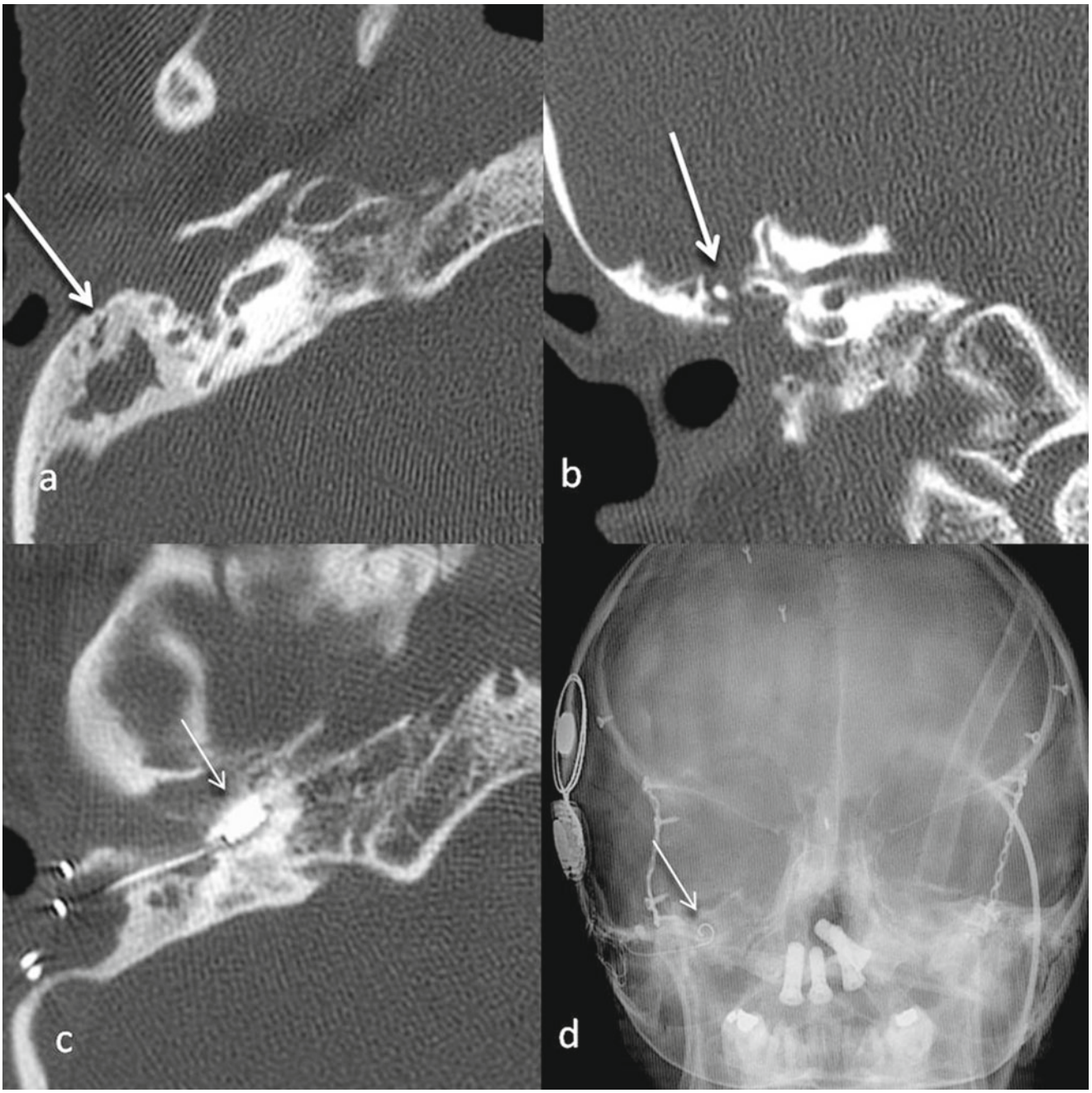

Fig. 4.8 Child with acquired hearing loss due to radiation therapy needed to treat nasopharyngeal rhabdomyosarcoma. Preoperative axial (**a**) and coronal (**b**) CT demonstrates extensive bone destruction (*arrows*) caused by radiation therapy. Satisfactory placement of electrode array within the cochlea (*arrows*) is seen on (**c**) axial CT and (**d**) frontal radiographic plain film

Congenital Cytomegalovirus (CMV)

Congenital CMV is now more frequently recognized as the cause of hearing loss in the pediatric CI population (Avettand-Fenoel et al. 2013; Park et al. 2014). Permanent childhood hearing impairment is the most common sequela of congenital CMV, if one considers both "symptomatic" and "asymptomatic" CMV infection (Manicklal et al. 2013). For children with congenital CMV, especially those more severely affected as evidenced by microcephaly, their medical condition should be assessed preoperatively and managed with great care as outlined above in Sect. "Developmental Delay and Multiple Handicaps" (Dollard et al. 2007; Cheeran et al. 2009). There are a number of radiological correlates for the neurological sequelae associated with congenital CMV. These findings include ventriculomegaly, cerebral atrophy, and intracerebral calcifications, which are known to correlate with the presence of senso-

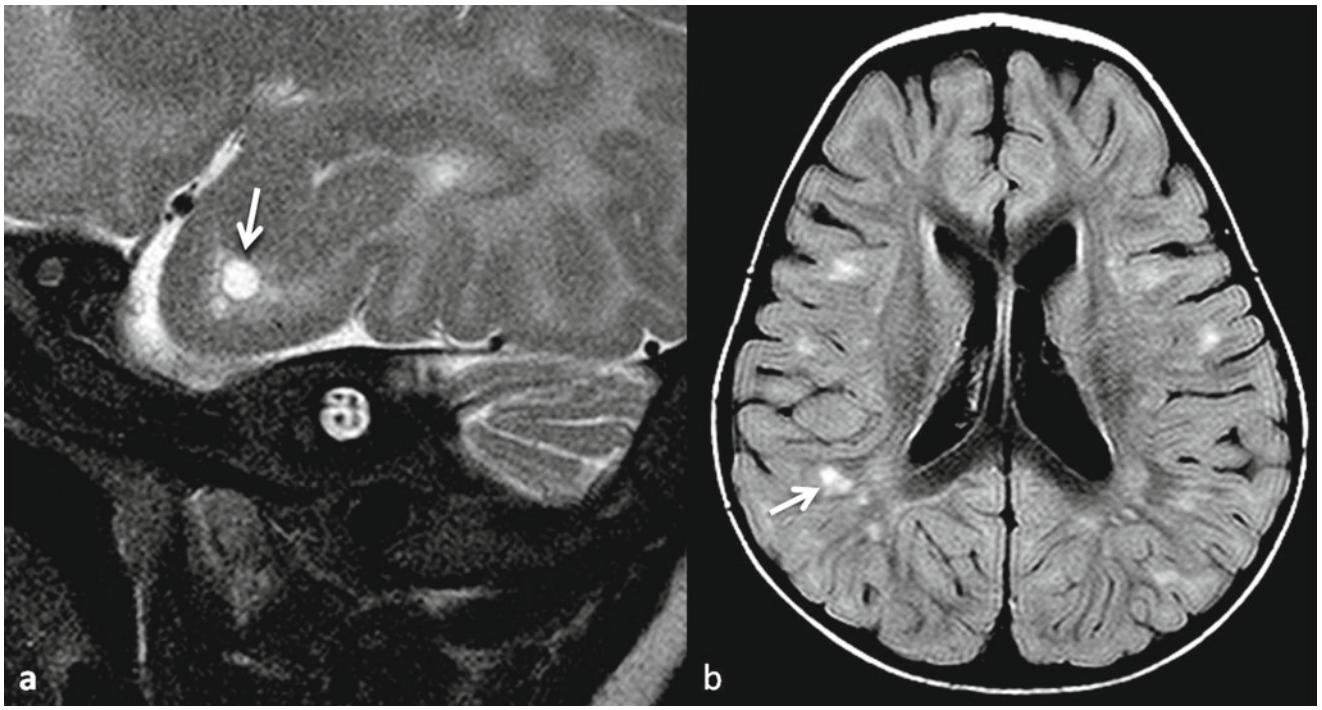

Fig. 4.9 Imaging of CI candidate with congenital CMV as etiology of sensorineural hearing loss. (**a**) MR T2 sagittal image at 4 months of age reveals typical cysts (*arrow*) in the anterior temporal lobe. The appearance of the nerves in the lateral IAC is normal. There are four nerve bundles seen of cross section: the facial nerve, cochlear nerve (the largest), superior and inferior vestibular nerves. (**b**) MR axial FLAIR image at 18 months of age demonstrates multifocal white matter lesions typical of late third trimester in utero exposure to CMV

rineural hearing loss (Fig. 4.9). In addition, these children may have microcephaly and/or positional plagiocephaly (flattening of the skull due to positioning). The latter problem is caused by motor weakness and poor coordination resulting in increased time spent lying supine which affects skull growth. The skull may precipitously slope away from the mastoid making positioning of the receiver stimulator critically important (Fig. 4.10a, b) (Barker and Briggs 2009). Surgical anatomy also may be affected by abnormal temporal bone development. The mastoid is often filled with marrow, resulting in increased bleeding and poorer visualization during surgery (Fig. 4.10c, d). These children also may have middle ear effusion that may need to be managed as outlined below in Sect. "Otitis Media."

Due to anomalies in temporal bone and skull development, implanting infants who cause of deafness is congenital CMV may be technically challenging. Fortunately many of these children become CI candidates when older, as many begin life with more hearing and over time experience progressive hearing loss. Finally, as mentioned above in Sect. "Vestibular Abnormalities," children with deafness due to congenital CMV often have vestibular impairment and poor balance which places their implant at increased risk of damage (Karltorp et al. 2014; Wolter et al. 2015).

Otitis Media

On the topic of otitis media with effusion (OME) and acute otitis media, there are specific management considerations both prior to, and following cochlear implantation. When considering the preoperative management, there is a large amount of both literature and opinion. However, no consensus has been achieved on the topic of managing children with otitis media with effusion prior to implantation. While we think that all surgeons would avoid implantation of an acutely infected ear that contains frank pus, there exists a spectrum of opinion when it comes to managing OME. As the age of implantation continues to decrease, more implantations are likely to take place during the ages when children are more prone to otitis media. Discussion of managing these children has been the topic of several panels and interesting manuscripts since the mid-1990s. There are proponents of placing ventilating tubes in children with underlying OME and then either removing them just prior to or even at the time of CI surgery versus leaving them in situ. Others avoid tympanostomy tubes altogether, unless middle ear disease is interfering with the audiological candidacy evaluation (Luntz et al. 2004, 2013). At the other end of the spectrum, a minority have advocated an aggressive approach that

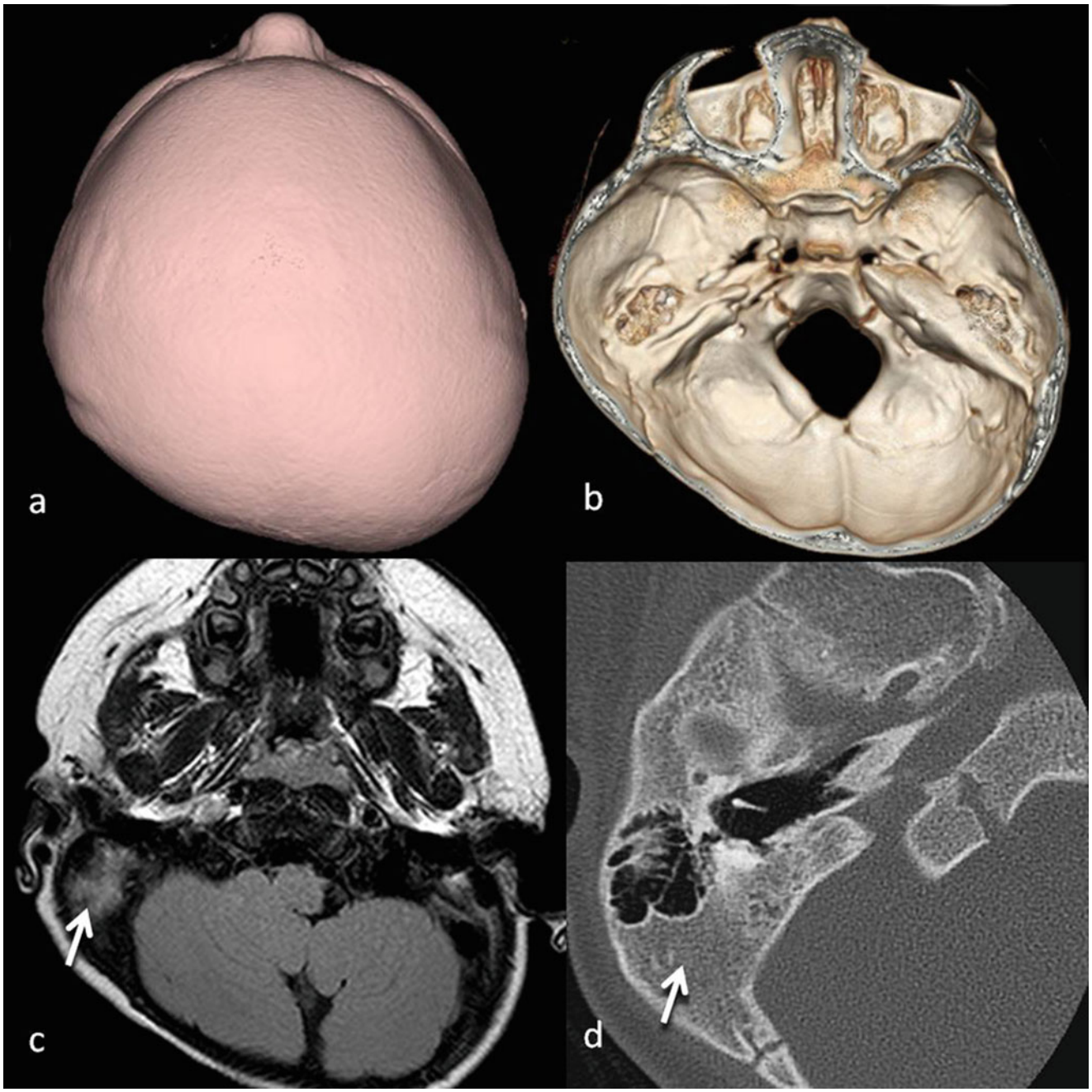

Fig. 4.10 Posterior flattening of the skull and marrow filled mastoid in an infant. (**a**) CT 3D skin surface and (**b**) skull base reconstruction shows flattening of the right side of the skull due to prolonged supine position. Marrow filled mastoid (*arrows*) is demonstrated on (**c**) MR axial FLAIR and (**d**) and axial CT images

includes a subtotal petrosectomy for management of recurrent otitis media in children undergoing cochlear implantation, although this is not a common approach (Free et al. 2013). The variability of practice reflects uncertainty as to the likelihood of infectious complications due to otitis media as well as concerns that ventilating tubes may also increase risk of subsequent infectious complications. Concerns regarding the bacterial colonization of ventilation tubes are highest in tubes that have been in place for a prolonged period of time, which is frequently the case when "long term" tubes designed to remain in place for years are present. We are particularly concerned about long shafted tubes due to our experience with a single case in which Pseudomonas aeruginosa was cultured from this type of tube at the time of CI explantation necessitated by chronic infection. For this reason our group removes this type of ventilating tube prior to CI surgery.

We believe it is reasonable to apply the same protocol to any ear intubated with a ventilating tube in which there has been recurrent otorrhea prior to implantation.

Cochlear implantation can be more challenging in an ear with persistent OME and associated extensive middle ear mucosal disease without evidence of active infection. In such instances, we employ a change in the sequence of the procedure. In children without extensive mucosal disease it is our practice to begin with a cortical mastoidectomy followed by creation of the receiver-stimulator recessed bed in the skull. We subsequently return to the mastoid cavity to perform the facial recess, cochleostomy and device insertion. In children with significant mucosal disease we follow a different sequence. We perform the facial recess approach immediately after the cortical mastoidectomy. This permits access the middle ear early in the procedure. Next the edematous mucosa is dissected off of the promontory. The mucosa is vasoconstricted by placing adrenaline (1:10,000) saturated cotton pledgets in the middle ear. The recessed bed for the receiver stimulator is then created, thus allowing the vasoconstrictor time to work. By following this approach we are usually able to proceed with cochleostomy creation or round window approach to electrode insertion with optimal exposure of landmarks.

Complications also may arise from acute otitis media following implantation. A large proportion of the children in our implant program are at a relatively high risk for acute otitis media (AOM) given the age of our implant candidates. The mastoidectomy itself, performed as part of CI surgery, may offer a protective effect against the frequency of development of AOM and its complications. However, when children who have undergone recent CI develop AOM, they may be more likely to develop spread of middle ear infection into the post-auricular soft tissue space behind the ear overlying the mastoid. Infection of this area is more likely to occur soon after CI when scar tissue formation is not complete (Rubin 2010) (Fig. 4.11). These patients' clinical presentation is similar to individuals with acute coalescent mastoiditis, although one can argue the spread of infection is due to an easy path for spread of otitis media in the recent post-surgical CI recipient who has a surgically created defect in the mastoid cortex. In 2010, a policy statement from the American Academy of Pediatrics outlined principles of management of AOM in children with cochlear implants (Osborn et al. 2013). This statement includes recommendations regarding the prevention, early recognition, and treatment of AOM and meningitis. However, there is a relative dearth of literature regarding treatment of post-auricular abscess/mastoiditis in an implanted ear. Based on our experience with eight patients, we proposed a treatment algorithm to achieve resolution of infection while minimizing the need for device explantation (Osborn et al. 2013). We recommend immediate institution of intravenous antibiotics without delay for the purposes of obtaining cultures. In our experience, if examination reveals a typical presentation, imaging may not be required. We believe that early surgical drainage of the mastoid to ensure rapid resolution is important to minimize the likelihood that explantation will be necessary. All children should be examined by an otolaryngologist, preferably one with considerable CI experience. Children with frank abscess in the post-auricular region should undergo incision and drainage, in conjunction with a myringotomy and tube. At our center, drainage is generally performed by opening the incision made during implantation. An alternate technique is large-bore needle aspiration, with careful direction of the needle away from the implant body. Damage to the device may occur during either technique, so it is essential to remain mindful of its location. In children without obvious post-auricular abscess, but with pus in the middle ear, we proceed with a myringotomy and tube. We proceed with opening of the post-auricular incision if they fail to improve or worsen (Osborn et al. 2013).

Seizure Disorders

Many of our medically complex children who undergo cochlear implantation may have a history of seizures, and given the increasingly young age of implantation, some will develop a seizure disorder subsequent to implantation. The main surgical consideration, in the setting of a known, preoperative history of seizure disorders, is protecting the device from possible repetitive trauma during generalized seizures. These children therefore may benefit from firm fixation of the device as described above in Sect. "Developmental Delay and Multiple Handicaps" Additionally, several children within our implant population either developed new-onset seizures or the frequency of their preexisting seizures appeared to increase following implantation. The question arose as to whether or not the implant may be playing a role in seizure generation. In light of this, we performed a review examining the prevalence and features of seizure disorders in our implant population (Shinghal et al. 2012). Overall, we demonstrated that the prevalence of post-implantation seizure disorders in our population (0.37 %) is lower than that of the overall population (0.5–1 %). We also described the use of implant triggered EEG as a useful tool in determining if there is any correlation between seizure activity and implant activation (Shinghal et al. 2012).

Ventriculoperitoneal Shunts

The presence of a shunt does not preclude cochlear implantation (Chadwick et al. 2014; An et al. 2011). Ideally, we avoid operating on the side of the head with a working shunt and instead implant the contralateral ear. Our philosophy is that shunt function takes precedence over the implant and therefore

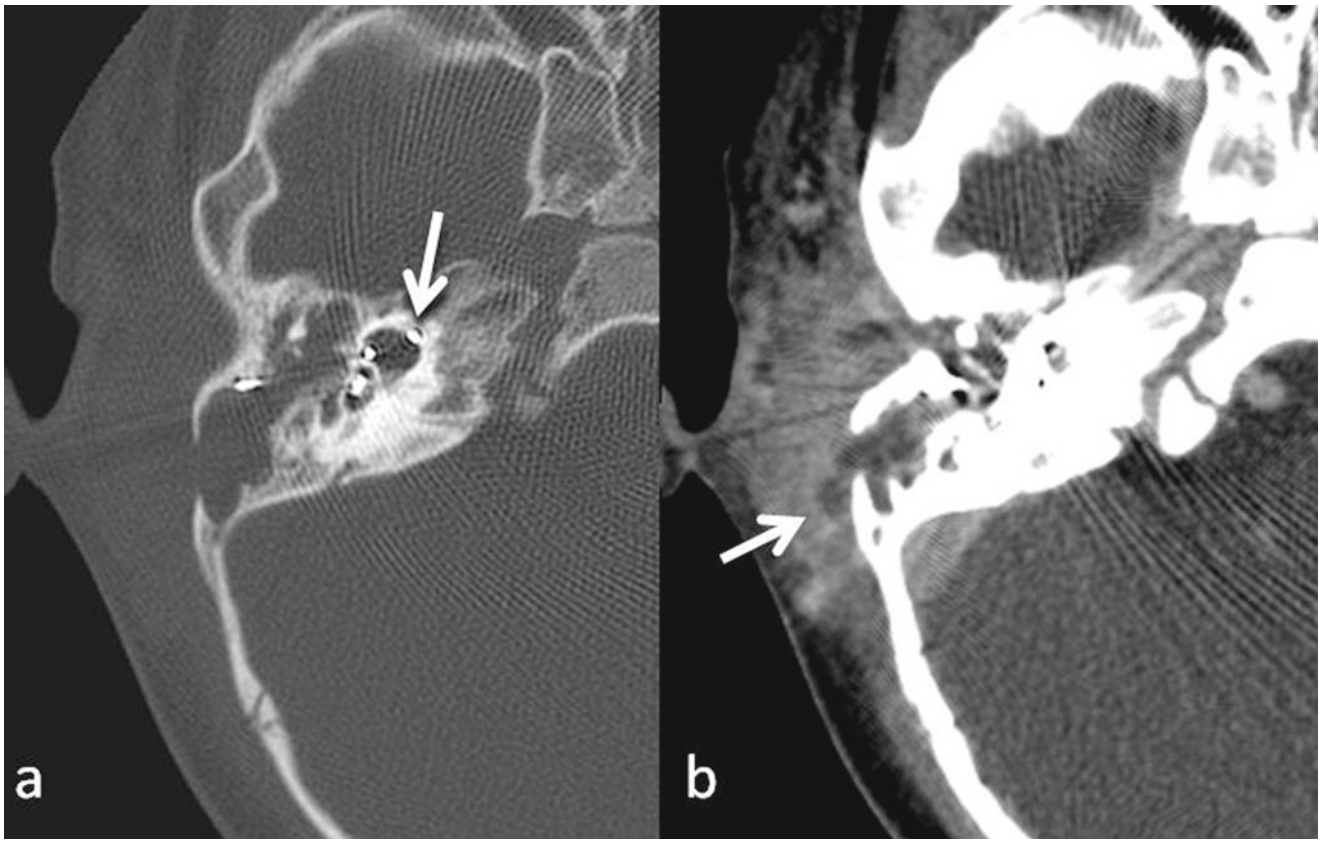

Fig. 4.11 Subperiosteal abscess 1 year after implantation. (**a**) Axial CT of right ear demonstrates swelling of peri-auricular soft tissue, surgical defect in cortical mastoid bone and opacified mastoid and middle ear. The electrode array (*arrow*) is appropriately positioned inside the cochlea. (**b**) Contrast enhanced axial CT reveals abscess and inflammation in the mastoid cavity extending into the peri-auricular soft tissue (*arrow*)

have avoided shunt revision/repositioning for the sole purpose of cochlear implantation. However, as children who require shunts over the long term typically do require periodic revisions, the future opportunity to direct repositioning of the shunt to allow for ipsilateral cochlear implantation may arise. An additional complicating factor is that many of the newer shunts are programmable with a magnet. These children may experience interference with shunt function when the external coil magnet is brought into proximity of the shunt valve (Wiet and El-Kashlan 2009).

Radiologic Conditions Affecting Cochlear Implant Candidacy, Surgical Technique or Outcome

Identifying Radiologic Anomalies of the Labyrinth and Temporal Bone

Since the advent of cochlear implantation, imaging of the inner ear has been an integral part of the evaluative process during candidacy assessment. Just as the devices we implant and the surgical techniques that we use have evolved, so have imaging techniques and our ability to interpret them. Early on, high resolution non-contrast computed tomography (CT) of the temporal bone was the primary imaging modality used to evaluate CI candidates. CT provides optimal evaluation of the bony anatomy and identification of anomalies of the inner ear. However, there are limitations to CT including its lack of sensitivity in identifying cochlear nerve deficiency. Direct imaging of the auditory nerve is one of the major advantages of magnetic resonance imaging (MRI) with high resolution three dimensional sequences of the temporal bone.

An additional limitation of CT is that it provides less useful information about the brain. In comparison, MRI is more likely to demonstrate a variety of finding that may point to a particular etiology of SNHL (e.g., CMV or hyperbilirubinemia). We discuss the specific imaging characteristics for each of these entities below in Sect. "Managing Radiologic Anomalies of the Labyrinth and Temporal Bone."

For a period of time we utilized both CT and MRI to assess implant candidates. To accomplish these studies, young children require sedation or general anesthesia. To

minimize the number of anesthetic exposures, these studies were done under that same anesthetic which necessitated transportation of the child from one area of the hospital to another. In light of the cost, logistics and safety concerns we conducted a study that enabled us to streamline our imaging protocol. This study, which included a blinded assessment of relevant clinical information obtained or lacking from both imaging techniques demonstrated that MRI alone was effective for all measured parameters (Trimble et al. 2007). CT was rarely required and its value could often be predicted based on the presence of certain diagnoses such as CHARGE syndrome (Fig. 4.12), meningitis (Fig. 4.13), bilateral atresia with SNHL. Another advantage of using MRI alone is avoidance of radiation exposure. Based upon our study findings, MRI alone is routinely done for preoperative CI evaluation of children. In

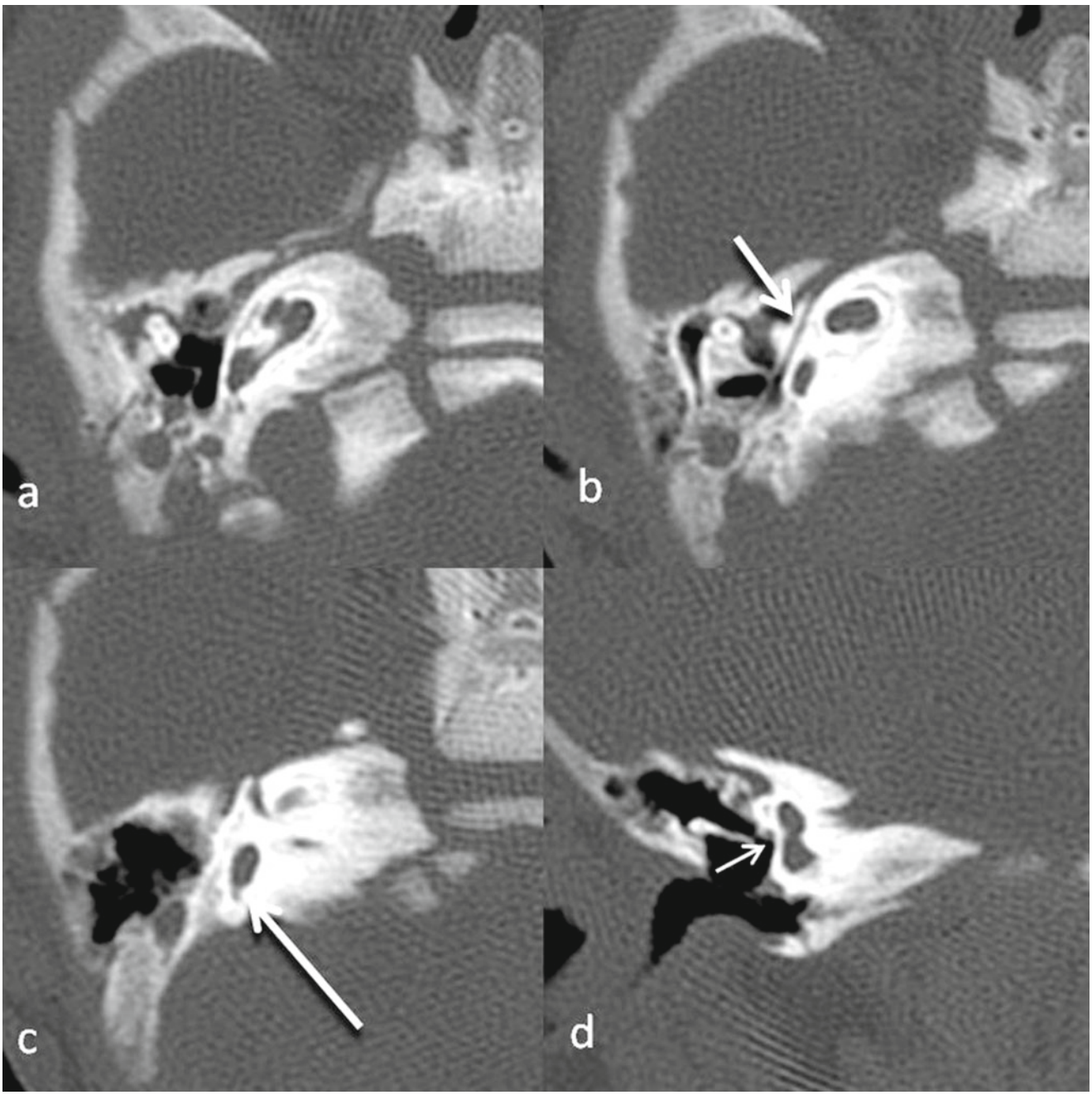

Fig. 4.12 CT of right ear of infant with CHARGE syndrome demonstrating (**a**) hypoplastic cochlea; (**b**) Contiguous slice reveals hypoplastic facial nerve (*arrow*); (**c**) a "millet-shaped" vestibule (*arrow*); (**d**) Coronal image confirms absence of the oval window (*small arrow*) with overlying horizontal segment of the facial nerve

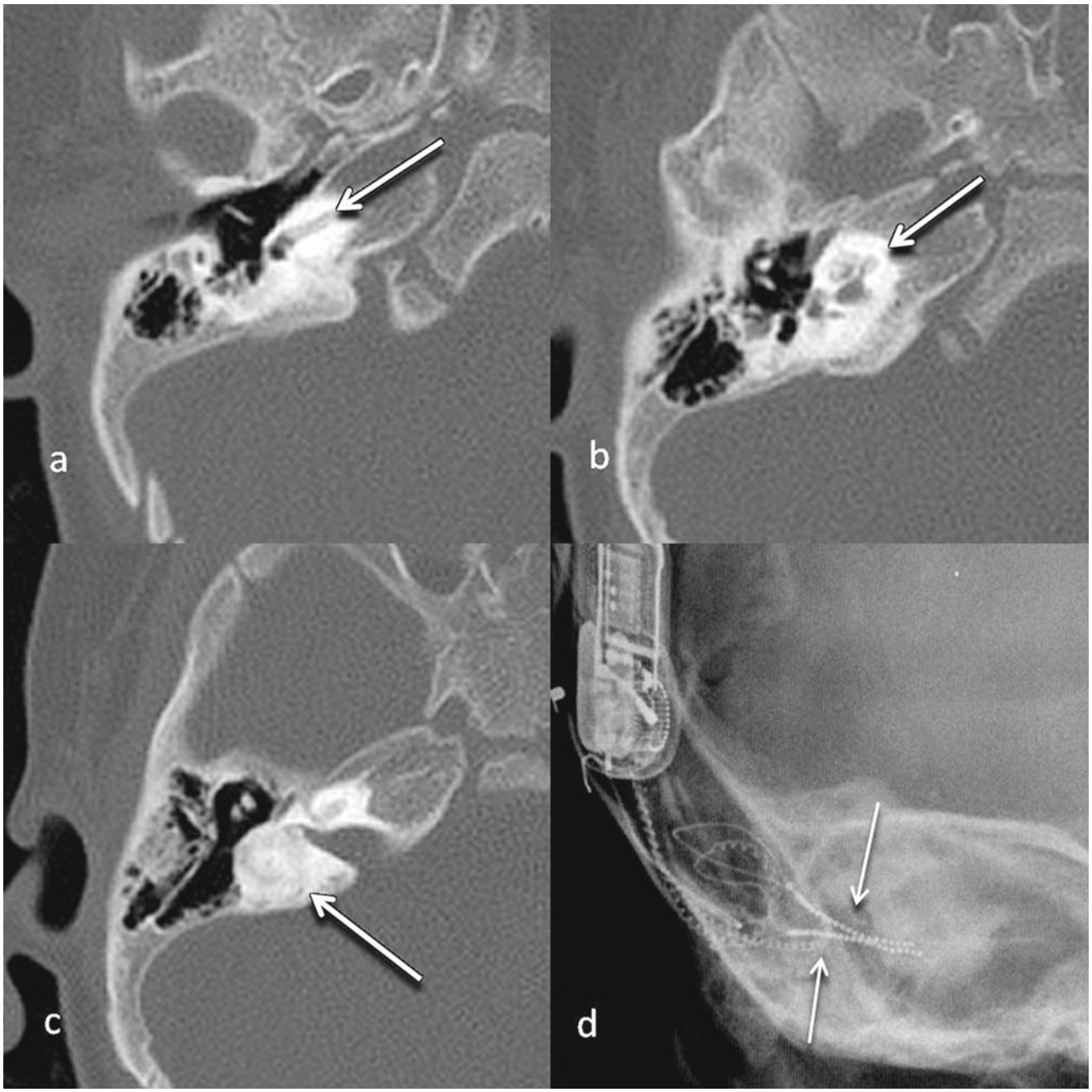

Fig. 4.13 Ossification of right cochlea secondary to bacterial meningitis. Axial CT images reveal ossification (*arrows*) of the (**a**) basal turn; (**b**) involvement of all turns of the cochlea; and (**c**) semicircular canals. Radiograph (**d**) demonstrates CI device with two electrode arrays (*arrows*). The distal portion of each array contains the active electrodes which lie within the ossified cochlea within surgically created channels

the presence of certain diagnoses where unusual surgical anatomy is expected, or when unusual temporal bone anatomy is of concern based upon MRI findings, a non-contrast CT temporal bone is also obtained using a low radiation dose technique. In our experience, less than 5 % of pediatric CI candidates require CT in addition to MRI.

To systematically identify and create a surgical plan for CI candidates, we review imaging studies during a weekly case conference attended by neuroradiologist(s) and implant surgeon(s) along with trainees. The results of imaging for each are assessed for evidence of cochlear nerve or internal auditory canal abnormalities, cochlear labyrinthine anomalies as well as abnormalities or features of the temporal bone that would affect access to the cochlea. We categorize labyrinthine abnormalities using a classification system proposed in 2002 (Sennaroglu and Saatci 2002). In addition we will look for any changes in the brain that may help us attribute the hearing loss to a specific etiology.

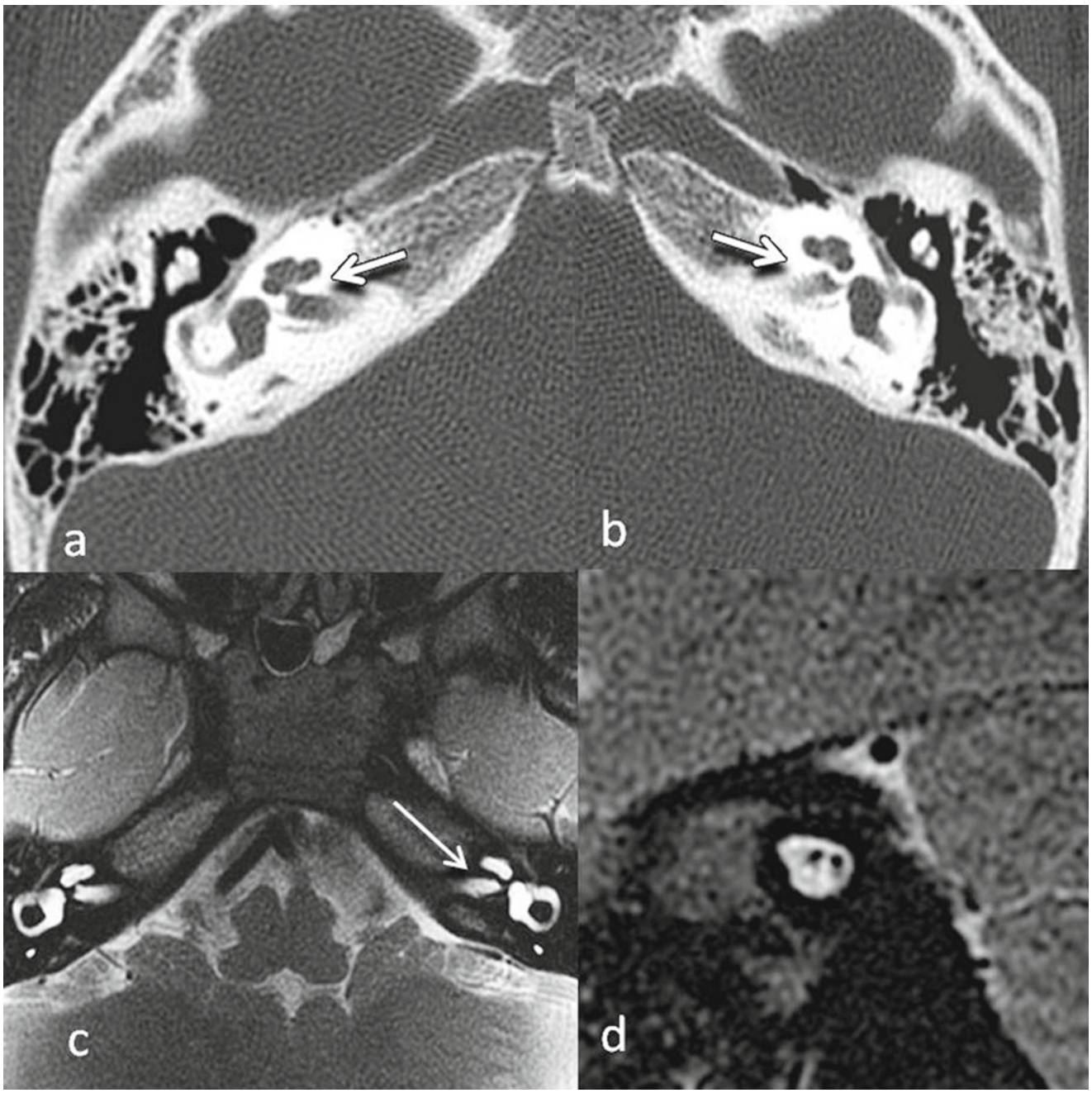

Fig. 4.14 CT and MRI findings demonstrating cochlear nerve deficiency in a 3 year old child with profound bilateral sensorineural loss. Axial CT demonstrating abnormal bony canal through which cochlear nerve leaves the IAC to enter the inner ear: (**a**) right cochlea with moderate stenosis (*arrow*) and (**b**) left cochlea with very severe stenosis (*arrow*). MRI findings: (**c**) axial T2 image without evidence of nerve entering the inner ear (*arrow*); (**d**) sagittal T2 image of left ear shows only two nerve bundles in the internal auditory canal; neither in the normal position of the cochlear nerve

Managing Radiologic Anomalies of the Labyrinth and Temporal Bone

Cochlear Nerve Deficiency

In our experience, the single most important radiologic abnormality that impacts auditory outcomes is the presence of cochlear nerve deficiency (CND), a term that includes nerve hypoplasia and aplasia (Fig. 4.14). CND occurs most commonly, although not always, in the presence of a narrow or absent cochlear nerve canal (Fig. 4.15) (Yan et al. 2013).

Children who present with labyrinthine agenesis (Fig. 4.16), Michel aplasia (Fig. 4.17) or an isolated otocyst remnant (Fig. 4.18) are not candidates for cochlear implantation as they have no inner ear in which to place the CI electrode array. Those with absent auditory nerve as determined by MRI have also been viewed as non-candidates.

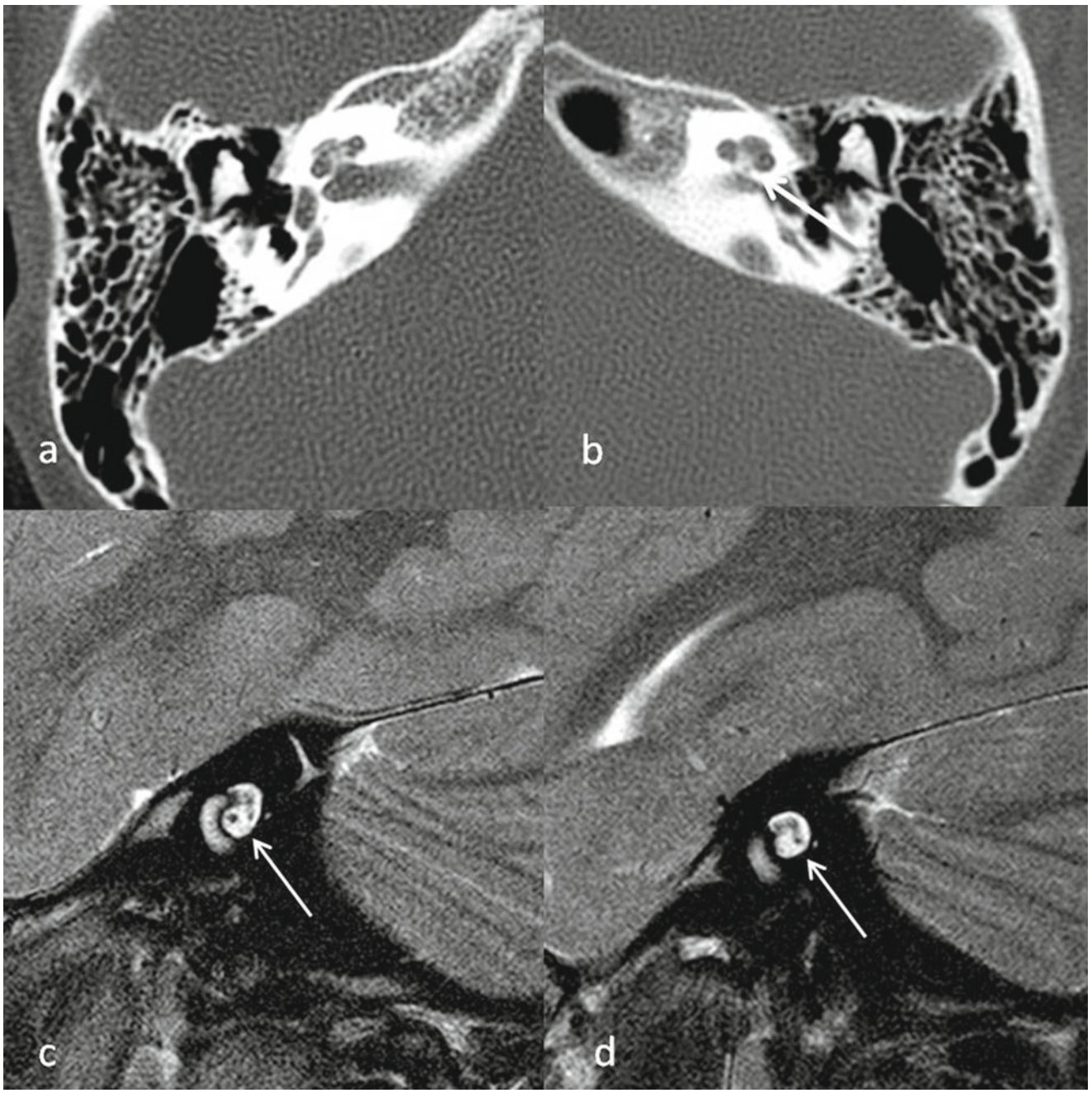

Fig. 4.15 Example of bony plate obstructing cochlear canal, the entryway into modiolus of cochlea. Axial CT: (**a**) right ear with normal cochlear nerve canal; (**b**) left cochlear nerve canal obstructed by a bony plate (*arrow*). MRI Sagittal views: (**c**) right IAC (*arrow*) with all four nerves bundles present; (**d**) left IAC (*arrow*) with only three nerve bundles due to absence of cochlear nerve

However, MRI resolution and quality may limit differentiation of complete aplasia from deficiency. If CND is identified, there is lack of consensus on when CI is appropriate. Some programs have attempted to use electrophysiological testing to predict outcomes when CND is present. However, an electrophysiologic response at the level of the auditory nerve or brainstem in response to CI stimulation in this population does not always predict good outcome as measured by behavioral speech measures (Valero et al. 2012). There are a number of published reports of reasonable performance in some implanted children with CND. Favorable outcomes in individuals with CND likely means that at least the minimum number of auditory fibers necessary to provide useful input to the brain are present (Young et al. 2012).

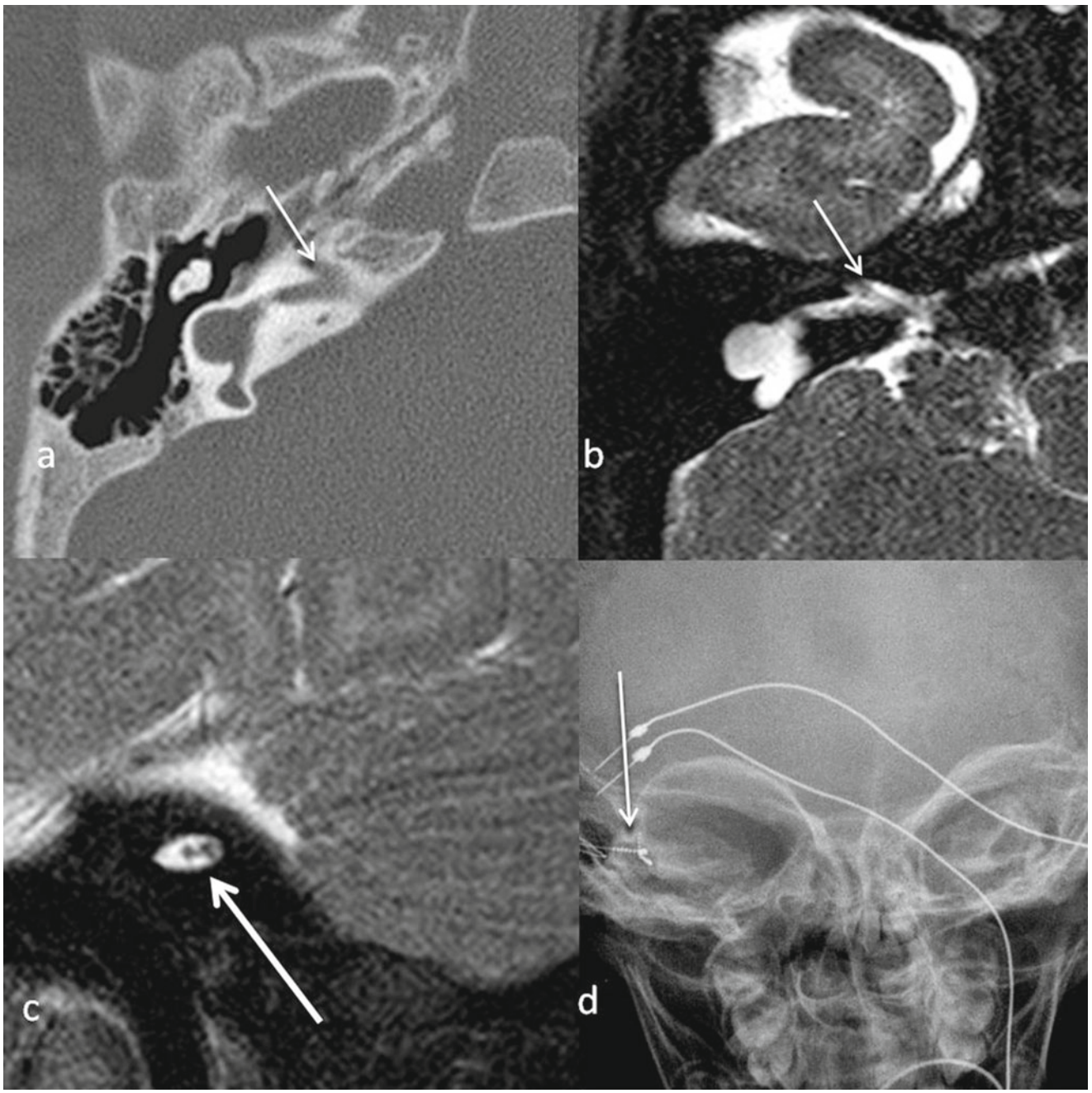

Fig. 4.16 Example of imaging of cochlear agenesis. Right ear axial (**a**) CT and (**b**) T2 MRI reveal medialization of the facial nerve (*arrow*), not an uncommon finding in labyrinthine dysplasia. The IAC is angled posteriorly toward a dysplastic vestibular labyrinth. The turns of the cochlea are absent. (**c**) Sagittal MR reveals only two nerves within the IAC (*arrow*). (**d**) Postoperative frontal radiograph of CI lead which appears buckled (*arrow*) with malformed labyrinth

As a general statement, it is fair to say that children with a CND perform significantly poorer than children with a normal size auditory nerve. Poorer performance had led some to consider children with a CND as candidates for auditory brainstem implants (ABI) (Colletti et al. 2014), while others recommend alternate modes of communication. A discussion of CND is present in Chap. 14, of ABI in Chap. 15 and the use of electrophysiological measures in Chap. 8.

Another consideration is the socioeconomic dimension of implanting a child with CND. Since the surgical risks are low in the hands of a capable surgeon, a "trial" of cochlear implantation maybe considered although the degree of

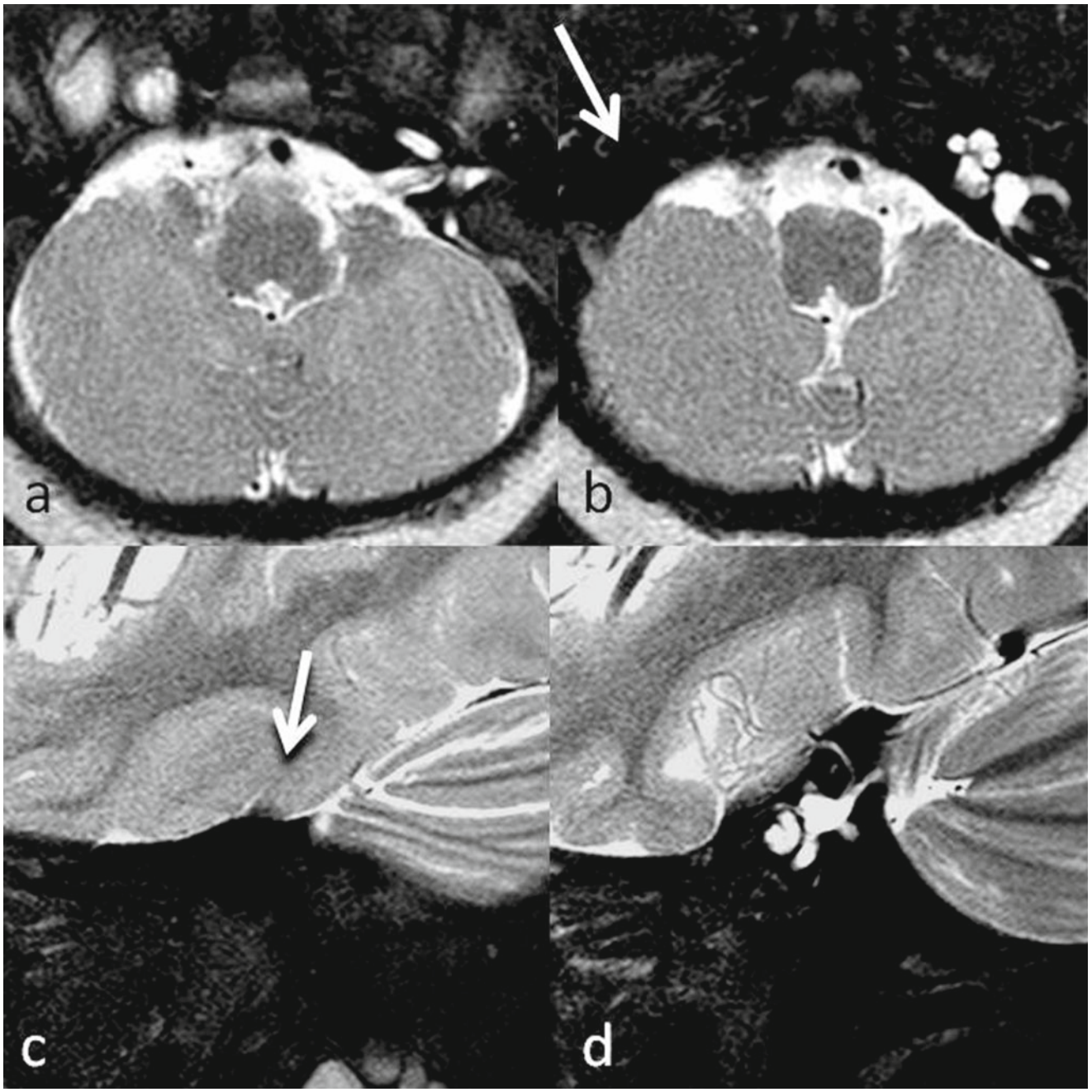

Fig. 4.17 Newborn with complete absence of the right inner ear (Michel aplasia) as demonstrated by MRI. (**a** and **b**) Axial views demonstrating lack of development of right petrous apex, labyrinth and IAC (*arrow*). The left IAC and cochlea are normal. (**c** and **d**) Sagittal T2 views in the same child at 6 years of age confirms hypoplastic petrous apex (*arrow*), lacking the normal petrous tip covering the superior semicircular canal seen in (**d**)

auditory improvement is uncertain. Although CI may not prove beneficial, good progress with a standard CI may avoid the additional cost and risk of an ABI. Perhaps with further research we will be able to better predict CI outcomes in this population either by more sophisticated imaging or electrophysiological techniques.

Cerebrospinal Fluid (CSF) Leak and Its Management

Successful intraoperative management of a CSF leak that may arise when implanting children with cochleovestibular anomalies is imperative to prevent complications of bacterial meningitis. In addition, the presence of a brisk CSF leak arising from within

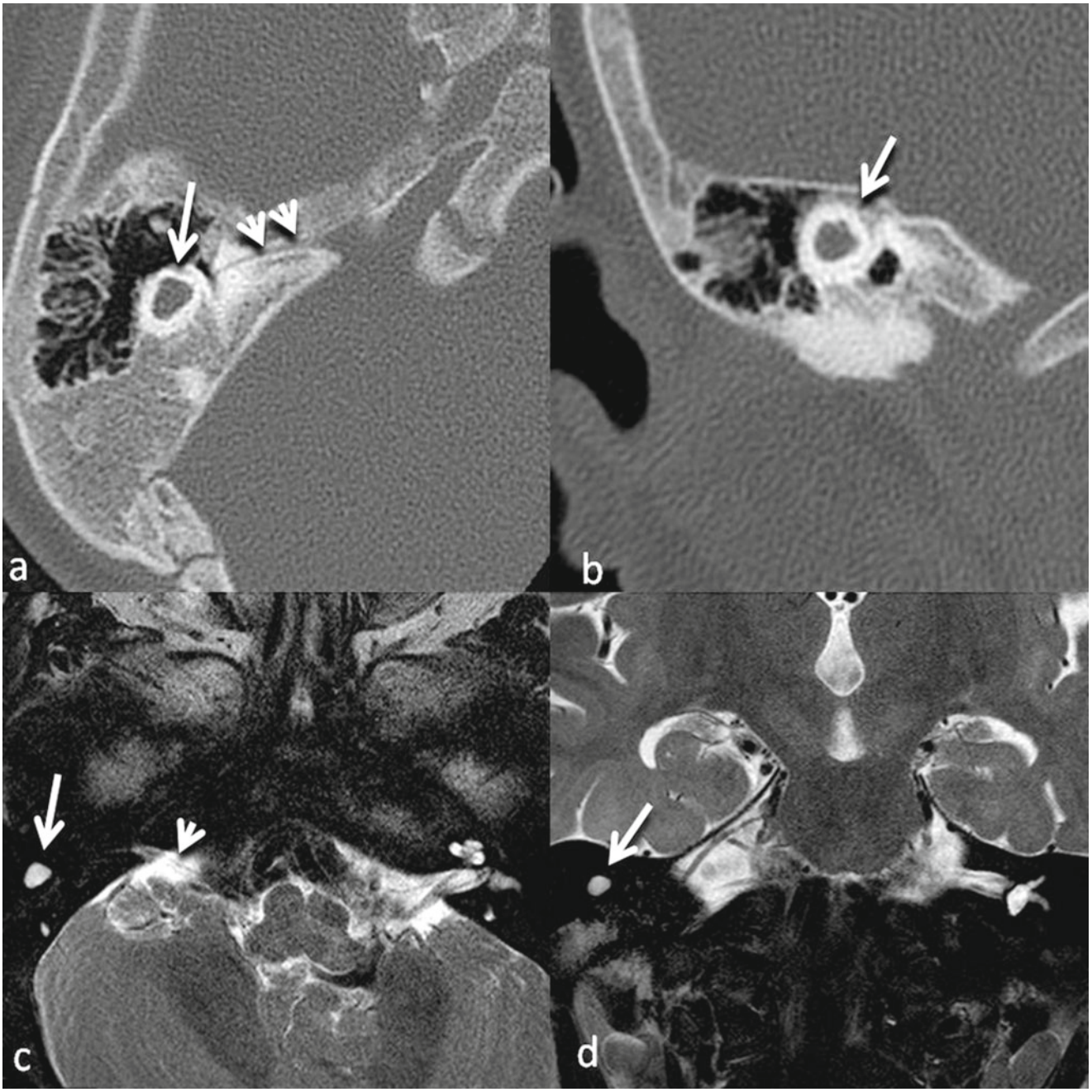

Fig. 4.18 Imaging findings in a 14 month old with right profound loss and facial nerve paralysis. A right isolated otocyst remnant is present and left ear has normal anatomy. CT: (**a**) Axial view isolated otocyst (*arrows*) remnant and extremely small facial nerve canal (*arrow heads*) is seen "pointing" to the remnant; (**b**) sagittal view of otocyst remnant (*arrow*). MRI (**c**) axial view of right otocyst remnant (*arrow*) and small canal for facial nerve (*arrow head*), with normal appearing left labyrinth and IAC; (**d**) coronal view of right otocyst (*arrow*)

the cochlea presents an additional technical challenge to precise surgical placement of the electrode array.

Based upon imaging, it is usually possible to predict when there is increased risk of CSF being present within the cochlea, and thus a CSF leak occurring during CI surgery. From a parental counseling and surgical planning standpoint, advanced knowledge is advantageous. The risk of a CSF leak is increased if cochleovestibular anomalies are present. It might seem logical to predict that the degree of dysplasia would relate directly to the likelihood of encountering a CSF within the cochlea; however, this is not necessarily the case. A CSF leak occurs as a result of a defect at the lateral end of the internal auditory meatus. This barrier can be deficient in some congenitally dysplastic ears allowing confluence between CSF and perilymph (Phelps et al. 1993; Papsin 2005). A defect in the modiolus may be very

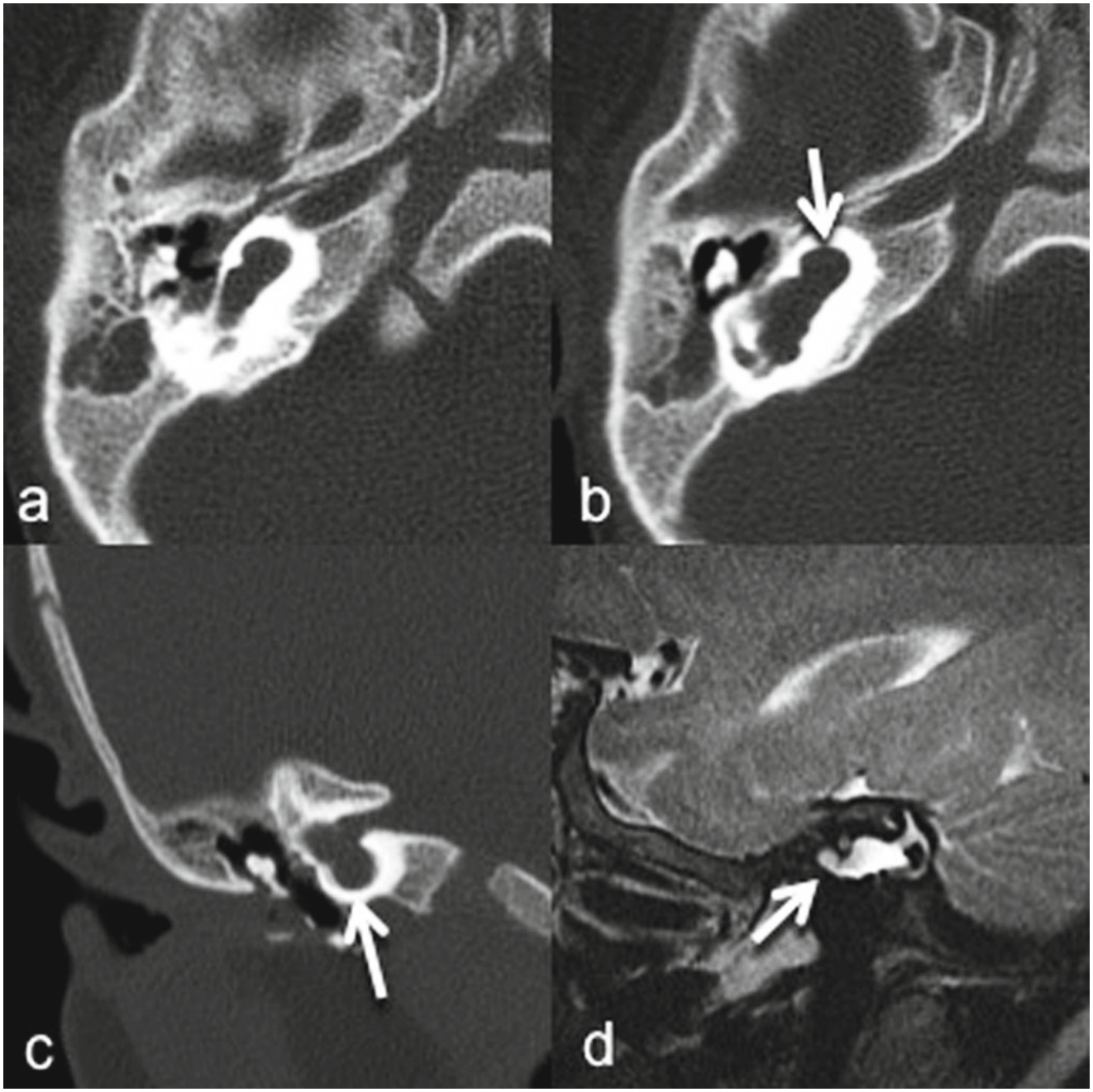

Fig. 4.19 Infant with common cavity malformation of right ear. CT at age 8 months: axial (**a** and **b**) and (**c**) coronal sections. The cochlear pole (*arrow*) is bulbous and there is a malformed vestibular pole with dilated vestibule and semicircular canals. (**d**) Sagittal T2 MRI at age 11 months again reveals the dilated cochleovestibular sac (*arrow* points to cochlear pole)

apparent on CT scan (Graham et al. 2000). However, in some cases the modiolar defect is subtle and not easily recognized. Modiolar defects may be present in children whose only labyrinthine abnormality is an isolated wide vestibular aqueduct, the most common cochlear malformation. CSF leaks may occur in common cavity deformity (Fig. 4.19), incomplete partition type I (IP-1) (Fig. 4.20), incomplete partition type II (IP-2) (Fig. 4.21), isolated enlarged vestibular aqueduct, cystic cochleovestibular anomaly with enlarged ductus reunions as well as in the setting of X-linked deafness with stapes gusher syndrome (Phelps syndrome) (Fig. 4.22) (Stankovic et al. 2010).

A number of techniques have been developed to help facilitate accurate electrode insertion and decreased the risk of post-op complications due to persistence of CSF leak. In cases where CSF leak is expected, we make the fol-

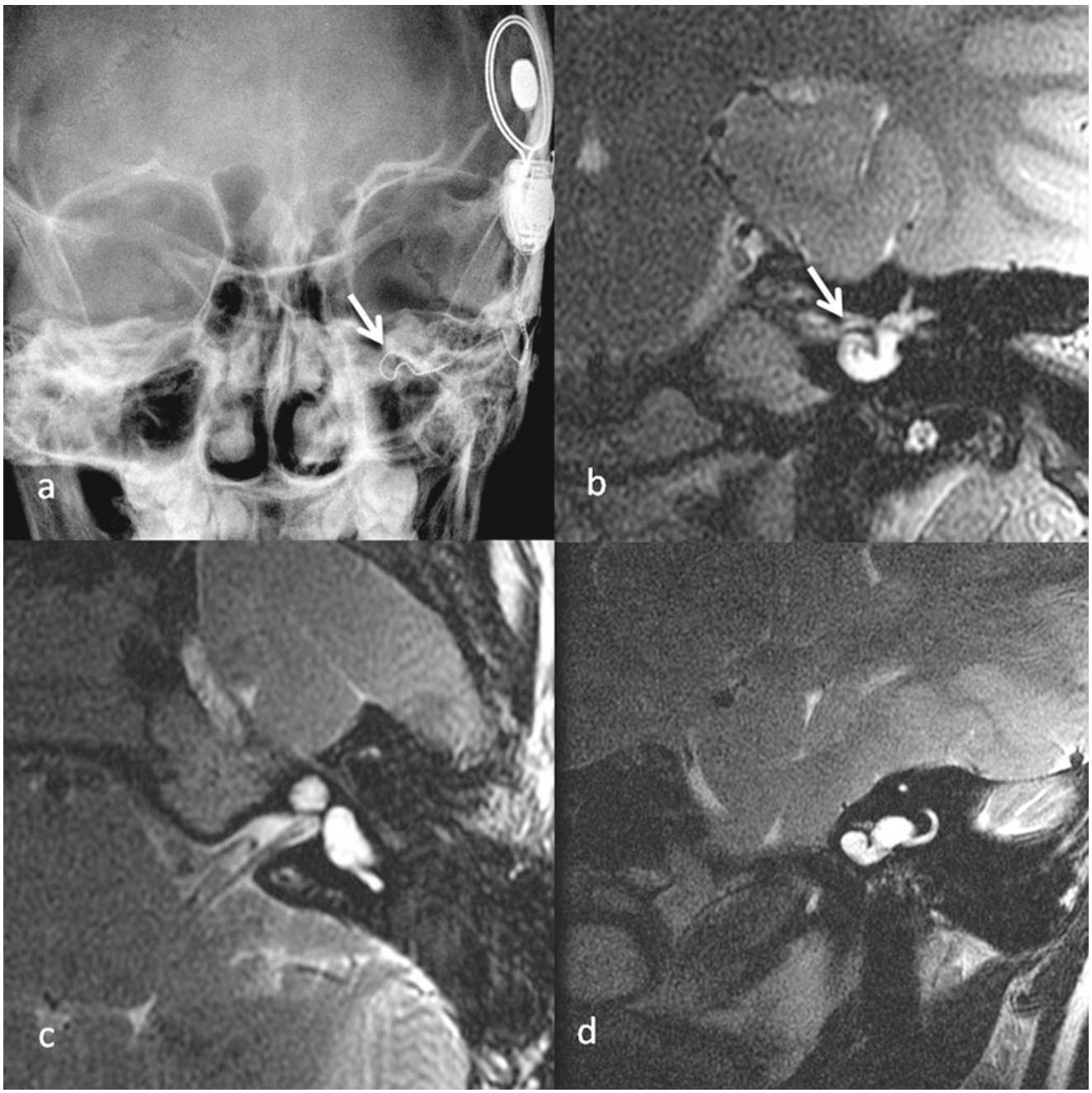

Fig. 4.20 Incomplete partition type 1 (IP-1) malformation. (**a**) Frontal plain postoperative radiograph with atypical appearance of CI electrode array (*arrow*). This finding is due to abnormal cochlear anatomy demonstrated by MRI T2 (**b**) coronal, (**c**) axial and (**d**) sagittal sections. (**b**) *Arrow* is pointing toward dilated cochlea (*arrow*). Axial (**c**) and sagittal (**d**) image demonstrate the amorphous cochlear and vestibular poles separated by a dilated ductus reuniens

lowing preparations in the surgical suite: (1) Second suction available in operative field; (2) Harvest of a large piece of temporalis fascia; (3) Request tissue glue be available. Visualization may be challenging during the case due to the brisk CSF flow. To facilitate visualization, we have an assistant hold the second suction, typically under the incus, and have found that this significantly improves our visualization of the cochleostomy. This maneuver creates a dryer field that permits more accurate electrode array insertion and more effective placement of the soft tissue packing to seal the leak. Many surgeons, including ourselves, create a larger than typical cochleostomy in the setting of a CSF leak (Phelps et al. 1993). A larger cochleostomy facilitates placement of soft tissue packing just inside of the cochlea

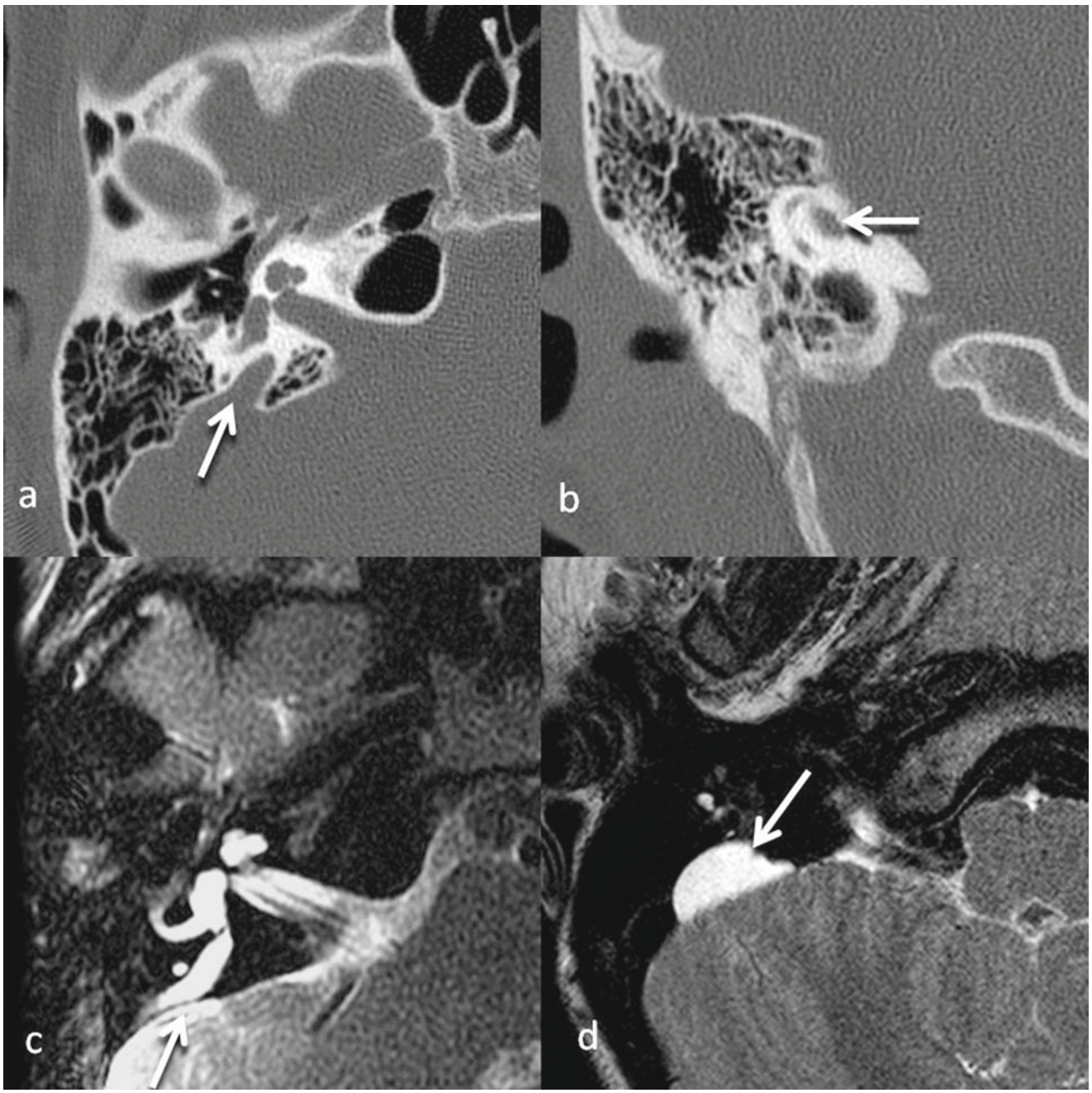

Fig. 4.21 Incomplete partition type 2 (IP-2) (Mondini) malformation. CT right ear (**a**) axial and (**b**) coronal images reveal enlarged bony vestibular aqueduct (*arrow*). Additionally, the modiolus is not well formed. MR T2 (**c** and **d**) axial images demonstrate a dilated endolymphatic sac (*arrow*)

surrounding the array, rather than only laying it over and around the cochleostomy within the middle ear. Placement of packing inside the cochlea takes advantage of the pressure head of CSF which assists in keeping the packing in place. We also prefer a larger, slightly keyhole-shaped cochleostomy to allow easier manipulation of the instruments used to pack the cochleostomy (Graham et al. 2000). Use of tissue glue in addition to soft tissue packing may be helpful. We also often fill the middle ear with soft tissue and tissue glue (Papsin 2005). We prefer to create a large cochleostomy rather than perform a pure round window insertion as we believe the former permits a more effective placement of soft tissue necessary to seal the leak. At the termination of the procedure, no visible continued leak of CSF should be present. Consultation with a neurosurgeon may be of benefit, especially if there is concern about adequacy of surgical containment of the leak solely via an otological procedure.

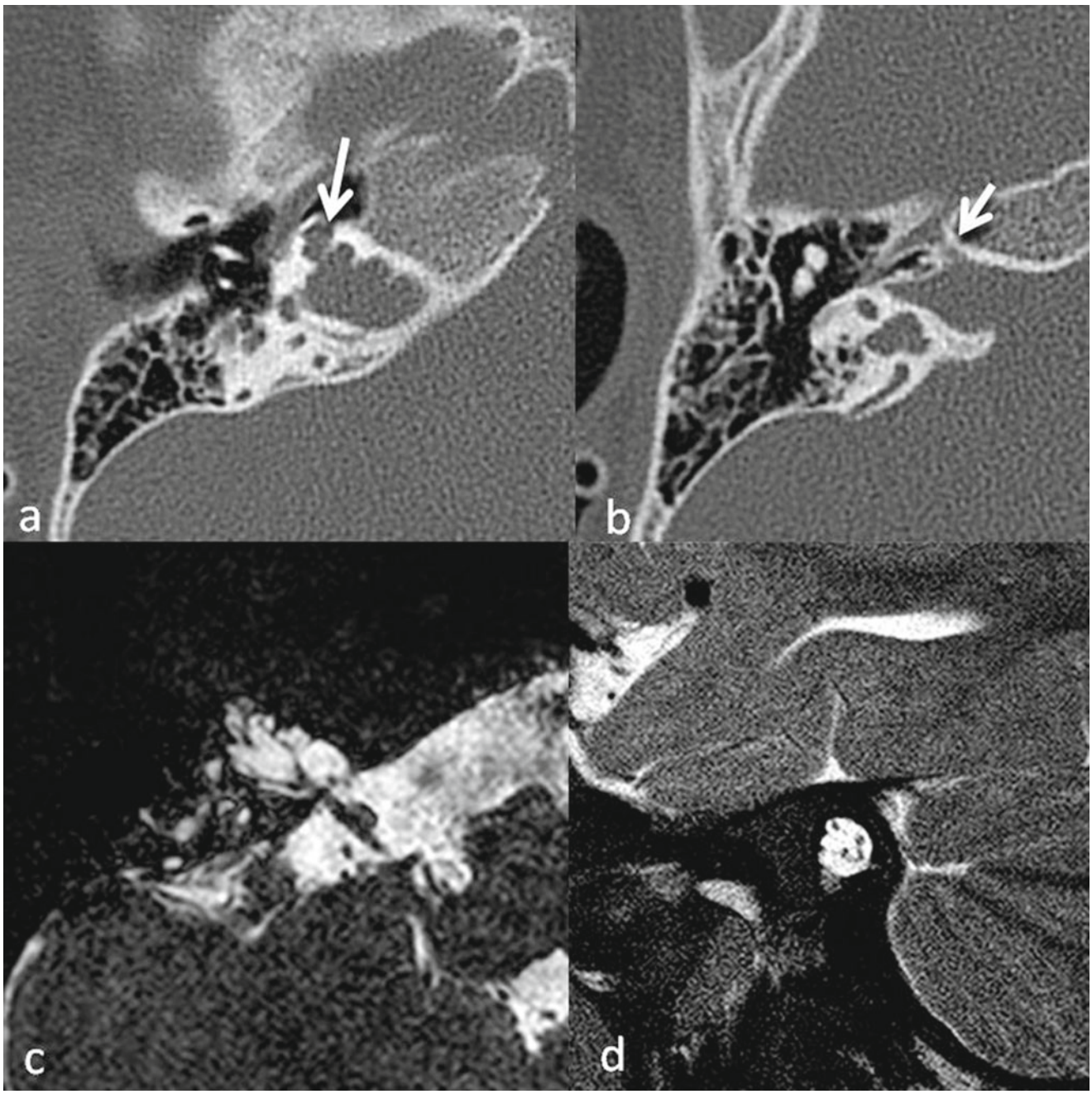

Fig. 4.22 Imaging of right ear of a 2 year old child with findings consistent with X-linked deafness with stapes gusher. CT (**a**) axial section demonstrates typical enlarged and scalloped IAC and hypoplastic cochlea (*arrow*). Contiguous (**b**) axial image demonstrates medial positioning of the facial nerve canal (*arrow*). MRI T2 (**c**) axial and (**d**) sagittal images show position of the facial, cochlear and vestibular nerves within the enlarged IAC (see Fig. 4.25 for additional imaging of this ear)

The literature contains numerous reports of additional techniques and maneuvers for intraoperative management of CSF from within the cochlea. For example, some surgeons will routinely place lumbar drains at the time of surgery, while others place them only in cases of persistent CSF leak. Others recommend use of hyperosmolar therapy at the time of surgery to reduce the flow of CSF (Loundon et al. 2008). A special cork shaped electrode array has been designed to simultaneously serve as a stopper or plug within the cochlea, although this design may limit insertion depth (Sennaroglu et al. 2014).

Common Cavity

In addition to management of a CSF gusher as outlined above, there are additional important considerations when implanting a child with a common cavity. The hearing improvement from a CI may be heavily influenced by whether or not the cavity is anteriorly or posteriorly oriented (Fig. 4.19) (Sennaroglu et al. 2006). Those with a completely posterior orientation are more likely to have no auditory nerve associated with the cavity and therefore outcomes in this group are often poor with respect to speech and language acquisition. Based upon imaging it is often difficult to determine if there is innervation of the common cavity by the auditory nerve. Although there are reports of electrophysiological testing being used to determine candidacy in children with common cavities, we have considerable experience in this area and have not found this type of testing reliable in predicting benefit. Therefore we have abandoned intraoperative electrophysiologic testing for this specific purpose.

Insertion of the electrode array through an opening made into the common cavity on the floor of the of the antrum, in the region where the lateral semicircular canal in normally located, was first described by McElveen et al. (1997). This elegant approach obviates the need for the facial recess approach and permits excellent visualization for insertion of the electrode array. In addition to differences in surgical approach, there are a wide range of electrode arrays that have been used for common cavities including some specially designed for this malformation. However, no study comparing effectiveness has been done in large measure because of the very low incidence of this malformation.

The Aberrant Facial Nerve

An aberrant facial nerve is more likely to be encountered when cochlear and middle ear anomalies are present (Sennaroglu et al. 2006; Song et al. 2012). In rare cases the surgical approach to the cochlea may need to be modified. For example, we have dealt with cases of CHARGE Syndrome in which it was safer to perform a round window insertion to avoid a facial nerve just inferior to the round window (Figs. 4.23 and 4.24). Careful review of the imaging preoperatively, including special attention to the course of the facial nerve, is an important aspect of surgical planning.

Electrode Considerations in the Abnormal Cochlea

The available selection of implant electrode arrays continues to increase. A major consideration in severe cochlear abnormalities such as the common cavity is the location of the neural elements to be stimulated. In an anatomically normal cochlea, the spiral ganglion cells of the auditory nerve will be located centrally within the modiolus. Given that the modiolus is absent in a common cavity, the supposition is that the nerve fibers are located on the lateral wall. With this in mind many surgeons will use a straight electrode array, rather than a pre-curved array designed to "hug" the modiolus, in order to maximize contact with the outer wall of the common cavity. Some surgeons prefer banded electrodes that fully encircle the array to maximize proximity of each electrode to the nerve fibers. However, despite this theoretical advantage, we prefer arrays with half -banded electrodes in order to avoid difficulty should explantation become necessary to address device failure. We have experienced scar tissue causing difficulty with removal of fully banded electrode arrays from the common cavity. Due to current spread, we have found no disadvantage to achieving effective neural stimulation with half banded electrodes. In children with common cavities or hypoplastic cochlea a less that complete insertion of all electrodes is not uncommon. This is not surprising as most electrode arrays were designed for the normal cochlea. Analysis of preoperative imaging often permits this situation to be recognized, thereby preventing kinking of the electrode array due to misguided effort to achieve full electrode insertion.

Another consideration as outlined above in Sect. "Cerebrospinal Fluid (CSF) Leak and Its Management," is that many cochlear anomalies (IP-1, IP-3, cystic cochleovestibular anomalies with ductus reunions) are associated with deficiencies of the modiolus. When defects are present, there is potential for the electrode array to be inadvertently inserted into the internal auditory canal (Todt et al. 2013). One tip-off that the internal auditory canal has been entered may be sudden onset of CSF leak during electrode array insertion. With this potential complication in mind, some surgeons prefer to use a pre-curved electrode array that may be less likely to enter the internal auditory canal as it is unlikely to be in contact with the lateral wall of the cochlea during insertion. Regardless of electrode preference, what is most important is keeping the problem in mind so that it may be recognized and addressed in a timely manner. Intraoperative fluoroscopy has been reported to be a useful tool to monitor electrode array placement during cases with challenging cochlear anatomy (Fig. 4.25) (Fishman et al. 2003). Postoperative findings that should raise concern about insertion into the internal auditory canal include poor auditory percepts and facial nerve stimulation.

Ossified Cochleae

Since the institution of pneumococcal vaccination during early childhood the incidence of meningitis induced deafness has significantly declined. For this reason, implant surgeons now encounter the ossified cochlea much less often. Ossification secondary to bacterial meningitis occurs at

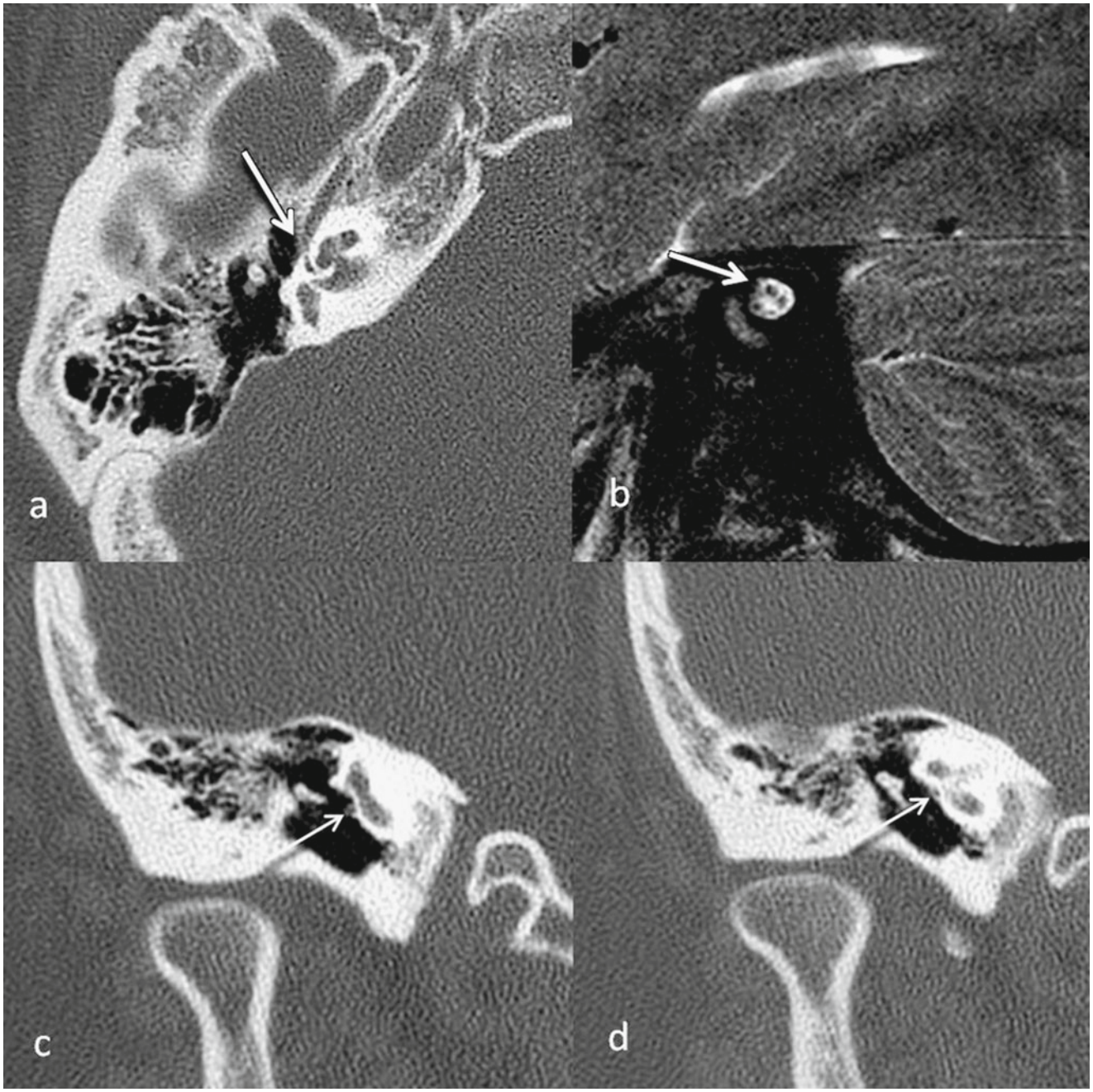

Fig. 4.23 Imaging findings, right ear, of teenager with CHARGE syndrome. CT (**a**) axial section demonstrates a narrow facial nerve canal. Note hypoplastic cochlea and absent semicircular canals. (**b**) MR T2 sagittal image reveals four nerve bundles within the lateral aspect of the IAC (*arrow*). (**c** and **d**) CT coronal sections demonstrate abnormal position of the horizontal segment of the facial nerve canal (*arrow*). The oval window is absent and the facial nerve canal courses over and below area where oval window would be expected in normal development

variable rates and to varying degrees. Some cochlea may be completely ossified (Fig. 4.13) within several weeks to months whereas other cochlea never become fully obstructed. In the presence of complete ossification alternative surgical approaches and specialized electrode arrays may be used. In cases of partial ossification it may be possible to removed ossified bone in the proximal basal turn of the cochlea and achieve full electrode array insertion. There are many approaches and philosophies regarding the ossified cochlea which are not addressed in this chapter (see Chap. 5 for further discussion). At our institution, we address the fully ossified cochlea by using a double array after creating two tunnels into the ossified cochlea. In light of the technical challenges of implanting an ossified cochlea

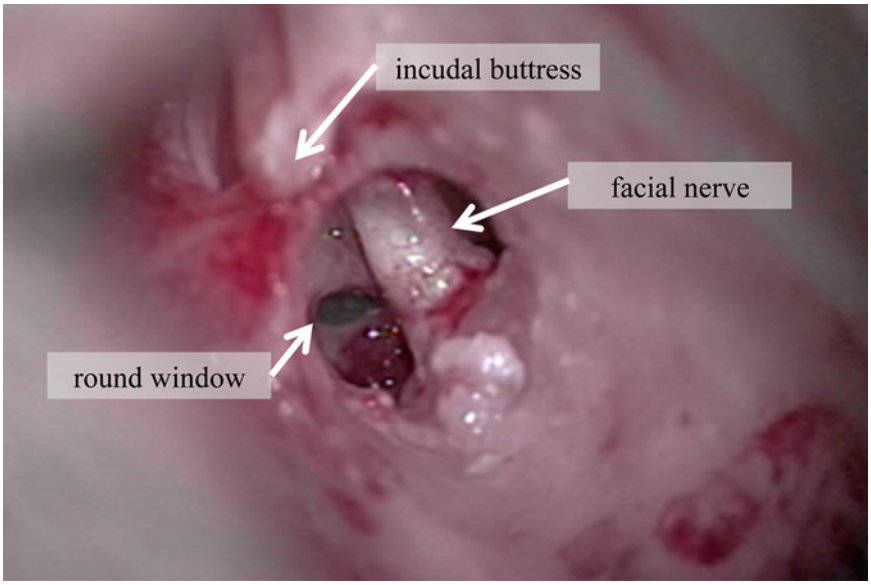

Fig. 4.24 Intraoperative view, right ear, through the facial recess in a teenager with CHARGE syndrome. Note the facial nerve running anterior and immediately adjacent to round window

and the poorer outcomes associated with this problem, it is ideal for these children to be implanted expeditiously, prior to onset of ossification. MRI has replaced CT as the imaging modality to identify and monitor cochlear obstruction after bacterial meningitis. The most sensitive imaging modality to identify early cochlear obstruction is a high resolution pre and post-gadolinium MRI of the temporal bone. A very early finding is the presence of T1-weighted post-gadolinium labyrinthine hyperintensities. It occurs prior the loss of the T2 signal within the labyrinth (Dubrulle et al. 2010). In our institution, all children diagnosed with bacterial meningitis are automatically evaluated by the audiology department in order to rapidly identify which children have developed hearing loss. Our audiology and imaging procedures have enabled us to implant children deafened by meningitis within 6 weeks of diagnosis.

The Sclerotic Mastoid

Many CI candidates evaluated later in childhood who have a history of frequent otitis media, and therefore Eustachian tube dysfunction, will have sclerotic mastoids. Sclerotic mastoids often have a low-lying tegmen and an anterior sigmoid sinus. These anatomical findings may significantly limit access to the mastoid antrum necessary to open the facial recess and find the round window niche. In some cases the anterior sigmoid may need to be uncovered and compressed to achieve adequate visualization. In addition, some sclerotic mastoids may have chronically thickened mucosal tissue that obscures anatomic finding (see Sect. "Otitis Media" regarding management). In these cases we sometimes find it useful to remove the incus or the incudal buttress to improve exposure. Additional anatomic challenges to surgical access may occur as a result of large venous channels, which we have encountered most often in children with CHARGE syndrome (Friedmann et al. 2012). Although other approaches including use of endoscopic techniques have been described (see Chap. 5 for further discussion), in our practice we have not found alternative approaches to be necessary.

Summary

Although the majority of children that we implant are medically and anatomically straightforward, some implant candidates do pose significant challenges in these domains. The experience that we have gained since the inception of our cochlear implant program enables us to provide safe and effective CI for these more challenging candidates. The key to safe and effective implantation for these children relies upon recognizing potential challenges and formulating a proactive plan to address each concern.

Acknowledgments Special thanks to Dr. James Robertson, The Hospital for Sick Children, for his input in outlining the anesthetic considerations in special populations of children undergoing cochlear implantation.

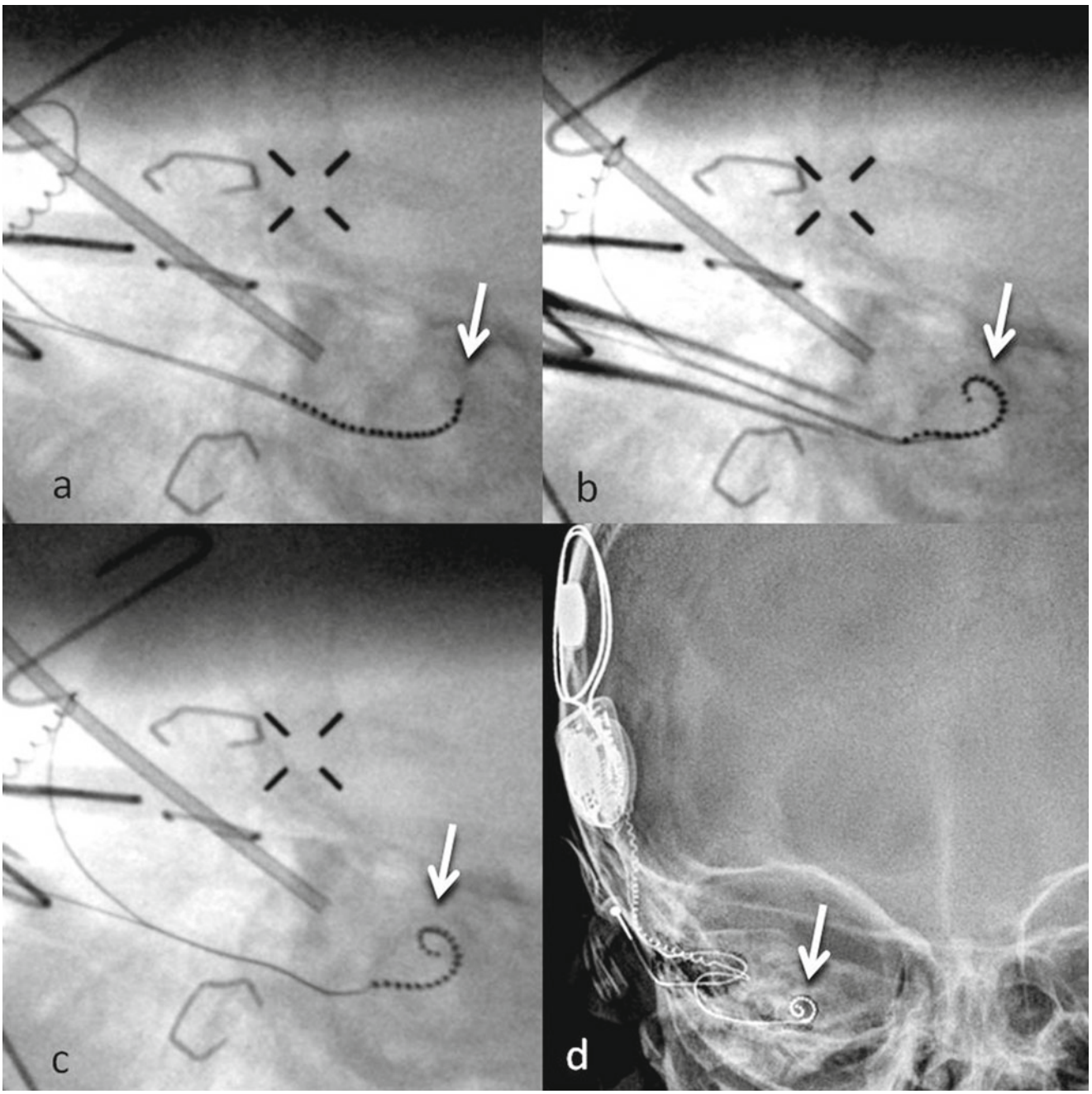

Fig. 4.25 Fluoroscopy images (**a**–**c**) obtained during image guided insertion of electrode array demonstrating the desired coiling of the electrode array (*arrow*) within the cochlea of child with X-linked deafness with stapes gusher. (**d**) Frontal radiograph confirms electrode array (*arrow*) in good position within cochlea

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Surgical Considerations

5

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Preoperative Considerations

Several important decisions need to be made prior to scheduling the patient for surgery. Many of these decisions will benefit from input from all of the members of the implant team.

The implant team must decide whether or not to implant one or both ears. Bilateral cochlear implantation may be done simultaneously or sequentially. In children deemed to be appropriate candidates, several advantages can be realized from bilateral implantation. Speech reception may be significantly improved in noise and, in some recipients, in quiet. In addition, the majority of bilateral cochlear implant (CI) recipients experience improved spatial hearing, an obvious personal safety advantage (Asp et al. 2012; Grieco-Calub and Litovsky 2010). Moreover, sound localization also aids speech understanding in noise (Galvin et al. 2007; Litovsky et al. 2006; Loizou et al. 2009; Buss et al. 2008). Most bilateral CI recipients assert that the second implant significantly reduces their "work of hearing." They expend less energy and concentrate less intensely in order to understand spoken language. Many bilateral recipients report this change to be the most significant advantage of having a second implant. Unfortunately, validated measures of "work of hearing," in children are not available (Rader et al. 2013). In addition, in terms of access to hearing, there is a significant practical advantage to bilateral instead of unilateral CI for those lacking useful aided speech perception in their unimplanted ear. It is also unlikely that problems which interfere with device use such as skin irritation at the transmitter site, internal or external device malfunction, will affect both ears simultaneously. Therefore, bilateral CI recipients are much less likely to find themselves in a situation in which they are no longer able to hear from either ear.

Despite many advantages of bilateral cochlear implantation, many parents and patients are concerned about saving one ear for future technology and medical advances such as hair cell regeneration. This concern should be weighed against the potential for reduced benefit from the second implant secondary to decreased capacity of the central nervous system to respond after longstanding sensory deprivation (Loizou et al. 2009). Hence, delay may reduce benefit. There is also the issue of degree of benefit from amplification. Current candidacy criteria permit implantation of children and adults with speech perception ability, albeit less that would be expected from a CI. For infants and young children it is not possible to determine speech perception ability, which may influence the team to recommend implantation of one ear, typically the poorer hearing ear, thus enabling the child to be a bimodal (CI plus hearing aid on opposite ear) user until more in-depth testing is possible. Some parents of children with useful unaided low frequency hearing do not wish to risk loss of unaided sound detection because of the potential safety advantage this may provide when their child is asleep or in other situations without their CI. Disadvantages of bilateral implantation also may include added expense, increased time to maintain external hardware for both ears and increased number of visits to the implant center. The additional commitment and complexity may be too much for some families, especially initially. Therefore, some children may be better served by sequential implantation. However, bilateral simultaneous implantation may be strongly preferred by families who desire bilateral CIs to avoid two separate anesthesia and surgical experiences. Anesthesia related risks of bilateral CI are not expected to increase significantly in carefully selected patients (Buss et al. 2008).

If the decision has been made to implant just one ear, then the team will need to decide which side to implant. If imaging studies or physical examination indicates that one side is more anatomically favorable than the other, then, other

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things being equal, the ear with better anatomy should be implanted. If both ears are anatomically normal but have different levels of retained residual hearing, the implant team must decide whether to implant the better or worse-hearing hearing ear. If the ear with better hearing has enough residual hearing to enable significant aided word recognition, implantation of the poorer ear is advantageous. This approach provides the opportunity to determine bimodal benefit which may include improved speech recognition in noise as well as the potential benefit of preserving acoustic hearing in the unimplanted ear. If both ears lack speech perception ability but one ear has better aided detection there is no consensus on whether the better or worse-hearing ear should be implanted. While arguments can be made either way, available data has not demonstrated any differences in outcome (Chen et al. 2001). Another consideration is the length of time that an ear has been profoundly deaf, as prognosis is better in ears that have not experienced long term auditory deprivation. A more recently deafened ear is more favorable than one with longstanding deafness and no aided benefit. If, however, the surgeon or patient prefers to proceed with implantation of a profoundly deaf ear with limited response to amplification, it is very important to determine the status of the eighth nerve with high resolution magnetic resonance imaging (MRI). Identification of auditory nerve deficiency, a highly unfavorable finding, is important in choosing the ear to implant.

Conservation of residual hearing in the implanted is now possible, and attempts to retain residual hearing are now widespread. Hearing conservation is believed to be one marker for reduced trauma to inner ear structures. It is theorized that minimizing trauma will not only conserve residual hearing but may retain the capacity of an ear to respond to future advances in treatment (Jayawardena et al. 2012).

There is evidence that even small amounts of residual acoustic hearing may be intrinsically beneficial and may result in improved receptive language (Gifford et al. 2013). If sufficient residual hearing is retained, the CI recipient may be fitted with a processor that permits both electrical hearing through the CI and acoustic amplification of lower frequency hearing, if needed, in the same ear. A recipient's ability to understand speech, especially in noise, may be significantly better using combined acoustic and electric stimulation in the same ear in comparison to either acoustic or electrical stimulation alone (Rader et al. 2013; Gifford et al. 2013). The synergistic effect of electroacoustic hearing on speech perception may be quite dramatic, especially in noise. In addition, recipients with electroacoustic hearing often have much better pitch perception, especially in the lower frequencies. Improved pitch perception permits clearer appreciation of the fundamental frequency of a speaker's voice and, consequently, significantly improved speech perception in noise. Research in adults has also demonstrated that improved pitch perception also allows much better melody appreciation. CI recipients who retain some low frequency residual hearing may have melody perception that is nearly normal. Improved melody recognition, in turn, enhances music appreciation (Adunka et al. 2013; Dillon et al. 2014).

Conservation of residual hearing has been quite successful in the short term but less successful in the long term (Carlson et al. 2011). It may take longer for CI recipients to achieve maximum benefit from an electroacoustic device; presumably because it takes longer for the central nervous system to learn to fuse the two disparate signals. The vast majority hearing preservation studies of CI recipients have been limited to adults with stable hearing. This population was used in studies that were the basis for FDA approval in 2013 for commercial use of the Cochlear Nucleus Hybrid [Cochlear Americas] in adults. The extent to which outcomes in studies of adults apply to children is unclear. One fundamental difference in the pediatric population is that many children who are CI candidates have progressive hearing loss. Therefore, any preservation of hearing after CI may be short lived due to the natural history of their loss. For this reason use of a device such as the Cochlear Nucleus Hybrid [Cochlear Americas] that contains a short electrode may result in the need to surgically re-implant children with a standard length electrode array. Hearing preservation using longer length electrodes that function equally well in ears with no residual acoustic hearing may ultimately prove to be preferable for the pediatric population, although more research is necessary.

Imaging

Medical imaging is an essential element in the preoperative pediatric cochlear implant evaluation process. Computed tomography (CT) of the temporal bone allows for visualization of the external auditory canal, middle ear, mastoid, fallopian canal, otic capsule (inner ear or labyrinth), and internal auditory canal. The latest CT scanners can acquire a high quality images in minutes and obviates the need for anesthesia in many pediatric patients. CT has a number of other advantages including lower cost, detailed visualization of the fallopian (facial nerve) canal, and bony cochlear nerve canal (internal auditory canal). Disadvantages of CT temporal bone studies include radiation exposure, lack of direct visualization of the eighth nerve, and limited accuracy to determine patency of the inner ear.

High resolution three-dimensional MRI provides detailed views of the otic capsule (inner ear or labyrinth), eighth cranial nerve (auditory nerve), brain, and also permits visualization of an enlarged endolymphatic duct and sac. Advantages of MRI also include the lack of radiation exposure and early identification of cochlear obstruction from inflammation

or fibrosis (Booth et al. 2013; Isaacson et al. 2009). Disadvantages of MRI include increased cost, need for general anesthesia in many children, longer image acquisition time, and less information about the course of the facial nerve canal. The authors have used MRI as the imaging modality of choice in pediatric patients since the 1990s for all the above mentioned advantages. To minimize episodes of anesthesia, at our center MRI has been safely coordinated with an auditory brainstem evoked response test or other necessary medical procedure (Parry et al. 2005).

A number of CI centers obtain both CT and a MRI for the evaluation of pediatric sensorineural hearing loss. The authors' center uses a more judicious approach. CT is not routinely obtained unless MRI has identified inner ear or other temporal bone malformations. For children known to be at high risk for cochlear malformations, such as those with CHARGE syndrome, MR and CT are scheduled simultaneously, rather than sequentially. We have found that our approach to imaging provides optimal surgical information about relevant surgical anatomy while minimizing the number of children who require CT scans.

Electrode Array Options

A large variety of electrodes are now available from the cochlear implant manufacturers (Fig. 5.1). Each manufacturer has at least two different electrode arrays and some offer many more choices. Implant centers that work with multiple manufacturers therefore have many electrode arrays to choose among. Available data indicating better outcomes with a particular electrode is currently limited. However, depending upon the ear anatomy and the surgeon's goal for the procedure regarding preservation of residual hearing, certain electrode designs may be advantageous.

A variety of factors should be considered by the surgeon when selecting the optimal electrode array. The anatomy of the cochlea may make one type of electrode more appropriate than another. In ears with cochlear hypoplasia and incomplete partition, a shorter electrode may be more desirable. In regard to the common cavity malformation preferences vary widely. Some surgeons prefer a shorter electrode, others a straight fully banded or a curved array of standard length, and yet others an electrode custom designed for each common cavity.

Normal cochleae vary in length with the majority between 25 and 32 mm. There is varying surgeon preference as to how deeply within the cochlea an electrode should ideally be inserted. Some surgeons prefer a longer electrode to achieve a "deep cochlear insertion" that allows stimulation well into the second turn of the cochlea (Boyd 2011) while others prefer a shorter 360° insertion which covers only the full basal turn. In terms of electrode array length, in most cases a 360° insertion can be achieved with a length of 24 mm. Some surgeons prefer to adjust the length of the electrode to fit the individual cochlea. The availability of electrode arrays between 15 and 31 mm in length makes this possible. Preoperative imaging of the inner ear is used to determine the preferred length. In the authors' experience, this judgment should take into account the planned surgical technique as a longer electrode length by 1–2 mm is beneficial when insertion is done through the round window.

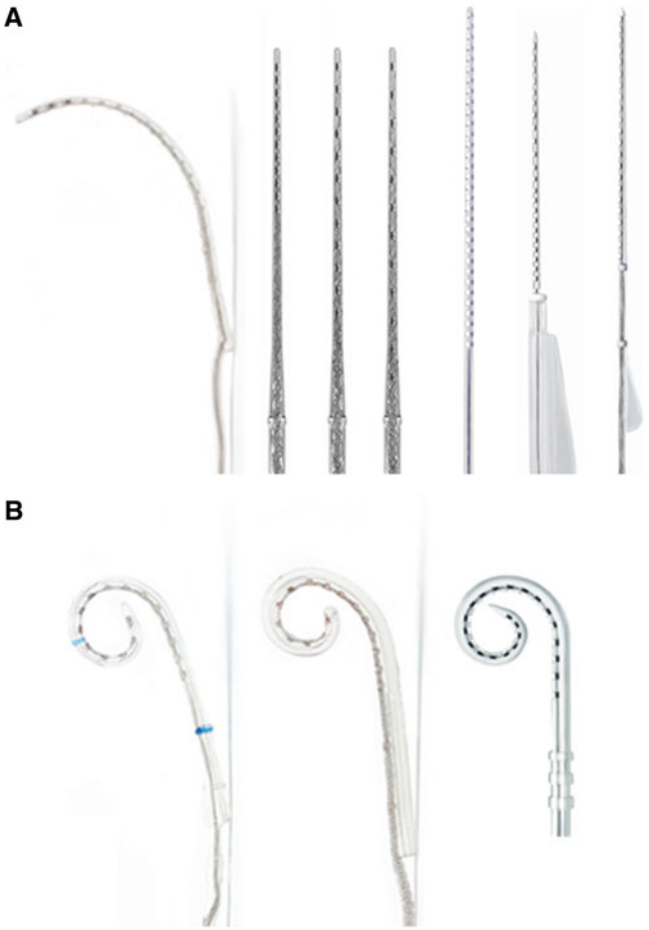

Fig. 5.1 (**a**) Current lateral wall electrode array options include short, long and thin options. (**b**) Perimodiolar curled electrode array options come packaged with a straightening stylet which is removed during insertion

Other approaches should be considered in the event cochlear obstruction with bone or fibrous tissue is encountered that make it impossible to achieve adequate electrode array insertion. One option is to use a "double" electrode array, consisting of two short arrays exiting one device. A superior and inferior tunnel within the ossified basal turn is created for each array. The superior tunnel is created 2 mm anterior to the anterior aspect of the oval window just inferior to the cochleariform process. The inferior tunnel is created using a peri-round window approach. When extensive ossification of the cochlea is present the double electrode array

approach may provide stimulation in the more superior aspect of the basal turn or the middle turn that cannot be reached by a partial drill out of the inferior basal turn (Isaacson et al. 2008). An alternative approach is a classic "lateral wall drill-out" with use of a single short or compressed electrode array (Cohen and Waltzman 1993; Balkany et al. 1996). A drill out procedure may necessitate complete removal of the lateral wall of the cochlea; however, preservation of a portion of the inferior basal turn promontory permits easier fixation of the electrode array. It is easier to identify the scala tympani and provide optimal electrode array placement before fibrous obstruction has progressed to dense bony ossification.

Electrode array design affects its position within the scala tympani. Some electrodes arrays are pre-curved so that they lie close or "hug" the modiolus (central bony wall of the inner ear adjacent to the spiral ganglion cells of the auditory nerve) while straight arrays will position themselves along the lateral wall of the cochlea. Yet others have been designed to take a midway position within the cochlea. These designs are based upon theoretical advantages. However, data is inconclusive as to which type of electrode produces a better outcome. There is some evidence that an electrode that lies in close proximity to the modiolus is less likely to produce facial nerve stimulation than a lateral wall electrode (Battmer et al. 2006).

Electrode insertion may be challenging in some circumstances. Every surgeon develops preferred strategies for different situations. Therefore, a surgeon's own experience is an important factor in determining the optimal electrode array for the recipient. Another factor the surgeon may consider is the likelihood that a repeated insertion may to be necessary. If so, the option of withdrawing and reinserting the same electrode is advantageous.

Perioperative Considerations

Antibiotic Prophylaxis

Although many CI surgeons prescribe antibiotics during the perioperative period, there is no specific research or consensus as to its use, including type or length of course. Most general guidelines on surgical antibiotic prophylaxis advise limiting antibiotic treatment to the first 24–48 h after surgery. Antibiotics, if used, should be initiated 30 min prior to the skin incision (Barker and Pringle 2008; Weber and Callender 1992).

Management of an Existing Tympanostomy Tube

Many children will have an existing tympanostomy tube in the tympanic membrane of the ear to be implanted. While some surgeons choose to remove the tube prior to implantation and wait for the tympanic membrane to heal, the literature supports leaving a clean, dry tube in place and proceeding with the implant procedure. If infection is present, the procedure should be delayed until this has resolved (Kennedy and Shelton 2005; Melton and Backous 2011).

Management of Perioperative Otitis Media

There is wide agreement that acute otitis media in CI recipients can be effectively managed with oral antibiotics alone. As otitis media commonly involves the mastoid to varying degrees, this treatment approach remains appropriate when imaging demonstrates mastoid involvement as long as there is no clinical evidence of a subperiosteal abscess. The usual oral antibiotics recommended for management of acute otitis media suffice in almost all cases. However, otitis media developing shortly after implant placement is an exception. Most of the literature recommends intravenous antibiotics in this circumstance, ideally using an agent which is highly concentrated in the middle ear, such as ceftriaxone. The presence of a subperiosteal abscess, on the other hand, is best managed with incision and drainage in addition to intravenous antibiotics (Rubin et al. 2010; Kempf et al. 2000; Luntz et al. 1996).

Positioning and Preparation

Patient positioning is an important aspect of cochlear implant surgery. The operating room table is typically reversed so that the patient's head is positioned at the foot of the bed. The table is then rotated 90–180° away from the anesthesiologist after the induction of anesthesia. Prior to turning the table, care must be taken to ensure that the intravenous catheter and endotracheal tube are adequately secured to the patient. Careful padding of the undersurface of the head, contralateral ear, and the extremities is necessary to prevent pressure ulcers.

Electric clippers may be used to remove hair close to the incision. Local anesthesia is used to infiltrate the planned incision site. Marcaine with epinephrine provides a longer duration of anesthesia which may assist in postoperative pain management. In young children it is important to ascertain the maximum permissible dose of local anesthesia to avoid anesthetic complications.

Intraoperative facial nerve monitoring is routinely used during CI surgery. Younger pediatric patients may be more at risk for a facial nerve injury given the more superficial location of the nerve along the mastoid segment and the higher incidence of facial nerve anomalies in children with congenital hearing loss. Paired needle electrodes are placed in the orbicularis oculi and orbicularis oris. The ground and stimulating electrodes are typi-

cally placed in the skin of the shoulder or chest. These electrodes are secured to prevent dislodgement. One common reason for facial nerve injury despite facial nerve monitoring is improper use of this monitoring equipment. Therefore, a number of precautions are helpful to ensure monitoring is ongoing. Careful placement and timing of infiltration of local anesthesia are also important to ensure monitoring is occurring. The latter is of particular importance in infants and young children whose facial nerve may exit the stylomastoid foramen from a more superficial location and therefore more likely be exposed to local infiltration. The impedances of the electrodes and the setting of the stimulation parameters should be checked prior to prepping and draping. In addition to tapping upon the face to elicit a response from the monitor prior to the incision, the authors recommend use of a bipolar or monopolar facial nerve stimulating probe during the procedure. Use of a probe permits stimulation of the facial nerve in its tympanic or mastoid segment when working within areas of the mastoid where this structure is most at risk. Good communication with the anesthesiologist is necessary to ensure that muscle relaxants are not given as these agents interfere with facial nerve monitoring.

Operative Considerations

Incision Placement

Incision length has become significantly smaller since CI surgery began. Surgeons often use incisions as small as 2–4 cm. Incision design includes a standard postauricular, a lazy S type incision, or a posterior postauricular incision (Fig. 5.2). These smaller incisions typically provide adequate exposure of surgical landmarks and result in less pain and faster healing. The incision should provide exposure of the mastoid cortex to the level of the superior bony external auditory canal. If a recessed bone bed for the receive stimulator is planned, the incision must permit exposure of this area as well.

Receiver-Stimulator Placement

Placement of the receiver-stimulator is an important but easily over-looked aspect of CI surgery. The surgeon should position the receiver-stimulator far enough behind the auricle so that an externally worn behind-the-ear processor will not rest against it. Avoiding contact between the processor and the receiver-stimulator minimizes the risk of irritation that may result in breakdown of the intervening skin. On the other hand, if the receiver-stimulator is placed too far from the auricle, the lead may not be long enough to allow for full insertion of the electrode array. A surgical template specific to each manufacturer's behind-the-ear processor is available to assist the surgeon with positioning. The receiver-stimulator is usually positioned just superior to the tight attachment of the periosteum to the occipital-parietal suture. A more anterior superior placement may be preferred for children with motoric problems such as severe cerebral palsy that limit head control. These children often require use of a head support when seated that may overly the receiver-stimulator and thus interfere with use of the transmitter coil. This problem will interfere with CI use and can be avoided with surgical planning.

A number of methods have been described for positioning and securing the receiver-stimulator. A tight subperiosteal pocket technique involves creation of a space just large enough to place the receiver-stimulator and not permit its free movement once fully seated (Balkany et al. 2009) (Fig. 5.3). A bone bed can be created for the implant to provide additional stability when using the subperiosteal pocket technique. If the subperiosteal pocket does not securely hold the receiver-stimulator in place, additional measures to secure the device are necessary. These may include creation of a bone bed and use of sutures to bone to prevent future displacement of the implant (Fig. 5.4). Other fixation methods have also been described including use of screws, mesh, and resorbable plates.

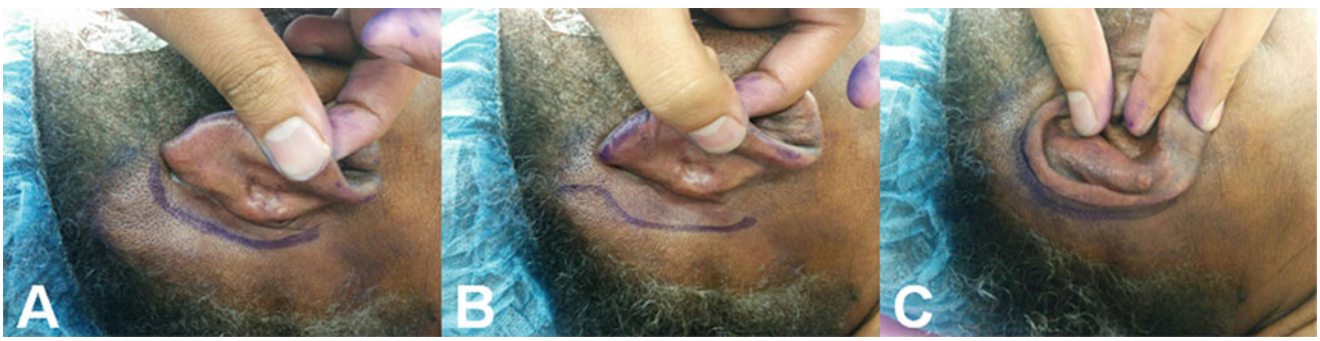

Fig. 5.2 (**a**) Standard postauricular incision. (**b**) Postauricular "lazy S" incision which allows for improved access for receiver stimulator placement. (**c**) Posteriorly placed postauricular incision which allows improved access for receiver stimulator placement

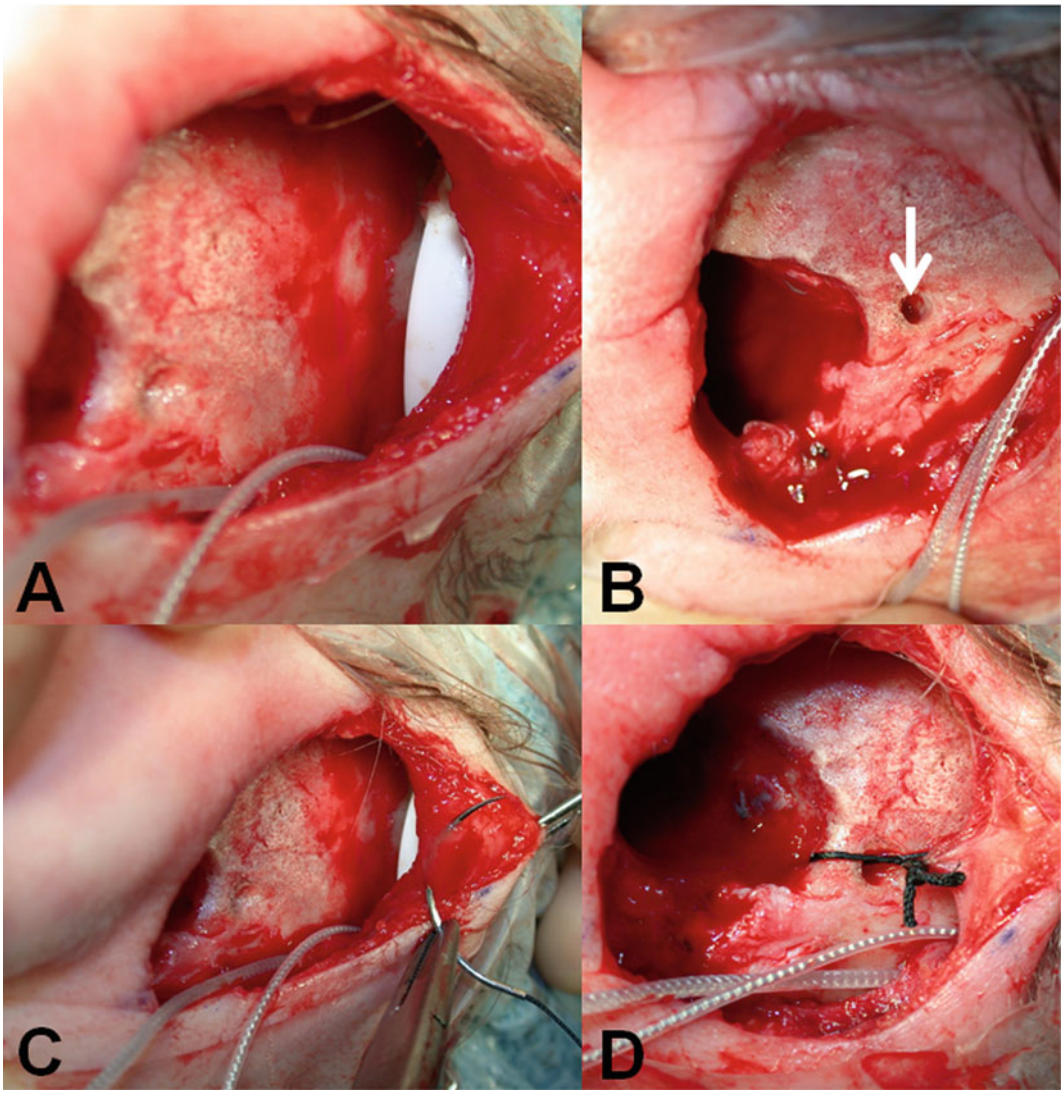

Fig. 5.3 (**a**) Receiver stimulator placed in tight subperiosteal pocket. (**b**) Fenestration created between cortical bone and mastoid cavity. (**c**) Suture placed in the posterior periosteum. (**d**) Suture passed through hole securing the implant in the tight subperiosteal pocket which also prevents anterior displacement

Approaches to the Cochlea

The traditional surgical approach described to access the cochlea is a canal wall up mastoidectomy with posterior tympanotomy via the facial recess to obtain access to the round window niche. The mastoid-facial recess approach, in the vast majority of cases, provides excellent exposure of the round window niche. The mastoidectomy requires identification of the mastoid tegmen and skeletonization of the posterior aspect of the external auditory canal until the aditus ad antrum is identified. The opening of the aditus ad antrum is extended anteriorly until the lateral semicircular canal and short process of the incus are identified. These two structures are excellent landmarks for identification of the vertical facial nerve and facial recess (Fig. 5.5).

The facial recess may be performed using two different methods: (1) Facial nerve identification approach or (2) Posterior canal thinning approach. The former approach entails opening the recess by removing the bone between the vertical segment of the facial nerve and chorda tympani nerve while leaving a narrow buttress of bone between the recess and the short process of the incus. The posterior canal thinning approach entails thinning the posterior canal wall up to the level of the incus buttress and lateral semicircular canal. Facial recess air cells are often encountered and are opened while working parallel to the facial nerve. For both approaches bone removal is done using small diamond burrs to reduce risk of damage to neural structures. The facial recess must be opened sufficiently to achieve visualization of the round window niche and to obtain the desired insertion angle for the electrode. Regardless of approach, the opening

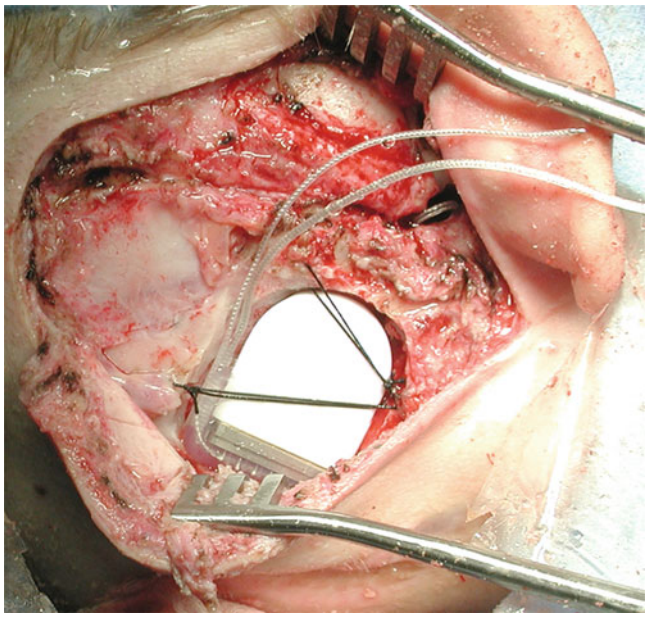

Fig. 5.4 Traditional well with tie-down sutures with large subperiosteal pocket

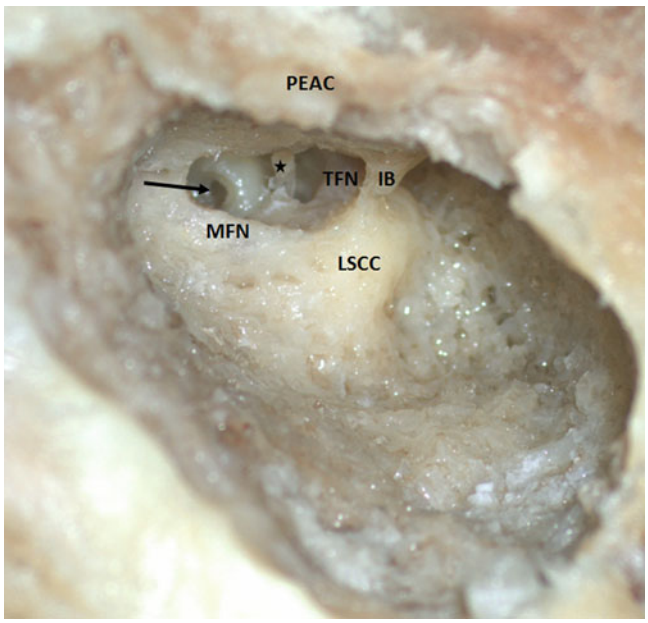

Fig. 5.5 Wide angle view of facial recess with visible incudostapedial joint (*black star*), tympanic facial nerve (TFN), mastoid facial nerve (MFN), incus buttress (IB), lateral semicircular canal (LSCC), posterior wall of the external auditory canal (PEAC), and round window (*black arrow*)

of the facial recess should permit visualization of the posterior ligament of the incus, stapedius tendon, oval and round window niches, and the origin of the chorda tympani from the facial nerve

Overhanging bone of the posterior external auditory canal, along with an anterior and laterally displaced mastoid segment of facial nerve can significantly restrict visualization of the round window niche. Removing the lateral aspect of the posterior canal wall in addition to skeletonization of the anterior aspect of the mastoid segment of facial nerve may improve exposure in cases where the round window niche is not readily visible. The edges of the mastoid cavity are typically left slightly overhanging (no saucerization) in order to enhance securing of the excess electrode lead that is coiled within the cavity (McRackan et al. 2012).

A transcanal approach to CI surgery involves placement of the electrode array through the ear canal after elevation of the posterior canal skin and tympanic membrane. It is not widely used in the USA. Unfortunately this approach is associated with a significant risk of breakdown of posterior ear canal skin overlying the lead wire, especially in children. Consequently in the unusual case where this approach is necessary, it is best combined with closure of the ear canal. Closure involves removal of the drum and medial canal skin with eversion and closure of the lateral canal skin. The authors have found the transcanal approach advantageous when there is significant distortion of mastoid anatomy. For example, we have used this approach in children with CHARGE syndrome who had mastoid venous lakes and abnormal facial nerve anatomy in whom a facial recess approach would have been very challenging. We have also found it helpful to use a combination of the transcanal and the traditional transmastoid facial recess approaches in situations where the round widow niche is not visible through the facial recess. The round window may be visualized transcanal and the electrode array placed through the facial recess for insertion. Because the lead is protected by the posterior canal wall, closure of the ear canal is not required as there is no risk of lead exposure within the canal.

Although rarely indicated, a middle fossa approach for the purpose of cochlear implantation has been reported (Colletti et al. 2000). This intracranial extradural approach uses the greater superficial petrosal nerve and geniculate ganglion as landmarks to identify the ascending and descending basal turns of the cochlea. It may be useful in ears in which the inner ear is completely obstructed by ossification or to avoid placing the device in a chronically infected ear refractory to management (Colletti et al. 2000). The middle fossa approach is rarely if ever necessary in the pediatric population as otitis media usually responds to medical management, placement of a pressure equalization tube or, when necessary, mastoid surgery.

A retrofacial approach is another method that may be employed to identify the round window niche. This approach necessitates skeletonization of the medial and posterior aspect of the vertical segment of the facial nerve as well as the ampullated end of the posterior semicircular canal, after performing a canal wall up mastoidectomy (Fig. 5.6). Indications for this approach include a lateral and anteriorly displaced facial nerve which obstructs the view of the round window niche via a facial recess approach (Liening et al. 1994).

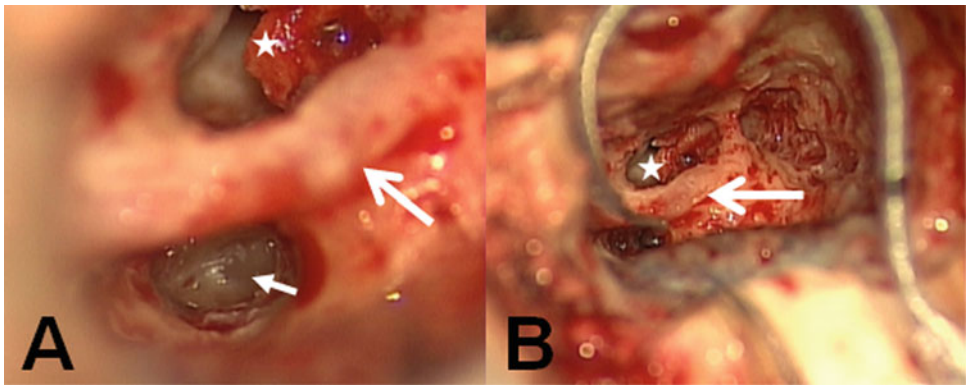

Fig. 5.6 (**a**) Retrofacial approach with a view of the round window membrane (*star*). The retrofacial approach is defined by the facial nerve laterally (*large arrow*), lateral and posterior semicircular canal superiorly, jugular bulb inferior and posteriorly. (**b**) Less magnified view of retrofacial approach with electrode traversing medial to the facial nerve (*large arrow*). The incudostapedial joint (*star*) is visible through the facial recess

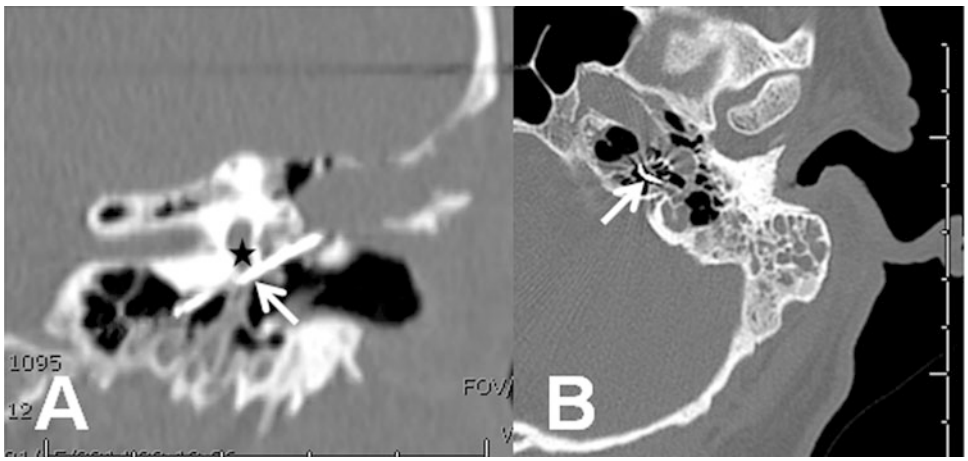

Fig. 5.7 (**a**) Coronal computed tomography demonstrating an extracochlear electrode array in the infracochlear tunnel (*arrow*) into the inferior petrous apex. The round window niche is seen just superior to the electrode array (*star*). (**b**) Axial computed tomography showing the tip of the electrode array in the inferior petrous apex (*arrow*)

Electrode Insertion into the Cochlea

The insertion of the electrode array into the cochlea is one of the most critical steps of CI surgery. Techniques to minimize insertion trauma are especially important when the goal is preservation of delicate inner ear structures or acoustic hearing. Ideally the array is inserted into the scala tympani. Regardless of technique, the round window membrane (RWM) is an important landmark which is rarely absent. Failure to identify the RWM increases the likelihood of inadvertent misplacement of the electrode array into the scala vestibule or entirely outside of the turns of the cochlea. Locations of inadvertent misplacement of the electrode array include the vestibule and semicircular canals, carotid canal, eustachian tube, and hypotympanic air cell tract (Ying et al. 2013) (Figs. 5.7 and 5.8)

The anterior and posterior bony overhang (pillars) of the round window niche obscure visualization of the RWM to varying degrees. The bone of the posterior portion of the round window niche (the posterior pillar) is removed until the majority of the RWM is visualized (Roland et al. 2007) (Fig. 5.9). There is often mucosal membrane overlying the RWM which can usually be removed atraumatically. The RWM appears darker and tenser helping to distinguished it from overlying mucosal (Fig. 5.10) The size and orientation of the round window membrane will determine whether the electrode array can be inserted through the RWM (a "pure" or "membranous" round window insertion) without any enlargement of the round window's bony orifice. When the pure round window approach is used in combination with an appropriately designed electrode, insertion trauma is minimized, in part because this approach avoids use of the drill to gain access to the inner ear. When performing a RWM insertion the electrode array should be inserted from a posterior superior to anterior inferior angle to help minimize trauma to the modiolus. One caveat is that electrode array insertion through the round window may in some instances cause conductive hearing loss in ears with preserved residual hearing (Roland et al. 2007).

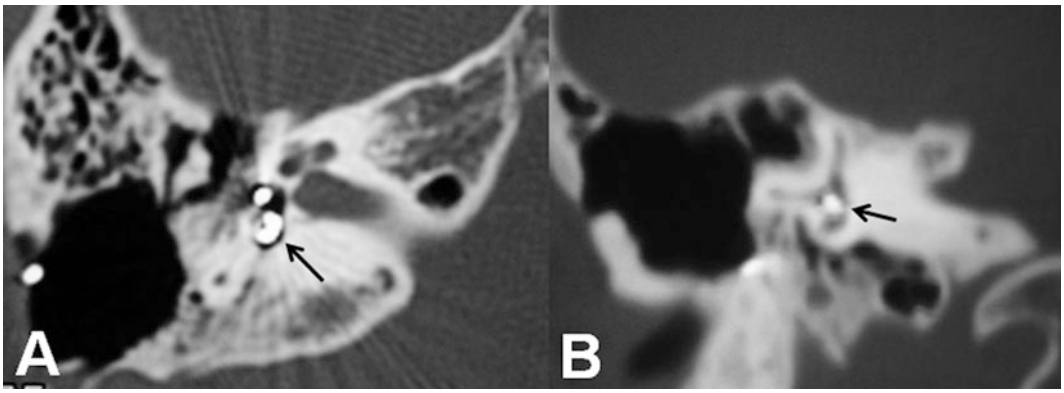

Fig. 5.8 (**a**) Axial computed tomography demonstrating an electrode array in the vestibule (*arrow*). (**b**) Coronal computed tomography showing the electrode array in the vestibule (*arrow*)

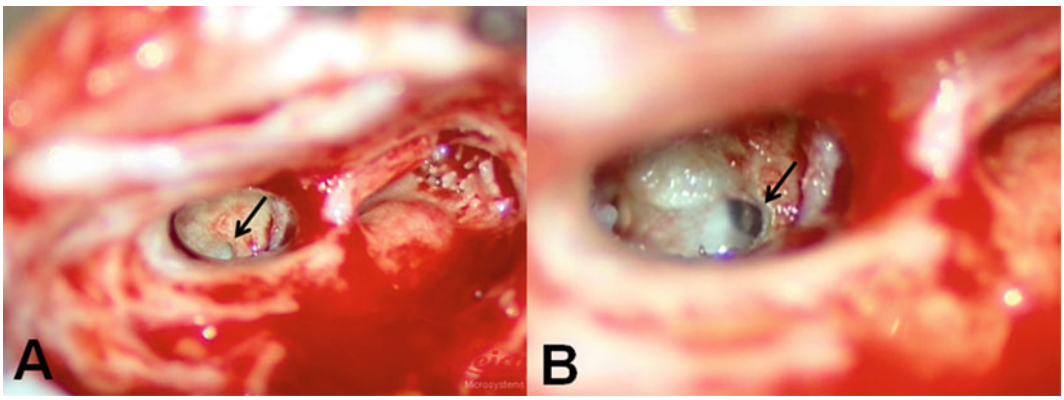

Fig. 5.9 (**a**) Round window niche with significant osseous overhang (*arrow*). (**b**) Round window membrane visualized after removal of osseous overhang (*arrow*)

Originally the cochleostomy was described as creation of a separate and distinct opening in the proximal inferior basal cochlear turn on the promontory just anterior inferior to the round window membrane. A more inferior location relative to the round window has become preferred to minimize the likelihood of damage to inner ear structures. Many surgeons make this opening at the inferior aspect of the round window creating an opening that is contiguous with the round window, rather than creating a distinct opening. Visualization of the round window membrane is necessary to create this opening in the proper location. The promontory cochleostomy is opened inferior to the round window using a 0.6–1.2 mm diamond burr depending on the diameter of the selected electrode array. Drilling anterior and superior to the round window is best avoided as it is likely to result in injury to the spiral ligament, basilar membrane with resultant electrode array placement into the scala vestibuli (Adunka et al. 2007).

As preservation of residual hearing and inner ear structure have increasingly become goals of CI surgery, "soft surgical technique" has become more important. These techniques include removing bone down to the endosteum (membranous lining of the inner ear) in order to prevent bone dust from entering the cochlea as well as greater care not to remove perilymph. Friction related to insertion of the array may be reduced by use of lubricants. Slow and steady insertion of the electrode array has also been shown to reduce trauma to delicate inner ear structures. After insertion of the array the site of entry is sealed by placing soft tissue around the electrode array to prevent perilymph loss and to reduce the risk of labyrinthitis (Giordano et al. 2014; Roland et al. 2005).

Electrode insertion through the round window avoiding any drilling to enlarge the bony margins of the round window orifice has also become increasingly popular, especially since more electrodes designed for this type of insertion have become available. However, a pure round window insertion is not always possible due to variations in the size and orientation of the RWM. This situation often can usually be remedied using a marginal round window cochleostomy technique. The goal of this technique is to sparingly enlarge the orifice of the round window as atraumatically as possible employing the least possible amount of drilling. The technique begins with exposure of the RWM. Using a 1 mm or

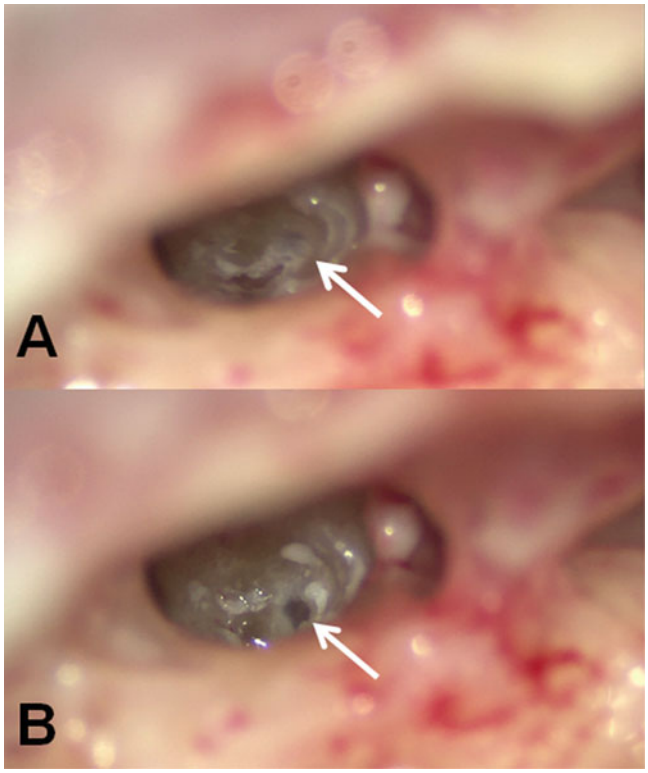

Fig. 5.10 (**a**) Poor visualization of the round window membrane secondary to a pseudo-membrane (*arrow*). (**b**) The round window membrane is visualized after removal of mucosal pseudo-membrane and the osseous margins of the round window niche

smaller diamond burr at reduced speed the anterior inferior osseous attachment of the round membrane (crista semilunaris) is removed exposing cochlear endosteum. The endosteum and round window membrane are then opened to permit placement of the electrode array. Drill entry into scala tympani is best avoided as it has been shown to create significant proximal inferior basal turn trauma which often results in new bone growth at the site of trauma (Richard et al. 2012).

The Malformed Cochlea

Implantation of the malformed cochlea may pose unique challenges from the standpoint of surgical approach, facial nerve abnormalities, electrode array selection, and cochleostomy technique. Most inner ear abnormalities are readily identified by preoperative MRI or CT. A number of centers now use MRI as study of choice in the cochlear implant evaluation process (Parry et al. 2005). However, the MRI finding of inner ear malformations more severe than incomplete partitioning type II (Classic Mondini) or vestibular malformations should prompt the surgeon to consider obtaining a CT to better define facial nerve abnormalities that more frequently occur in the presence of these anomalies (Pakdaman et al. 2012). CSF leak, often described as a "gusher," is also more frequently encountered within malformed inner ears. The surgeon should be prepared to manage CSF leaks at the time of CI surgery (Wootten et al. 2006).

The transmastoid labyrinthotomy approach to the common cavity (severe malformation in which normal architecture of vestibular and cochlear apparatus are absent) was first reported by McElveen in a series of four patients (McElveen et al. 1997). This approach requires identification of the otic capsule posterior to the facial nerve. It is found in the vicinity of where the lateral semicircular canal would typically be expected in normal ears. A full insertion was achieved in three out of four patients using this approach and 16 of 22 electrodes were placed within the cochlea in the remaining patient. Auditory perception was noted by all four patients in this series (McElveen et al. 1997).

Intraoperative fluoroscopy has been used to confirm the correct placement of the electrode array within the common cavity (Coelho et al. 2008). However, the additional cost and radiation exposure must be weighed against benefit, especially if used for cases in which the likelihood of electrode malposition is very small.

Beltrame described the double posterior labyrinthotomy technique in three patients with a common cavity malformation in whom he used a specially modified Med-El electrode array and reported favorable results (Beltrame et al. 2005). The array is modified by the addition of a silicone coated platinum wire fixed to the end of the electrode array and may be custom ordered from the manufacturer. The surgical technique involves creating two labyrinthotomies 4 mm apart. The specialized electrode array is placed into the superior labyrinthotomy and retrieved from the inferior opening using a platinum ball attached to the modified silicone carrier. The electrode array is then advanced simultaneously into both openings which pushes it into the anterior aspect of the common cavity adjacent to the internal auditory canal. The labyrinthotomies are both sealed with fascia to secure the electrode array into place (Beltrame et al. 2005).

The Ossified Cochlea

Ossification of the cochlear scala may occur within weeks after bacterial labyrinthitis, a problem that is seen most commonly as a consequence of pneumococcal meningitis. The cochlear aqueduct has been theorized to serve as a portal of entry for inflammation and bacteria between the subarachnoid space and the perilymph space. The proximal inferior basal turn of the scala tympani is the most common initiation point for ossification. Early identification of labyrinthitis with gadolinium enhanced MRI allows the surgeon to fast track the patient to surgery (Kopelovich et al. 2011). MRI enables the early cochlear obstruction due to fibrosis that

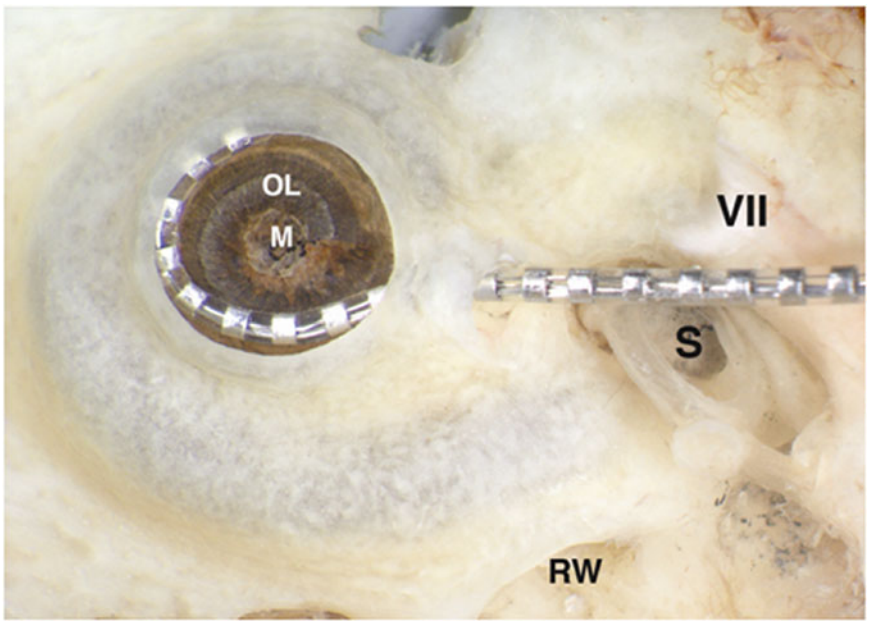

Fig. 5.11 Middle turn cochleostomy with an electrode in situ in the scala vestibuli (*M* modiolus, *OL* osseous spiral lamina, *RW* round window niche, *S* stapes footplate, *VII* distal tympanic facial nerve). Courtesy of Charles G. Wright

precedes ossification to be identified, whereas CT is only able to verify obstruction during the later stages of ossification (Booth et al. 2013; Isaacson et al. 2009). Early implantation in the setting of labyrinthitis increases the likelihood that the surgeon can fully insert an electrode array. Delays between labyrinthitis and cochlear implant surgery can result in dense ossification of the cochlear scalae that will necessitate special surgical techniques in order to maximize the number of electrodes inserted.

A proximal inferior basal turn drill out often allows for full insertion of an electrode array because the basal turn may be patent distal to the obstruction that is removed. The new bone filling scala tympani can usually be distinguished and removed from the surrounding dense otic capsule using the drill or micro-instruments. A depth gauge can be used to assess for more distal patency once the neo-osteogenic bone has been removed. An electrode with a stylet is often helpful in these cases as they are stiff enough to push through soft tissue scalar obstruction. Ossification that progresses into the ascending basal turn will require additional measures in order to maximize electrode insertion depth (Isaacson et al. 2009).

A combined inferior basal turn and middle turn cochleostomy is a viable alternative in patients with extensive basal turn ossification that limits the number of electrodes inserted. This technique entails performing a standard or round window cochleostomy in conjunction with removal of ossification along the inferior basal turn up to the transition of the ascending basal turn. The incus and stapes superstructure will need to be removed to perform the middle turn cochleostomy. The middle turn cochleostomy is performed 1–2 mm anterior to the anterior margin of the oval window niche just inferior to the cochleariform process (Fig. 5.11). A specialized double array cochlear implant is then used to insert separate electrodes arrays into the inferior basal turn and middle turn cochleostomies (Isaacson et al. 2008).

Scala vestibuli is more likely to be patent in the setting of labyrinthitis ossificans given the location of the cochlear aqueduct fundus in scala tympani. A cochleostomy opened just superior and anterior to the round window niche may permit access to scala vestibuli and allow for a full insertion of the electrode array is some patients (Gulya and Steenerson 1996).

A circumodiolar drill out procedure can be used in cases of extensive cochlear ossification involving the scala tympani and vestibuli. The technique entails a round window or standard cochleostomy followed by removal of the neo-osteogenic bone while preserving the lateral cochlear wall along the inferior basal turn. Preservation of the lateral cochlear wall along the inferior basal turn permits better retention of the electrode array once it is inserted. The lateral cochlear wall is then removed along the ascending and descending basal. The neo-osteogenic bone within the scala has a distinct appearance from the surrounding otic capsule bone and is removed. The surgeon must pay particular attention when opening the proximal ascending basal turn since the first genu of the petrous carotid artery is in close proximity to the cochlea in this area. The electrode array is then placed into the cochleostomy and is secured into place along the ascending and descending basal turns using fat or muscle plugs (Balkany et al. 1996). A large tympanomeatal flap or modified Rambo ear canal over closure, in addition to the standard transmastoid facial recess approach, is often necessary when performing a circumodiolar drill out to open the ascending and descending basal turns (Balkany et al. 1997).

Complications

Major complications as a result of cochlear implant surgery such as facial nerve injury causing facial palsy or paralysis, CSF leak and meningitis are fortunately quite rare. Other complications include: diminished taste due to chorda tympani nerve injury, electrode array malposition, bleeding/hematoma, and vestibular dysfunction. Loss of residual hearing may also be viewed as a complication if its preservation was a goal. Other complications that may occur subsequent to surgery include pain, device malfunction or complete failure, facial nerve stimulation, displacement of the receiver stimulator or electrode array and infections including meningitis (Venail et al. 2008).

Significant blood loss can occur and accumulate in the surgical field especially in cases in which a large subperiosteal pocket is elevated, although this problem is less common in children. As a general principle, meticulous hemostasis is of paramount importance prior to placing the implant as hematoma may increase the risk of surgical site infection. Use of drain in the surgical site is best avoided as it may increase the risk of infection (Filipo et al. 2010). Hemostasis in infants and small children is also important to minimize perioperative anesthetic complications secondary to hemodynamic instability. Once the implant is in the surgical field the monopolar cautery is turned off and any additional hemostasis should be obtained with suture ligatures or hemostatic sealants. Bipolar cautery use near the device is preferably avoided in order minimize risk of damage to the implant.

Facial nerve injury causing permanent facial paralysis is rare and typically occurs secondary to direct injury during opening of the facial recess. It may also occur due to heating of the nerve from rotation of shaft of the burr during drilling of the facial recess or cochleostomy (Venail et al. 2008). Heat injury to the nerve often presents with delayed facial palsy or paralysis and has a better prognosis than immediate facial paralysis. The latter presentation of facial paralysis warrants immediate facial nerve exploration to determine if the nerve is visibly injured and to perform a repair if warranted. Facial nerve monitoring may help reduce the risk of injury but does not replace a thorough understanding of temporal bone anatomy and use of appropriate surgical techniques. In addition, monitoring will not alert the surgeon to thermal injury. Patients with cochlear malformations have a higher incidence of facial nerve anomalies and therefore are at greater risk of facial nerve injuries (Pakdaman et al. 2012). As noted earlier in this chapter many centers use MRI as the preferred imaging modality in the evaluation of a cochlear implant candidate (Parry et al. 2005). Although MRI readily demonstrates cochlear malformations it does not provide as much information regarding the course of the facial nerve. Therefore, availability of a CT temporal bone study may be beneficial for children with cochlear malformations. This is especially true for when the lateral semicircular canal is abnormal or absent as these finding are more often associated with abnormal facial nerve anatomy (Ellul et al. 2000). During surgery, identifying the lateral semicircular canal and incus, as well as thinning the external auditory canal are essential steps in identifying the facial nerve and safely and accurately opening the facial recess. Copious irrigation and drilling parallel to the facial nerve with small diamond burrs allows the surgeon to identify the facial nerve and reduces the risk of direct and thermal injury.

Facial stimulation due to current spread from electrodes within the cochlea does occur in children. It can usually be easily managed successful by the audiologist (see Chap. 7) by turning off an electrode(s) lying in proximity to the horizontal segment of the facial nerve. There are rare patients with abnormal temporal bone anatomy or bone disease that experience facial nerve stimulation that is challenging to manage and may preclude successful device use. The best example of a bone disorder causing this type of problem is far advanced otosclerosis, which fortunately has not been reported in the pediatric population (Frijns et al. 2009). Perimodiolar electrode arrays may reduce the risk of facial stimulation. Eliminating the offending electrodes often decreases or eliminates facial stimulation, but may reduce auditory performance (Battmer et al. 2006).

Chorda tympani nerve injury is not uncommon after cochlear implant surgery especially in patients with a narrow facial recess. The altered sense of taste is typically self-limiting but can last up to a year. Great care should be taken to preserve at least one chorda tympani nerve in patients undergoing bilateral CI surgery (Venail et al. 2008).

Vestibular dysfunction is usually a self-limiting event that occurs in up to 10 % of cochlear implant recipients and may result from: perilymph fistula, suctioning perilymph, serous labyrinthitis, lateral semicircular canal fenestration, electrode displacement into the vestibule, perilymph/endolymph mixing from electrode trauma, or endolymphatic hydrops (Venail et al. 2008). Surgical fenestration of the semicircular canal is rare and can easily be avoided using proper mastoidectomy techniques. Correct cochleostomy and electrode array insertion usually prevents inappropriate electrode array positioning. Packing the cochleostomy after insertion of the electrode array reduces the risk of perilymph fistula as well as the potential for suppurative labyrinthitis and meningitis in the setting of otitis media.

Electrode malposition or migration is fortunately rarely seen after implant surgery (Connell et al. 2008). Careful identification of the round window membrane, cochleostomy placement, and electrode array insertion are critical to avoid inadvertent placement into the eustachian tube, carotid canal, infracochlear air cell tract, internal auditory canal, or vestibule. Inserting the electrode array from a posterior superior to anterior inferior direction usually ensures ideal positioning of

the electrode array within the scala tympani. The use of intraoperative fluoroscopy, X-rays, computed tomography or electrical compound action potential testing available in each commercially available CI system may be used to confirmation correct electrode array placement, if necessary (Coelho et al. 2008). In addition it is useful to keep in mind that in some cases a fully inserted electrode may become displaced at some point after surgical insertion. Partial electrode extrusion has been reported in cases of progressive cochlear ossification. Electrode extrusion may also occur in the normal patent cochlea. It is more likely to occur when a straight electrode has been used. Therefore, it is important to pay meticulous attention to secure the lead in the facial recess and mastoid.

Cerebrospinal fluid leak may occur by a number of mechanisms during cochlear implant surgery. CSF leak is unavoidable if CSF is already present within the cochlea because of a congenital defect permitting CSF to communicate with the inner ear. It may also occur due to exposure and injury to dura during creation of a recessed bone bed for the receiver-stimulator, during mastoidectomy, or due to injury of the modiolus during either drilling of the cochleostomy or as a result of electrode insertion. The need to create a recessed bone bed with its risk for CSF leak may be eliminated by using other techniques to secure the receiver stimulator such as a tight subperiosteal pocket. Tegmen and posterior fossa plate injury with underlying dura are rare during cochlear implant surgery but usually easy to identify and repair at the time of the procedure with allografts (muscle, fat, cartilage, bone), or artificial materials such as bone cement. Allowing CSF drainage for several minutes after opening the cochlea, in the setting of a gusher, often reduces the flow rate and facilitates sealing the cochleostomy thus eliminating the leak. Creating a central opening in a dry piece of fascia which the electrode array is passed through is another method that can be used to repair a CSF gusher. The author's preferred technique to manage a leak involves creating an opening in a piece of fascia through which the electrode array is threaded so that it may be used to seal the cochlea around the electrode array once it has been completely inserted. Alternatively fascia, fat or muscle may be used after electrode insertion to seal the cochleostomy. Persistent CSF leak after CI surgery may necessitate placement of a lumbar drain or further surgery which may include a subtotal petrosectomy with over closure of the ear canal and obliteration of the eustachian tube (Wootten et al. 2006; Free et al. 2013).

Meningitis is a potential life threatening infection that may occur in CI recipients. A recipient presenting with fever, and meningeal signs should undergo an emergent evaluation for meningitis including lumbar puncture with cultures. Initiation of empiric antibiotic therapy should be initiated immediately after cultures are obtained to hopefully avoid the potentially devastating complications of meningitis. The implant team should carefully follow the Centers for Disease Control guidelines with respect to vaccinations for cochlear implant candidates and recipients in order to minimize the risk of bacterial meningitis (see Chap. 3).

Stimulator-receiver displacement is a rare event after CI surgery. Trauma during the perioperative healing period is the most common cause of implant displacement. Delayed traumatic implant displacement is much less common due formation of a tight fibrous capsule around the receiver-stimulator. The subperiosteal pocket fixation technique has more potential for receiver-stimulator displacement during the immediate postoperative period; however, this risk is reduced over time by osseous remodeling and capsule formation around the implant (Balkany et al. 2009). Displacement of the receiver-stimulator toward the pinnae may result in difficulty wearing a BTE processor because of skin irritation. Significant displacement of the implant also may cause partial or complete extrusion of the electrode array. Signs of electrode extrusion include the most proximal electrodes demonstrating abnormally high impedance and loss of evoked compound action potential responses. Lack of progress or a decline in performance may be noted. These findings may be confused with device malfunction (Chung et al. 2010). Once suspected, electrode extrusion can be confirmed by high resolution CT of the temporal bones. If the implant contains a magnet that is removable, the magnet may become dislodged. It can usually be replaced successfully with a minor surgery (Ellul et al. 2000).

Post-implantation Magnetic Resonance Imaging

Issues surrounding MRI studies after cochlear implantation should be discussed with the family. At present two of the three manufacturers of CI devices that are FDA approved for commercial use in the USA do not have FDA approval that permits them to recommend that patients with their devices containing a magnet undergo MRI. To address the potential need for MRI in CI recipients these two manufacturers' (Cochlear Americas and Advanced Bionics) current implants are designed to permit magnet removal with subsequent replacement. However, there is evidence that implant recipients with these devices may undergo a 1.5 T MRI with the removable magnet remaining in place if a firm head wrap is used in order to minimize magnet displacement and movement of the receiver stimulator (Broomfield et al. 2013; Crane et al. 2010). These CIs have CE Mark approval (requirement for products sold to the European Market) for use in this manner, and therefore, this practice is common in many countries where FDA approval is not required. One potential problem is that repeated MRI studies in patients with a CI may result in reduced strength of the internal

device magnet (Broomfield et al. 2013). However, if the device has a removable magnet, replacement with a new magnet may be performed to address this issue. Another issue for consideration is that images of the head region will contain substantial artifact if they are obtained with the magnet in place, whereas studies of the remainder of the body should not be similarly impacted.

When the device model allows for magnet removal and replacement, a spacer is placed within the silastic pocket that holds the magnet. In children these procedures usually requires anesthesia (Migirov and Wolf 2013). Although minor, these procedures introduce the risk of infection and damage to the silastic pocket which may preclude magnet replacement. Therefore, the authors prefer to obtain MRI without magnet removal unless its elimination is necessary to obtain a clinically useful MRI of the brain. Magnet removal with subsequent replacement is not an option for CI's manufactured by Med-El. However, in 2013 this manufacturer introduced devices that are FDA approved for use in 1.5 T MRI.

Conclusion

Just as devices have improved, CI surgery has become more sophisticated and preservation of residual hearing and delicate inner ear structure are now possible with careful selection of the electrode array and use of more atraumatic surgical techniques. CI surgery may be more challenging in children than adults at times. Infants and very young children have variable anatomy as their mastoids are still developing. Their physiology is also less mature. Hence the importance of vaccinations to reduce CI related infection and use of pediatric anesthesia techniques appropriate to this age group. The increased incidence of congenital cochlear and temporal bone malformations in the pediatric population also needs to be born in mind because of the increased the likelihood of abnormal facial nerve anatomy and CSF leak. However, CI surgery in children may be performed safely and effectively using modern surgical and anesthetic techniques when appropriate perioperative preparation and procedures are followed.

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Cochlear Implant Programming for Children

6

Holly F.B. Teagle

Introduction

Cochlear implant speech processor programming is a dynamic process that requires astute observation skills, patience, and working knowledge of the current (and past) cochlear implant software and hardware systems. For children with no previous experience with sound, the programming audiologist is setting the stage for all future auditory experiences. The child with a congenital hearing loss is a blank slate, so to speak, primed to begin the journey of mapping sound to meaningful experience. For children with progressive or acquired hearing loss, the remapping of this new percept to their previous knowledge of sound is a fresh beginning that will lead to better hearing with a cascading impact on social skills and educational opportunities. In any case, expertise is required to afford each child the means to achieve his or her full listening potential. The audiologist has the privileged opportunity to be intimately involved as the child's auditory abilities originate and evolve.

Children with hearing loss who receive cochlear implants are a highly diverse population. Specific steps in creating speech processor programs are common to the process but optimizing settings for an individual requires an appreciation of each child's special needs and stage of development, as well as the family, social, and educational environments. The cochlear implant is a tool, not a cure, for minimizing the effects of deafness. Many factors will affect outcomes. The best possible result for any individual child begins with managing this tool to ensure it has been set to exploit all the technological advantages it has to offer.

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Working Assumptions

Before delving into the steps and considerations of device programming, some working assumptions about the process should be asserted. This includes the assumption that a team approach is being used to provide patient care; there is an existing interdependence of roles and responsibilities among team members with the common purpose of providing comprehensive care for the child. Device programming is one component of the overall management of the child with a cochlear implant. It hinges on and matriculates with surgical management and overlays and interacts with therapeutic services. Another working assumption is that the audiologist has knowledge of the child from both a social and a medical perspective. Finally, assuming the team approach is in place, and the audiologist has interacted with the child with some level of rapport, there is the assumption that decisions about cochlear implantation have been made from a point of knowledge and informed discussion.

A Team Approach

The importance of a team approach to pediatric cochlear implantation cannot be overstated. During the evaluation process, all aspects of the child's development are assessed and reviewed. The speech-language pathologist, audiologist, and early interventionist or educator contributes information and perspective on the child's development, strengths, and needs. The surgeon provides perspective on the child's medical status. There may be additional professionals who offer information and support as needed, such as psychologists, neurologists, geneticists, social workers, and other health or educational specialists. During the evaluation period, information from the team is shared to determine candidacy, to provide the family with appropriate expectations, and to make plans for habilitation. Postimplantation, the team approach is essential for monitoring progress and navigating concerns if they arise. Figure 6.1 illustrates the dynamic

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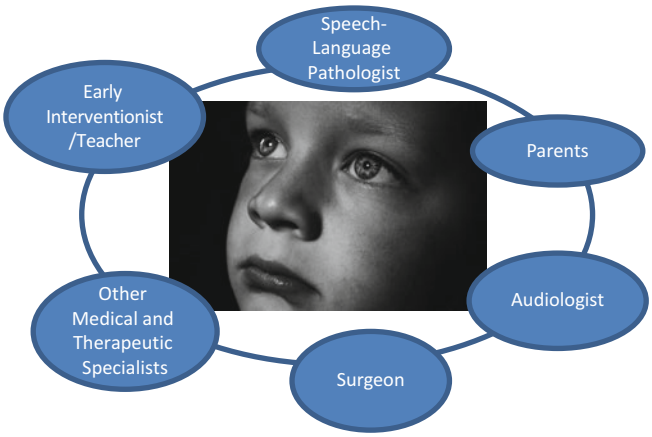

Fig. 6.1 Collaboration among medical and educational professionals is essential during the cochlear implant evaluation, after surgery and the initial stimulation of the device, and on an ongoing basis to ensure each child meets his or her potential in communication development

interaction among all team members, with the focus being the child's unique strengths and needs.

Relative to device programming, consistent and candid communication between the audiologist and surgeon is critical for a number of reasons. Before surgery, decisions about device selection and electrode choice are made based on the patient's anatomy and residual hearing and whether the child has any special physical considerations related to device options. (As an example, special consideration of device manufacturer and processor style should be given for a child who spends many hours a day in a wheelchair with head support. A processor that offers different wearing options and also has an alarm that alerts a caregiver to the coil falling off would be particularly helpful.) During surgery, results of intraoperative testing of device function and the ear's responses to electrical stimulation confirm successful placement and can be useful for future programming decisions. (For example, an electrode array that is not fully inserted due to the child's anatomy will result in electrodes outside of the cochlea that should not be stimulated.) The surgeon and audiologist continue to confer as needed if concerns about a child's health and progress emerge over time. Likewise, the relationship between the programming audiologist and the child's therapists and teachers is essential to navigate hardware concerns, to plan for successful use, to assess the effectiveness of device settings, and to monitor progress.

Knowledge of the Child

Ideally, the audiologist who is involved with the initial and ongoing programming of the speech processor will have established a relationship with the child and family during the evaluation process. Candidate assessment and presurgical counseling presents the opportunity for the programming audiologist to learn about the child and family and establish that most important foundation of the provider/patient relationship, trust. Counseling that takes place as the family is learning about cochlear implantation should include a discussion of goals for the child. The audiologist must gain some insight about the family's acceptance of the diagnosis and the stage at which they are entering the decision-making process. Are they responding from grief or anger? Have they idealized the process and created expectations of normal hearing? Are they cognizant of other developmental or medical issues the child might have and does the audiologist appreciate what these might be? Is the family's preference for communication mode realistic and are services in place to support this plan?

Informed Decision Making

A final working assumption is that the family has been properly counseled and has been central to the decision-making process regarding whether to implant, when to implant, and which device to use. Part of counseling and decision making includes discussion of the family's engagement in habilitation and their knowledge and consideration of therapy and educational needs. There is a plethora of information available to families via the internet, including the manufacturer websites and social networking sites. Support groups and other cochlear implant recipients can also share personal experience and perspective. It is important to note that not all sources of information will provide accurate and unbiased information. In the interest of preparing effectively for surgery and device programming, and for achieving outcomes that meet the family's expectations, counseling from members of the cochlear implant team and shared decision making among the team members and the family is essential.

Fig. 6.2 (**a**) The cochlear implant audiologist has the privilege and responsibility of making the "hearing birthday" positive and memorable for the family. (**b** and **c**). Important elements of this initial stimulation visit include use of a high chair for good positioning and visibility of the child and a pilot cap to keep the processor and coil in place on the child's head and programming cables out of the way. A test assistant can help engage and condition the child and parents should be welcomed to interact and provide insight on how the child is responding

Setting the Stage

The physical environment for the initial stimulation is worthy of considered preparation as it requires planned positioning of seating and props. It is important to acknowledge and prepare for the emotional environment of the event as well. The "hearing birthday" is often remembered and celebrated by some families for years. The typical scenario is that, once past the worry of surgery, family members are excited and have variable expectations about what will occur during the appointment, depending on their level of involvement with and memory of preoperative counseling. Most parents or other family members present have heard about or have seen a video of a child hearing for the first time with a cochlear implant. They may have envisioned it to be miraculous and happy. It is helpful to provide some structure to the process in aspiration of at least approaching these expectations (Fig. 6.2a).

The physical environment should allow the audiologist good access to the programming hardware and software and to the child. To date, a wireless connection from the programming computer and the speech processor does not exist, though this is surely on the horizon. Managing connecting cables is a consideration since it is important that the speech processor stay on the child and connected during programming. To keep the child positioned for engagement and observation, use of a high chair for a very young child (Fig. 6.2b), or a small table and chair for a toddler, or a well-positioned, comfortable chair for an older child or teenager is needed. Choices of toys and reinforcers should be available and ample. Toys for distracting and toys and games for engaging in listening in a conditioned response task should be on hand. For toddlers and preschool age children, having a test assistant to distract the child with interesting toys and activities, or to engage the child in conditioned response activities, is essential.

The preparation for device programming with children should begin with a review of the goals of the initial programming session, which are acceptance, comfort, and audibility. Subsequent programming sessions will be needed to fine tune the program over time. A successful initial stimulation is made possible when expectations are clear; this will also set the tone for future programming sessions. It is important to impart that the programming of the cochlear implant is a process that will evolve over time. Counseling should prepare parents to be aware of the range of responses from the child as the device is first activated and to invite the sharing of their observations of the child's reactions (Fig. 6.2c). Responses from the child may range from subtle to the sublime. Once the device has been programmed, it is crucial to have ample time to review device components, settings, and plans for use. Parents need to feel confident in their ability to manage the hardware and to monitor the child for comfort and acceptance of the device.

Determining Speech Processor Program Parameters

Each cochlear implant manufacturer produces dedicated software and hardware for the purpose of programming speech processors. Multiple generations of technology exist for each company and continued change and development is inevitable. Staying current with new technology and remembering the idiosyncrasies of older systems can be challenging for the audiologist. Training and support from the manufacturers is usually readily available.

A speech processor program is created for a new recipient by opening a patient file in the programming software and then choosing various stimulation options. Some parameter choices are made based on the implant manufacturer and the generation of the cochlear implant system. The primary parameters that dictate how sound is coded include: (1) speech

coding strategy, (2) stimulation mode, (3) pulse width and electrical charge or current levels, (4) stimulation rate, and (5) the number of channels and their frequency boundary settings. Many options exist but all three cochlear implant systems used in the US suggest default settings for some parameters based on in-house research and, generally, these offer a good starting place. Because of brain plasticity, most children will learn to use the signal generated by the chosen parameters and perform well, but the audiologist should be prepared to make changes if responses and progress are not as expected.

There are other decisions to make when programming the speech processor that relate to how the instrument will detect, mix, and deliver sound. These include microphone settings, direct input mixing ratios, and enabling output settings, such as t-coil and blue tooth detection. The utility of these features will likely change over time as the child becomes a more sophisticated listener. They will be addressed in a later section.

Speech Coding Strategy

Contemporary cochlear implant systems have developed overtime based on the performance outcomes of increasing numbers of cochlear implant recipients using a variety of speech coding strategies. The speech coding strategy defines how acoustic information is analyzed, filtered, and then represented and delivered electrically to the cochlea. The brilliant evolution of strategies since the early clinical use of cochlear implants is a testament to the dynamic interaction in the field between engineers, clinicians, and researchers. Single channel implants coded intensity and envelop cues through amplitude modulated stimulation. Early speech coding strategies for multichannel devices added spectral information by taking advantage of the tonotopic organization of the cochlear and through varying stimulation rates. With continued research and development, a variety of strategies now exist, most focusing on coding of temporal cues. Describing them all is beyond the scope of this chapter. However, as related to working with pediatric cochlear implant recipients, it is noteworthy that not all speech coding strategies are FDA approved for use with children. This means the manufacturers have not included children in the clinical trials conducted to demonstrate efficacy claims. The various coding strategy options are accessible in the programming software and it is left to the clinician's discretion as to whether to use particular strategies with children. This may be unsettling when audiologists strive to make clinical decisions grounded in evidence-based research. Experience with adult recipients and older children, as well as consideration of peer-reviewed studies of patient performance, may increase the audiologist's comfort level for using some of the strategies. As with other aspects of patient care, a decision to use a particular strategy depends on the individual's response. The audiologist's knowledge of the patient and the expected outcomes, the routine use of test batteries to track performance, and consultation with therapists and parents to monitor progress drive decisions to make and keep changes in speech coding strategy.

Stimulation Mode

Stimulation mode refers to how current, or electrical charge, is directed and grounded to create electrical circuits in the implant. The ability to manipulate stimulation mode varies by manufacturer. This feature is often set by manufacturer default settings to a monopolar mode and, in the majority of cases, does not need to be altered. Children with cochlear malformations or implanted devices that are not typically placed may require some creative changes in stimulation mode. Future generations of cochlear implant electrode arrays and speech coding strategies may employ varying stimulation modes that can be selectively changed during processor programming.

Pulse Width and Electrical Charge or Current Level

Determining the amount of electrical charge needed to excite the auditory system is fundamental to cochlear implant programming and the process of determining these values is discussed in detail later in this chapter. Pulse width and electrical charge or current levels are two parameters that interact dynamically to create the electrical dynamic range. In general, lower electrical charge creates the percept of soft sound and greater electrical charge enables loudness growth. Depending on the programming software, pulse width can be set to adjust automatically with increased charge needs, or it can be set at a fixed level by channel. When set automatically, as charge or current levels increase, the pulse width broadens based on the system compliance (relationship between electrode impedance and the conductive environment in the cochlea) with the goal being to afford the most efficient use of current. When pulse width is fixed, the audiologist must monitor current levels, to ensure they are within system compliance, and change pulse width as needed. Children with compromised anatomy may require more electrical charge to stimulate neural tissue. Often, this can only be achieved by using a broader pulse width (Buchman et al. 2011). The amount of charge needed to establish an optimal electrical dynamic range is unique to individual ears and can affect other parameter settings, such as stimulation rate and number of channels made active. This has implications for bilateral recipients. With a fixed pulse width

and rate of stimulation, the audiologist is assured that both ears have more similar parameter settings. A recipient programmed with automatically increasing pulse widths could end up with very different rates of stimulation between ears. This may or may not have importance for optimizing performance but is something that the programming audiologist should be well aware.

Stimulation Rate

Pulse width has a reciprocal relationship with stimulation rate; the wider the pulse width, the slower the rate of stimulation. Conventional thinking has been that a faster rate may be better so that the acoustic sampling and delivering of stimulation can be more sensitive to temporal changes in sound. However, this is yet another situation where the individual and the unique aspects of anatomy and physiology and experience with sound affect decision making. Studies of the effects of rate suggest that individuals have subjective preferences for rate of stimulation and often demonstrate differences in performance with different rates (Skinner et al. 2002; Balkany et al. 2007). These studies have been performed with adults, many of whom have memory of natural hearing and a reference for sound quality judgments. Most children who hear well for the first time with a cochlear implant are not able to critically compare speech processor programs of varying rates, adapt in a short period of time, and then express preferences. Evaluating different programs after brief exposure is difficult because performance is influenced by experience with a given strategy. Once again, it is critical to strategize and make changes to programming parameters, including rate, if a child fails to make the expected progress when all other components for success are in place.

Number of Channels and Frequency Boundary Settings

The brilliance of multichannel cochlear implants is the ability to mimic the natural tonotopic organization of the cochlea and provide frequency-specific information along the base to apex. Sound quality and speech recognition can be altered by manipulating the number of active channels and the frequency boundary assignments among them. Default parameters are set in the programming software and typically require little change. However, systematic attention to error patterns children exhibit in their speech perception, and in many cases their speech production, may provide insight to adjustments for specific channels. This highlights the need to monitor speech perception and speech production over time when making programming decisions. An understanding of speech acoustics provides the basis for making such changes. Here is a scenario as an example: a child is not easily alerting to soft high-frequency speech sounds, such as /s/ and /f/. When measuring electrical thresholds on basal electrodes it is demonstrated that detection is elevated relative to other channels, and/or the child does not report loudness growth on that channel. By disabling the most basal channel and shifting the frequency boundaries down, more neural elements are stimulated when those speech sounds occur. A team approach to management of the child makes these kinds of observations and resulting modifications possible.

Suffice to say that while manufacturer default settings guide the choice of speech processor parameters, the audiologist's knowledge of the patient and consideration of individual differences determine how and when changes in parameters should be made. Changes can and should be made with consideration and discretion. Parameters selected for the initial speech processor program may need to be changed as the child adapts to the signal and begins to demonstrate understanding of speech.

Creating Speech Processor Programs

Depending on the child's age and cognitive and physical development, the audiologist gages how much reliance to place on behavioral measures provided by the child and/or objective measures that can be obtained through the programming software and are not dependent on a behavioral response. Both measures are essential to creating and optimizing a program. However, the young child who has little experience with sound may not provide clear responses initially. Engaging the child in an activity and then observing him or her as stimulation levels are gradually increased is usually a successful approach to obtaining the first indicators of sound awareness. Often a young child who has had no or limited hearing will adapt to the electrical signal, or cease to respond when it is no longer novel. It is important not to miss the "first hearing" behaviors. Once a response has been observed, the audiologist and test assistant can condition the child's response. A detection response, which may reflect a threshold or a suprathreshold percept, can be obtained through a number of behavioral responses. Once programming parameters have been determined during the initial stimulation of the implant, behavioral or objective measures are used to set current levels and create the electrical dynamic range of the program. With time and experience, the child's development of audition skills will dictate how speech processor programs can be further optimized to yield the best possible performance. Verifying settings through ongoing assessment and monitoring is a critical aspect of device programming.

Behavioral Measures

Behavioral testing is the cornerstone of diagnostic pediatric audiology (American Academy of Audiology 2012) and these techniques are also applied when programming the speech processors of children with cochlear implants. Developmentally appropriate methods, depending on the child's age, cognition and motor skills, are used to determine electrical threshold and comfort levels, which define the electrical dynamic range of the speech processor program. Behavioral measures are often corroborated by objective measures and, as a child ages and develops, will have a larger role in device programming.

Electrical Threshold Measures

Electrical threshold responses can be obtained in a number of ways:

- Behavioral Observation is used with infants or with children who have significant developmental delays and are a supplement to objective measures. In truth, the programming audiologist should be continually observing the child for signs of sound awareness or discomfort during device programming. Often, the indicators of a change in auditory environment are demonstrated by subtle changes in behavior, such as cessation of activity, eye widening, or startling that is time locked to the onset of stimulation. As in conventional audiometric testing, electrical threshold and comfort levels that are determined based on behavioral observation should be supplanted by conditioned behavioral responses when the child is developmentally able to participate.
- Visual Reinforcement Audiometry (VRA) techniques can be used effectively during device programming in a fashion similar to diagnostic testing. Device programming is never done is a sound booth so the programming office, which should be child friendly and comfortable, must be oriented with a reinforcement device that can be controlled by the programming audiologist or test assistant.
- Conditioned Play Audiometry (CPA) as used in diagnostic audiology can be used to determine electrical threshold levels. While Med El and Advanced Bionics systems do not require threshold measurements, they can be useful, and they are essential for Cochlear Corporation device programming. Traditional CPA involves conditioning a child to perform a task when sound is detected. These tasks should include using manipulatives, such as puzzles and games, that are age appropriate and of interest to the child.
- Traditional or volunteered responses for children who do not require toys to maintain interest and attention usually consist of verbal responses or descriptions of sound. To ensure electrical thresholds are audible, the child can count the number of stimulations.

Electrical Comfort Measures

Electrical comfort levels are set to ensure sound does not exceed the child's tolerance for sound. The goal in device programming is to determine a sufficient electrical dynamic range such that sounds are perceived as soft, moderate, and loud in keeping with acoustic correlates (Davidson et al. 2009). Behavioral observation and feedback from the child dictates the magnitude of the dynamic range. Young children will not understand the concept of loudness growth until they gain experience and language skills. Often a fixed range relative to electrical thresholds is used to set electrical comfort levels until a child can reliably scale loudness on individual channels. For older children, loudness scaling is often accomplished using pictures that represent a range of sound percept from soft to uncomfortably loud. Depending on the child's abilities and experience a three-point scale (soft, ok, too loud) up to a 10-point scale (barely audible…uncomfortably loud) can be used, as shown in Fig. 6.3a. The notions of soft sound and loudness growth can be illustrated with graphics of parallel concepts as shown in Fig. 6.3b. The amplitude and pulse width of current levels interact when determining electrical comfort levels, as previously discussed.

Ordering and Eliminating Channels

Another aspect of device programming that requires a behavioral response is pitch ranking. Maturity and previous hearing or considerable experience with listening through a cochlear implant is needed for children to reliably pitch rank. Many adult recipients struggle with this task. Loudness, pitch, and quality are often confused by young listeners. Orienting them to the concept through use of a keyboard is helpful. The goal in pitch ranking is to eliminate or reorder channels that do not match the expected tonotopic ordering of sound. When modifications are made to channel order, it is very important to document performance before and after to validate the change in settings.

In addition to loudness scaling and pitch ranking, further optimization of programming parameters includes identifying aberrant electrodes that produce a poor quality of sound or do not produce growth in loudness with increased current levels. A sophisticated listener may be able to identify a channel of stimulation that has poor quality relative to other channels.

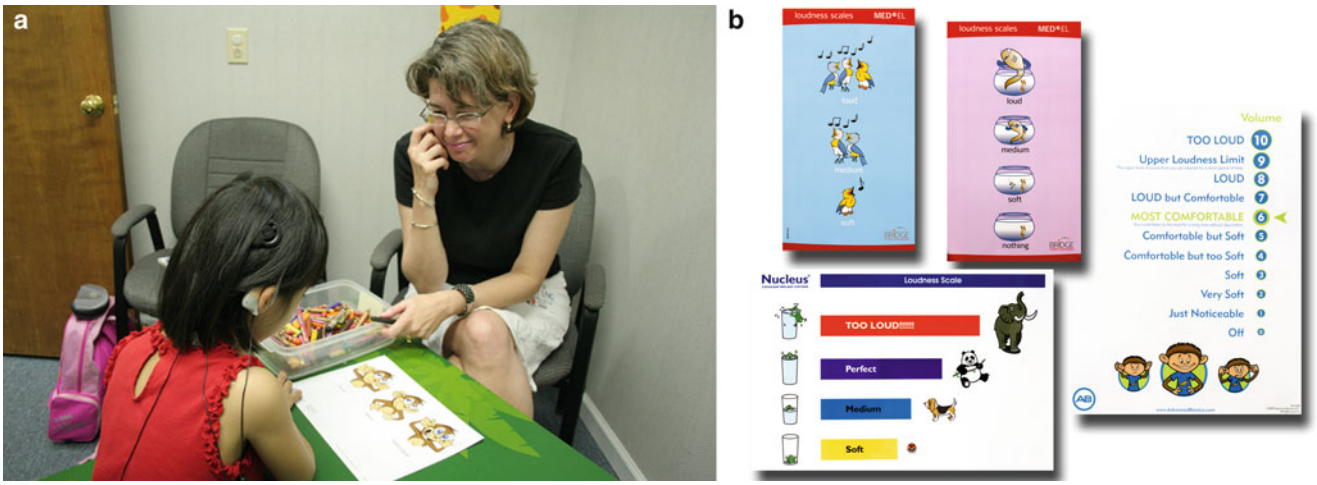

Fig. 6.3 (**a**) Teaching the concepts of soft and loud sound with images or signs can begin at an early age and, with practice, children can be reliably engaged in the process. (**b**) A variety of tools for loudness scaling are available to the programming audiologist from the cochlear implant manufacturers

With multiple programming slots on modern speech processors, a comparative trial between programs can be undertaken with monitoring from parents and therapists to determine preferred settings. Assessment of performance with speech perception tests adds objective evidence for optimizing program parameters.

Objective Measures

Currently, all manufacturers offer the ability to perform objective measures through the cochlear implant system, but early generations of cochlear implants do not have this capability. Objective measures useful for device programming include impedance telemetry, the electrically evoked compound action potential, and the acoustic reflex.

Impedance Telemetry

Electrode impedance refers to the capacity of each electrical contact to conduct current. This measurement is indicative of the status and environment of the electrical contact. Impedance measurements are performed through the programming software and should be collected at each programming session. In fact, the manufacturer programming software is created so it is essentially impossible to avoid this. The primary utility of this measure is to identify electrodes that have high impedance or are shorted together or are open circuits. Repeated measures of impedance telemetry reflect the stability of the electrode array and the interface to neural elements. The status of the electrode array can change for a number of reasons and it is important to ensure that each channel is effectively stimulating neural tissue. It is not uncommon for impedance values to be higher during intraoperative monitoring due to recent placement and air bubbles within the cochlea and then after the electrode settles in, to show improved capacity to conduct current with lower impedance values. Stability is desirable, so electrodes that vary significantly in impedance should be turned off in the speech processor program to minimize the possibility of odd percepts. With use, impedance values typically lower and remain stable. Elevated impedances across the electrode array, relative to previous measures, are often indicative of nonuse of the device. If the contacts are not stimulated, debris collects and makes them less conductive. Other reasons for variance in electrode impedances may include hormonal changes or disease processes within the cochlea. Finally, changes in electrode impedance can be a sign of impending internal device failure. For a child who does not have the experience or developmental skills to report odd acoustic percepts that may be related to device integrity, impedance measurements are of great value and should be monitored at every opportunity.

Electrically Evoked Compound Action Potential (ECAP)

The electrically evoked compound action potential or ECAP threshold assessment is a measure of auditory nerve function that is obtained by stimulating individual electrodes through the device programming software and using nearby electrodes to record neural activity generated from that stimulation. ECAP software has a unique name and calculation algorithm for each cochlear implant manufacturer.

For Cochlear Corporation, the measurement is called Neural Response Telemetry or NRT; Advanced Bionics Corporation software is called Neural Response Imaging or NRI; and for the Med El Corporation, this tool is called Auditory Response Telemetry or ART. The ECAP threshold is typically a stable measure over time, which is the core of its value and utility (Hughes, et al. 2001). Change in the measure for any individual may represent change in auditory nerve response. The correlation between the ECAP response and behavioral electrical current levels varies for each company. Depending on the manufacturer, the ECAP can contribute information that supplements behavioral measures but in no case does it completely supplant the need to perform behavioral measures of electrical threshold and comfort. Still, the presence of the response ensures the implant is effectively stimulating neural elements within the cochlea. It does not indicate that auditory processing is occurring at higher levels in the auditory system.

The ECAP response is typically acquired fairly quickly and easily because the child does not have to be sedated to record the response and there is no interference from myogenic activity. While ECAP measures can be collected at any time postimplant, intraoperative measurement of ECAP thresholds is particularly useful for ensuring proper placement of the electrode array and function of the internal device. The recording of the response is not only reassuring to the audiologist and surgeon, but also provides a level of relief to parents, which may make anticipation of the initial stimulation less stressful. Obtaining ECAP responses while the child is sedated is also desirable because the level of stimulation required to elicit the responses may be too loud or frightening for a child to collect during the initial stimulation, which is contrary to the goals of acceptance and comfort. Once a child has acclimated to wearing the speech processor, collecting ECAP responses is less threatening.

The ECAP response is obtained using a stimulation rate that is slower than the speech processor program rate, making the relationship between ECAP response levels variable relative to the electrical dynamic range of the program (Brown et al. 2000; Franck and Norton 2001; Holstad et al. 2009; Hughes et al. 2000). For most recipients the ECAP response will fall within the mid to upper end of the dynamic range (Brown et al. 2000; Hughes et al. 2000). During the early stages of programming, this provides the audiologist with a reasonable point to practice the conditioned response with a child, which can lead to determining reliable electrical threshold levels. In some cases, it may be necessary to rely more heavily on this objective measure for very young children or children who are developmentally delayed and unable to provide consistent behavioral results. Studies of adult performance on speech perception tests using programs created with ECAP-based programs compared to behavioral response-based programs suggest small differences in performance overall (Seyle and Brown 2002; Smoorenburg et al. 2002). Until settings can be optimized based on the child's behavioral responses and evaluation of progress over time, ECAP-based programs offer a reasonable starting point.

Electrically Evoked Stapedial Reflex Threshold (ESRT)

The electrically evoked stapedial reflex threshold is another objective measure that can be useful in device programming. It is recorded in the same manner as acoustic reflexes using an immittance bridge but elicited by stimulating channels or groups of channels through the cochlear implant programming software. Unlike ECAP, the response is not manufacturer or device dependent, and recipient program parameters for stimulation can be used to elicit the response. It does require a relatively still child with healthy middle ears and some coordination between the audiologist and test assistant or parent. Early studies of the utility of the ESRT indicated it was measurable in about 70 % of cochlear implant recipients but, when present, was highly correlated to behavioral electrical comfort levels in recipients who could scale loudness (Battmer et al. 1990; Hodges et al. 1997). If used with children, it should be collected using an ascending approach to avoid an aversive reaction. The level of stimulation necessary to elicit the response may be above comfortable program levels, particularly for a new recipient. Newer generations of programming software offer tools to incorporate visual markers of the ESRT into speech processor programs.

Program Optimization

The determination of speech processor program settings is a dynamic process which is influenced by the auditory system's adaptation to the electrical signal and the child's ability to provide information. Many programming sessions are required during the first 6 months of device use and ongoing programming should be completed to fine tune or adjust settings because of physical and/or sensory changes in the child. The appropriateness of settings is realized with observable changes in the child's sound awareness and meaningful use of sound. Settings should be verified frequently through sound field hearing tests and progress should be measured through informal and formal speech perception measures and speech and language assessments at regular intervals. Introducing multiple programs that can be used to optimize listening in various sound environments and situations is more effective once basic parameters have been determined and become stable.

Verification of Audibility

Skinner et al. (1997, 1999) first demonstrated the importance of using sound-field thresholds as a guide for fitting cochlear implant processors in adult recipients. Research has suggested that detection levels that are less than 30 dB HL across frequencies from 250 Hz to 6 k Hz are optimal (Firszt et al. 2004; Davidson et al. 2009). For children who may not be able to report on sound quality, a detection audiogram is critical to ensure speech is audible at a soft conversational level in a quiet environment. Flat detection levels between 250Hz and 6 KHz at 25–30 dB HL, when the child is seated about 1 m from the speaker, confirm acoustic features of speech in the frequency and intensity domains are available to the listener.

Speech Perception Assessment

Beyond detection, a CI recipient's resolution of sound should be assessed through speech perception assessment. For very young children, this can be done informally in collaboration with parents and therapists by observing the child's developing repertoire of speech sounds. An analysis of words and phonemes that are used in therapy, for example, while practicing speech babble (Estabrooks 1994; Pollack et al. 1997) may reveal a pattern of errors. This may suggest the need to alter frequency boundaries or stimulation levels that correspond to the formant frequencies of vowels and consonants.

In addition to ongoing informal assessment, a battery of speech perception tests should be used that is age appropriate and sensitive to changes over time. Preoperatively tests are used for candidacy determination. Postoperatively repeated measures will be useful to document progress and to identify therapy needs. Consensus on a standard pediatric test battery has yet to be achieved; it is important that tests are administered in a controlled environment and are appropriate for the child's age and development. Recorded test materials should be used when possible. Assessment should be completed on an annual basis or more frequently if there are concerns about progress or if changes to program settings need to be evaluated. A battery of age appropriate tests that assess hearing at the word and sentence level and in quiet and in noise is recommended. For a thorough discussion of speech perception measures and considerations in developing and implementing tests, see Chaps. 3 and 13.

Personalization of Settings

Nearly every generation and style of speech processor has different features that can be adjusted by or for the recipient to improve the listening environment. The audiologist can create programs employing various ways to filter and compress sound, add gain or volume, or change the sensitivity and directionality of the microphones. Multiple program slots on the speech processor provide the recipient with options to select a program with these various parameters adjusted. In theory, user-driven decision making to select a program for the purpose of optimizing listening is reasonable and desirable. However, because children are not able to make such decisions and manipulate settings in real time, the audiologist in conference with the parent should determine what program options are made available. Use of a remote control that can offer visual feedback to confirm processor settings is helpful. Newer technology incorporates algorithms that perform acoustic scene analyses that lead to automatic adjustments of microphone function and filtering of sound. To date, however, there have been no verification studies of the effectiveness of this technology with children. In truth, many children do not like their perception of sound to change. Selective use of these features should be introduced when the child is old enough to consider and evaluate sound quality. Until then, the audiologist can create programs based on knowledge of the child's typical environments that parents can select and use while monitoring the child.

Another way of personalizing program settings is to consider the use of various input sources in a program. Modern speech processors can be coupled to a variety of other hearing assistive technology (HAT) and assistive listening devices (ALDs) to enhance sound quality and to address the common problem of hearing in noise. Use of systems that carry frequency modulated (FM), digital modulated (DM), infrared (light), electromagnetic (telecoil), or blue tooth transmission to the processor can be very effective to improve listening in noise and from a distant sound source. Programs that mix the input from the speech processor microphone and another microphone or sound source can provide the child better access to music, video entertainment, and telephone use. Though important, verification of settings can be challenging so once again, a child who can provide feedback on sound quality and preferences is the better candidate to use these personalized options (Wolfe and Schafer 2010; Schafer et al 2013).

Patient Management Issues

For children, parents or guardians are central to success with a cochlear implant. They nurture the child's development by being competent and confident in managing the tool and by providing the environment and models necessary for children to learn spoken language. For some, navigating the cochlear implant hardware comes naturally; for others, it can be stressful and confusing. Being comfortable with and adept at device maintenance and troubleshooting, as well as adhering to a regular schedule of follow-up is helpful.

Device Orientation and Retention

Confidence and diligence in managing the external hardware are essential for the best outcomes after implantation. For children, parents bear the responsibility for device care and maintenance but children who are old enough to participate should be involved in the orientation to the device as well. Typically, the components and considerations for daily use are reviewed after the initial stimulation. Orientation to the device should include a review of these points:

- User controls on the processor and remote control
- Battery use—disposable and rechargeable, expected life,
- Program settings
- Device retention
- Troubleshooting—knowing who to call and when
- Warranty information and how to obtain replacement parts
- Contraindications to use
  - Medical contraindications including MRI and monopolar cautery
  - Considerations for protecting the implant and speech processor during sports activities, when traveling, around water

While all of these topics are important, device retention is often the immediate need to address after the initial stimulation. The speech processor and coil must be worn comfortably and securely. Parents should be counseled to check the magnet site on a regular basis to ensure the retention is not creating a pressure sore. Keeping the coil in place is probably the most common frustration for parents with a young child. While the connection should be adequate, too much compression of the skin could lead to the need to remove the processor for healing, which in turn delays the time to optimize program settings. When the speech processor is stabilized, through use of an earmold, mic-lock, huggie, headband, or double-sided tape, the coil is less likely to fall off or be removed by little hands. Many creative solutions are available; the audiologist can help the family explore these to find alternatives for various situations and activities.

Maintenance, Support, and Information

All manufacturers provide written manuals as well as electronic documents and demonstrations through their websites on how to use and care for the external hardware. A number of other publications have been developed to support families through the various stages of a child's acclimation and eventual reliance on the cochlear implant. The manufacturers have increased, expanded, and improved their recipient support over the years and so it is no longer necessary for the audiologist to be the interface between the patient and the manufacturer. However, expert advice and support remains an essential part of patient care.

External hardware must be maintained for the best performance. Normal wear and tear results in the need to replace all components at some point in time. Secure retention and safe storage will help minimize replacement of parts. Methodical inspection of each component should take place on a regular basis to ensure children have the best sound quality. Troubleshooting guides are available for all makes and generations of speech processors through the cochlear implant manufacturer websites. Spare parts, warranties, and service contracts to maintain hardware are essential for children.

Follow-Up Schedule

The first year following the initial stimulation includes frequent device programming visits to optimize the program and ensure audibility is maximized. The typical child adapts to the electrical signal over time; tolerance increases and as experience in hearing grows the child can play a larger role in providing feedback about hearing. At minimal, the following schedule is recommended for children:

- Initial stimulation (IS)
- 2 weeks post-IS
- 1 month post-IS
- 3 months post-IS
- 6 months post-IS
- 9 months post-IS
- 1 year post-IS
- Semiannual visits thereafter

A detection audiogram, as described previously, should be attempted at all visits occurring after the initial stimulation to guide programming decisions and validate settings. For most children, it should be possible to obtain an audiogram within 3 months of the initial stimulation. Ideally, a stable response can be documented by 6 months postinitial stimulation. If this is not the case, interim appointments should be scheduled to practice the conditioned response and obtain repeatable responses. As mentioned, speech perception tests should be completed after the audiogram as an ongoing record of progress and device stability. More comprehensive assessments, including speech and language and a speech perception test battery, should be performed at semiannual intervals to ensure progress in auditory and communication skills is being made as expected.

Typical and Atypical Outcomes

For a child with no other significant medical or behavioral issues who is implanted early and has appropriate intervention and family support, the potential outcome of cochlear implantation is age equivalent communication skills. However, there are many known and some unknown variables that may preclude this desirable outcome. External hardware must function properly at all times. Internal device issues are relatively rare but certainly can be the cause of a poorer than expected outcome.

Factors Affecting Performance

Factors that have been shown to be predictive of outcome are shown in Fig. 6.4 and include:

- aspects of the child, including anatomical, physiological, cognitive, developmental, and behavioral function;
- hearing history, including onset of hearing loss, degree and type of loss and age at diagnosis, and age at implantation;
- the use of technology, including the age at hearing aid fitting and cochlear implantation, consistency of use, and maintenance;
- the family and environment, including social, economic, cultural, commitment, resources, and goals;
- the educational and therapeutic methods used, including the consistency of therapy, the expertise and experience of therapist, family support for carryover and expansion of skills, emphasis on auditory learning, and integration of audition for communication.

Among these many factors that interact and interrelate, age at implant is the most remarkable variable that repeatedly correlates to performance outcomes. In long-term studies of various aspects of performance among large groups of children, age at implant has a pervasive impact on results. This is true for studies of speech and language development, speech perception ability, and electrophysiological studies. It also factors into results related to benefit from bilateral cochlear implantation and bimodal hearing device use (Eisenberg et al 2006; Niparko et al 2010; Sharma et al. 2002; Nicholas and Geers 2007; Ching et al 2014).

Diagnosing Soft and Hard Failures of the Internal Device

If concern about a child's hearing with a CI develops, the external hardware should be checked and replaced as needed as a first step. Indications that the internal components of the cochlear implant are faulty may be evidenced when monitoring electrode impedance and compliance values over time or by intermittency or lack of stimulation or changes in sound quality. Internal device malfunction should be considered for a child who fails to make progress in auditory skills development in the presence of other factors known to be necessary for success. The integrity of the internal device can be assessed through measures of device tolerances that are not available in the clinical software but can be evaluated using routines typically performed by a manufacturer clinical specialist. A hard failure is diagnosed when evidence of device malfunction can be documented in these ways or by obvious loss of communication with the internal device. Soft failures

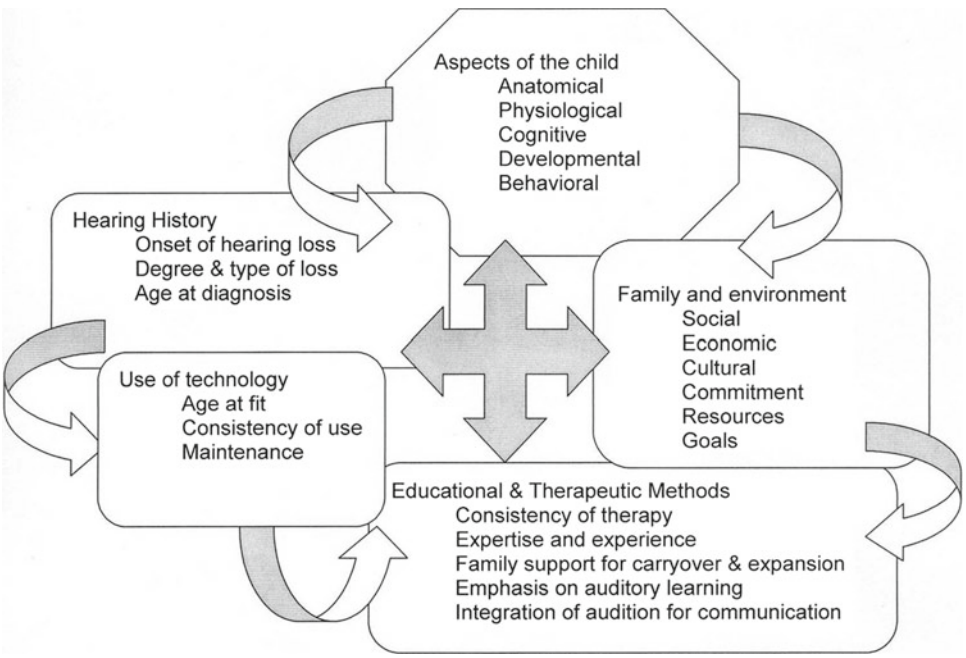

Fig. 6.4 Individual differences are the basis of variability in outcomes among cochlear implant recipients and there are a number of variables that lend themselves to infinite combinations of individual outcomes. Reproduced with permission of Taylor and Francis Group LLC Books (Teagle and Eskridge 2010)

are more difficult to determine as children may not be able to report on changes in sound quality or intermittency and the reference for a change in performance may not be well established. Validation of a soft failure may only be possible after cochlear implant revision. In general, cochlear implant survival rates are high among implantable medical technologies (Soli and Zheng 2010) and children typically tend to resume and exceed their level of performance after revision with time (Marlowe et al. 2010) (see Chap. 7.

Conclusion

The comprehensive management of pediatric cochlear implant recipients is an art and a science. The audiologist must draw on many aspects of professional training, including diagnostic skill and intuition, knowledge of the anatomy and physiology of hearing, psychophysical measurements, electrophysiological measures, speech acoustics, normal child development, and all facets and features of hearing technologies. Skills are further developed through experience with the wide variety of recipients and the unique aspects of their medical, social, and educational situations and environments. The practice of cochlear implant programming has evolved as the technology has become increasingly sophisticated and as the indications for use have expanded. The concept central to success for pediatric cochlear implant programming is to keep the unique aspects of the individual in mind. These are made known through the gathering of regular speech perception assessment and ongoing monitoring of speech and language development. It is also vital to work as part of a collaborative team and partner with parents to empower them with the knowledge and skills they need to facilitate the best outcome possible for the child.

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Diagnosis and Management of Cochlear Implant Malfunctions

7

Teresa A. Zwolan and Casey J. Stach

Introduction

One of the most important aspects of management of patients with cochlear implants (CIs) is monitoring of device function; failure to diagnose a malfunction could result in months or even years of reduced performance. The responsibility for this falls with the CI team as a decision to explant a device is often made based upon input from the surgeon, audiologist, and speech–language pathologist, and often includes input from the device manufacturer.

There are several reasons that a CI may be explanted. A fault of the internal device that results in immediate, insufficient auditory input to the patient is often referred to as a hard failure (Balkany et al. 2005). Such failures are easily confirmed by the device manufacturer and are rather straightforward in terms of their identification and rectification as they are manifested as an inability to communicate with the internal device, resulting in a complete loss of sound for the recipient. In these cases, explant with reimplantation is clearly indicated to return hearing to the recipient. Soft failure is a term that refers to CI malfunction interfering with clinical outcomes that cannot be proven with currently available means prior to explantation. Soft failures are often challenging to recognize because the recipient has improved hearing compared to preimplantation and many factors are known to affect growth of auditory skills. Soft failures may present with performance that unexpectedly plateaus or deteriorates over time, or is poorer than one would expect based on patient history. Unlike hard failures, manufacturer testing often fails to provide conclusive in vivo confirmation of device malfunction (Balkany et al. 2005). Identification of a soft failure is often challenging because of other non-device-related variables that may impact performance and rate of progress. Prior to recommending explantation with reimplantation of a new CI, it is important to evaluate, and where possible ameliorate, other factors that may be contributing to poorer than expected outcomes.

Fortunately, only a small portion of patients who receive a cochlear implant will experience a device failure. Reports in the literature regarding prevalence of device failures vary from a low of 1.9 % (Masterson et al. 2012) to a high of 14 % (Parisier et al. 1991). Additionally, many studies report a higher prevalence of failures among children than adults (Wang et al. 2014; Sunde et al. 2013).

Diagnosis of malfunction of the surgically implanted portion of the CI system typically begins with clinical documentation of signs that may be indicative of device malfunction such as changes in electrode impedances and inability to maintain consistent connection with the internal receiver, as well as reduced clinical benefit. When a device failure is suspected, the manufacturer is contacted and in vivo integrity testing is performed (see a later part of this chapter for a description of integrity testing). If the results of the integrity testing are inconclusive, definitive evidence of device malfunction may be possible only after the device has been explanted and a detailed analysis has been performed by the manufacturer. However, prior to recommending explantation for suspected soft failure, clinicians must also consider the possibility that reduced performance may be due to factors other than device malfunction and that reimplantation may result in no change or even a decline in performance. In rare cases, analysis of an explanted device from a patient with clinical improvement subsequent to reimplantation may not identify a cause of malfunction.

A review of the literature indicates that although some investigators report reduced speech recognition skills following reimplantation (Henson et al. 1999), most investigators report that patients perform at least as well after CI replacement when compared to performance with the initial device (Lassig et al. 2005; Balkany et al. 1999). Improved performance may occur for a variety of reasons including

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improvements in CI technology (internal or external) and improved electrode placement. Infants, young children, and those with developmental delays may be unable to participate in formal testing to document a change in performance after reimplantation. In all cases, the decision to replace a device should be made carefully. Patient performance with the existing device and the potential risk for reduced performance need to be considered. If replacement is not recommended, careful monitoring of CI function and performance is necessary.

There are several steps that can be taken throughout the cochlear implant process to assist with management and diagnosis of device malfunction. The following chapter will provide information regarding clinical procedures that can be used to assist with this important aspect of patient care.

Preoperative Procedures

Preparation for the possibility of device malfunction begins in the early stages of the implant process. Preoperative counseling should include discussion of the potential for device failure and discussion regarding reliability data for internal and external components. Additionally, clinicians should evaluate the outcome of various preoperative tests in order to develop an estimate of anticipated outcome. This evaluation includes the case history, imaging results, speech and language assessment, neuropsychological testing, and preoperative assessment of speech perception. Clinicians should review the results of such tests with patients and parents in order to develop a realistic estimate of expected performance. Such an estimate is valuable as it plays a strong role when considering the cause of atypical clinical responses. For example, it is not uncommon for a child who presents with an abnormal cochlea to have elevated psychophysical responses, increased pulse widths, and deactivation of several electrodes due to the presence of facial nerve stimulation (Francis et al. 2008). On the other hand, it is not typical for an implanted child with a normal cochlea to present with such symptoms. Therefore, in the latter situation, a higher index of suspicion is warranted regarding possible device malfunction. If such symptoms do occur in any recipient, the clinician should closely evaluate possible causes, including electrode position with the cochlea, the status of the mapping, changes in the child's medical condition, as well as device malfunction. Thus, the child's case history should be considered when clinical judgments are made regarding device function.

Preoperative counseling should also include discussion of the essential role consistent device use plays in performance, as well as participation in a program of habilitation that includes development of auditory skills. The importance of attendance at all postoperative appointments should be stressed as such appointments provide opportunities to monitor function of external and internal components and to evaluate and optimize patient performance. The recent introduction of data-logging software has made it possible for clinicians to obtain objective information about CI use. This data makes it possible for clinicians to better understand the degree to which device use may be a contributor to poorer than expected performance.

Perioperative Testing

Several objective measures are available that can be used either intraoperatively or postoperatively to verify gross function of the device. Objective measures that may be performed in the operating room include impedance telemetry, electrically evoked stapedial reflexes, electrically evoked auditory brainstem response, and electrically evoked compound action potentials (ECAP) (Hughes 2013). Performing such tests at the time of surgery provides a quick check of device function and provides objective information that can serve as a baseline for future comparison if concerns about performance or device function arise. In our experience, a radiograph to document the position of the electrode array within the cochlea is quite valuable. This type of imaging is associated with low radiation dosage. It may be obtained in the operating room or in the ambulatory setting. Our clinical practice includes obtaining a radiograph prior to device activation and again if there is reason to suspect device malfunction. The radiograph provides baseline information regarding electrode placement and may identify problems such as a kink in the array, a fold-over of the tip of the array, an over-insertion, or partial insertion of the electrode array (Fig. 7.1). The baseline radiograph may be helpful when interpreting subsequent radiographs done to determine if progressive electrode extrusion has occurred. Electrode extrusion is a problem that may be mistaken for device malfunction as it may cause changes in performance or psychophysical responses. Therefore, imaging is an important component of the evaluation process. In addition, early identification of electrode extrusion provides an opportunity to turn off the extra-cochlear electrodes which may lead to improved performance.

Postoperative Appointments with the Audiologist

Postoperative counseling should include the importance of monitoring the skin between the internal magnet and magnet within the headpiece. Parents should be advised to contact the CI center if they have concerns about changes in the skin overlying the receiver–stimulator or proptosis of the ear, the

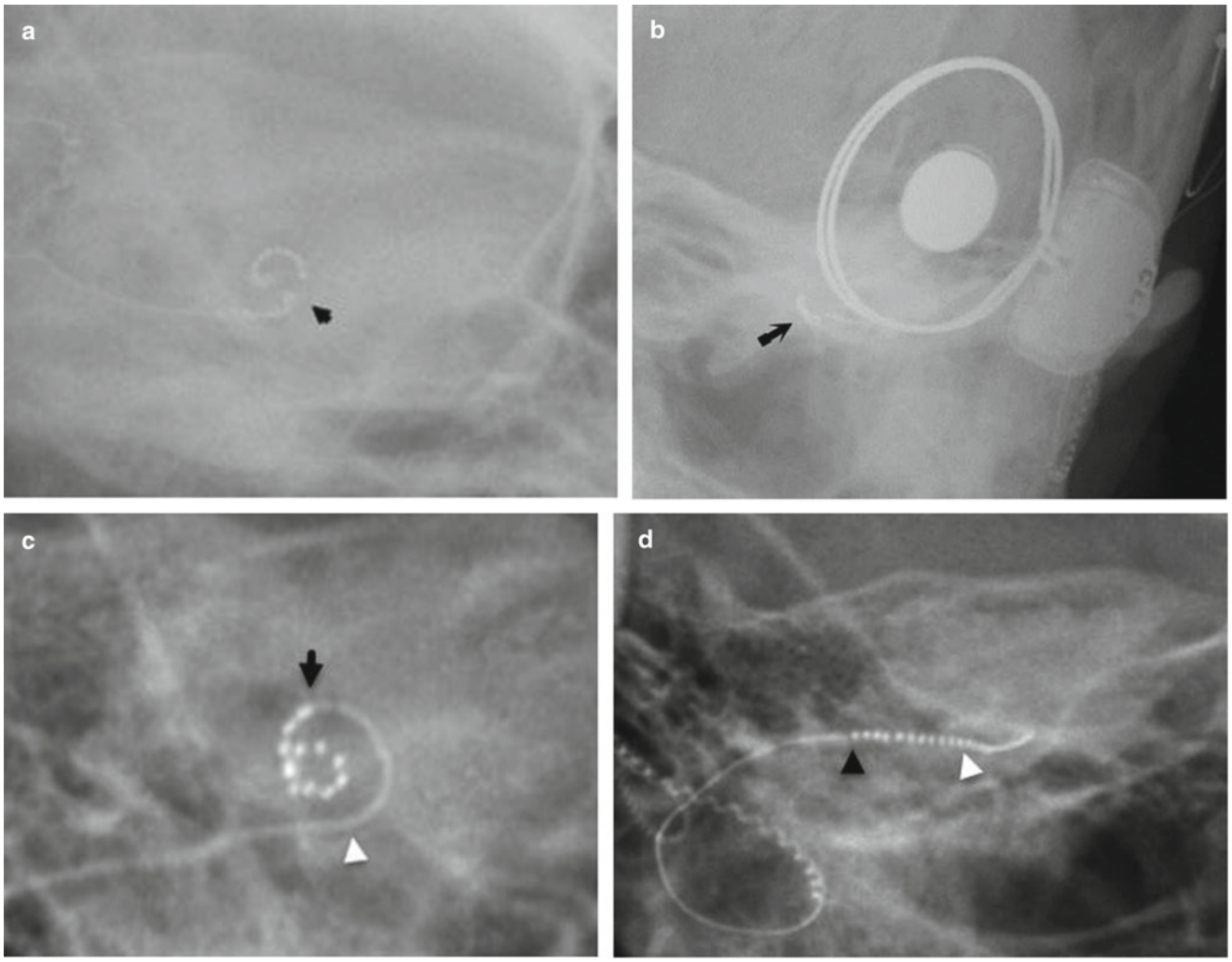

Fig. 7.1 Example of anterior posterior skull radiographs used to confirm placement of various electrode arrays. Although the exact point at which the array becomes intracochlear cannot be identified using this type of radiograph, normal positioning within the cochlea, general insertion depth, and status of the electrode array can usually be confirmed. Radiographs performed at device activation can also provide an important baseline for comparison with a repeat radiograph if shift in position of the electrode array is suspected. The following problems were detected by these radiographs: (**a**) a kink (*arrow head*) in the electrode array, (**b**) a fold-over (*arrow head*) of the tip of the array, (**c**) an overinsertion of the array, placing it deeper into the cochlea than desired, resulting in absence of electrodes in the proximal basal turn of the cochlea where high-frequency information is typically delivered (*white arrowhead* is placed at approximate entrance into basal turn of cochlea; the space between *arrows* signifies the area of the basal turn where electrodes are typically placed) and (**d**) partial insertion of the array (*white arrow head* at approximate location where array enters the cochlea; electrodes between the two *arrowheads* are outside the cochlea)

latter being a sign of infection within the mastoid causing swelling behind the ear. Device activation and subsequent appointments should include regular examination of the skin behind the pinna overlying the mastoid to look for possible signs of swelling or infection and to look for signs of irritation caused by use of the speech processor. The area beneath the magnet should be examined to ensure proper magnet strength as factors such as changes in weight or skin thickness can affect the strength of the magnet needed for proper adhesion of the coil. Occasionally, children experience trauma to the head that will affect the skin near the implanted device. When trauma to the skin near the implant does occur, a recommendation to temporarily stop using the device until the skin heals is necessary in order to prevent additional damage. Failure to address such issues in a timely manner may lead to infection and skin breakdown. These complications may require CI explantation.

At each appointment, external equipment should be inspected for proper function as issues with external equipment may mimic symptoms associated with an internal device malfunction. If undetected, external equipment issues may cause intermittent sound or reduced sound quality which can result in months or years of reduced performance. Inspection of external equipment should always include a

listening check of the speech processor microphone. All components should be carefully scrutinized, including the speech processor, battery, cable, coil, and magnet. Contemporary speech processors have the ability to alert the parent or recipient of component malfunctions. These alerts may include indicator lights on the processor or a visual display on the remote control that identifies the broken part. Correct use of these tools facilitates quick identification and remediation of external equipment problems.

Unfortunately, alerting systems will not identify all external equipment problems. One important example is dirt obstructing the microphone filter which may reduce sound quality. This problem may occur gradually over time resulting in progressive decrease in the recipient's ability to recognize speech. In order to detect malfunctions, parents of young children should perform a daily listening check of the speech processor microphone that includes evaluation of the child's awareness to speech sounds using the Ling Six Sound Test (Ling 2002). This test provides a quick assessment of a child's ability to hear speech sounds that range from low to mid to high pitch as detection of such sounds is essential for developing spoken language. Additionally, parents should visually inspect all speech processor components daily. Microphone filters should be changed at the recommended time intervals, and recommendations provided by the device manufacturer regarding regular maintenance of the device should be followed.

At the beginning of each appointment, the clinician should question the patient/family about the child's consistency of device use, discuss any problems encountered by the patient, inquire about any changes in performance, and review data logs of device use, if available. Such logs provide important information regarding daily average time of device use, which program has been used most often, the sound environment the recipient is most often in, which volume settings are most often used, and number of times the recipient has experienced signal intermittency. If feedback from the family or data logging raise concerns about device use, the family and child should be questioned further to determine the reason for such problems. For example, recent head trauma, a change in health status, or the person supervising the child or equipment problems may impact device use. This discussion may help the clinician and family to better understand the reason for inconsistent CI use and to develop a plan to address the problem.

Impedance Telemetry

Programming of all contemporary speech processors typically begins with impedance telemetry. This test provides information about the properties of the tissue surrounding the electrode array, determines if appropriate current is being delivered by each electrode, and alerts the clinician when problems with the electrodes occur, such as open or short circuits (Hughes 2013). Programming software alerts the clinician when impedance values reach a level indicative of either an open or short circuit. It is important to identify these problems so that the affected electrode(s) may be removed from the patient's map. Open circuits are indicated by high impedance values and may result from a variety of issues, such as a break in the lead wire or a damaged electrode contact, air bubbles around the electrode contact, and electrodes that may be positioned outside of the inner ear (due to incomplete insertion or delayed extrusion) (Carlson et al. 2010). Short circuits are represented by low impedance values that typically occur when two or more intracochlear electrodes share a common electrical course (Carlson et al. 2010). Partial short circuits may also occur, meaning that impedances decrease over time but fail to reach a value that will be flagged by the software as a short circuit. Impedance telemetry measurements are not available with early devices, such as the Nucleus 22 (Cochlear Corporation) and Clarion (Advanced Bionics Corporation) CI systems. With these older devices, electrodes with open or short circuits are often identified during psychophysical testing; electrodes with an open circuit typically demonstrate no response to stimulation while electrodes with a short circuit typically demonstrate static, buzzing, or intermittent sound. When this occurs, the electrode in question should be deactivated as they have the ability to create erratic electrical fields (Mens 2007).

It is important for clinicians to perform impedance testing prior to each programming session and to evaluate possible changes in impedance values over time. Typically, impedance measurements will be lowest at the time of surgery, increase slightly following surgery, decrease following device use, and stabilize after a few weeks or months of device use (Hughes 2013). When performing impedance testing, the clinician should be aware of electrodes that have been eliminated from the patient's map, as impedance values typically increase when an electrode has not been stimulated. Thus, in the case of a deactivated electrode, an increase in impedance value of the deactivated electrode is not cause for concern. Impedance results should be evaluated for changes over time, as atypical patterns may indicate problems with an influx of fluid into the array that results in either zig-zag, flat low, or random impedance patterns (Cullington 2013; Zwolan et al. 2012). Abnormal findings on impedance testing may also be indicative of issues with the tissue environment surrounding the electrode array (Hughes 2013), such as when cochlear ossification is present or when the recipient has structural abnormalities of the cochlea. Thus, clinicians need to be aware of conditions in the cochlea that may affect impedance test results. Examples of various electrode impedance test results for various devices are provided in Fig. 7.2.

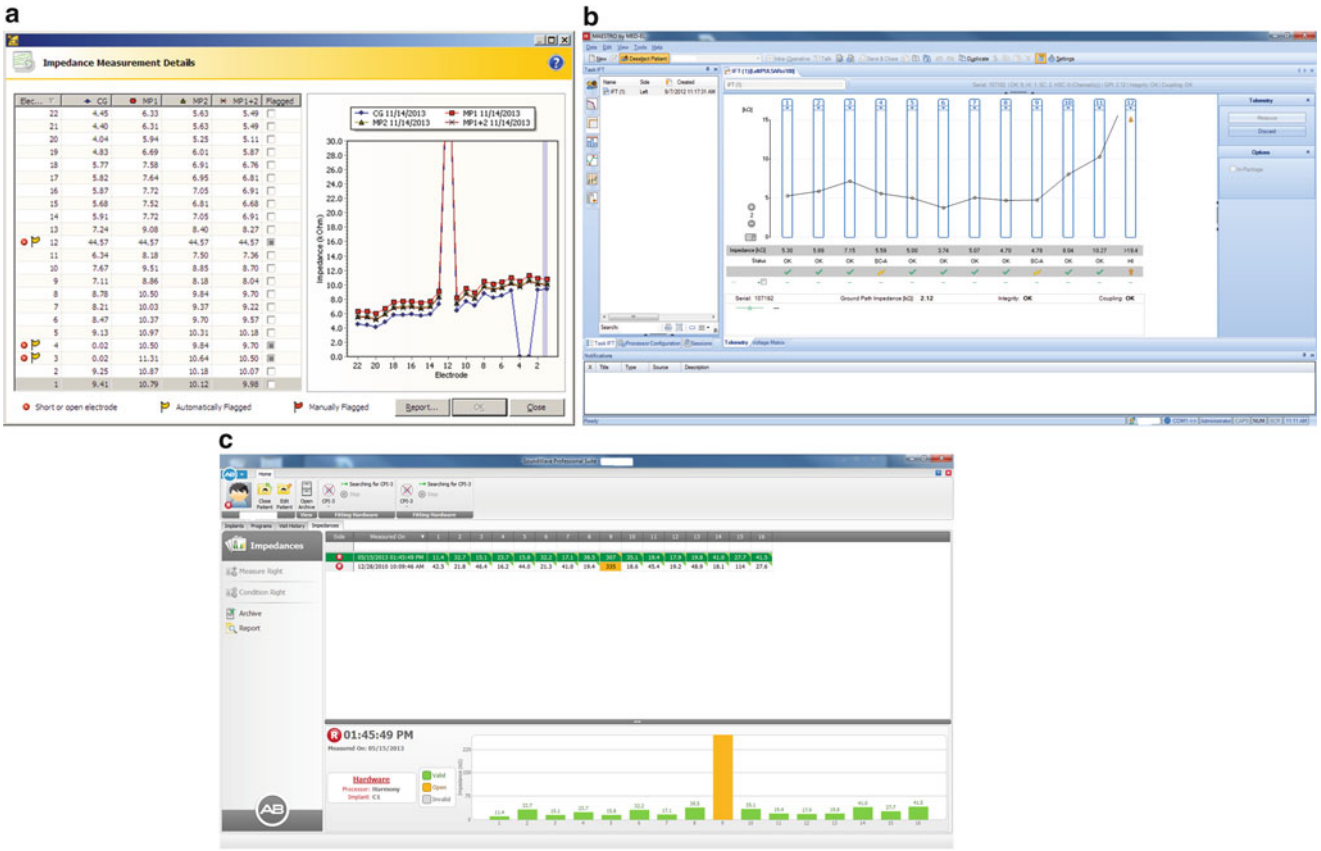

Fig. 7.2 Electrode impedance test results demonstrating open electrode for the following device models (**a**) Nucleus CI24RE (Cochlear Corporation), (**b**) Pulsar (Med-EL Corporation), and (**c**) HiRes 90 K (Advanced Bionics Corporation)

Speech Processor Programming

Speech processor programming involves the setting of numerous parameters by the clinician followed by assessment of various psychophysical measurements. The two most commonly obtained psychophysical measurements include threshold (T) and comfortable (C) or most comfortable (M) levels. Each measurement is obtained for electrodes included in the patient's map. The precise definition of threshold varies for the different manufacturers (Wolfe and Schafer 2010) but typically refers to the lowest level of stimulation resulting in detection of the electric signal. The C or M level also varies depending on device, but typically refers to the upper level of stimulation and represents either a loud but comfortable (C) or a most (M) comfortable loudness level.

In most clinics, patients are typically seen 6–10 times during the first year of device use for programming of the speech processor. The schedule for programming children typically followed at our facility is provided in Table 7.1. During these appointments, threshold and C/M levels are reassessed. Typically, threshold levels decrease slightly over time while C/M levels increase as the patient is able to tolerate louder sounds provided by the implant. T and C/M levels typically stabilize following about 12 months of device use and, in most cases, remain stable for years (Zwolan et al. 2008). After 1 year of device use, our clinic recommends adult patients be seen once a year and that pediatric patients be seen twice a year (or more often if needed) to reassess and modify threshold and C/M levels and to evaluate speech perception. If levels are not set correctly or are not adjusted when needed, a patient may fail to reach their full potential for understanding speech with the device. Additionally, abnormally high, abnormally low, or unexplained changes in threshold or C/M measurements may be indicative of a problem with the device. Changes in behavioral responses should be compared to changes in impedance telemetry measures and also compared to baseline objective responses (i.e., ECAP thresholds) obtained intraoperatively or at device activation. It is important for clinicians to routinely monitor the stability of threshold and C/M levels. Changes in these levels may be a sign of device malfunction. The reader is referred to Zwolan et al (Zwolan et al. 2008) for information regarding typical T, C, and M levels.

Table 7.1 Schedule for Evaluation and Programming by Audiologist for Cochlear Implant Recipients at University of Michigan

Adult recipients	Pediatric recipients
Activation	Activation
1 week post	1 week post
2 weeks post	2 weeks post
1 month post	1 month post
3 months post	3 months post
6 months post	6 months post
12 months post	9 months post
Annually thereafter	12 months post
	Every six months thereafter

Increases in T or C/M levels may necessitate changes to other mapping parameters, including an increase in pulse width, reduction in overall pulse rate, reduction in maxima, or increases in power level. Changes in these parameters may affect the quality of the sound reported by the recipient and may result in reduced speech recognition. Thus, if large mapping changes are made, it is often wise for the clinician to reevaluate speech perception ability following a period of use to determine if and how the changes have affected performance.

Increases in electrical levels may result in some electrodes causing nonauditory percepts which may include stimulation of the facial nerve. Older children and adults may report abnormal sensations. However, often young children and those with limited language exhibit behavioral changes but do not directly report what they are experiencing. Therefore, these types of problems are often more difficult to diagnose in young children. For this reason, the child's face should be closely watched during psychophysical testing to identify whether stimulation of an individual electrode causes facial nerve movement or aberrant behaviors due to abnormal/unpleasant sensations. Facial nerve stimulation (FNS) typically presents as a twitch around the eye, mouth, or neck on the side of the implant and may or may not be uncomfortable for the patient. It most often occurs in patients requiring high levels of electrical stimulation, such as those with abnormal anatomy or cochlear ossification (Francis et al. 2008). Although rare, FNS may also be observed in patients with normal cochlea. It may be possible to alleviate problems associated with FNS via manipulation of pulse width, by reducing the level of stimulation, by changing the rate of stimulation, or by deactivating the electrode(s) that is causing the response. When FNS affects several electrodes imaging is necessary to evaluate the position of the electrode array. Active electrodes lying within the internal auditory canal, middle ear, or facial recess may cause FNS.

There are symptoms that older children and adults may report that may indicate a problem with the internal device. Such symptoms should always be investigated. These symptoms may include changes in loudness growth, reduced pitch discrimination across electrodes in the array, elevated or fluctuating threshold or C/M levels, intermittent popping sounds, presence of constant static, pain or discomfort at the implant site, and unexplained decrease or fluctuation in sound field detection thresholds. As stated previously, speech recognition skills with a CI should be stable and should not decrease over time. Therefore, reductions in speech recognition scores should always be investigated.

Symptoms associated with device malfunction are particularly difficult to diagnose in young children. Changes in behavior such as sudden refusal to wear the device, reports of reduced hearing, reduced speech intelligibility, or reduced performance at school should be cause for careful examination of all possible contributing factors. This evaluation should include examination of external equipment, a review of the child's daily routine related to device use at home and at school including consultation with the child's educators and speech–language pathologist regarding device use and performance. Evaluation should include auditory thresholds with the CI, speech perception, and speech and language skills. Impedance telemetry should be measured and speech processor programming evaluated and modified as deemed necessary. If the child is not currently enrolled in aural habilitation or speech–language therapy, it may be beneficial to enroll him/her in such therapy for a brief period of time to determine if performance improves with intervention. If concerns persist, an in-depth evaluation of the device which will likely include integrity testing by the manufacturer should be performed. Prior to integrity testing a medical evaluation performed by the CI surgeon and imaging to ensure proper positioning of the electrode array is often beneficial.

With patients who receive bilateral implants, a significant difference in the perceived quality of sound between the two ears may signify a problem with the inferior-sounding device. This situation can be particularly difficult to diagnose in a child who receives a sequential bilateral implant following years of unilateral device use. In our clinic, we have encountered three adolescents who reported poor sound quality in the ear that had been sequentially implanted following more than 10 years of profound loss without auditory stimulation. Our initial impression was that the reduced sound quality was due to auditory deprivation in the later implanted ear. Further evaluation was undertaken after these recipients continued to report poor sound quality and persistent static despite months to years of device use. Telemetry and integrity testing did not conclusively indicate device malfunction but a recommendation to explant the device was made based on the presence of consistent poor sound quality. In all three cases, explant with reimplantation of a new device resulted in improved sound quality, elimination of static, improved speech recognition skills, and consistent device use. Additionally, analysis of all three explanted devices confirmed malfunction of the receiver-stimulator.

Thus, if we had failed to question the functionality of the internal device, we would have missed the opportunity to provide valuable binaural hearing to these three adolescents.

Conversely, another of our recipients received his first implant at age 2 and his second implant at age 12. He performed significantly better with the second ear, despite its longer period of auditory deprivation. This outcome was unexpected and inspired us to closely evaluate the first device. Although manufacturer integrity testing and psychophysical responses appeared normal, we recommended reimplantation. The patient demonstrated greatly improved speech recognition skills after receiving a new CI. Manufacturer evaluation of the explanted device confirmed the presence of device malfunction. These cases demonstrate the complexity of diagnosing suboptimal device function that merits CI replacement in a recipient whose hearing has been improved by their CI. Astute clinicians should listen to the child and caregivers, seek input from other professionals who work with the child, maintain records of telemetry and speech processor programming, and consistently evaluate speech recognition and speech and language ability over time. Doing so will increase the likelihood that clinically relevant internal device problems are identified and rectified, making it possible for CI recipients to meet their full potential.

Postoperative Assessments

While preoperative assessment of speech recognition provides a baseline for postoperative comparison (and helps determine expected levels of performance), regular sound field detection thresholds, speech processor programming, and regular speech perception and speech–language testing are essential tools for monitoring the function of an internal device. Such tests help determine if the patient is demonstrating appropriate detection skills, if he/she meets expected levels of performance, if performance has decreased over time, or if reasons other than a device malfunction can explain why performance is poorer than expected or has declined.

Speech Detection Thresholds

Sound field detection threshold assessment should be performed regularly to evaluate the patient's ability to detect sound with the speech processor. Such testing ensures the device is providing the patient with access to important speech information and should include functional gain assessment for test frequencies ranging from 250 to 4000 Hz. Patients with contemporary devices typically respond at approximately 15–20 dB HL at these frequencies, but responses may be poorer in certain circumstances including some anatomically abnormal cochlea. The sound field detection thresholds should be stable over time. Responses that are worse than expected can signify a problem with the external equipment, may indicate a need for reprogramming, or may be indicative of a problem with the internal device.

Speech Perception

It is important for clinicians to understand factors that will influence performance so appropriate expectations can be set. It is also important for clinicians to understand the typical progression of postimplant speech recognition so that deviations from the norm can be recognized. Typically, if children are provided with appropriate therapy and implanted early, children with normal cochlea or isolated wide vestibular aqueducts and normal cognitive skills demonstrate great gains in their speech perception and speech–language skills. Children and adults should demonstrate either stable performance or improvements over time in regards to their speech perception skills. Decrements in speech perception may indicate device malfunction and should always be cause for concern and further evaluation.

With both children and adults, performance with the implant is influenced by several factors, including age at onset of deafness, age at implantation, age at identification of hearing loss, parental involvement, etiology, status of the cochlea, communication mode, preoperative residual hearing, nonverbal cognition, and parental socioeconomic status, to name a few (Connor et al. 2006; Yanbay et al. 2014; Varga et al. 2014; Freeman et al. 2013; Miyamoto et al. 1994; Harrison et al. 2005; Cowan et al. 1997; Fryauf-Bertschy et al. 1997; Sarant et al. 2001; Dowell et al. 2002). Additionally, the specific measures used to evaluate performance vary depending on the patient's age, language level, and listening skills. For both adults and children, the results of postoperative testing can be compared to the individual's preoperative scores to ensure improved performance. Performance that is poorer than preoperative with hearing aids is extremely rare and causes including device malfunction should be considered. Performance should also be continuously compared to expected outcomes based on the preoperative medical, radiological, and audiological assessment and case history.

Speech perception testing is essential for monitoring device function. Such testing should be performed at least annually and should include standardized tests that are presented in an auditory only setting while the patient utilizes the CI. Such testing is typically performed in a sound booth with recorded test stimuli presented at a level of 60 dB SPL (Firszt et al. 2004). For children, hierarchies of listening

skills are assessed, beginning with the simple task of detection. Testing then progresses to evaluate discrimination, identification, and comprehension of speech (Kirk and Choi 2009).

Test batteries may include both closed- and open-set tests, depending on the child's demonstrated ability. Closed-set tests provide the listener with a set of possible responses. The chance score for a closed-set test will vary depending on the number of choices. Often, closed-set tests are tailored to the vocabulary and language levels of small children and are also good to use with adults who demonstrate very limited speech recognition skills. Closed-set measures used with children include Early Speech Perception Test (ESP) (Moog and Geers 1990), Pediatric Speech Intelligibility (PSI) test (Jerger and Jerger 1984), Word Identification by Picture Identification (WIPI) (Ross and Lerman 1979), to name a few. Closed-set measures used with older children may include vowel or consonant recognition measures or various measures of the Minimum Speech Test Battery (Luxford and Ad Hoc Subcommittee of the Committee on Hearing and Equilibrium of the American Academy of Otolaryngology-Head and Neck Surgery 2001). Open-set tests are more difficult than closed-set tests as they do not provide the listener with a set of possible responses. Open-set tests used with children include the Multi-Syllabic Lexical Neighborhood Test (MLNT) and the Lexical Neighborhood Test (LNT) (Kirk et al. 1995), HINT-C sentences (Nilsson et al. 1994), CNC Monosyllabic Words (Peterson and Lehiste 1962), and Pediatric AZBio Sentences (Spahr et al. 2014).

Both open- and closed-set tests can be presented in quiet or in the presence of background noise. Administration of test materials in noise increases the level of difficulty for the listener and should be considered if the patient demonstrates a ceiling effect on selected measures presented in quiet. Presenting stimuli in the presence of noise may provide additional information regarding the impact that various changes, such as changes in mapping parameters or removal of electrodes, has on performance. Testing in the bilateral or bimodal (CI plus hearing aid in opposite ear) can provide valuable information, especially when testing hearing in noise.

The Role of the Speech–Language Pathologist

The speech–language pathologist (SLP) who has experience with auditory skill and spoken language development of implanted children may be very helpful in the diagnosis and management of device malfunction. Preoperatively, the SLP helps determine the expected levels of postoperative performance based on information obtained during the case history and based on the child's speech and language skills prior to implant. Postoperatively, children seen in our clinic are encouraged to enroll in weekly speech/language therapy with one of our Auditory-Verbal/LSLS certified speech language pathologists. This enables them to have access to a trained professional who closely monitors their ability to make use of the sound provided by the CI, serving both a diagnostic and a therapeutic purpose. Information obtained by the SLP during therapy is shared with the child's CI audiologist. Their input helps to guide speech processor mapping and to identify children who are not progressing as expected. The SLPs input often assists the audiologist in uncovering a problem with the child's CI system.

Postoperatively, the SLP on the CI team evaluates the speech–language skills of the child and compares performance to that obtained by other similar children with implants as well as to the child's same-aged peers with normal hearing. These evaluations typically include assessment of voice quality and intelligibility, receptive and expressive vocabulary, receptive and expressive language, and reading and auditory comprehension skills. Such evaluations can be combined with the results of speech perception testing to obtain information about the benefits the child receives from using the device. Such outcomes should be compared to preoperative performance and to the expected outcome to determine if the device appears to be functioning properly. Performance on the speech–language measures should improve with time and plateaus or decrements in performance should not occur. If plateaus or decrements do occur, device function should be questioned as the cause or a possible contributor. SLPs experienced in working with young children who receive CIs are also adept at identifying other problems that may benefit referral to occupational therapists, physical therapists, and child psychologists. Such referrals may help the implant team determine if other factors may be contributing to less than optimal performance with the device.

Medical Evaluation

Concerns about device placement, infection, and the health of the skin overlying the lead wire or receiver–stimulator should be brought to the attention of the implant surgeon. In addition, if a recommendation is made to explant a failed device or one suspected of malfunction, the surgeon will discuss surgical risks with the family.

Manufacturer Integrity Testing

When a clinician suspects a problem with the internal device, it is essential that the device manufacturer be contacted. The manufacturer may be aware of other cases where patients

Table 7.2 Clinical tests performed by manufacturers to evaluate the function of an implanted device

Advanced bionics	Cochlear Americas	Med-El
Electric field imaging evaluates current flow, helps detect open or short circuits	*Intermittency* presents continuous stimulation, enabling the tester to check for missing pulses, dropped pulses, or unexpected variations in the waveform	*Impedance and Field Telemetry (IFT)*—comprehensive evaluation of implant function. Verification of the external to internal coil coupling, assessment of short circuits between electrodes, and identification of open circuits
Bionic ear integrity test evaluates device functionality such as amplitude growth and pulse width modulation	*Common ground, bipolar, pseudomonopolar, monopolar1, and monopolar 2* These tests check the function of the electrodes using various modes of stimulation	*Extended expert telemetry mode*: adds the capability to carry out measurements with differing instructions in order to observe commensurate changes in output from the implant. This mode also provides the ability to observe electrode impedances in real time over a period of time to observe intermittency
Surface potential test is a far field measure of the ICS and also evaluates electrode function	*High and low rate RF* presents stimulation at high and low stimulation rates to check if the receiver–stimulator functions normally. The RF transmission range is additionally tested	*Examination of voltage table* assesses the electronics and communication between the device and the DIB coupling
Capacitor test measures current leakage across the DC output capacitors on the electrodes	*RF Power Up* determines how quickly the implant will power up after a series of RF pulses are applied	
Link test evaluates the integrity of the electrical connection between the internal and external device and determines if there is sufficient energy to power the device at all times	*Current level* checks the response of the receiver–stimulator to varied current levels	
	Pulse width checks the response of the receiver–stimulator to varied pulse widths	

have presented with similar symptoms, and they may be able to provide suggestions that will help resolve the issue. In addition, if the problem cannot be resolved by the clinic, the device manufacturer has specialized equipment needed to perform integrity testing of the internal device. Typically, a representative trained in such testing will travel to the implant clinic to perform the integrity testing. Such testing involves placement of surface electrodes on the patient's head in order to record the responses generated by the internal device. Tests included in the various integrity test batteries are summarized in Table 7.2.

The results of integrity testing may be helpful in determining if device explant is warranted. Occasionally, results of the integrity test may be used to guide speech processor mapping. For example, electrodes may be identified that should be deactivated, thereby removing them from the patient's map. When this occurs, performance should be closely monitored to determine if changes improved performance. With small children, aural habilitation and speech–language therapy provided by an experienced therapist can be helpful to closely monitor of progress after map changes are made, as results of formal speech perception testing may yield limited information. In some cases, the implant team may recommend explantation of a device based upon the overall clinical presentation, despite integrity testing that does not identify device malfunction.

Device Explant/Reimplant

If the patient has experienced a hard failure of the device, the decision to recommend device explant followed by reimplantation of a new device is usually straightforward. The decision to explant a device that is functioning and has limited or no measurable evidence of malfunction (soft failure) should only be made following careful consideration. In most cases, device explant/reimplantation can be done safely and successfully. Risks associated with this procedure are typically similar to those of a primary implant surgery. Although rare, complications such as inability to fully remove the electrode array have been reported (Kang et al. 2009). In addition, factors such as unusual anatomy including risk of cerebral spinal fluid leak and other underlying medical conditions may be important considerations in some cases. Although most investigators report improved or stable performance following reimplantation, some investigators have reported a reduction in performance with the subsequent device (Henson et al. 1999). Therefore, the possibility of diminished benefit following explant of a device that is functioning must be considered. In addition, parental perception of past implant benefit and financial considerations may influence parental decision making once a recommendation to explant a device has been made.

In the United States, clinics are required to report device failures to the Food and Drug Administration (FDA). The FDA maintains a publicly accessible database of cochlear implant adverse events called the Manufacturer and User Facility Device Experience (MAUDE) (Manufacturer and User Facility Device Experience Database (MAUDE). http://www.fda.gov/MedicalDevices/DeviceRegulationandGuidance/PostmarketRequirements/ReportingAdverseEvents/ucm127891.htm Accessed 19 Nov 2014). Additionally, clinics return explanted devices to the implant manufacturer so analysis can be performed and the cause of the failure determined. Such information is used by device manufacturers to improve the reliability of future devices. The manufacturer typically classifies the device failure as (1) confirmed (the device failed one or all of the tests performed as part of the failure analysis), (2) a failure of unknown origin (device failed the in situ test but passed the postexplant failure analysis), (3) other reason for explant (device passed the in situ test but was explanted for a clinical reason and performance improved following reimplantation, or (4) medical reason for explant (such as infection, extrusion, improper placement of the original device, or electrode migration, to name a few).

In our clinic, activations following reimplantation for device failure typically occur 1–2 weeks after surgery, compared with 4 weeks after a primary implantation. Expediting the activation in order to decrease the time the child is without sound is often possible as the second surgery typically involves less drilling and reduced healing time than the first surgery. Expediting this appointment is especially valuable when the child does not have an implant in the contralateral ear.

Reimbursement Issues

Manufacturers of currently available devices provide comprehensive warranties for both external and internal cochlear implant components. All three manufacturers of commercially available CI systems offer a 10-year warranty for the internal device that begins the day the device is implanted. If the device fails prior to expiration of the warranty, the manufacturer will provide a new internal device at no cost to the implant center or patient. If a new model of implant has been introduced since the patient received the first device, the manufacturer may provide the updated model and may also provide the most current model speech processor free of charge. It should be noted that the second device is only warranted until the end of the 10-year warranty that came with the initial device. Typically, the implant center will preauthorize the costs associated with surgical explant/reimplant of the device with the patient's insurer (with the cost of the internal device excluded if the failure occurs while the device is under warranty). In some cases, the manufacturer may provide financial assistance to the patient if such costs are denied by the insurer. Each manufacturer has employees who specialize in working with patients and their families to assist with the aspects of care when a device malfunction occurs.

Increasing Awareness and Consensus

There are several groups that have been formed to assist with terminology and to recommend procedures that can be used to evaluate CI device failures. In 2005, a panel of experts from across the United States representing the fields of audiology, otolaryngology, speech–language pathology, communication science, and engineering was organized to prepare a consensus statement regarding CI soft failures (Balkany et al. 2005). Around the same time, 11 international experts from across Europe organized a group called the "Global Consensus Group on Cochlear Implant Reliability." This group proposed an international classification of reliability for implanted CI (Battmer et al. 2010). Presently, the United States Food and Drug Administration (FDA) is working with the Association for the Advancement of Medical Instrumentation (AAMI) to bring together device manufacturers, otolaryngologists, audiologists, and engineers to publish a document that summarizes output characteristics and performance requirements of CIs (AAMI/CDV-1 CI86, Cochlear implant systems -Safety, performance and reliability 2015). This document was submitted for public review in May 2015 and will be published as "*Cochlear implant systems - Safety, performance and reliability"*—a voluntary standard and recommended practices related to CIs. This document is important as it represents the first published U.S. standard and will include items such as guidelines for manufacturers to follow when evaluating and reporting device failures, and recommended procedures for clinicians to follow when questioning the integrity of an internal device.

Conclusion

Great strides have been made in recent years in regards to diagnosis and treatment of CI device failures. In many cases, diagnosis of the failure is complex, rarely straightforward, and requires input from multiple professionals, including the surgeon, audiologist, speech–language pathologist, educator, parent, and device manufacturer. Although rare, failure of a cochlear implant can have severe ramifications for the recipient, particularly if the failure goes undetected for long periods of time. Definitive diagnosis of hard failure as well as well carefully considered determination of soft failure often leads to the recommendation that a new CI be surgically implanted.

In most instances, this action will return recipients whose device had completely failed to at least their prior level of CI performance. Those individuals with soft failures often experience resolution of their complaints and improved performance. Of course if the diagnosis of soft failure was incorrect and other nondevice issues underlay poorer than expected performance, the results will likely be disappointing. In addition, there is a small risk that reimplantation will not be successful and that performance may be reduced. Therefore, a decision to explant should only be made following careful consideration of all of the risks associated with revision surgery, and only after the professionals involved have exhausted all other options to try and improve performance.

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The Role of Electrophysiological Testing in Pediatric Cochlear Implantation

8

Karen A. Gordon

Recording Electrophysiological Measures

Although electrophysiological measures are commonplace in audiology clinics globally, and are a central component of the test battery for identification of hearing loss, they are not as widely used in the management of CI recipients. Their underutilization in CI settings occurs despite the long-standing availability and knowledge regarding these techniques as objective tools for assessment of function in CI recipients (Kileny et al. 1997; Shallop 1993). One explanation may be concerns regarding equipment setup and troubleshooting. For this reason, this section will outline some key features to keep in mind as you attempt to record and analyze electrophysiological responses from children using CIs.

Neural Generators of the Recorded Responses

Electrophysiological responses are generated from groups of neurons responding in synchrony. Depending on the alignment of the neurons, the individual responses will sum into an electrical field sufficiently large to be detected and recorded some distance away in a process known as electroencephalography (EEG). The electrophysiological tests and interpretations of the recorded response will be limited to the populations of neurons that are stimulated and recorded. This principle is no different when stimulating through the CI as when acoustic sound is used to evoke electrophysiological responses. Responses from the auditory nerve and brainstem are generated by neurons aligned in the same orientation. An acoustic click-evoked auditory brainstem response (ABR) measured with surface recording electrodes on the head from a 7-year-old child with normal hearing is shown in Fig. 8.1a. Amplitude peaks emerge over time after stimulus onset (latency) as neural activity ascends through the auditory nerve (waves I and II) to the cochlear nucleus in brainstem (wave III) and up the lateral lemniscus (waves IV and V) to the midbrain (reviewed in Chap. 4, (Picton 2010)). The electrically evoked auditory brainstem response (EABR) from a 2 year old with 9 months of unilateral CI use in the right ear, shown in Fig. 8.1b, has similar waves, although the earliest peak, wave I, cannot be seen because of the large stimulus artifact in the early part of the recording time and wave IV is not clear. Often, the EABR amplitude peaks are larger and sharper than the ABR peaks. These differences are likely due to an increase in neural synchrony in the brainstem driven by the electrical pulses from the CI compared to acoustic clicks (van den Honert and Stypulkowski 1986). The EABR waves occur at earlier latencies than the ABR because the electrical pulse stimulates the auditory nerve directly whereas acoustic sound must travel through the ear. This acoustic delay is measured by the latency of the ABR wave I as shown in Fig. 8.1a (~1.5 ms). As will be discussed in more detail in the section, "*Minimizing CI stimulus artifact,*" CI telemetry systems employ effective methods to remove stimulus artifact so that the homologous response from the auditory nerve may be recorded. This electrical compound action potential (ECAP) response, recorded by electrodes along the CI array, is shown in Fig. 8.1c. It is a biphasic response with the earliest amplitude peak occurring at ~0.30 ms—very rapidly after the onset of the electrical pulse delivered by the CI.

Whereas earlier latency responses are generated from the auditory nerve and brainstem, responses at latencies 10–50 ms (middle latency responses, MLRs) and later latencies (>50 ms) have thalamo-cortical generators. As shown in Fig. 8.2, responses recorded at the vertex position on the head (termed Cz) can be very similar in both children using CIs and normal hearing peers for both middle and later latency responses. The examples in Fig. 8.3a are of immature and mature middle latency responses and exam-

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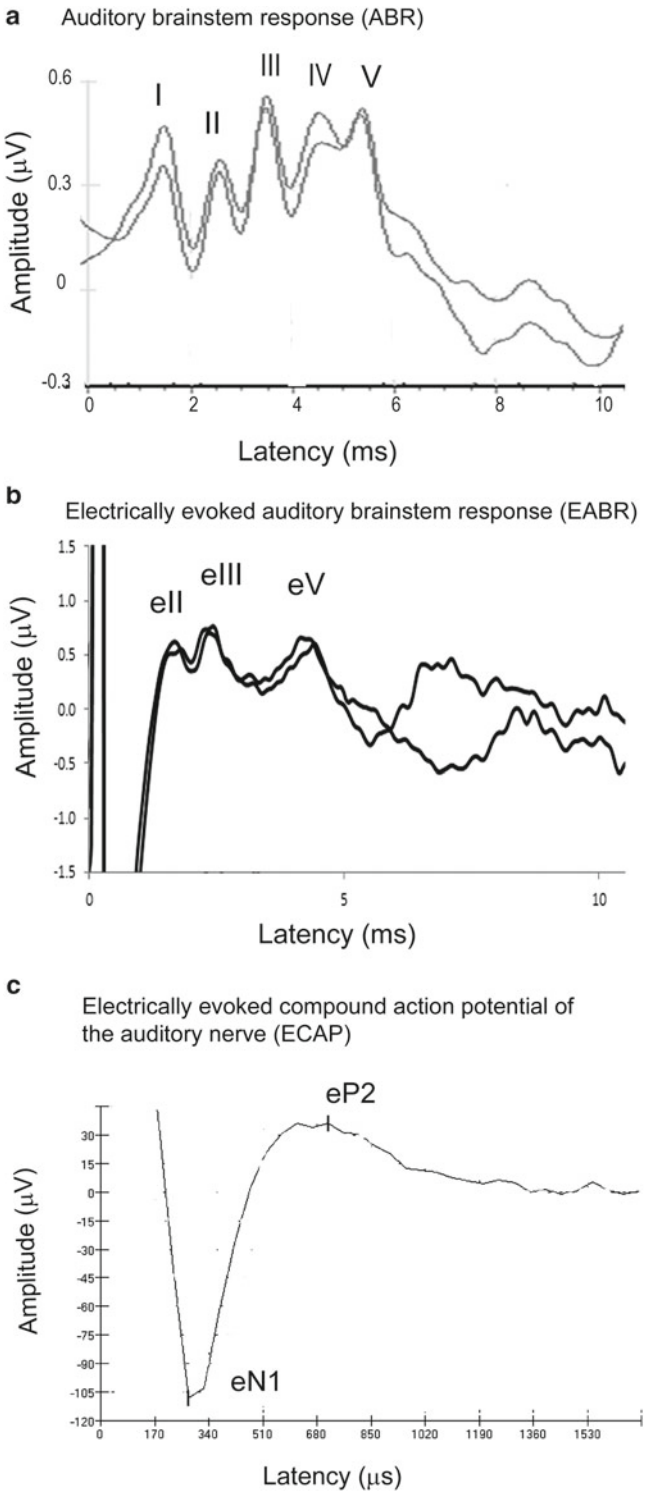


Fig. 8.1 (**a**) An example of an auditory brainstem response (ABR) measured from the centre mid-line of the head (Cz) referenced to the ipsilateral earlobe from a 7-year-old child with normal hearing. Clicks were presented at comfortably loud levels. The five characteristic peaks of the response are indicated. (**b**) The electrically evoked auditory brainstem response (EABR) from 2-year-old child with 9 months of right CI experience (recorded at Cz referenced to the ipsilateral right earlobe (A2). Electrical pulses were presented at comfortably loud levels. The stimulus artifact obscures the earliest peak but later peaks are visible. The 'e' in the nomenclature notes the electrical stimulus. (**c**) The electrically evoked compound action potential response (ECAP) measured from the auditory nerve using the telemetry system from the cochlear implant manufacturer from a 12-year-old child who received bilateral implants sequentially and had used this CI for 2 years

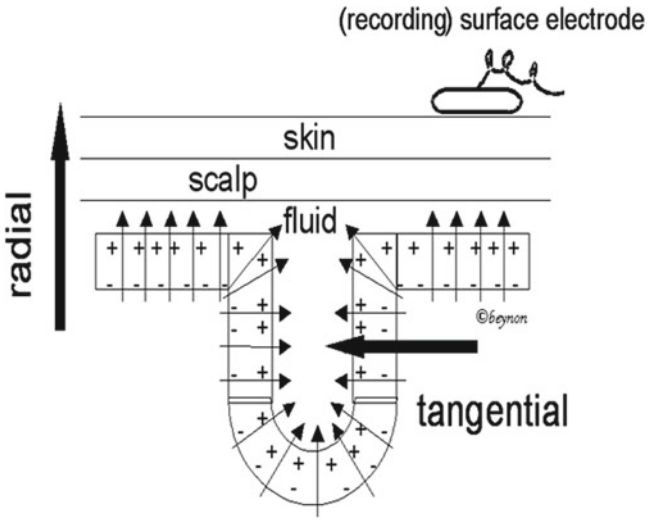


Fig. 8.2 Radial electrical fields point out of the head and are picked up by electrodes on the surface of the head. Tangential electrical fields run within the head and are thus missed by EEG recordings. (Figure by Andy Beynon, used with permission)

ples in Fig. 8.3b are of immature and mature cortical responses. The CI users were implanted at young ages and had used the devices for most of their lives. Yet, despite the similarity in the surface recordings shown in Fig. 8.2, there are important differences in the strength of the underlying neural generators in the left and right auditory cortices between the unilateral CI users and their peers with normal hearing (Gordon et al. 2013b).

Information specific to the neural generators underlying these responses can be obtained by recording the evoked electrical fields of activity from multiple positions on the head to view the activity from many different directions. Locating these neural sources can be complicated; multiple neural populations may be responding at the same time in different locations and at different orientations relative to the recording electrodes on the surface of the head. As shown in Fig. 8.3 (with permission from Beynon), electrical fields from cortical gyri may be oriented radially or directly out of the head. The direction of these fields permits them to be recorded by surface electrodes. By contrast, neurons in cortical sulci that produce fields tangentially oriented to the surface of the head will be missed by recording electrodes. It should be noted that these *electrical* fields also generate associated *magnetic* fields that run perpendicular to the electrical fields. As a result, the magnetic fields from tangential sources project out from the head and can be measured by surrounding sensors in a process known as magnetoencephalography (MEG). Locating sources of EEG and MEG recordings requires accounting for changes in orientation and pathway of signals as they pass through the brain lining (i.e., dura), scalp, and skull in different sizes of heads (Mosher et al. 1999; Roth et al. 1993). Possible locations of the neural activity are compared to the surface recordings using a variety of different methodologies such as Brainstem Electromagnetic Source Analysis (BESA) (Scherg and Picton 1991), standardized Low Resolution

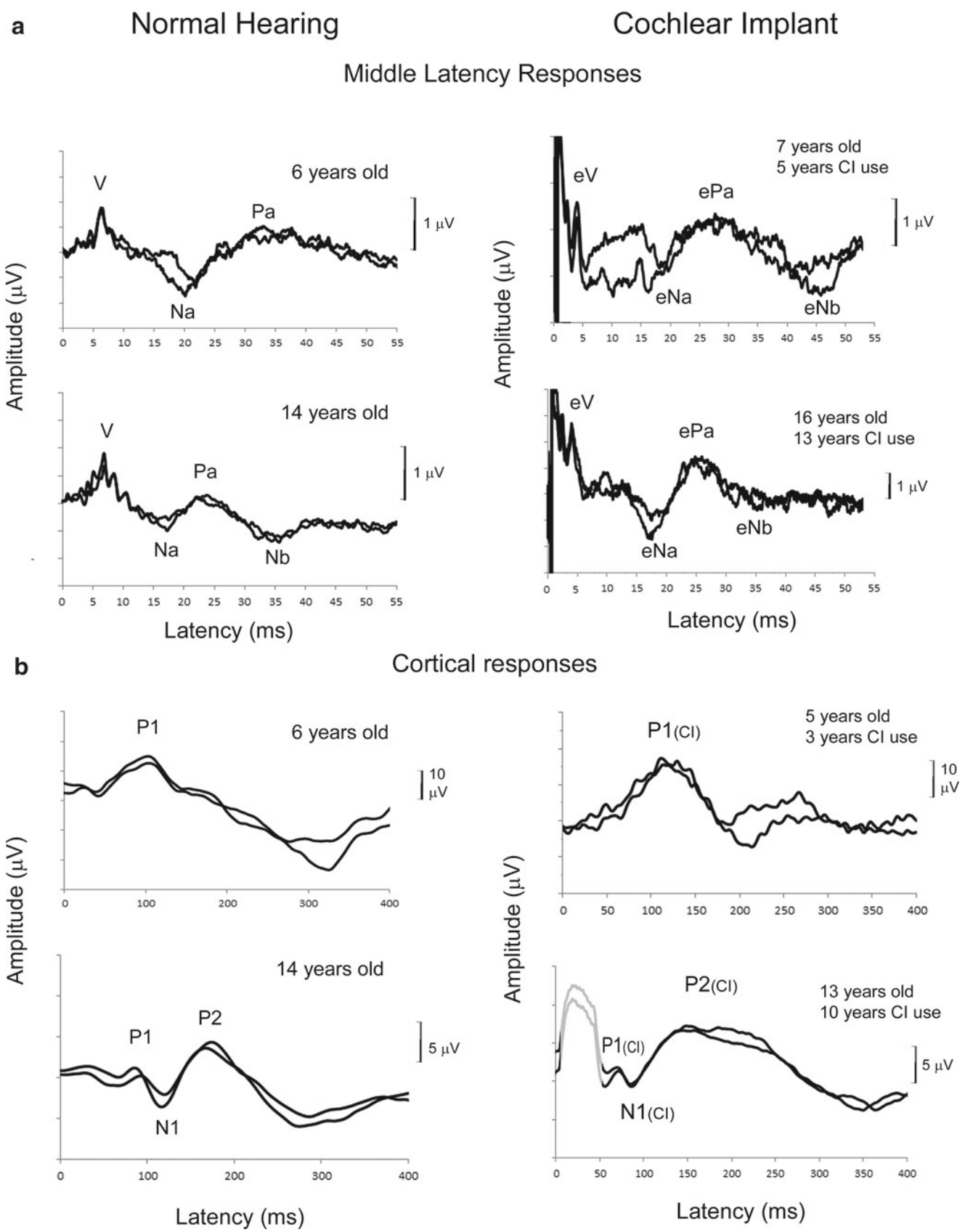


Fig. 8.3 (**a**) Middle latency responses in normal hearing children of 6 and 14 years of age, shown on the left, have characteristic peaks: Na, Pa, Nb. The ABR peak V can also be seen in the early part of the recording window. As shown in the responses on the right, the same peaks are clear in children who have grown up using a unilateral CI. The 'e' is used to indicate the electrically evoked response. (**b**) Cortical responses are shown for normal hearing 6- and 14-year olds on the left. The immature response is dominated by a positive peak, P1, whereas the 14 year old has peaks found in the mature response: P1, N1, P2. Responses from children with similar ages/durations of hearing are shown on the right. A 5 year old with 3 years of CI use has an immature response. The peak is noted as P1(CI) to indicate it was evoked by a CI and to acknowledge that it may have different generators from normal. The adolescent (13 years of age) with 10 years of CI use has a response with similar peaks to the mature response. The nomenclature of the peaks reminds us of the distinctiveness of the response: P1(CI), N1(CI), P2(CI)

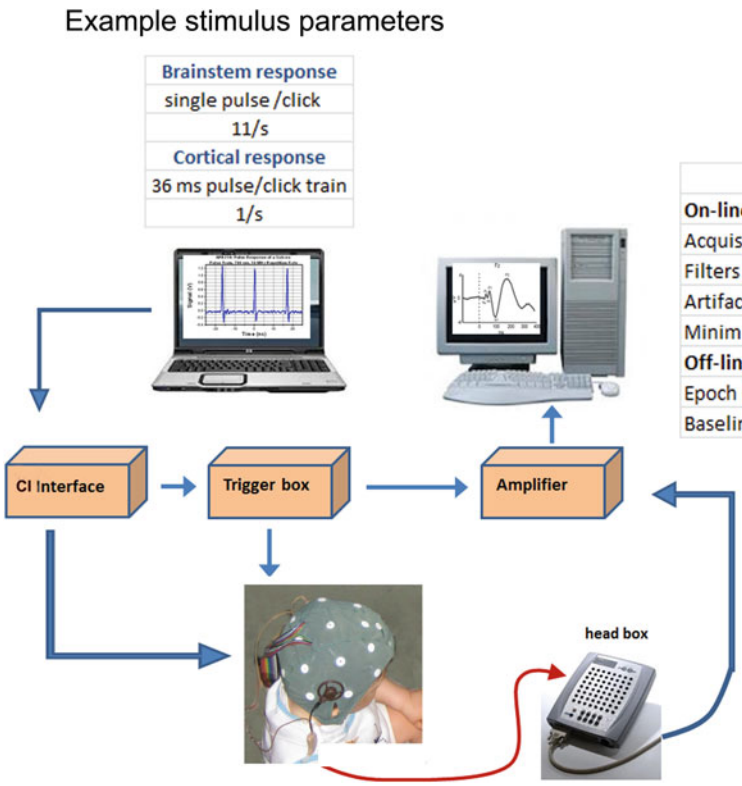
Electromagnetic Tomography (sLORETA) (Pascual-Marqui et al. 1994), and NME-Python (Gramfort et al. 2013). A particular spatial filter (linearly constrained minimum variance beamformer) has been adapted to locate areas of cortical activity in children using CI (Gordon et al. 2010, 2013b). The sLORETA technique has also been used to assess responses in children with CIs (Gilley et al. 2008; Henkin et al. 2004).

It is important to keep in mind which sources of neural activity are the targets to be recorded during electrophysiological recording. This knowledge will help to troubleshoot equipment problems (which can and do arise), as will be discussed in the next section, and interpret recorded data used in the management of children with CIs as discussed in the last part of this chapter.

Recording Electrical Fields of Neural Activity

There are several setups which can be used to record electrophysiological responses from the brainstem and thalamocortex in children. Cortical electrophysiological responses in children using CIs are often evoked by acoustic stimuli delivered through loudspeakers in the soundfield (e.g., Dinces et al. 2009; Gilley et al. 2008; Henkin et al. 2004, 2008; Sharma et al. 2005). The children typically wear their CIs at settings used daily. Alternatively, cortical responses can be evoked by stimuli presented directly to the CI through the appropriate accessory cable provided by the manufacturer. These methods are not often used to evoke auditory nerve or brainstem responses because the high degree of temporal synchrony required to record these higher frequency responses could be smeared by the child's movement relative to the loudspeaker in the soundfield and/or by the active processing in the CI speech processor. Note also that the stimulus delivered to the child will depend on the settings programmed into the speech processor. Instead, electrical pulses can be delivered by specific CI electrodes at defined rates (direct CI stimulation) to evoke both brainstem and thalamocortical responses (e.g., Gordon et al. 2003, 2005a, 2008, 2010, and 2011a; Jiwani et al. 2013; Sparreboom et al. 2014).

A schematic of equipment needed to directly stimulate the CI for electrophysiological measures is shown in Fig. 8.4. The baby with CIs is wearing a cap with integrated electrodes, but individual clinically available surface electrodes will work as well. Single channel recordings typically use a midline electrode (e.g., Cz or Fz) referenced to the ipsilateral earlobe or mastoid (A1 or A2). Multichannel recordings can range from only a few to over 100 electrodes with references



Example recording parameters

	Brainstem response	Cortical response
On-line parameters		
Acquisition A/D rate (Hz)	2000	1000
Filters (Hz)	10-3000	0.15-100
Artifact reject (uV)	± 30-40	± 100
Minimum accepted trials	300	400
Off-line parameters		
Epoch (ms)	-5 to 30	-200 to 800
Baseline correction (ms)	-5 to 0	-200 to -10

Fig. 8.4 The cochlear implant programming system directs CI pulses to be presented to the child through a CI interface which typically connects to the CI processor. The transmitting coil is placed in the correct location over the implanted CI receiver–stimulator. A trigger is required to synchronize the electrophysiological recording with the CI pulse presentations. In this case, the CI interface connects to a trigger box which connects to the amplifier of the recording system. Many systems require only a trigger cable between the CI interface and the amplifier. A set of example stimulus and recording parameters are provided for acquiring brainstem and cortical responses. The infant shown in the picture is wearing a cap of electrodes which are connected in to the head box of the recording system which is, in turn, connected to the recording amplifier

at different locations or averaged across recording electrodes. Example stimulus and recording parameters for brainstem and cortical response collection are shown. The stimulus equipment is often the same as that used for programming the CI. It includes a computer with the CI software and an interface to the CI. Standard electrophysiological recording equipment is also needed. The key addition needed to perform electrophysiological testing is that the two systems must be linked. Typically this is done using a trigger pulse which is sent by the CI system to the recording equipment through a cable. The trigger pulse marks when each electrical stimulus is delivered through the implant in the recording time window. The need to time lock the two separate systems means that the electrophysiological recording equipment must be able to accept an external trigger pulse. It is also recommended that the recording equipment be flexible enough to allow some manipulation of the criteria for artifact rejection. Using the example criterion for artifact rejection shown in Fig. 8.4, all recordings will be rejected if this includes the initial recording period during which the often very high amplitude stimulus artifact will be contained. It is possible to block the initial 2–2.5 ms of the recording period to alleviate this problem but this will remove the stimulus artifact from view, thus eliminating potential advantages of using the artifact to monitor CI function. Some recording systems provide an option of setting the time window during which the artifact rejection criteria are employed. In that case, the time window can be set following the end of the stimulus artifact to keep the CI artifact while removing noisy epochs resulting from participant movement and/or environmental noise.

It is beyond the scope of this chapter to review all aspects of electrophysiological recording (see Picton 2010 for a comprehensive and readable review). In brief, however, the example stimulus and recording parameters shown in Fig. 8.4 remind us that cortical responses occur at lower frequencies and longer latencies than brainstem responses and thus require lower frequency stimulation to evoke one response before stimulating another identical response. Recordings with the parameters shown in Fig. 8.4 will capture the transient onset response which can be viewed in an average of responses to repeated presentations of the same stimuli. Longer stimuli, however, evoke sustained responses such as the auditory Steady State Response (ASSR) (Picton et al. 2003) and the sustained component of the speech-evoked ABR (Kraus et al. 2009). These responses are assessed by analysis of amplitude and timing/phase which can be compromised by stimulus artifact. Methods for attenuating or accounting for CI stimulus artifact, discussed in the Section "*Minimizing CI stimulus artifact,*" are thus essential to make sustained responses viable for clinical use in cochlear implantation.

Analyses of transient onset responses in children using CIs will be easiest if the recorded response contains little noise. Minimizing noise can be particularly challenging when obtaining recordings from children using CIs. First, there are many electrical cables (recording electrodes and electrophysiological recording equipment, stimulating equipment, CI external device, trigger cable), which all may contribute unwanted electrical activity to the recording. Separating as many cables from the recording electrodes as possible is important to reduce electrical noise. Keeping the child as still as possible will help to lower myogenic activity as well as movement of the cables. Sedation is not needed to obtain clear EABR recordings at suprathreshold levels in most children (Gordon et al. 2003, 2004). In addition, the impact of sedation upon state of arousal will negatively impact recording of cortical responses (Campbell and Colrain, 2002; McGee et al. 1993). For these reasons, sedation is not used for electrophysiological recordings of CI recipients at our center. Recording in awake children is often possible although at times challenging. Success is more likely if a second tester helps during testing to: (1) keep the child distracted during recording; (2) ensure that equipment, including the CI, is properly in place; and (3) observe the child's voluntary/involuntary behavioral reactions to the CI stimulation. A typical recording session is shown in Fig. 8.5; one person sits with the child (Fig. 8.5a) while another operates the recording equipment (Fig. 8.5b). Stimulus presentation should be stopped immediately if a child provides any signs of discomfort to this input during testing. While movements uncorrelated to the CI stimulation will be reduced by averaging multiple recording trials of the same stimulus, movements which occur with every presentation of CI stimulus could affect the recorded response. For example, the occurrence of eye blinks time locked to CI stimulus presentation will contribute to the averaged signal. The presence of this type of reaction from the child in response to CI stimulation also provides clinical evidence that the stimulation level may be uncomfortably loud.

Averaging across multiple sweeps of recording to the same stimulus remains the most common method to reduce uncorrelated activity from the constant time-locked neural response. The averaged response can often be viewed while it is being collected during the recording process. Although there are guidelines for how many sweeps to accept when recording a response it will be necessary to acquire more sweeps when increased electrical noise is present.

Analyzing Recorded Responses in Children with Cochlear Implants

Both on-line and off-line analyses of data will require a similar set of questions to be asked:

1. Can a replicable response be detected over an acceptably low noise floor? If repeated recordings do not produce a replicable response, ensure that the recording equipment

Fig. 8.5 Recording electrophysiologic responses from awake children typically requires two testers. (**a**) One person works to distract and keep the child still while watching for behavioral reactions to the input. In this case, the infant is seated on the mother's lap. (**b**) One tester operates the recording system while the other works with the child. In this case, the child was able to sit alone

is working properly, that the recording and stimulating equipment are properly coordinated and that audible stimuli are being presented.
2. Do replicated responses respond to stimuli as expected? In other words, is the response collected the response that was expected? Keep in mind which part of the auditory system you intend to measure and whether you have recorded an auditory or nonauditory response, such as a myogenic response.
3. Is the stimulus artifact obscuring the response you wanted to measure? This issue is discussed in the following section.

If we apply these questions to the data shown in Fig. 8.6 (Davids et al. 2008a, b), we find that clear and replicable brainstem responses and middle latency responses were evoked by pulse trains of increasing widths. In Fig. 8.6a, all responses synchronized to the *onset* of the stimuli (i.e., 0 ms latency) are shown in the left panel. The middle latency response appears in the same latency window for all widths of electrical pulse trains, indicating that this response occurs to the onset of the train regardless of its length. As the stimulus artifact lengthens, responses in the early latencies are obscured. On the right panel in Fig. 8.6a, the same responses are shown but now relative to the *offset* of the pulse train. In this view, it becomes clear that the wave eV of the EABR is visible beyond the last pulse presented in the train. This finding indicated that the brainstem responds to each pulse in the train, including the last pulse. On the other hand, we now see the middle latency response shifting in latency, since this response occurs to the onset of the first pulse in the train instead of each pulse or the last pulse. In Fig. 8.6b, responses to the single electrical pulse are shown as stimulus current is decreased. Amplitudes decrease with latencies increasing slightly as expected until a response is no longer visible above the noise floor. The latency changes are reduced from the acoustically evoked response, likely reflecting the higher synchrony in the electrically evoked response (Gordon et al. 2003). The visual threshold is indicated in Fig. 8.6b. Thresholds can also be predicted by analysis of the rate of amplitude decline with decreasing current level using a regression line. This technique is available for "automatic" threshold prediction of the auditory nerve response in several manufacturer software systems.

Minimizing CI Stimulus Artifact

The CI stimulus artifact can be several 100 times larger than the auditory response being measured. There are at least two components: (1) a high-frequency artifact from the electrical pulses delivered by the CI electrodes and (2) a low-frequency direct current (DC) "pedestal" artifact which is related to the amplitude of the pulses. This latter component can be caused by mismatches in impedance between recording electrodes (Mc Laughlin et al. 2013). The artifact obscures early waves of the EABR as well as later responses depending on the type and duration of stimuli used. Moreover, it disrupts analysis of sustained neural responses to long duration stimuli. Discussed below are a number of solutions that have been proposed.

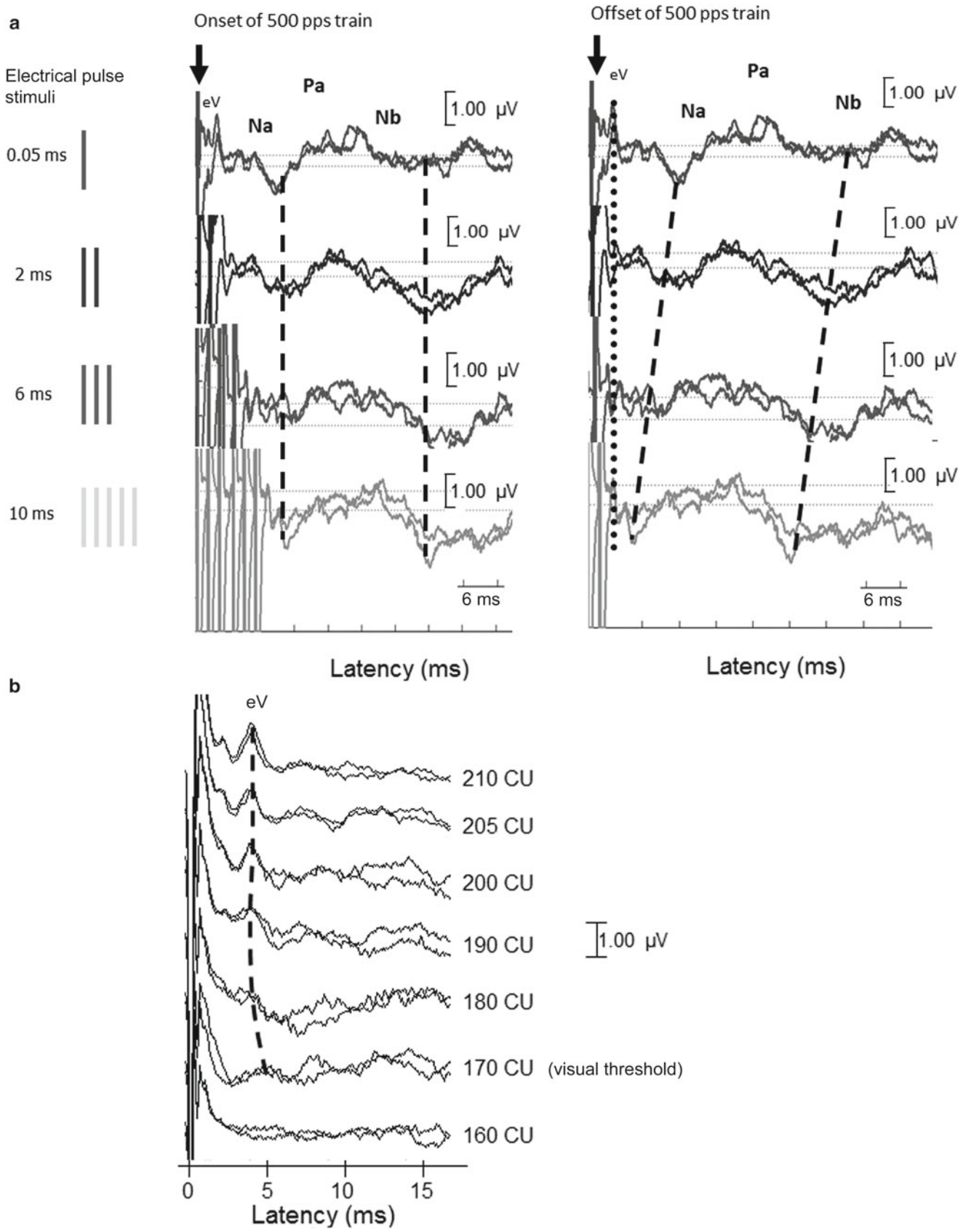


Fig. 8.6 A series of responses can help to identify replicable peaks in responses recorded in children using CIs. For example: (**a**) Replicable electrically evoked auditory brainstem responses (EABRs) and middle latency responses (EMLRs) were evoked in a child using a unilateral CI to single pulses or pulse trains of increasing duration (data included in Davids et al., 2008b). EMLRs occur at the same latency when the onset latency occurs at the beginning of the pulse train (*left panel*) indicating that they were evoked by the onset of the pulse train. By contrast, the EABR wave eV peaks occur at the same latency with decreasing latencies of EMLR peaks when onset latency occurs at the offset of the pulse train (*right panel*), indicating that the EABR occurs to each pulse in the train. (**b**) Replicated EABRs are shown with decreasing current intensity from a child using a unilateral CI. Amplitudes decrease until a response cannot be detected visually over the noise floor. The previous response was noted to be the threshold

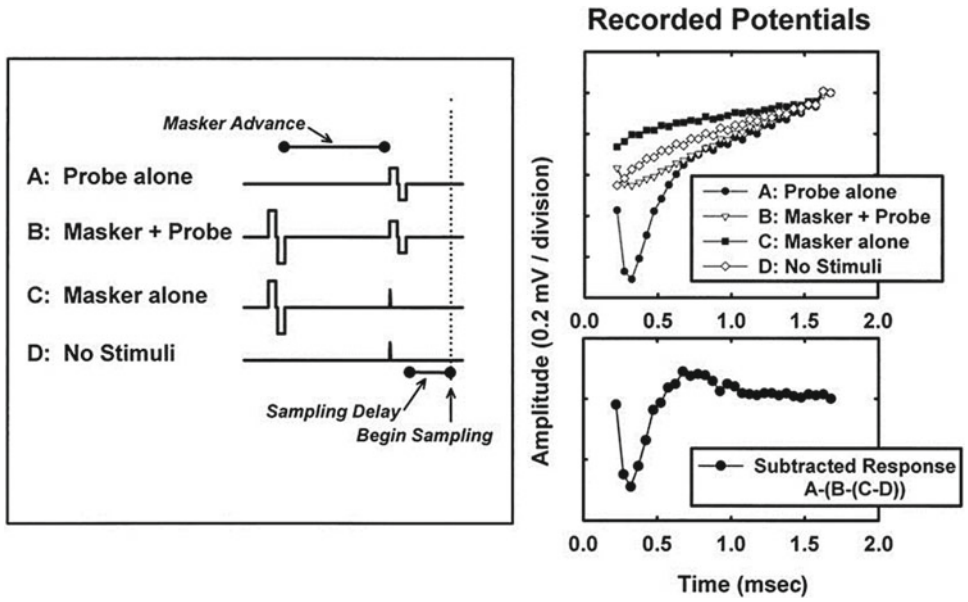


Fig. 8.7 Left panel illustrates the four stimulus conditions presented to the intracochlear stimulating electrode for use in the subtraction method. *Top right panel* depicts the respective waveforms obtained for each condition. *Bottom right panel* depicts the resultant waveform after all subtractions have been made. (Permission granted from Wolters Kluwer Health: Brown et al. (2000). "The relationship between EAP and EABR thresholds and levels used to program the nucleus 24 speech processor: data from adults." Ear Hear 21(2): 151–163, Fig. 2)

Alternating the polarity of the stimulating pulse will reduce the artifact if single pulses are being used to evoke the response (common for auditory nerve and brainstem responses) (Hofmann and Wouters 2010). This technique assumes that the response to the anodic first phase is the same as the cathodic first phase. However, the assumption underlying this technique is not entirely true (Miller et al. 2000). Therefore, a different approach, the forward-masking subtraction paradigm was proposed (Brown and Abbas 1990; Miller et al. 2000). This technique is now commonly used in CI telemetry systems to record the auditory nerve response in CI users. As shown in Fig. 8.7, a masker pulse is provided shortly before the target pulse so that most neurons will be in a refractory period and unavailable for stimulation. This response thus contains the artifact from the target pulse with little neural response. It can then be subtracted from a recording which contains both the artifact and the neural response, leaving only the neural response (Abbas et al. 1999; Brown et al. 2000). Although other procedures have been proposed for reducing the artifact from auditory nerve responses (Bahmer et al. 2010; Klop et al. 2004), the forward masking subtraction technique is most common and is employed in CI manufacturers' telemetry systems for recording auditory nerve responses. The forward masking subtraction method has also been used to remove artifact from trains of CI pulses from cortical responses with some success (Friesen and Picton 2010). One disadvantage to this method is the extra recordings needed. The doubling of recordings and thus recording time is not substantial for auditory nerve responses which are high frequency and can be recorded quickly and at high amplitudes with no external equipment through the CI telemetry systems; however, recording time will be longer for cortical responses which are low frequency and require a longer recording window/time.

Another suggestion for CI artifact removal in single channel recordings has been to move the reference electrode around the head until it is placed at a location which has the same stimulus artifact as the recording electrode (Gilley et al. 2006). More recently, it has been recommended that recording electrode impedances be matched within 1 kOhm and that any remaining stimulus artifact be attenuated from cortical recordings by a two-step procedure in which the high pass artifact is reduced by a low pass filter and the DC component be estimated and then subtracted (Mc Laughlin et al. 2013)

It is more common to use multiple recording channels to assess both the neural generators of the response and the stimulus artifact. Independent Component Analysis (ICA) has been used to separate artifact from neural components based on how the response appears at different recording locations. This technique has been used to reduce artifacts in both auditory nerve responses (Akhoun et al. 2013) and cortical responses (Debener et al. 2008; Gilley et al. 2006; Viola et al. 2012). Example components of cortical responses in children using CIs identified through independent component analysis are shown in Fig. 8.8a. Both individual sweeps as well as the average recording are shown, demonstrating some of the jitter from trial to trial which can occur. Clear differences in the responses recorded across the head (scalp topography) are present between the four identified components related to: (1) 60 Hz noise, (2) eye blinks, (3) a left CI stimulus pulse presented at 1 Hz, and (4) the evoked cortical response which also occurred at 1 Hz. The difference in recorded locations on the scalp and waveform between the stimulus artifact and neural response made it possible to distinguish between these two in this case, but this distinction would be more difficult if the differences in spatial distribution were smaller (Friesen and Picton 2010). Figure 8.8b provides an example of a cortical

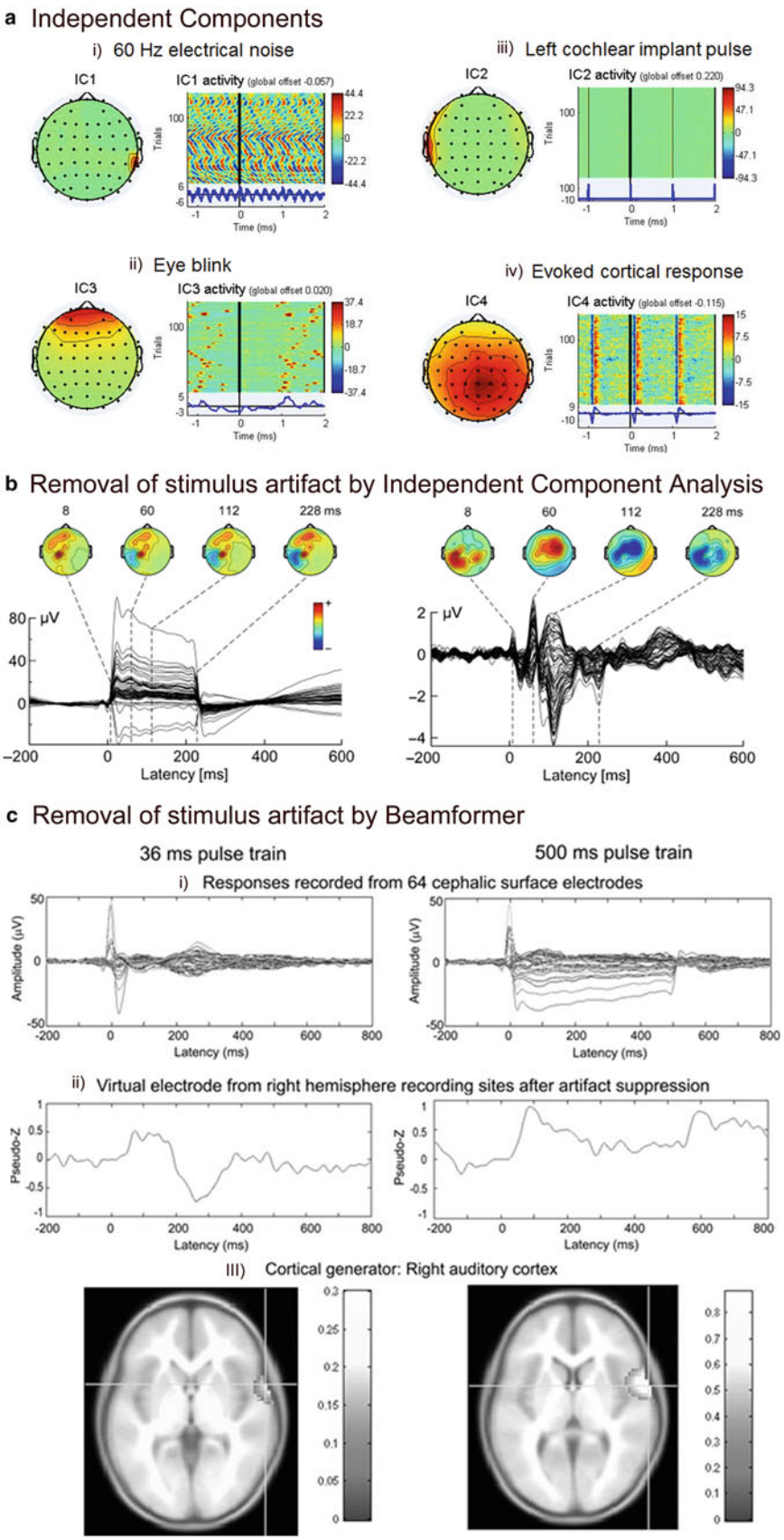


Fig. 8.8 (**a**) Independent components can be separated by their different scalp morphology shown by the heads in each of the four panels. The voltage (μV) of the component in each trial is shown to the right of each head as well as in the average of all trials (blue response below). (*i*) 60 Hz electrical noise occurs in this case on the right. (*ii*) Eye blinks (frontal field) occur at random times from trial to trial with effects on the average. (*iii*) A left CI pulse occurs at the same time and voltage in all trials, and appears clear in the average. The time window is long enough to capture more than CI stimulus presentation. (*iv*) The evoked response is time locked to the stimulus, occurring consistently from trial to trial and thus is clear in the average. (**b**) Stimulus artifact is separated from the neural response using Independent Component Analysis (Fig. 1 (a and c) from "Source localization of auditory evoked potentials after cochlear implantation." Debener et al. (2008). Psychophysiology 45(1): 20–24). The responses from all recording electrode channels shown on the left indicate the scalp morphology of the stimulus artifact which is the same at all latencies. On the right is the response once the component with the CI artifact had been removed. The scalp morphology shows changes over time suggesting a change in the underlying neural generators. (**c**) The TRACS (Time-Restricted, Artifact and Coherent source Suppression) beamformer was used to remove artifact from a 36 ms and 500 ms electrical pulse train (Fig. 3, Springer, Brain Topography, **24**(3–4), 204–219, "Use It or Lose It? Lessons Learned from the Developing Brains of Children Who are Deaf and Use Cochlear Implants to Hear." Gordon, K. A., Wong, D.D., Valero, J., Jewell, S.F., Yoo P., and Papsin, B.C.). (*i*) The averaged responses from 64 recording channels. (*ii*) The recording electrodes on the right hemisphere were assessed. One the artifact was isolated and suppressed, the activity from one space in the brain (voxel) and represented by a "virtual channel" has no evidence of the artifact. The activity is from the voxel shown by the cross hairs in (*iii*). The lighter regions shown are those voxels which had significantly higher signal than noise at ~100 ms

response obscured by stimulus lasting 220 ms which was revealed by using the ICA method (Debener et al. 2008). The scalp topography is shown above the responses and is dominated by the stimulus artifact coming from the left at 8, 60, 112, and 228 ms. After the component with the stimulus artifact is removed, the response can be seen with changes to the scalp morphology at different latencies. Another method uses a beamformer, a spatial filter, which has been adapted to isolate the CI stimulus artifact (TRACS beamformer method) (Wong and Gordon 2009). As shown in Fig. 8.8ci, this method was used to locate and measure cortical generators evoked by a short (36 ms) or long (500 ms) electrical pulse train from a right CI in the same child (Gordon et al. 2011b). The response peaks are obscured by the longer duration stimulus. The beamformer suppresses the contribution from the stimulus artifact as shown in Fig. 8.8cii. The axial view of the cortex in Fig. 8.8ciii shows the voxels (areas of the brain) with significant activity over the noise floor. The two stimuli evoke the same area of cortex, although a slightly increased area is active in response to the longer stimuli. Activity at the voxel indicated by the cross-hairs has the same peak strength, measured in pseudo-Z, at a similar latency of ~100 ms.

A simple method for avoiding stimulus artifact is to use short duration stimuli as shown in Fig. 8.8c (Gordon et al. 2011b). This approach may be successful if the aim is to measure cortical detection of CI input; however, it will not suffice if we wish to know how the child processes longer and more complex stimuli such as speech. It is also not sufficient to record certain types of electrophysiological responses. For example, when recording electrically evoked auditory steady state responses (EASSRs) it is necessary to use long trains of CI pulses. Reduction of CI stimulus artifact is required in order to permit analysis of the amplitude and phase of these neural responses. Artifact reduction may be achieved by first recording the response to electrical trains of alternating polarity with high sample rates in order to see each stimulus artifact clearly and then replacing each artifact with interpolated data (Hofmann and Wouters 2010).

The CI artifact is a complicating factor for measuring electrophysiological responses in children but is not prohibitive. The effort spent on development of the solutions reviewed here is evidence of the importance placed upon solving this problem. These efforts in turn reflect the belief in the value these measures may provide for monitoring CI use in both children and adults.

Present and Potential Clinical Uses

It is clear that electrophysiological responses in children can be collected but what are the purposes for these recordings? Just as these measures have been helpful for identifying the loss of auditory function in children, they can be instrumental in confirming that function has been regained through cochlear implantation. This information is very useful at the time of surgery. In addition, baseline recordings can be useful for comparison if concerns arise later. In general, the choice of which electrophysiological test(s) to be conducted at any point in time will depend upon the question being asked. If the goal of the assessment is to confirm CI internal device function, measures particular to the stimulation delivery of the particular CI system in question can be performed. It is also important to ensure that the auditory system remains responsive to CI input. With that goal in mind, comparing auditory nerve thresholds and rate of amplitude growth with the responses previously recorded from that child is valuable. In other situations, there may be questions about the child's innate ability to respond to sound through their CI(s). In those cases, measures assessing distinct parts of the auditory pathways including the brainstem and thalamo-cortex can be considered.

Confirming Auditory Function in Special Populations of Children

There are some populations of children who may be particularly at risk for poor CI use. As shown in Fig. 8.9, auditory nerve and brainstem responses can be variable in children with hypoplastic auditory nerves (Valero et al. 2012). On the one hand, responses shown in Fig. 8.9 provide evidence of responses to the CI. On the other hand, however, many of these responses are different from those expected and thus may not be generated from the auditory pathways, in part or in whole. Based on these concerns, we assessed the behavioral speech perception measures in these children and found that most had limited speech perception ability (Valero et al. 2012). We suspect their atypical responses reflect abnormal auditory brainstem function or nonauditory activity.

The question of auditory function has also been raised in children with Auditory Neuropathy Spectrum Disorder (ANSD). Despite having absent or abnormal brainstem responses to acoustic input, some children with ANSD have clear cortical responses to acoustic input (Michalewski et al. 2009; Rance et al. 2002; Sharma et al. 2011). These measures can be important to decide whether or not to provide a CI to a child with ANSD, particularly given the variability in behavioral thresholds to pure tones observed in this population. Excluding the children with radiological evidence of auditory nerve deficiency, ANSD and the associated finding of abnormal auditory nerve and brainstem responses is thought to be caused by poor synaptic coupling between the inner hair cells and auditory neurons (Harrison 1998; Moser et al. 2013). The aim of the CI is to bypass these lesions (Rance and Barker 2008; Zeng et al. 2005). With this reasoning in mind, it can be argued that the presence of the electrically evoked auditory nerve response is the most important initial measure to confirm CI effectiveness and establish

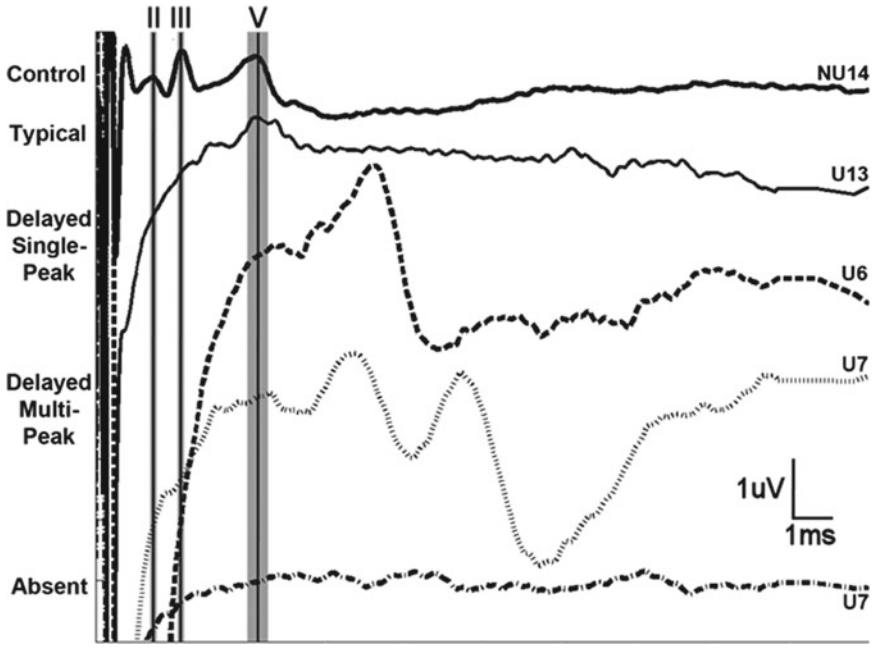


Fig. 8.9 Children with hypoplastic nerves had different EABRs ranging from "Typical" responses to "Absent" responses. A response from a child with a normal auditory nerve based upon radiologic findings is shown as an example of typically expected EABR peaks (Fig. 3, Valero et al. 2012). "Electrophysiologic and behavioral outcomes of cochlear implantation in children with auditory nerve hypoplasia." Ear Hear 33(1): 3–18)

that synchronous brainstem function has been restored. Indeed, those children with ANSD who had expected EABRs after implantation had better speech perception outcomes than their peers with absent/abnormal EABRs (Jeon et al. 2013; McMahon et al. 2008). Cortical measures have been used to determine whether the impairments in temporal processing associated with the dyssynchronous activity in the auditory nerve are alleviated by the CI. Impaired temporal abilities have been captured by electrophysiology after CI by measuring the latency of the cortical response (Campbell et al. 2011) or the jitter in latency/phase from one trial to another (intertrial coherence) (Nash-Kille and Sharma 2014). Impaired ability to hear small gaps in sound have been shown in individuals with ANSD (Zeng et al. 2005) and a particular variant of the cortical response, called the evoked cortical change complex, has provided an objective tool to measure this (He et al. 2013). In this technique the obligatory cortical response is measured to a stimulus which contains at least one change. A change in the cortical response indicates detection of both stimuli. He and colleagues created change in a train of electrical pulse trains by adding a silent gap in the middle (2013). Those children with ANSD receiving CIs who had cortical change responses only when large gaps were present had poor speech perception test scores with their CIs (He et al. 2013). The cortical change complex was also used to measure discrimination by children with ANSD between stimulation coming from different CI electrodes (He et al. 2014). The group with good speech perception scores had larger amplitudes of cortical change responses than the group with poorer scores. This was interpreted as poorer neural synchrony in the latter group despite electrical stimulation.

The use of electrophysiology in special populations of children using CIs reminds us that the response chosen to be recorded after implantation will depend on the question being asked. For example, do we want to know whether the CI can stimulate a hypoplastic nerve or bypass the lesion in a child with ANSD. Or are we interested in the temporal processing of sound in these children. Both are appropriate questions that may be relevant at different times after CI. Therefore, we recommend electrophysiological recordings to examine several different levels of the auditory pathway at different times after implantation. The initial priority is to obtain information about detection of CI stimulation by assessing the ability of the auditory system to respond to electrical pulses from the CI. This information may be obtained by evaluating auditory nerve responses through ECAP recordings which may be done through the telemetry system of all currently available devices in North America without need for external recording equipment. Measurements of auditory brainstem function (EABR) can help to confirm ECAP findings. Cortical responses will be useful to investigate questions about more central processing of CI input. Along with assessment of progress through observation and behavioral measures, this battery of electrophysiological testing will help to determine whether auditory function has been restored by CI in populations of children at risk for poor auditory stimulation and/or development.

Programming CI Stimulation Levels

One of the main goals after implantation is to provide appropriate levels of stimulation for each electrode along the array

within each inner ear. Using behavioral measures alone to accomplish this task in young children with little hearing experience and limited attention span can be difficult and time consuming. Children with developmental delays and other complicating medical conditions may be more challenging to reliably program using techniques solely relying on behavioral responses. Electrophysiological responses have long been looked to for help in finding appropriate levels of stimulation to program CIs, especially for these children.

Although electrophysiological measures are not sufficiently predictive of individual behavioral measures to be used in isolation to set CI devices in children (Brown 2003), they can be used as a helpful first step. In general, auditory nerve and brainstem thresholds are evoked at current levels which are audible (Gordon et al. 2004; Hughes and Abbas 2006; Jeon et al. 2010). The electrophysiological thresholds can be combined with some behavioral observations for programming. First, the clinician must ensure that the child is comfortable listening to these levels of current. In some devices, stimulation levels are determined from maximum comfortable levels measured at as many electrodes as possible. Current levels can be increased from the electrophysiological threshold until any behavioral signs of intolerance emerge. Other devices require minimum stimulation levels to be set; the gold standard has been to use behavioral thresholds to determine these levels at individual CI electrodes. The electrophysiological measures can be used to help the child establish a conditioned response to the audible CI input so that the behavioral thresholds can later be measured across electrodes.

Because monopolar stimulation has a wide spatial range of excitation, it is possible to extrapolate measures across a range of CI electrodes (Zhu et al. 2012). This means that there may be very little difference in the neural population stimulated by one CI electrode and that stimulated by the neighboring electrode (Long et al. 2014). More focused patterns of stimulation (e.g., bipolar, tripolar) offer potential benefits for pitch perception (Arenberg Bierer 2010) but would also require more focused measurement of required current levels for each stimulating CI electrode (Goldwyn et al. 2010; Long et al. 2014; Zhu et al. 2012). The extrapolation technique might then not be possible. Thus, the monopolar stimulation used in present CIs provides a practical benefit because it is difficult to obtain behavioral measures in up to 44 electrodes (22 electrodes in 2 devices), even in older children. Electrophysiological measures such as the ECAP and EABR can be used to determine whether there are electrodes which evoke markedly different responses than their neighbors. Their use for this purpose could become increasingly important if more focused stimulation strategies are adopted into clinical CIs.

Electrophysiological measures are sensitive to changes in CI technology. Auditory nerve thresholds were found to decrease with evolutions in CI arrays of one manufacture (Gordin et al. 2009; Gordon and Papsin 2013). This change likely occurred because of the desire to provide more focused stimulation by use of precurved arrays intended to place the electrodes in close proximity spiral ganglion cells adjacent to the modiolus. However, decreases in threshold were not consistent across the array, suggesting inconsistencies in distance of the electrode array from the modiolus (Gordin et al. 2010; Long et al. 2014). Because electrode placement is essential for effective CI use (Holden et al. 2013) and possibly for hearing preservation (Havenith et al. 2013), it is important to be able to monitor subtle differences between CI electrodes. Importantly, auditory nerve thresholds were found to be more sensitive to these changes than the behavioral thresholds (Gordon and Papsin 2013). This finding means that electrophysiological measures might provide functional corroboration for structural imaging studies and a method to monitor effects of new CI electrode arrays.

In sum, electrophysiological measures indicate audible CI stimulation levels and can be used to monitor differences in levels between electrode arrays and between individual electrodes on a single CI array. Electrophysiological measures are best used in combination with behavioral measures to provide children with optimal stimulation levels.

Potential for Programming Bilateral Cochlear Implants

Programs for bilateral cochlear implants should evolve to include methods to provide accurate binaural cues. Benefits could be realized when the 2 CIs are matched in pitch and level (Goupell et al. 2013). Without software to deliver current to targeted electrodes in the two devices simultaneously, each device must be programmed individually. This means that bilateral input could be mismatched in pitch and level which (Goupell et al. 2013). Traditional behavioral measures used to program CIs may not accurately predict current levels which do not produce a weighted perception to either the left or right CI (i.e., are "balanced") when both devices are used together (Gordon et al. 2012a). Providing "balanced" current levels might be helped by electrophysiological measures (Gordon et al. 2016). It is common for children to need different current levels in each implant to achieve a balanced perception of bilateral input. Both ECAP and EABR measures can help to determine which current levels to use. Growth in amplitude can be measured on both sides using ECAPs as shown in the example in Fig. 8.10a and EABR as shown in the example in Fig. 8.10b. The growth in amplitude of the example ECAP responses from Fig. 8.10a is plotted in Fig. 8.10c, revealing that the maximum amplitude of CI-1 is greater than CI-2, and CI-1 has a threshold which is ~15 dB CU higher. At ~175 CU,

amplitudes of the two responses are similar. In Fig. 8.10d, we assessed the rate of amplitude increase in the EABR from a pair of apical electrodes in a group of children and found little difference between sides (Salloum et al. 2010). The difference in levels between the two CIs needed for balanced hearing was predicted by the difference in ECAP

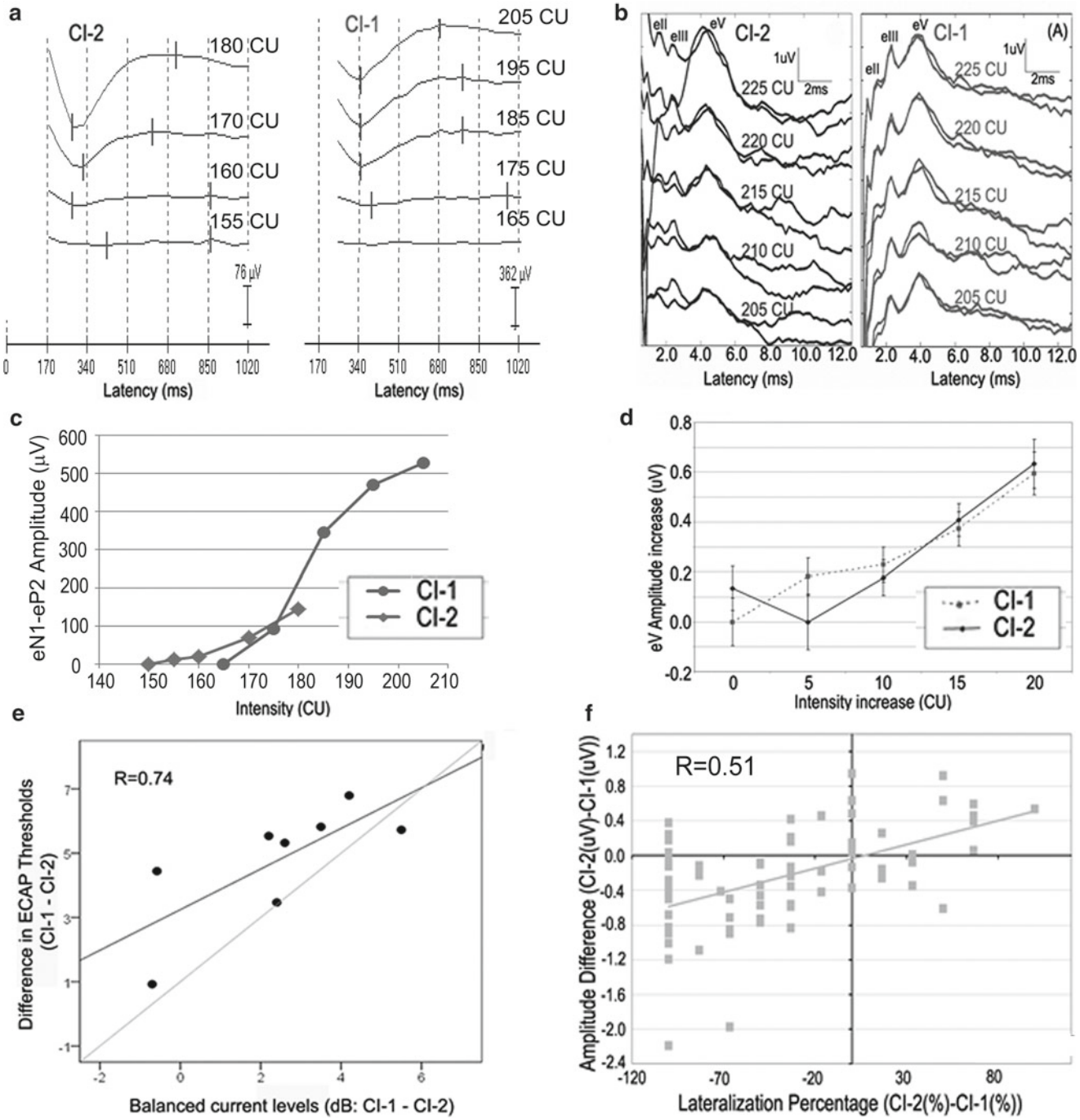


Fig. 8.10 Responses with stimulus intensity decreases are shown for (**a**) ECAPs and (**b**) EABRs for each CI in bilateral users (Fig. 1, Salloum et al. 2010). Amplitude growth curves are shown for the ECAPs evoked on each side in (**c**) and the EABRs evoked on each side in (**d**) (Fig. 3, Salloum et al. 2010). ECAPS and EABRs can help predict the levels at which children hear bilateral input as "balanced" (neither lateralized to the left nor the right side of the head: (**e**) Differences in ECAP threshold predict "balanced" levels (Fig. 4a in Gordon et al. (2012a). "Toward a method for programming balanced bilateral cochlear implant stimulation levels in children." Cochlear Implants Int 13(4): 220–227). (**f**) The difference in wave eV amplitude predicted the side to which children perceived bilateral input to be lateralized (Fig. 7, Salloum et al. (2010). "Lateralization of interimplant timing and level differences in children who use bilateral cochlear implants." Ear Hear 31(4): 441–456, Figs. 1, 3, 7 (b, d, e, respectively))

thresholds (Gordon et al. 2012a), as shown in Fig. 8.10e. Similarly, the amplitude of the wave eV of the EABR predicted on which side of the head children heard bilateral input (Salloum et al. 2010), as shown in Fig. 8.10f. Auditory nerve and auditory brainstem responses could therefore be helpful in setting stimulation levels in bilateral CIs. The ECAP might be particularly useful because it does not require external electrodes or equipment to record. The ECAP has also been found to predict the rate of loudness growth in children using CIs (Steel et al. 2014) and ECAP thresholds are typically audible for children; ECAP thresholds fall between threshold and maximum comfortably loud levels (Gordon et al. 2012a). Thus, there may be potential to incorporate ECAPs and/or EABRs into a protocol for setting optimal stimulation levels for children using bilateral CIs.

Identifying Unwanted Myogenic Responses

Whether one or two CIs are used, the primary aim of the electrical stimulation is to evoke auditory responses. However, nonauditory responses to the electrical stimulation can occur. Figure 8.11 provides an example of one such response. The response was confirmed to be myogenic as it was eliminated once an anesthetic agent with reversible paralytic was administered (Cushing et al. 2006, 2009). Once this response was removed, the characteristic peaks of the auditory brainstem response (EABR) were visible at expected latencies and interwave latencies. Further analyses of these types of responses have revealed that they can be evoked in most children using CIs at comfortable listening levels given slow rates of pulse presentation (Cushing et al. 2006). Using monopolar stimulation, no one particular CI electrode was more prone to evoking these responses than another (Cushing et al. 2006). At the threshold levels of these myogenic responses, most children could not feel anything when asked, but when current level increased, children began reporting sensations behind the eyes, around the mouth, or in the neck ipsilateral to the stimulating CI. These areas did not visually appear to twitch/move/respond until current was further increased. Thus, the electrophysiological responses were more sensitive to unwanted effects of CI stimulation than the more typical observation of the child's face.

Predicting and Monitoring CI Outcomes

It has been over two decades since CIs were provided to children; there have been remarkable outcomes shown in many behavioral measures, but variability remains. Structural and functional analyses of the auditory pathways are important components to help understand this variability. With these tools, we find that deafness in childhood has important implications for future auditory development and plasticity. Prior to implantation, the auditory brain can be imaged using a number of techniques, including electrophysiological measures. Structural studies through magnetic resonance imaging (MRI) reveal abnormal changes to

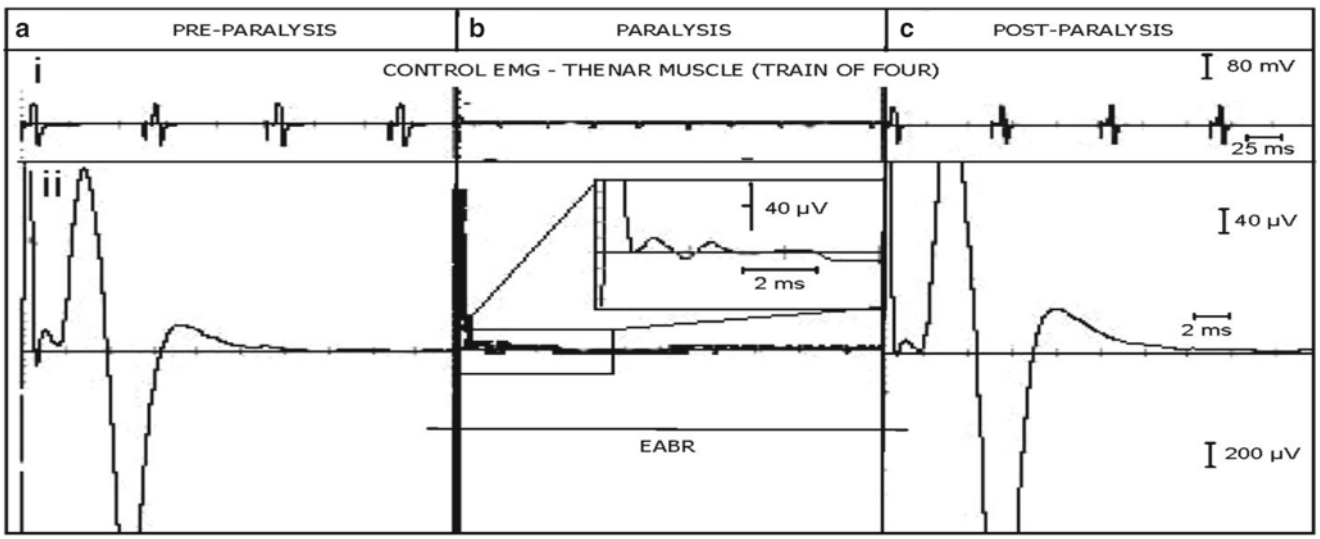


Fig. 8.11 Intraoperative recording demonstrating a myogenic response from the facial nerve during recording of the evoked auditory brainstem response (EABR) (Fig. 1, Cushing et al. 2006) (*ii*) channels (**a**), is obliterated by the presence of muscle relaxant allowing the underlying EABR to emerge (**b**). The myogenic, facial nerve response returns on reversal of the muscle relaxant (**c**). Control electromyography (train of four) of the thenar muscle was also performed (*i*). (Permission granted from John Wiley and Sons: Fig. 1 Cushing et al. (2006). "Incidence and characteristics of facial nerve stimulation in children with cochlear implants." Laryngoscope 116(10): 1787–1791, John Wiley and Sons)

the planum temporale, Heschl's gyrus, the frontal lobes, Broca's area, and visual cortices in adults with deafness from early childhood (Li et al. 2012; Shibata, 2007, Smith et al. 2011; Lepore et al. 2010; Penicaud et al. 2013). Functional imaging studies using positron emission tomography (PET) of resting brains of deaf children have also demonstrated effects of deafness in childhood (Lee et al. 2001, 2007). There are limitations to use of MRI of the brain after implantation because of the magnet in the device. MRIs of 1.5 T (Gubbels and McMenomey 2006) to 3.0 T (Med-El communications) have been reported as safe for CI use with the magnet in place. However, concerns regarding movement of the CI magnet remain even for MRIs done at relatively weak magnet strength (Cuda et al. 2013; Hassepass et al. 2014; Migirov and Wolf 2013). Moreover, the CI magnet produces a large artifact that often impacts imaging of the auditory cortex (Majdani et al. 2008, 2009). PET scans are possible in CI users but require the injection of radioisotopes and thus are not commonly used in children.

Despite having poorer spatial resolution than either PET or functional MRI, electrophysiological measures can be used to image activity in the auditory system of children with and without CIs, as shown in Fig. 8.8c. At present, these measures have been most effectively used in research to describe group effects (Gilley et al. 2008; Gordon et al. 2010, 2013b; Henkin et al. 2004).

Electrophysiological measures obtained on the first day of CI activation provide a unique opportunity to assess function of auditory pathways which, for many children, will have had no significant input until this moment. Although there is considerable variability in responses at this initial time point (Gordon et al. 2011a), the etiology of deafness provides some explanation. In particular, auditory nerve responses evoked by stimulation of apical and basal CI electrodes are more similar in amplitude in children whose deafness is associated with mutations in the GJB-2 gene (which codes the protein Connexin 26) than in children with other etiologies of deafness, perhaps reflecting the similar effects of Connexin 26 depletion at different places along the cochlea (Propst et al. 2006). Cortical responses, might also be classified by etiology of deafness; response morphology was more similar in a group of children with GJB-2 mutation than in a group of peers whose deafness was due to other causes (Gordon et al. 2011a).

The goal of the CI is to promote auditory plasticity and drive development toward a mature, normally functioning hearing system. Development of the implanted child's auditory system is commonly monitored using behavioral measures. Electrophysiological responses provide an important way to study the extent to which auditory plasticity has been compromised and differences in CI-driven activity from normal auditory function. These objective measures may provide insight into variability in outcomes including lack of expected benefit. They could also serve as a way to measure the impact of management techniques. Latency changes in cortical-evoked potentials with implant use have been shown after unilateral CI use in both the auditory brainstem (Gordon et al. 2003, 2006) and cortex (Ponton et al. 1996a). Latency changes are reduced or truncated in late implanted, suggesting limited auditory plasticity (Ponton and Eggermont 2001; Ponton 2006; Sharma et al. 2002, 2005). This finding is consistent with the suggestion that the auditory cortex becomes reorganized during the period of deafness and no longer available for hearing through a CI (Finney et al. 2001; Kral and Sharma 2012; Lee et al. 2001; Lomber et al. 2010). In general, early implantation is advantageous to limit the period of auditory deprivation impacting brain development. Many have advocated for implantation of congenitally deaf children before 3.5 years of age; however, implantation by this age is not a panacea nor should older children uniformly be viewed as poor candidates. Infants with profound loss and little access to sound from amplification may require very early implantation to avoid the effects of deafness on auditory pathway development. Older children who had acoustic hearing due to later onset or progressive hearing loss and/or effective acoustic amplification may experience auditory development that will be highly beneficial for CI use. Activity recorded using PET shows that functional changes to specific areas of the brain can be better predictors of speech perception with a CI than age at implantation alone (Lee et al. 2007), confirming the importance of an individualized approach. There is some evidence that noninvasive electrophysiological measures may be useful in predicting outcomes as well. For example, persistently abnormal cortical responses are indicative of low speech perception test results (Gordon et al. 2005b, 2008). On the other hand, we have to ask: do normal obligatory electrophysiological responses always reflect normal function?

More complex measures of auditory processing such as discrimination can be determined using a variety of measures. A difference in responses to a frequently occurring stimulus and an infrequent stimulus can be measured. If the participant is asked to listen for the infrequent stimulus, a P300 can be recorded whereas the mismatched negativity (MMN) can be recorded to the same stimuli during passive listening. The MMN has been recorded in adult CI users to assess possible precognitive cortical processing of input through the CI (Kraus et al. 1993; Lopez-Valdes et al. 2013; Ponton and Don 1995; Ponton et al. 1996b, 2000; Sandmann et al. 2010; Timm et al. 2014; Wable et al. 2000). By contrast, there are limited studies of this response in children using CIs. One group has used the P300 to assess vowel discrimination (Henkin et al. 2004) and others have used the cortical change complex to assess vowel discrimination (He et al. 2014) to study cortical discrimination of stimulation at different CI electrodes.

As indicated in the introduction of this chapter, electrophysiological and behavioral measures will provide the most accurate and useful answers when used in combination

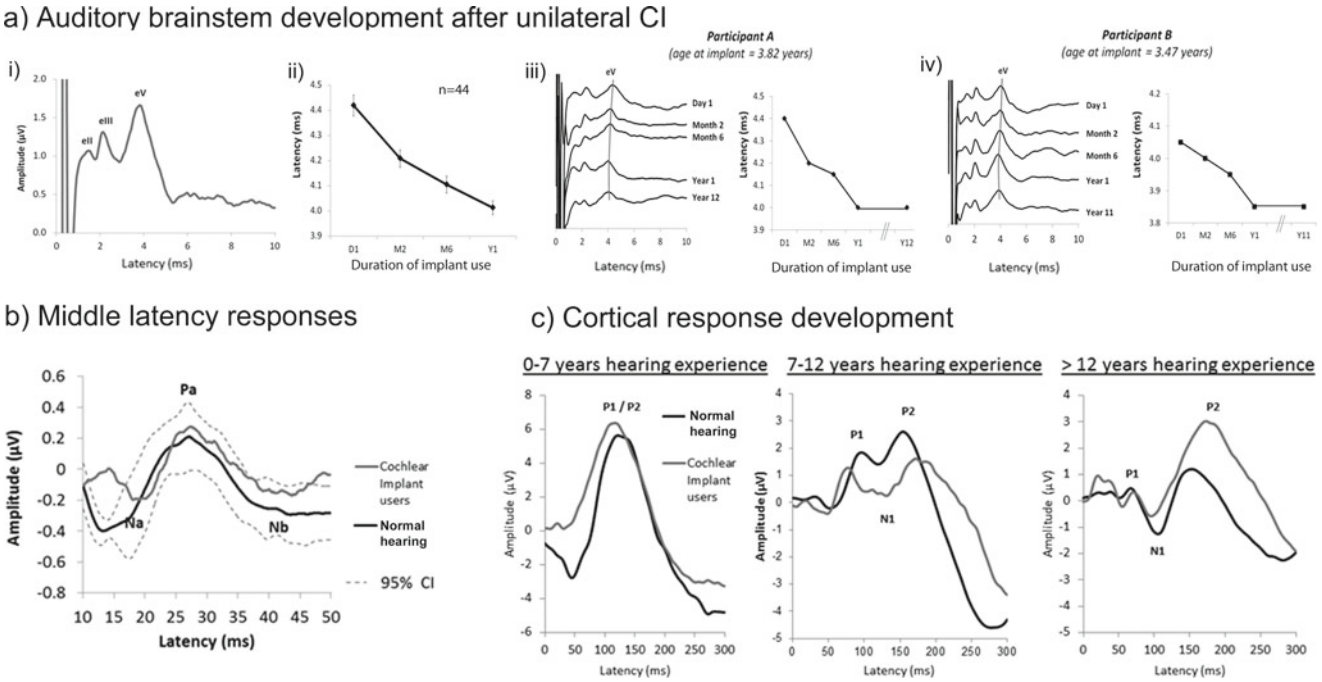


Fig. 8.12 (**a**) Latencies of wave eV significantly decrease over the first year of implant use (Fig. 1, Gordon et al. 2013a). These latencies do not significantly change over more than a decade after the first year of CI use as demonstrated by participants **A** and **B**. (**b**) Middle latency responses from children using CIs (shown in *red*) fall within the range of a group of peers with normal hearing (shown in *black*). (**c**) Cortical responses from children implanted early match those from peers with similar durations of hearing experience. Cortical responses in both groups take over a decade to develop into a response with mature peaks (Fig. 2, Gordon et al. (2013a). "Benefits and detriments of unilateral cochlear implant use on bilateral auditory development in children who are deaf." Front Psychol 4: 719)

(i.e., in a test battery) to assess auditory function. Having now followed children who have used CIs for over a decade, it is clear to the author that many children who are implanted early experience a maturing of the electrophysiological responses shown in Figs. 8.1 and 8.1. As shown in Fig. 8.12a, much of the developmental change in the auditory brainstem response occurs during the first year of CI use. Latencies at 1 year of use were similar in two children to those measured a decade later (Gordon et al. 2013a). The electrically evoked middle latency response (EMLR) of adolescents who had long periods of unilateral implant use was very similar to their normal hearing peers as shown in Fig. 8.12b. The EMLR was consistently present by 6 months of unilateral implant use in a large cohort of children (Gordon et al. 2005a). The cortical response, on the other hand, takes approximately a decade of CI use to mature; changes to the response in children who received CIs early compared to their peers with normal hearing are shown in Fig. 8.12c. The consistency between the CI and normally expected responses is encouraging but does not mean that unilateral CIs will achieve normal auditory function in these children. Dipole source analysis using the TRACS beamformer measured sources from the left and right auditory cortices independently is shown in Fig. 8.13a. Analyses of the dipoles demonstrated that greater than 1.5 years of unilateral CI use promotes abnormal strengthening of activity from the stimulated ear through the brainstem (Gordon et al. 2007, 2012b) and to both auditory cortices (Gordon et al. 2010, 2013b). This abnormal strengthening is associated with decreased ability to regain function of the opposite ear through a CI (Gordon et al. 2013b).

The unique plasticity promoted by CIs in children may be captured by electrophysiological measures and understood in context with behavioral responses. As we strive to provide children who are deaf with better and timelier access to sound, we continue to use and develop tools to monitor their auditory development. At present, analyses of electrophysiological measures provide an important way of capturing this remarkable progress.

Conclusions

Electrophysiological measures are important components to use as part of the battery of tests to program and monitor the progress of children with CIs. A number of issues regarding recording electrophysiological responses in children using CIs have been discussed in this chapter. In some respects, the issues discussed can be reduced to the following questions:

1. Is the response present?
   If not, determine the origin of the problem, whether due to technology or neural.
2. Can the response be optimized?

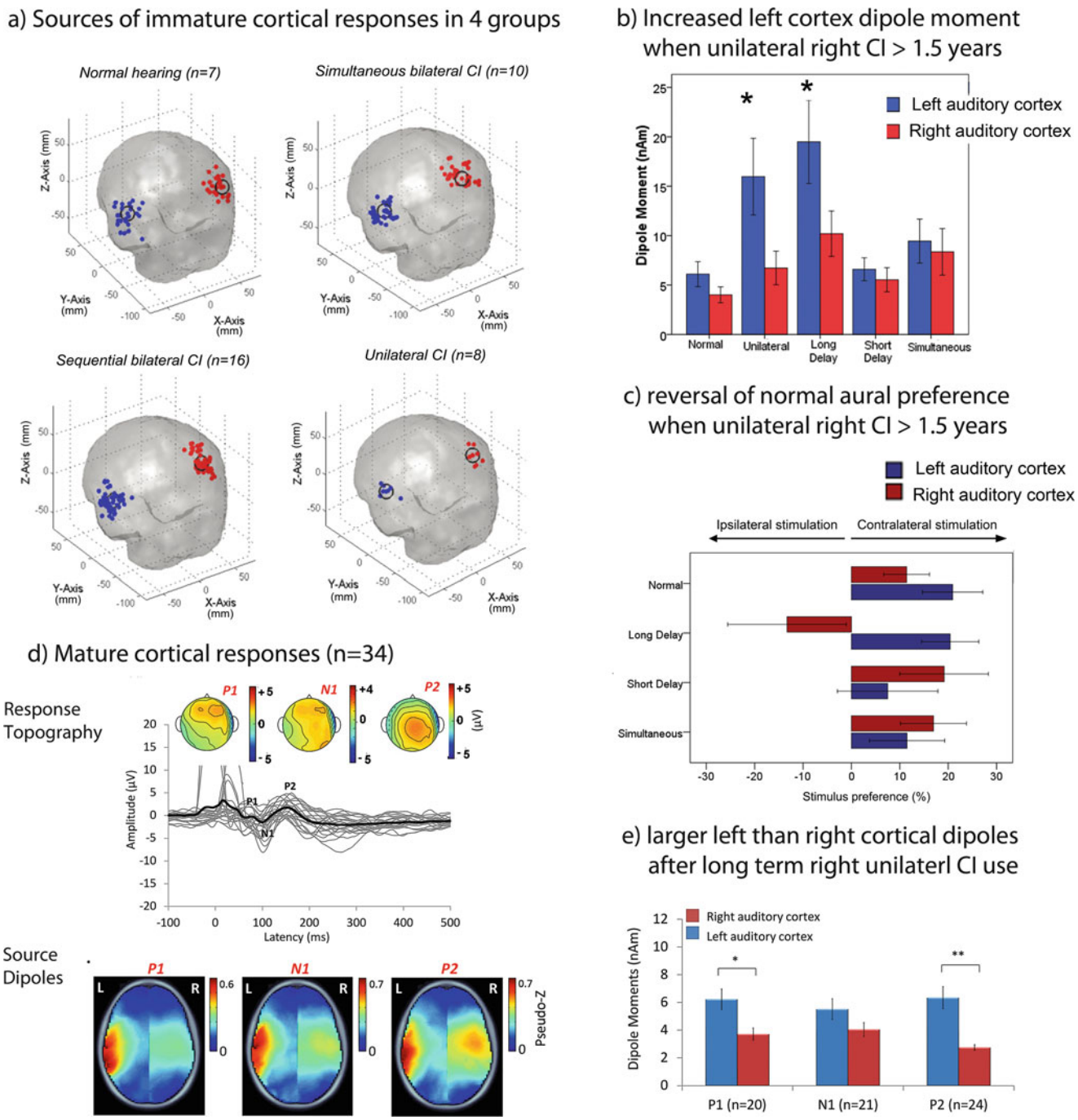


Fig. 8.13 (**a**) Dipole locations from children with immature cortical responses (Fig. 2, Gordon, K. A., D. D. Wong and B. C. Papsin, "Bilateral input protects the cortex from unilaterally-driven reorganization in children who are deaf." Brain, 2013, 136(Pt 5): 1609–1625, by permission of Oxford University Press). (**b**) Dipoles of cortical sources were larger when evoked by right side stimulation in those children with >1.5 years of unilateral CI use/deprivation (Fig. 7c., Gordon, K.A., D.D. Wong, and B.C. Papsin, "Bilateral input protects the cortex from unilaterally-driven reorganization in children who are deaf." Brain, 2013, 136(Pt5):1609–1625, by permission of Oxford University Press). (**c**) Dipoles in the left and right cortices were compared for aural preference/stimulus dominance [(contralateral-ipsilateral/contralateral+ispilateral) × 100], revealing a reversal from normal in the right (ipsilateral) cortex for children with >1.5 years of unilateral CI use/deprivation (Fig. 8, Gordon, K. A., D. D. Wong and B. C. Papsin, "Bilateral input protects the cortex from unilaterally-driven reorganization in children who are deaf." Brain, 2013, 136(Pt 5): 1609–1625, by permission of Oxford University Press). (**d**) Cortical responses from adolescents who had long-term right unilateral CI use show mature cortical peaks which have strong dipoles of activity in the contralateral left cortex (Fig 3a and 3b, Jiwani, S., B.C. Papsin, K.A. Gordon, "Early unilateral cochlear implantation promotes mature cortical asymmetries in adolescents who are deaf." Human Brain Mapping, 2016, 37: 135–152, Wiley Periodicals, Inc.). (**e**) Dipoles in the left cortex are larger than in the right in these children (Fig 3c, Jiwani, S., B.C. Papsin, K.A. Gordon, "Early unilateral cochlear implantation promotes mature cortical asymmetries in adolescents who are deaf." Human Brain Mapping, 2016, 37: 135–152, Wiley Periodicals, Inc.)

Answering this question will help to program unilateral and bilateral CIs.

3. Does the response change?

   Multiple measures are helpful for identifying expected and unexpected responses within one recording session as well as over time.

4. Does the response fit with the behavioral changes?

A battery of electrophysiological and behavioral measures should be used to assess, fit, and monitor children using cochlear implants. Because additional information is gained by electrophysiological measures, the use of these measures in a breverse may be true as well.

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Part III

Cochlear Implant Outcomes in Children

Speech Perception and Spoken Word Recognition in Children with Cochlear Implants

9

Karen Iler Kirk and Michael Hudgins

Introduction

The first child to receive a cochlear implant in the United States was implanted with a House/3M single-channel cochlear implant by Dr. William House in 1980. Eventually, 256 children were implanted with the House/3M device by Dr House and his co-investigators as part of a FDA clinical trial (Berliner 1990). These early pediatric implant recipients were generally totally deaf, often due to meningitis. The input they received from hearing aids was insufficient for them to understand spoken language or to develop intelligible speech. Pediatric implantation was quite controversial at that time; many clinicians and researchers felt single-channel implants were too crude to offer substantial speech perception and spoken language benefits to pediatric recipients. Opponents of pediatric implantation contended that placing an electrode array in the cochlea would damage existing cochlear structures, possibly preventing future implantation with more sophisticated technology. In contrast, Dr. House felt that waiting for more advanced cochlear implant systems would significantly prolong auditory deprivation in these children, making it difficult for them to benefit from future technological advances. Longitudinal studies of these children revealed that on average, they were able to recognize environmental sounds, discriminate speech on the basis of timing and duration cues (i.e., display pattern perception) (Eisenberg et al. 1983), and combine auditory input with speechreading cues to enhance speech understanding (Eisenberg 1985). Some of these children even demonstrated limited open-set speech understanding (Berliner and Eisenberg 1987). However, the majority of children with single-channel cochlear implants relied on spoken and signed English for communication at home and in the classroom. These modest early gains marked the beginning of a revolution in the medical management and education of children with profound, prelingual hearing loss.

Since those early days, we have seen remarkable technological advancements in electrode design and signal processing strategies, broadening of cochlear implant candidacy to include infants and those with some residual hearing, and an expansion in educational options for children with cochlear implants. As of 2013, approximately 38,000 children in the United States have received cochlear implants. Children with cochlear implants now obtain substantial open-set speech understanding, often demonstrate age-appropriate speech and language skills, and can be educated in mainstream classrooms with their typically hearing peers (A. E. Geers and Hayes 2011; A. E. Geers et al. 2009; Hayes et al. 2009; Moog and Geers 2010). Recent outcomes are highly encouraging, but challenges remain. As in adults, cochlear implant outcomes in children are variable, and most recipients have difficulty understanding speech in noise (Davidson et al. 2011; Gifford et al. 2011; van Wieringen and Wouters 2015). Understanding these individual differences and developing interventions to improve outcomes for low-performing children are priorities for cochlear implant researchers and clinicians. To address these challenges, recent work by Pisoni and colleagues has focused on the role of information processing in speech perception and spoken word recognition outcomes (see Chap. 17).

Speech recognition remains the most direct outcome of cochlear implantation, and provides the foundation for developing effective speech production, language, and literacy skills. As such, assessing speech perception is an important component in cochlear implant evaluations and in measuring outcomes in children with cochlear implants. In this chapter, we will review speech perception assessment

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procedures for children from infancy through school age, current speech perception outcomes in this population of cochlear implant recipients, and factors that impact cochlear implant outcomes.

Speech Perception in Infants and Toddlers: Assessment and Outcomes

Since the year 2000, cochlear implantation has been FDA approved for children as young as 12 months of age, but many children even younger than that have been implanted off label. Multiple studies have found that earlier-implanted cochlear implant users, including infants implanted before 12 months of age, have better long-term speech perception and linguistic outcomes than those implanted later (Colletti et al. 2011; Davidson et al. 2011; Kirk et al. 2002; Lee and van Hasselt 2004; Niparko et al. 2010; van Wieringen and Wouters 2015; J. L. Wu and Yang 2003; Zwolan et al. 2004). As Eisenberg and colleagues have pointed out, assessing performance in infants and young children is challenging. They advocate using a hierarchical approach which includes both subjective (i.e., parent report scales) and objective measures of performance (Eisenberg et al. 2006) to assess auditory and speech perception skills ranging from detection through comprehension (see Fig. 9.1).

Parent Report Scales

Parent report scales typically are employed to assess auditory skill and speech perception development in children between the ages of 1 and 3 years. The use of these instruments is widespread in both clinical and research settings. In the United States, two of the instruments used most commonly are the Infant-Toddler Meaningful Auditory Integration Scale (IT-MAIS) (McConkey Robbins et al. 2004; Zimmerman-Phillips et al. 2000) and the LittlEars (Kuehn-Inacken et al. 2003). Both have been adapted for use in a variety of languages (Kosaner et al. 2013; Taitelbaum-Swead et al. 2005; Weichboldt et al. 2004). The IT-MAIS has been used widely in Food and Drug Administration (FDA) pediatric clinical trials of cochlear implant systems. The IT-MAIS asks parents 10 questions about the auditory behaviors their child demonstrates in daily activities. The questions probe a hierarchy of behaviors from detection through speech recognition and vocal development. Each probe receives a score of 0–4 depending on how frequently the child demonstrates the behavior. The LittlEars test battery was developed to assess subjective performance outcomes for children with hearing aids and cochlear implants. It has two components, the LittlEARS Auditory Questionnaire and the LittlEARS Diary. The Auditory Questionnaire consists of 35 yes/no questions about a child's auditory behavior (e.g., detection, responses to sound, and vocalizations). The Diary allows parents to document their observations of the child's development over the course of 6 months after cochlear implant fitting. More detailed information about these and other parent report scales can be found in Chap. 2.

Objective Assessments

Speech Discrimination

Objective measures of speech perception in infants can be used in either clinical or research settings. In clinical settings, objective assessment of infant speech perception is focused on detection. Objective measures of speech discrimination (i.e., the ability to tell if two sounds are the same or different) and speech recognition may require technologically advanced methodologies, and greater time for test administration; thus, these procedures more typically are employed in research studies. By observing an infant's

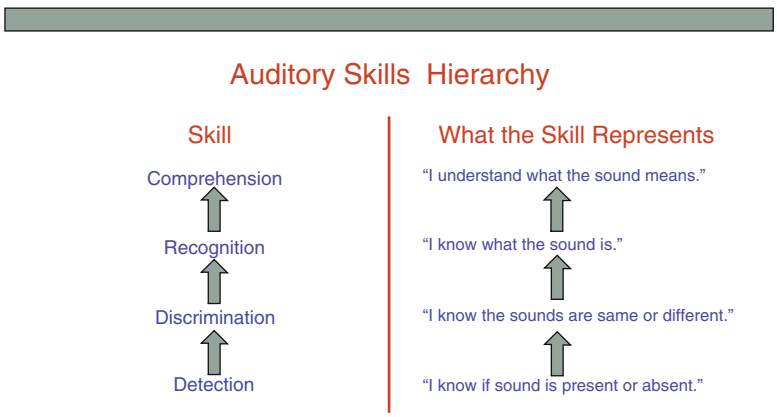

Fig. 9.1 The auditory skills hierarchy

behavior in response to speech sounds, researchers are able to test hypotheses about how infants perceive speech.[1] The techniques used to assess speech discrimination and spoken word recognition in infants are built upon techniques used clinically to assess infants' sound detection abilities, including behavioral observation audiometry and visual reinforcement audiometry (VRA). As shown in Fig. 9.1, speech sound discrimination is a higher level ability than sound detection.

One methodology used to assess speech discrimination in infants with CIs is the conditioned head turn procedure. In this technique, infants are conditioned to respond to changes in patterns of speech stimuli. First, the infant is presented with one speech stimulus (e.g., a repeating vowel sound). Next, a different speech stimulus is presented along with a visual reinforcer as in VRA. The experimenter gradually introduces a delay between the onset of the stimulus change and the onset of the visual reinforcer until the infant begins to anticipate the onset of the visual reinforcer upon recognition of the stimulus change. Finally, control trials (no stimulus change) and test trials (stimulus change) are randomly presented. Looking longer to the visual reinforcer on test trials than on control trials suggests the infant can discriminate the stimuli. This technique has been used in prelinguistic infants with normal hearing to assess their ability to discriminate speech features or speech sounds (Kuhl 1979).

Because the conditioned head turn response is such a useful method for assessing speech discrimination in infants with normal hearing, researchers have tested its feasibility as a means of assessing infants with cochlear implants. Eisenberg et al. (2012) utilized a conditioned head turn response to assess infants' discrimination of speech features contrasts. The Visual Reinforcement Assessment of the Perception of Speech Pattern Contrasts (VRA-SPAC) was adapted from Boothroyd's Speech Pattern Contrast Test for adults (Boothroyd 1984). The VRA-SPAC assesses an infant's ability to discriminate a novel or different stimulus within a repetitive series of VCV syllables. Eisenberg and colleagues showed that both infants with normal hearing and infants with hearing loss could consistently discriminate the speech feature of vowel height; infants with hearing loss were less consistent than their peers with normal hearing at discriminating the speech feature of vowel place. Consonant feature discrimination was highly variable in both groups of children (Martinez et al. 2008). They concluded that the VRA-SPAC is a useful objective tool for assessing vowel perception in infants and toddlers younger than 2 years of age.

Uhler et al. (2011) used the VRA-SPAC to monitor the acquisition of speech perception skills in three infants following cochlear implantation, and to compare their performance to that of seven infants with normal hearing who served as controls. Two of the infants with cochlear implants were age matched to one child in the control group. The cochlear implant recipients were implanted between 12 and 16 months of age. Prior to implantation, the children with hearing loss were unable to master discrimination of any phonemic contrast. Following 2 or 3 months of cochlear implant use, the pediatric implant recipients had mastered three out of five phonemic contrasts. The results suggested a similar trend in the development of speech feature perception for the pediatric implant recipients and their age-matched controls.

The visual habituation procedure is another methodology that can be used to assess an infant's ability to discriminate two sounds on the basis of various auditory or auditory-plus-visual cues. In this procedure, infants are seated on a caregiver's lap in front of a visual monitor and loudspeaker (see Fig. 9.2 for an illustration of the testing setup). In the habituation phase, the infant's gaze is directed toward the monitor and a visual stimulus, such as a checkerboard, is presented while an auditory speech sound is repeated. The experimenter records how long the infant looks at the display. When the infant looks away for longer than a predetermined amount of time, the trial and stimulus presentation ends. Successive trials with the same stimulus are repeated until the infant reaches a predetermined habituation criterion. During the test phase, control trials using the habituation stimulus and experimental trials using a novel auditory stimulus are presented. Longer looking times to the novel stimulus compared to the stimulus used during habitation suggest successful discrimination.

Houston and colleagues were among the first to use the visual habituation procedure to assess speech discrimination in children with cochlear implants (Houston et al. 2003). They employed contrasting speech sounds that allowed them to assess discrimination of stress and duration cues ("ahh" vs. "hop hop hop"), prosody (rising vs. falling /i/), and sound detection (speech vs. silent trials) in infants with normal hearing and infants with cochlear implants. Infants with normal hearing looked significantly longer at the checkerboard display during trials with sound. Although the infants with CIs could detect the speech sounds (i.e., the sounds were presented above their auditory detection thresholds) and were trending toward a preference for the sound trials over the silent trials, their performance in the two conditions did not yield a statistically significant difference. Not surprisingly, infants who had been using their CIs the longest did show a greater preference for sound trials than those with less cochlear implant experience.

More recent research concerning infants' perception of infant-directed speech is in line with the hypothesis that infants with CIs are less attentive to speech than their NH peers. Infant-directed speech is characterized by higher

[1]We are grateful to Derek Houston, Ph.D., for providing information about infant speech perception research methods. For a more detailed review of these methodologies, see D. M. Houston et al. (2012).

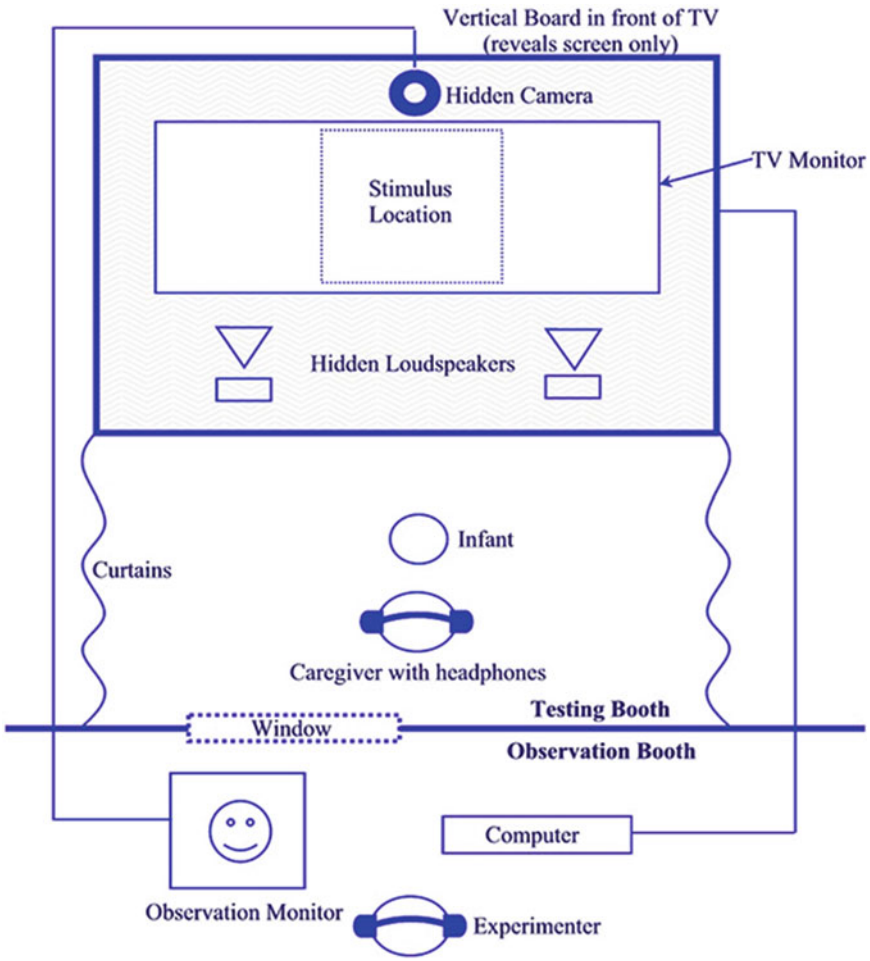

Fig. 9.2 An illustration of the Visual Habituation testing paradigm. The child is seated on the caregiver's lap facing a large computer screen. The caregiver is masked via headphones. The experimenter and observer are outside the testing suite

pitch, more varied pitch, more repetition, and more varied amplitude than adult-directed speech. Infant-directed speech is thought to help children identify word boundaries in connected speech. Robertson et al. (2013) found that normal-hearing infants and infants with hearing loss both attended longer to infant-directed compared to adult-directed speech. This suggests that they both discriminate the prosodic differences in the two types of speech. However, the hearing impaired infants exhibited shorter looking times to both types of speech. Houston and colleagues propose that a preference for infant-directed over adult-directed speech may not arise in infants with CIs until they have had extensive hearing experience (Houston and Bergeson 2014; Segal and Kishon-Rabin 2011).

Spoken Word Recognition

Long before they utter their first words, infants can recognize familiar words and associate them with referents in the environment. For example, 5-month-olds prefer to listen to their own names than to similar-sounding or contrasting names (Mandel et al. 1995). At approximately 6 months of age, infants with normal hearing show an ability to recognize words that represent familiar objects in the environment such as "mommy" and "daddy" (Tincoff and Jusczyk 2012). At the same age, infants also demonstrate the ability to recognize words that represent general categories of objects such as "hand" and "feet" when viewing videos of adult hands and feet (Tincoff and Jusczyk 2012). These studies demonstrate that infants are able to recognize words and, to some extent, understand what they mean.

The preferential looking paradigm has been used to assess spoken word recognition abilities in infants with CIs. In this technique an infant is seated on a caregiver's lap in front of two displays (or one split-screen display) with different visual stimuli on each side. With both visual stimuli on display, an audio track corresponding to only one of the visual displays is presented (see Fig. 9.3 for an illustration of the testing setup). Subsequent trials contrast the infant's looking time to the two pictures when auditory stimuli representing each of the visual displays are presented one at a time. Infants look longer at pictures they have learned to associate with a given auditory stimulus in the familiarization phase. This

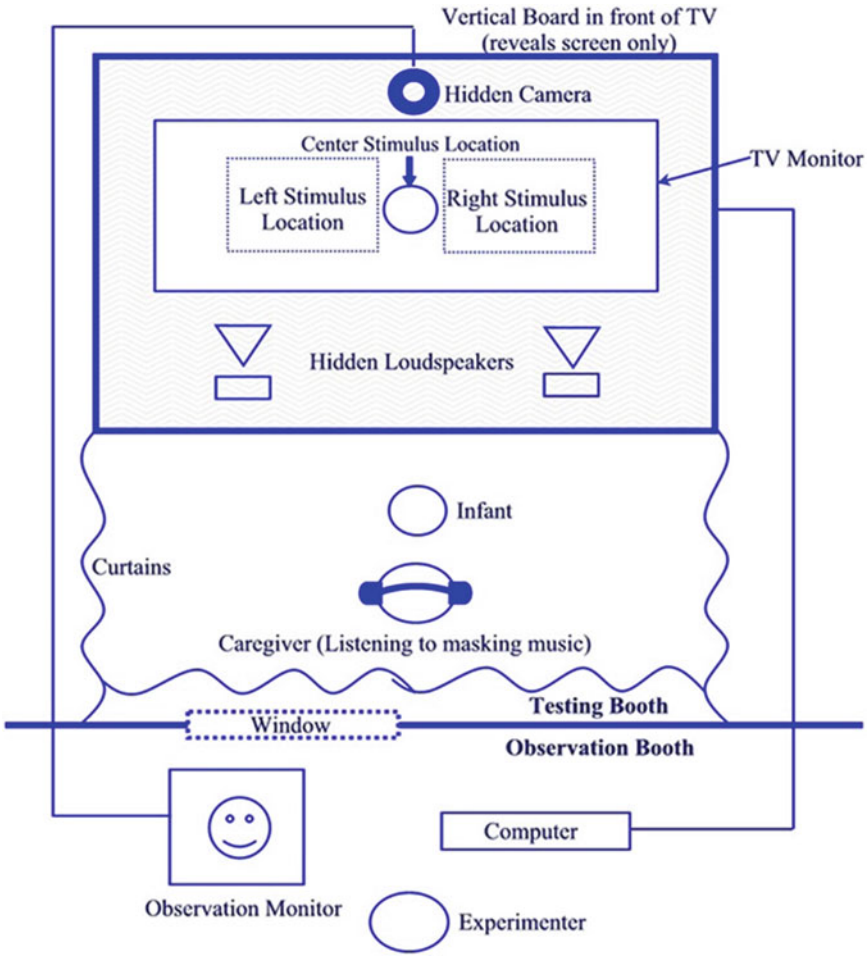

Fig. 9.3 An illustration of the Preferential Head Turn Procedure testing paradigm. The child is seated on the caregiver's lap facing two images on the monitor(s). The caregiver is masked via headphones. The experimenter and observer are outside the testing suite

allows objective measure of speech recognition long before children can make behavioral responses. For example, Kuhl and Meltzoff (1982) simultaneously presented footage of a person producing an /a/ sound and an /i/ sound, and found that infants with normal hearing looked longer at the video display that matched the presented auditory signal.

Bergeson et al. (2010) used the preferential looking paradigm to examine whether infants and toddlers with bilateral hearing loss utilize both auditory and visual speech cues. Specifically, they examined whether infants with cochlear implants exhibited a preference for congruent audiovisual stimuli as would be expected from their peers with normal hearing. Infants with normal hearing as young as 2 months are able to match phonetic information with corresponding lip and face movements (Patterson and Werker 2003). Bergeson et al. employed videos of two words with visually contrasting articulation patterns ("judge" vs."back"). The two infant groups showed the opposite recognition patterns: infants with normal hearing preferred to look at the matching auditory-plus-visual stimuli in the first test block, but not in the second. Infants with cochlear implants showed no preference for the matching stimulus on the first block, but did so on the second. The authors suggested that children with normal hearing may have habituated or lost interest in the task in the second block of trials. The results of this study suggest that very young children with cochlear implants require more exposure to auditory-plus-visual stimuli to integrate the multimodal speech signals than children with normal hearing.

Speech Perception in Preschool and School-Aged Children

Test Battery Selection and Administration

A battery of tests is typically employed to assess speech perception and spoken word recognition in young children with hearing loss who can participate in formal testing. When developing a speech perception test protocol, one must consider a variety of factors, including participant and test characteristics. Figure 9.4 illustrates relevant child and test characteristics that should be considered in test selection and administration.

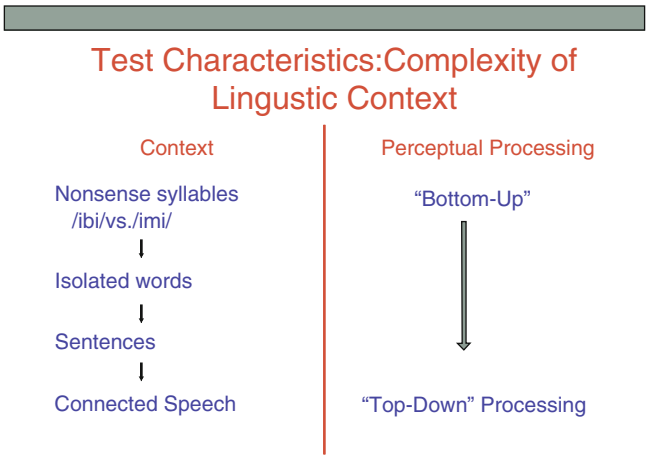

Fig. 9.4 The contributions of higher-order processing as a function of test stimulus type

Child Characteristics

The child's age and developmental level will impact familiarity with test vocabulary and ability to attend to the task. This in turn will influence selection of both stimulus and response parameters. Administering objective tests of speech perception and spoken word recognition usually is not possible until a child approaches his or her third birthday. Prior to that, parent questionnaires are used clinically to document auditory skill development prior to and after cochlear implantation. When a child reaches the age of 3 years, more formal testing becomes possible. Young children or those with a developmental delay may have limited vocabularies, and this is exacerbated in young children with severe-to-profound hearing loss. Children with normal hearing may be able to repeat unfamiliar words or sentences. However, children with hearing loss have to "fill-in-the-blanks" in the degraded auditory signal they receive from a sensory aid, which is difficult if their vocabulary is severely delayed. A vocabulary test such as the Peabody Picture Vocabulary Test (Dunn and Dunn 2007) or parent report (e.g., MacArthur-Bates) (Fenson et al. 2007) can be administered to determine a child's vocabulary age. This should give the clinician some idea of whether test vocabulary is appropriate for that child. If necessary, test vocabulary can be reviewed with the child prior to testing.

The goal of speech perception testing is to assess listening skills. Speech perception test items should be administered in an auditory-only format or an auditory-plus-visual format without sign language. However, children who use some form of sign language in conjunction with spoken language should be provided instructions and allowed to respond in their preferred communication mode.

As noted above, stimulus characteristics such as vocabulary level can affect performance on a given test of spoken word recognition. The way in which the stimuli are presented also impacts test difficulty. Finally, the response format employed will influence a child's ability to perform the task, as well as its ecological validity (i.e., how well performance on the test reflects listening skills in daily situations). In this section we review major stimulus and response characteristics to consider in test selection and administration. Participant factors and test characteristics interact. Both should be considered when selecting and administering a speech perception test.

Presentation Characteristics

Ideally, spoken word recognition tests should be presented using recorded stimuli because performance can vary as a function of the talker producing the stimuli. Recorded test materials allow for longitudinal comparisons within a given child or cross-sectional comparisons across children. Of course, this is only true if the same recorded versions of the tests are used over time or at different centers. Introducing a different recording presents the same problems for reliability as using live-voice testing. Live-voice testing may be preferred when testing very young children. Typically, live-voice presentation occurs when clinicians are seated at a table with a young child, engaging them in, and monitoring their attention to the task. This allows the clinician to present stimuli when the child is engaged and ready to listen. Most children can participate in recorded testing by the age of 4 or 5 years.

For many years recorded test stimuli used to assess spoken word recognition in children with cochlear implants were presented at 70 dB SPL. However, this presentation level is more intense than conversational speech and may overestimate a child's speech recognition ability. In a study of adult cochlear implant users' spoken word recognition, Firszt et al. (2004) compared speech recognition when stimuli were presented in quiet and in noise at 50, 60, and 70 dB SPL. In quiet, 60 and 70 dB presentation levels yielded similar speech recognition scores; performance did not decline until presentation level was reduced to 50 dB SPL. However, in noise, performance at 60 dB SPL was significantly poorer than at 70 dB SPL. More recently, Davidson et al. (2011) demonstrated similar stimulus presentation level effects in children. Most current speech recognition tests recommend stimulus presentation levels of 60 or 65 dB SPL.

Traditionally, spoken word recognition tests employed a single male talker producing the stimuli using carefully articulated speech. Examples include the Phonetically Balanced Kindergarten Word List (PBK) (Haskins 1949) and the Hearing in Noise Test for Children (HINT-C) (Gelnett et al. 1995), two of the tests used most widely in the United States to assess speech perception in children with cochlear implants. Such tests with highly constrained stimulus characteristics may overestimate daily listening skills. More

Table 9.1 Closed-set tests of spoken word recognition for children

Test	Stimulus format	Presentation mode	Test condition	Stimulus mode	Response set	Test age
ESP-Low verbal[a]	1-, 2-, or 3-syllable words, Spondees	Monitored-live-voice (MLV) or Recorded	Quiet	Audio	Objects	2+ years
ESP-Standard[a]	1-, 2-, or 3-syllable words, Spondees	MLV or Recorded	Quiet	Audio	Picture	3 years+
WIPI[b]	Monosyllabic words	MLV or Recorded	Quiet	Audio	Pictures	4 years+
PSI[c]	Monosyllabic words, sentences	Recorded	Quiet	Audio Visual Audiovisual	Pictures	3 years+
CRISP[d]	Spondees	Recorded	Adaptive testing in quiet or in noise	Audio	Pictures[e]	4 years+
CRISP-Jr[f]	1- or 2- syllable words	Recorded	Adaptive testing in quiet or in noise	Audio	Pictures[e]	2.5–3 years

[a]Moog and Geers (2012)
[b]Ross and Lerman (1979)
[c]Jerger et al. (1980)
[d]Litovsky (2005)
[e]Test words are familiarized before testing. The words that are not easily identified by a child are eliminated from the closed-set choices
[f]Garadat and Litovsky (2007)

recent tests of open-set word recognition, such as the Pediatric AZBio test (Spahr et al. 2012), employ multiple talkers using a conversational speaking style. Other tests have been made available in both a single-talker and multiple-talker version. For example, the Multisyllabic Lexical Neighborhood Test and the Lexical Neighborhood (Kirk et al. 1995) are available in a male single-talker version and in a multi-talker version using both male and female talkers (see Table 9.2).

Auditory-only speech recognition remains the gold standard for assessing cochlear implant outcomes. However, not all children achieve high levels of auditory-only speech recognition, especially in noise. Furthermore, in daily listening situations, children have access to both auditory and visual speech cues. Visual speech cues provide information about place of articulation that complements the manner of articulation and voicing cues well-conveyed by a cochlear implant. Sumby and Pollack (1954) were among the first to demonstrate that adding visual speech cues substantially improves speech understanding in difficult listening situations. Whenever possible, both auditory and auditory-plus-visual assessment of speech understanding in noise should be conducted.

Response Format

Response parameters must be suitable for the child's age, attention span, and speech intelligibility skills. Closed-set tests, wherein children are given a limited number of response alternatives from which to choose a response, reduce vocabulary demands. Most closed-set tests assess perception of isolated words, although tests of closed-set sentence recognition are available (e.g., the Pediatric Sentence Intelligibility Test [Jerger and Jerger 1982; Jerger et al. 1980]). When a closed-set response format is used, the child does not have to search his or her entire mental lexicon. Thus, closed-set tests limit the demands on higher-order processing skills and linguistic knowledge that influence open-set spoken word and sentence recognition (see Fig. 9.5).

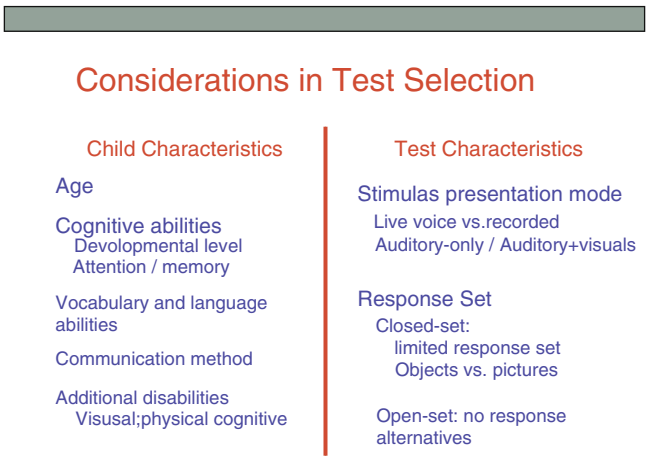

Fig. 9.5 Child and test characteristics that influence speech recognition test outcomes

Closed-set tests generally employ pictures or objects, which helps maintain interest and attention in the task. Closed-set tasks allow for pointing responses which is helpful in young children or those who cannot provide intelligi-

Table 9.2 Open-set speech perception tests for children

Test	Stimulus format	Presentation mode	Test condition	Stimulus mode	Lists	Test age
MLNT[a]	2- or 3-syllable words	Recorded 1 male talker or 2 male and 3 female talkers	Quiet	Audio	2 lists of 24 words	3 years+
LNT[a]	Monosyllabic words	Same as MLNT	Quiet	Audio	2 lists of 50 words	4 years +
PB-K[b]	Monosyllabic words	Recorded	Quiet	Audio	4 lists of 50 words[c]	5 years+
CAVET[d]	1-, 2-, or 3- syllable words	Recorded (CD-ROM, VHS)	Quiet	Audio Visual A+V	3 lists of 20 words	7–9 years+ children with profound prelingual hearing loss
BKB[e]	Sentences	Recorded	Quiet	Audio	21 lists of 16 sentences	6 years+
HINT—C[f]	Sentences	Recorded	Quiet or in Noise (Adaptive)	Audio	13 lists of 10 sentences	5 years+
AV-LNST[g]	Sentences	Recorded (QuickTime movie files)	Quiet	Audio Visual A+V	6 lists of 8 sentences	4 years+
Pediatric AzBio[h]	Sentences	Recorded	Quiet or in multi-talker babble	Audio	16 lists of 20 sentences	5 years+
MLST-C[i]	Sentences	Recorded (.avi video files)	Quiet or in speech-shaped Noise	Audio Visual A+V	21 lists of 8 sentences	3 years+

[a]Kirk et al. (1995)
[b]Haskins (1949)
[c]Although there are four lists developed by Haskins (1949), only lists 1, 3, and 4 are similar in difficulty (Meyer and Pisoni 1990)
[d]Tye-Murray and Geers (2001)
[e]Bench et al. (1979)
[f]Gelnett et al. (1995)
[g]Holt et al. (2011)
[h]Spahr et al. (2012)
[i]Kirk et al. (2012)

ble verbal responses. The difficulty of closed-set tests depends on the foils presented with the target words. The larger the number of foils, and the more phonemically similar they are to the target word, the more difficult the task. Some tests, such as the Minimal Pairs Test, use foils that are similar to the target in all but one speech feature. Because these tests emphasize assessment of sensory input while minimizing top-down processing, they are useful in identifying speech features that are well conveyed by a sensory aid. Table 9.1 lists some commonly used closed-set tests of speech perception and their test characteristics.

Although closed-set tests are useful with young children and children with limited speech perception skills, as in the early stages of cochlear implant use, they do not reflect "real-world" listening demands. As soon as possible, open-set tests of spoken word recognition, wherein the child must repeat the word or sentence without response alternatives, should be employed. Open-set tests require the child to produce some sort of verbal, written, or signed response. Open-set tests of both word and sentence recognition are widely used. Table 9.2 lists auditory-only tests of spoken word recognition that have been used widely to assess cochlear implant outcomes.

Speech Perception Outcomes in Children with Cochlear Implants

Over the last 20 years, there have been numerous longitudinal studies examining spoken word recognition in children with cochlear implants, as well as factors that affect performance. It has been well documented that children with multichannel cochlear implants obtain substantial levels of speech understanding in quiet. Furthermore, they leverage this skill to produce speech that is highly intelligible to naïve listeners (Chin et al. 2003), and to acquire receptive and expressive language skills (Davidson et al. 2014; Niparko et al. 2010). A variety of factors have been shown to influence these gains; outcomes are enhanced when children are implanted at a young age (Barnard et al. 2015; C. C. Dunn

et al. 2014; Eisenberg et al. 2002; A. E. Geers et al. 2000; Kirk et al. 2002; Miyamoto et al. 1989; Peterson et al. 2010), are in home and educational environments where spoken language is emphasized (Dettman et al. 2013; A. Geers et al. 2003; Kirk et al. 2002; Peterson et al. 2010), and have greater residual hearing prior to implantation (Gratacap et al. 2015; Holt et al. 2005). Family characteristics also have been shown to influence outcomes (Holt et al. 2013; Marnane and Ching 2015; Peterson et al. 2010; D. Wu et al. 2015). In this section, we will review long-term speech perception outcomes in children with cochlear implants and examine factors that are associated with successful outcomes in several longitudinal studies.

In the early days of pediatric cochlear implantation, the performance of children with cochlear implants was compared to that of their peers who used hearing aids (A. E. Geers 1997; Hesketh et al. 1991; Miyamoto et al. 1995; Osberger et al. 1991; Svirsky and Meyer 1999). Such studies produced converging evidence that cochlear implants yielded more benefits than hearing aids in children with profound deafness, and led to a broadening of cochlear implant candidacy to include children with greater residual hearing. Many of the children with residual hearing also obtained substantial speech perception gains after implantation (Carlson et al. 2015; Holt et al. 2005; M. L. Hughes et al. 2014). Another approach was to use each child with a cochlear implant as his or her control, and to compare performance longitudinally over time. More recently, researchers have begun to compare auditory, speech, and language development in children with cochlear implants to that of their typically developing peers with normal hearing. In this section, we will review long-term speech perception outcomes in children with cochlear implants and examine factors that are associated with successful outcomes in several longitudinal studies. Children in these studies were considered "traditional" cochlear implant candidates and were not identified as having auditory neuropathy spectrum disorder. Cochlear implant outcomes in children with auditory neuropathy spectrum disorder are presented in Chap. 14, which discusses implantation and outcomes in "non-traditional" pediatric populations.

Longitudinal Outcomes

One of the most comprehensive longitudinal studies to examine cochlear implant outcomes was conducted by Geers and colleagues (Davidson et al. 2011; A. Geers and Brenner 2003; A. Geers et al. 2003; A. E. Geers et al. 2011a, b; Tobey et al. 2011). Between 1996 and 2000, 181 children were enrolled in the initial study. They were between the ages of 8–9 years at the time of study entry and had received a cochlear implant prior to age 5 years. All children were prelingually, profoundly deaf and had between 4 and 7 years of cochlear implant use. Children and family member(s) attended a 3-day "cochlear implant research camp" during the summer that included educational and social activities for the child and his or her family. Children were administered a battery of tests and parents completed questionnaires to assess speech perception, speech production, language skills, reading, and psychosocial development. A primary purpose of this study was to evaluate the impact of educational setting on cochlear implant outcomes in children. As Geers and Brenner (2003) pointed out, a number of pre-existing factors can impact outcomes separately from educational setting and these had to be accounted for in their data analyses. Therefore the study documented child, family, and educational characteristics in their sample. The children were drawn from throughout the United States and Canada. On average, families had higher socioeconomic status than the general population. Initially, children were enrolled in a wide range of educational options, with similar distributions across public and private schools, special education classrooms, and mainstream classrooms. Both children who used Oral Communication and children who used Total Communication (i.e., the combined use of signed and spoken English) were enrolled. With increased duration of cochlear implant use, children were more likely to be in educational environments that emphasized the use of listening and speaking skills in the classroom. Geers et al. (2003) reported speech perception results for children in the study. On average, children demonstrated 50 % speech recognition on open-set tests, and this increased to 80 % when both auditory and visual speech cues were available. Good speech perception skills were significantly associated with greater nonverbal intelligence, smaller family size, longer use of the most recent cochlear implant processing strategy, a fully inserted electrode array, greater dynamic range between threshold and maximum comfort stimulation levels, and a greater growth of loudness with increasing intensity of the stimulus. After controlling for these factors, Geers and colleagues found that the primary rehabilitative factor associated with good speech perception was placement in an educational setting that emphasized oral-aural communication.

In the second part of this longitudinal study, Geers and her colleagues examined cochlear implant outcomes in 112 of the original study participants when they were between 15 and 18 years of age and had used their cochlear implant for an average of approximately 13 years (Geers et al. 2011a). The participants and their families again attended a research camp where they were administered standardized tests of speech, language, and reading, as well as measures of executive function and working memory. The participants were asked to complete questionnaires intended to probe their social development, and affinity with the hearing community, the Deaf community, or both communities.

Geers et al. (2011a) reported that 72 of the participants in the first study did not return for the second study. Of these, 22 families could not be located and 49 did not wish to travel to St. Louis for study participation. Characteristics of the participants that returned and those that did not were compared. They did not differ on age at implantation, performance IQ, or communication mode rating. However, the participants who returned demonstrated higher open-set sentence recognition, reading, and speech production intelligibility at 8–9 years of age than the participants who did not return. This illustrates one challenge in longitudinal studies: families of children who are performing well are more likely to remain in the study than families of children making more modest communication gains. In this second study, the authors compared the performance of children with CIs to that of children with normal hearing in two ways. First, when they administered standardized tests that were normed on children with normal hearing, they analyzed implant recipients' standard scores. When non-standardized tests were used, a control group of 46 age-matched children with normal hearing was administered the tests. The control and cochlear implant groups were similar in terms of age at testing, socioeconomic status, and family size.

The second study analyzed speech perception skills in this sample of 112 pediatric participants (Davidson et al. 2011). Specifically, they examined: (1) changes in open-set word and sentence recognition between elementary and high school, (2) changes in the ability to distinguish words on the basis of fine spectral cues, (3) the impact of a degraded auditory signal on speech perception performance in high school, (4) changes in the benefit provided by the addition of visual cues to the auditory speech signal, and (5) the relationship between open-set speech recognition performance and speech and language skills. Results revealed that both open-set word and sentence recognition improved significantly between elementary and high school. Mean word recognition scores were 50.6 % and 60 %, respectively, at the elementary and high school testing periods. Mean sentence recognition scores improved from 63.2 to 80.3 % over the same time period. However large variability was noted in both word and sentence recognition scores, with scores ranging from 0 to 100 % across word and sentence tests. In order to examine the use of fine spectral cues in speech recognition, the authors compared performance on the easy and hard words on the Lexical Neighborhood Test (Kirk et al. 1995). Words in the easy category have few phonemically similar words, whereas words in the hard category are phonemically similar to many words. Thus hard words require finer spectral distinctions for correct identification. At both time periods, easy words were recognized with significantly greater accuracy than hard words. To examine the impact of a degraded auditory signal, children in the second study were administered a test of open-set word recognition in quiet at 70 dB SPL and 50 dB SPL. An open-set sentence test was administered at 70 dB SPL in quiet and at 60 dB SPL in multi-talker babble with a signal-to-noise ratio of +10 dB. As stimulus presentation levels were reduced, word recognition declined significantly from 60 to 47 %. Sentence recognition declined from 80 % in the quiet condition to 52 % when presentation intensity was reduced and multi-talker babble was introduced. The addition of auditory cues to visual cues provided significant speech perception benefit at both the elementary and high school test intervals, with gains of 43 % and 45 %, respectively. Finally, the second study revealed that improvements in speech perception scores between elementary and high school were closely associated with concomitant changes in speech production and language, up to an age-equivalent language score of 10 years. Furthermore, those children with better vocabulary and syntax skills in elementary school demonstrated the most advanced literacy skills in high school (Geers et al. 2011b).

The Childhood Development after Cochlear Implantation (CDaCI) (Fink et al. 2007; Niparko et al. 2010; Wang et al. 2008) headed by Dr. John Niparko and a team of investigators is a multicenter, longitudinal national cohort study that compared pediatric cochlear implant recipients with aged-matched hearing peers. Eligibility criteria for the study included enrollment prior to age 5 years, normal cognitive and motor development, and commitment to educating children in English-speaking schools. Both unilaterally and bilaterally implanted children were eligible. Children were excluded from the study if they did not meet the above criteria, or if they had postsurgical complications, had families that could not participate, or if English was not spoken at home. As described by Fink et al. (2007), the primary outcome measure was oral language development. Assessment of secondary outcomes included spoken word recognition, cognitive processing (attention and problem-solving skills), behavioral and social skills, social adjustment between parent and child, and health-related quality of life and cost-effectiveness. Participants were tested every 6 months for 3 years after enrollment. A total of 188 children with cochlear implants and 97 children of similar age with normal hearing were enrolled in the study. Participant characteristics including age, gender distribution, ethnicity, socioeconomic status, and parental education were tracked. For participants with cochlear implants, etiology of hearing loss, type of communication method, and type of preschool or therapy program were also tracked.

Because of the age range of the children and the longitudinal nature of the study, it was necessary to use a variety of speech perception assessment materials. Test administration followed a speech recognition hierarchy ranging from parental reports through pattern perception (i.e., speech recognition based on duration and stress cues) and word and sentence tests in closed- and open-set response formats (Wang et al.

2008). Test selection for a given child was based on his or her age and functional hearing abilities. For each level in the test battery hierarchy, children had to reach a criterion level of performance before moving on to the next level. Testing was discontinued when a child reached ceiling on two consecutive intervals. This approach minimizes floor and ceiling effects while reducing test time and child frustration.

A speech recognition cumulative index was calculated to represent each child's speech perception performance in quiet (Wang et al. 2008). At baseline, the children with normal hearing demonstrated ceiling performance on the parent report scales and nearly all had reached ceiling on tests of open-set word recognition. One-third of the children with normal hearing were administered tests of open-set word recognition in quiet, resulting in a mean score of 94 % sentences correct. In contrast, at baseline the children with cochlear implants lagged far behind on parent report scales. Twenty-five percent of the cochlear implant users were administered a test of pattern perception, resulting in a mean score of 58 %. No child with a cochlear implant was administered a word or sentence recognition test at baseline.

Data from the 24-month study interval revealed that both groups had made gains in their speech perception abilities compared with baseline performance. The control group of children with normal hearing achieved a mean percent correct on the Multisyllabic Lexical Neighborhood Test or the Lexical Neighborhood Test (Kirk et al. 1995) of 60–70 % at 2.5 years of age, and approached 100 % correct by 6.5 years of age. Results for children with cochlear implants varied, but most could be administered the MLNT or LNT after 24 months in the study. Wang et al. concluded that the majority of children implanted at younger ages demonstrated similar growth trajectories in speech recognition skills as the children with normal hearing, but age at implantation was not reported.

Niparko et al. (2010) subsequently reported on outcomes from the CDaCI study after 3 years. The primary outcome was spoken language production and comprehension as measured by the Reynell Developmental Language Scales (Reynell and Gruber 1990; Reynell and Huntley 1985). The reported mean age at enrollment was 2.2 years and 2.3 years for the cochlear implant and control groups, respectively. Children with cochlear implants were stratified by age at implantation as follows: <18 months ($n = 72$, or 38 %), 18–36 months ($n = 64$, or 34 %), and >36 months ($n = 52$, or 28 %). The children's speech recognition abilities showed similar trajectories to that of their peers with typical hearing. However, a different pattern emerged for language comprehension, the most difficult skill on the auditory comprehension hierarchy (see Fig. 9.1). Although the cochlear implant group made significantly greater gains in spoken language over the 3-year period than would have been predicted from their preimplant baseline scores, they remained delayed when compared to their peers with normal hearing. Younger age at implantation was associated with significantly steeper trajectory in language comprehension and a smaller gap in performance between the children with CIs and those with normal hearing. The performance gap remained consistent for children implanted before 18 months of age. In contrast, the gap continued to widen for children implanted after the age of 18 months.

Niparko et al. used a multivariate analysis to examine factors that were associated with improved outcomes. Gains in speech recognition were significantly associated with improvements in verbal language. Greater preimplant residual hearing, higher rates of parent-child interactions, and higher socioeconomic status were associated with greater gains in language expression and comprehension. Improved language comprehension was not associated with gender, congenital onset of sensorineural hearing loss, baseline cognitive performance, or the exclusive use of spoken language at baseline. Bilaterally implanted children performed similarly to unilaterally implanted children after adjusting for the effect of other variables. Children who had a longer period of time between onset of hearing loss and implantation showed reduced levels of language improvement, whether or not they used a hearing aid during that time.

The above longitudinal studies demonstrate that early implantation has a significant impact on speech recognition and language skills. However, there are other conflicting findings. Gantz and his colleagues examined the effects of age at implantation on speech recognition and language abilities in a group of 83 children who received a cochlear implant by age 4 years. All children had prelingual hearing loss. They were divided into two groups: those implanted prior to age 2 years and those implanted between age 2 and 3.9 years. Data were obtained at annual intervals over a course of 5–13 years for speech recognition performance and 7–11 years for language and reading skills. The results suggested that age-at-implant effects evident in the early post-implant period may diminish with increased device use. There was no significant effect of age at implantation for language comprehension by 8 years of age, for expressive language by 10 years of age, and for reading by 7 years of age. There was no significant difference in speech perception scores between the two age-at-implant groups at ages 7, 11, and 13 years. The authors pointed out that younger children showed ceiling effects, and that a stronger age-at-implantation effect may have been evident over time if more difficult speech perception materials had been introduced. Communication method also impacted speech perception outcomes. Children who used oral communication showed higher speech perception scores than children who used Total Communication. Language comprehension and reading did not differ significantly as a function of communication mode. Similar language performance between the two

groups is not surprising given that children who used Total Communication were tested using both signed and spoken English. Dunn et al. reported that 69 % of the children implanted before 2 years and 50 % of the children implanted between 2 and 3.9 years used oral communication in this sample of children followed over 7 years of device use.

Bilateral Cochlear Implantation Outcomes

Broadening of cochlear implant candidacy and technological advances have allowed children with cochlear implants to achieve unprecedented levels of speech recognition and spoken language processing. However, challenges remain and chief among these is speech perception in noise. Bilateral implantation was introduced in the hopes of improving sound localization and speech perception in noise. In the next section, we will briefly review bilateral cochlear implant outcomes. A more thorough review can be found in Chap. 10.

The benefit of binaural input for normal-hearing listeners in speech recognition-in-noise tasks is well established, and can be at least partially attributed to the head-shadow effect, interaural level differences (ILDs), interaural time delays (ITDs), and binaural summation (van Hoesel and Tyler 2003). The rationale behind bilateral cochlear implantation is that bilateral cochlear implant users may be able to obtain some of the binaural benefits of normal-hearing listeners. The body of evidence concerning speech perception outcomes for bilateral cochlear implant users continues to grow as children undergo either simultaneous or sequential bilateral cochlear implantation. As this population ages, it is becoming more feasible to examine the long-term speech perception abilities of these bilaterally implanted children. Specifically, it is now possible to compare outcomes for children with unilateral and bilateral implants at multiple stages in their speech and language development.

One factor that may influence the benefit of bilateral cochlear implants is duration of use in each ear. CIs are often implanted sequentially, and depending on the delay between the first and second implantation, the child will have more or less experience with his/her first cochlear implant and a longer or shorter duration of deafness in the later-implanted ear. Most children who receive sequential implants have a positive attitude toward their second implant, and use the bilateral configuration full time (Galvin et al. 2014). Parents of children with bilateral CIs reported that their children performed better at sound localization, required less repetition in an auditory task, and were more responsive to sound in general (Galvin et al. 2014). These subjective reports are in line with studies that have shown a bilateral over unilateral cochlear implant advantage in perceptual tasks like melody localization (Beijen et al. 2007), speech recognition in noise (Wolfe et al. 2007), and speech localization (Galvin et al. 2008).

Johnston et al. (2009) conducted a meta-analysis of bilateral pediatric cochlear implantation, including 13 studies examining speech recognition in noise. Of those 13 studies, 11 reported that pediatric bilateral cochlear implant users had superior speech recognition in noise compared with their unilaterally implanted peers. In the two studies that did not report this benefit, the participants had longer delays between sequential implantation than children in the other studies. Although this suggests that duration of deafness in the later-implanted ear may impact outcomes, Johnston et al. noted that this association was not consistently reported across studies.

Sparreboom et al. (2015) reported long-term outcomes in a cohort of 30 children with sequentially implanted bilateral cochlear implants. All children were implanted with a unilateral cochlear implant before age 3 years and received the contralateral cochlear implant by the age of 8.5 years. None of the children had any additional handicapping conditions, or ossified or malformed cochleae. After 5–6 years of bilateral implantation, 25 of 30 children continued to use both CIs; one had a soft device failure and the remaining four voluntarily chose not to use the second implant. Speech and language outcomes in the remaining children were compared with a group of unilaterally implanted children matched by age at first CI, age at testing, duration of first cochlear implant use, and age at entering mainstream education. None of the unilaterally implanted children used a hearing aid in the contralateral ear. All children in both groups spoke only Dutch. Measures of localization, phoneme recognition in quiet and in noise, and vocabulary recognition were administered. At the time of testing, the mean duration of first or only cochlear implant use across the two groups was 8.5 years. The speech recognition and language scores were subjected to linear regression analyses with entered predictor variables of group (bilateral vs. unilateral), educational placement (mainstream vs. a school for the deaf), and duration of first or only cochlear implant use. Results revealed that 22 of 24 children with bilateral CIs were able to lateralize sound location at significantly greater than chance levels. After 8.5 years of first or only cochlear implant use, the adjusted bilateral advantage on speech recognition performance in noise was 9.5 %. Across both groups of children, speech recognition in noise was higher for children educated in mainstream classrooms than for children educated in schools for the deaf. Both group and educational placement significantly impacted receptive vocabulary language performance. Bilaterally implanted children and children in mainstream educational settings had higher language skills. Speech recognition performance in noise was not a significant predictor of vocabulary recognition.

Phoneme, word, and sentence recognition tasks represent the gold standard of audiological evaluation, but they may not entirely account for the speech recognition benefits of bilateral cochlear implantation. Hughes and Galvin (2013) exam-

ined the listening effort expended by adolescent and adult cochlear implant users in a word-perception-in-noise task. Bilateral cochlear implant users were tested when using two implants vs. one. Although cochlear implant users demonstrated similar phoneme recognition scores when tested bilaterally vs. unilaterally, they expended less effort in the bilateral condition as demonstrated in a dual task paradigm employing a visual matching task. This result suggests that tests of phoneme perception fail to measure an important benefit of the second implant, namely reduced cognitive load during auditory processing. If speech recognition using bilateral cochlear implant input imposes less cognitive demands than unilateral cochlear implant use, then perhaps bilateral cochlear implant users have more processing resources to devote to other perceptual and cognitive activities (Sarant et al. 2014). The lighter cognitive load may also contribute to the improved language learning outcomes (Boons et al. 2012) that have been associated with bilateral cochlear implantation.

Some of the variation in bilateral cochlear implant speech recognition outcomes may be attributed to differences in physiological maturation between central auditory pathways that process information from the first and second cochlear implant. Auditory evoked cortical responses (e.g., the P1 latency) of children with a unilateral cochlear implant who were implanted before age 3.5 years are similar to those of their normal-hearing peers, suggesting similar development of the central auditory pathway (Sharma et al. 2002). A more recent study measured these auditory evoked cortical responses in 29 children with bilateral CIs. P1 responses were measured separately in each ear after 12 and 24 months of bilateral cochlear implant use (Sparreboom et al. 2014). P1 latencies were longer in the second-implanted than in the first-implanted ear. This suggests that there is a delay in central processing for the second CI, but longer-term assessments are necessary to determine if this delay persists later in life.

Taken together, the current body of evidence surrounding speech recognition with bilateral cochlear implants seems to suggest a bilateral advantage over unilateral cochlear implant use in certain situations (e.g., speech recognition in noise). It is important to note that outcomes for bilateral cochlear implant users are very heterogeneous. The benefit of the second implant may be mitigated by a number of factors, including age at implantation and duration of auditory deprivation in each ear.

Combining Electric and Acoustic Input on Contralateral Ears

With broadening of cochlear implant candidacy to include children with greater residual hearing, some children are now using a unilateral cochlear implant combined with low-frequency acoustic information provided by a hearing aid on the contralateral ear (bimodal configuration) or in the same ear (electroacoustic stimulation, or EAS). A number of investigators have examined the potential benefits of bimodal fitting.

Ching et al. (2001) investigated sound localization and speech recognition in children with unilateral CIs who used a hearing aid in the contralateral ear. They also examined whether hearing aid fitting had to be adjusted when the hearing aid was combined with a cochlear implant in the contralateral ear. Children were tested in four different aided conditions: cochlear implant with hearing aid as it was typically worn, cochlear implant alone, hearing aid alone, and cochlear implant with the hearing aid adjusted for the individual child. They found that it was necessary to adjust the gain of the hearing aid in order to loudness balance with the cochlear implant signal. Average results indicated that children obtained significant benefit in speech perception and localization when tested in the bimodal configuration with the gain-adjusted hearing aid as compared to the condition in which the hearing aid fitting was not adjusted to balance loudness.

Holt et al. (2005) conducted a study to determine whether pediatric cochlear implant users with residual hearing benefitted from using a hearing aid in the non-implanted ear. They also examined the time course over which potential benefits of bimodal implant use might emerge. The children all had preimplant severe-to-profound sensorineural hearing loss in the implanted ear and severe sensorineural hearing loss in the contralateral ear. Onset of hearing loss in both ears was prior to age 3 years. Additional inclusion criteria included no additional disabilities and use of current cochlear implant processing strategies. Twenty-two children met these criteria. Ten of the children continued to use a hearing aid in the contralateral ear after implantation and 12 did not. The children were administered the Phonetically Balanced Kindergarten Word List (Haskins 1949) and the Hearing in Noise Test for Children (Gelnett et al. 1995) prior to cochlear implantation and at approximately regular 6-month intervals for 1–2 years after device activation. The bimodal cochlear implant users were tested in three conditions: cochlear implant alone, hearing aid alone, and cochlear implant plus hearing aid. No adjustments were made to the children's hearing aid fitting, and loudness balancing was not carried out. Performance did not differ significantly among the three conditions at 1-year postimplantation. After 2 years of cochlear implant use, significant speech perception differences emerged as a function of cochlear implant configuration. The largest differences were noted when the HINT-C was administered in noise at +5 dB SNR. Performance in the CI-plus-HA condition was approximately 30 percentage points higher than in cochlear implant alone condition, and approximately 40 percentage points higher than the HA alone condition. Thus, even when hearing aids were not adjusted, children obtained substantial bimodal benefits, especially in noise.

More recently, Mok et al. (2010) investigated the effect of using a hearing aid in bimodal configuration or a second cochlear implant on speech recognition in noise. They also investigated the ability to perceive phoneme groups of different frequencies in the children fit with bilateral or bimodal cochlear implant configurations. Nine bimodal cochlear implant users and four bilateral cochlear implant users participated. All 13 participants had a severe-to-profound hearing loss that was assumed to be congenital. The bimodal group's mean ages at implantation and testing were 6.5 and 11.8 years, respectively. Mean ages at implantation for the bilateral group were 1.9 years for the first cochlear implant and 8.9 years for the contralateral cochlear implant. The average age at testing was 10.2 years for the bilateral group. Both groups completed loudness balancing tasks prior to speech perception assessment and adjustments to either the hearing aid or second cochlear implant were carried out as necessary. Speech recognition testing utilized the Consonant-Nucleus-Consonant Word Test presented from the front with four-talker babble at a SNR of +10 dB presented from the front, and at 90 and 180°. All subjects were tested in ear independent and then with the two devices in use. For the bimodal users, scores in the cochlear implant only condition were significantly higher that scores in the HA alone condition. For the bilateral cochlear implant users, performance with the first cochlear implant was greater to, or at least equal to, performance with the second cochlear implant. To evaluate the benefits of binaural input, performance in the best single-ear condition (cochlear implant only for bimodal users, and first cochlear implant for bilateral users) was used for comparison. The results demonstrated a significant binaural advantage for speech perception in noise for bimodal users as a group; six of the nine participants showed a significant bimodal advantage when individual data were analyzed. The results suggested a greater binaural advantage for participants with better aided thresholds at 250 and 500 Hz in the hearing-aided ear. Average results from the bilateral group showed a significant binaural advantage when noise was presented on the side of the first cochlear implant; two of the four individual participants showed this pattern of performance. None of the bilateral cochlear implant users experienced any decrement in performance when the second cochlear implant was added to the first. Mok et al. concluded that pediatric cochlear implant users should be fit with bimodal or bilateral cochlear implant configurations.

Conclusions

The majority of children who are born deaf or lose their hearing early in life have hearing parents who want their children to communicate orally and be a part of their hearing community. Prior to the advent of pediatric cochlear implantation, children with profound hearing loss struggled to acquire spoken language via input from a hearing aid. Today, children with severe-to-profound hearing loss who receive a cochlear implant demonstrate substantial gains in speech perception and they use this input to acquire speech production, language, and literacy skills that were previously unprecedented. It is clear that early implantation can prevent or minimize communication delays, and that children benefit from educational environments that emphasize the development of listening and speaking skills. Bilateral and bimodal cochlear implant use can improve speech recognition in noisy environments such as classrooms, but they do not restore normal spatial hearing or spoken language processing. Continued research is needed into individual differences in outcomes and intervention strategies to maximize cochlear implant benefit in all children.

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Binaural and Spatial Hearing in Implanted Children

10

Ruth Y. Litovsky

Introduction

Children with normal hearing (NH) utilize information that arrives at the two ears in order to perform a multitude of tasks in their everyday listening environments. In the field of audiology, the question regarding provision of auditory input to one vs. two ears has been around for many years. Several decades ago, questions revolved around bilateral amplification with hearing aids, and researchers generally thought that children should be fitted with amplification in both ears in order to maximize the stimulation of the right and left auditory pathways. Additional benefits that were of interest, but only measured in patients with usable hearing in both ears, were related to binaural benefits. The potential benefits from having two usable ears will be discussed in detail below as they relate to the binaural cues that are available to listeners when using acoustic hearing, or electric hearing through cochlear implants (CIs).

In the past decade there has been a steep increase in the number of children who are deaf and implanted bilaterally. At the start of the millennium, this clinical approach was somewhat novel and considered to be lacking in evidence regarding benefits. However, the clinical trend has shown momentum towards bilateral stimulation, with justification revolving around several main issues. First, if hearing is usable in both ears, and if the inputs arriving at the two ears are coordinated in the time domain, then the auditory system uses binaural hearing. That is, the brain receives crucial information regarding the location of sound sources, enabling listeners to localize sounds of interest. In this ideal scenario, the brain also compares inputs from the two ears in order to segregate speech from background noise. Second, under less ideal conditions, with inputs to the two ears arriving in an uncoordinated fashion, the auditory system receives bilateral hearing. There are crucial auditory cues that allow a listener to gain access to the target speech signal and to localize sounds in a fairly crude manner. Nonetheless, the access to this information can lead to improved hearing in everyday listening situations. Third, regardless of whether binaural or bilateral hearing is utilized, there are dual-implant assurances; the fact that both ears are stimulated has important benefits including assurance that the better ear was implanted, which is crucial for language acquisition, and also assurance that if one of the CI devices fails to operate the child will not be "out of sound." This chapter will first review binaural hearing and acoustics that can provide binaural and/or bilateral inputs. Second, this chapter will describe the methods that are used to evaluate bilateral and/or binaural benefits in children. Third, this chapter will review the measured outcomes as indicated through behavioral testing.

Binaural Cues

Throughout development, in most social and learning environments infants and children are faced with auditory signals that arrive from multiple locations; it is important to understand how acoustic inputs give rise to spatial cues when sound sources reach the ears. Sounds that occur in the horizontal plane and reach the ears from the side will naturally create differences in time of arrival between the ears, because sounds reach the near ear before the far ear. In addition, the near ear will have a greater intensity than the far ear. For example, as shown in Fig. 10.1a, for a sound arriving from 90° to the left, an adult head will have ~0.7 ms interaural timing difference (ITD) favoring the left ear. In particular, ITDs play a role at low frequencies (<1500 Hz). At high frequencies an acoustic "shadow" is created which results in interaural intensity (or level) differences (IIDs or ILDs)

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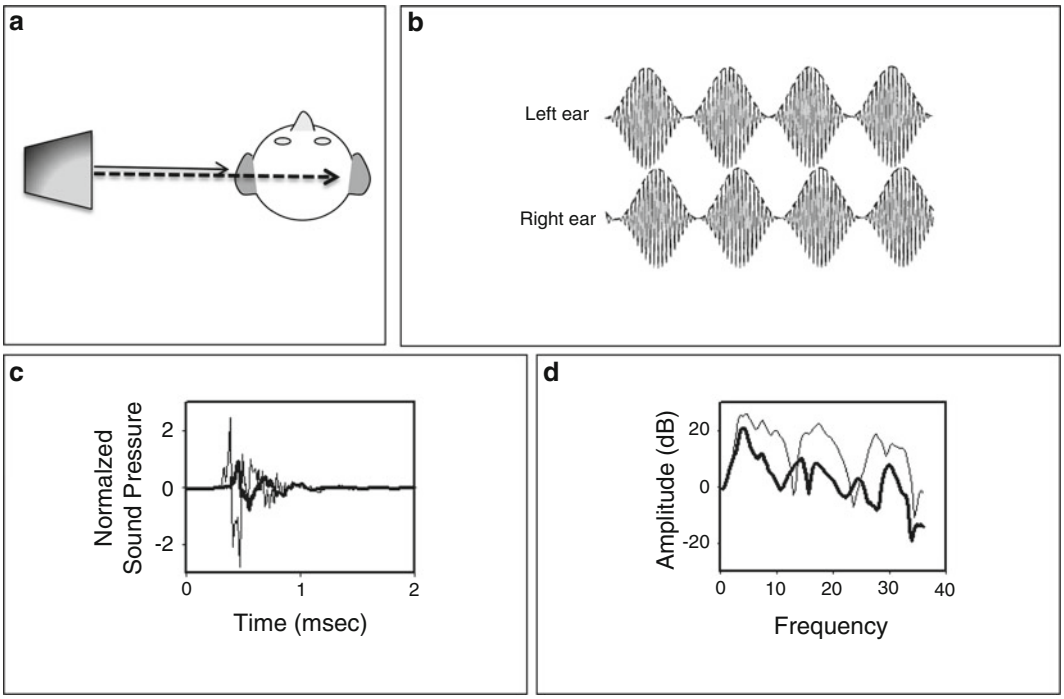

Fig. 10.1 Localization cues are depicted for a sound arriving from 90° to the left. (**a**) The head of an adult is shown with sounds arriving at the two ears, with a ~0.7 ms interaural timing difference (ITD) favoring the left ear. (**b**) An example of a high frequency stimulus with an amplitude modulation is shown, whereby ITD cues are also available from differences in the timing of the envelopes of the stimuli. (**c**) Stimuli reaching the two ears are shown on the same graph, to depict the interaural time difference between the *thinner* and *thicker lines* (left and right ears, respectively). The same stimuli, reaching the two ears, are shown to depict the difference in amplitude across the two ears

between the ears. IIDs or ILDs are frequency dependent but can be as large as 20 dB. For amplitude modulated sounds (e.g., speech) ITD cues are also available from differences in the timing of the envelopes (slowly varying amplitude) of the stimuli, as shown in Fig. 10.1b. Stimuli that reach the auditory system after entry through the ear canals, and that are coordinated across the ears in the time domain, will provide listeners with binaural cues. Examples of these cues are depicted in panels C and D, where differences between the ears are shown in the time domain (C) and in the spectral domain (D). It should also be noted that a different set of cues helps in the localization process for sounds that occur in the vertical plane. Those cues arise from directionally dependent filtering of sounds by the head and pinna. Thus, when sounds vary location in elevation, their spectral content is "shaped" differently for sources arriving from above, in front or below. CI processors inherently have degraded spectral resolution, and high frequencies are cut off above ~8000 Hz, rendering the availability of vertical-plane cues minimal or absent. Thus, the current chapter focuses on perceptual effects that have been studied in the horizontal plane. More detailed reviews of localization cues can be found in Blauert (1987), Middlebrooks and Green (1991).

One important note regarding development in early childhood pertains to the fact that head size changes as children grow, particularly during the first few years of life. In fact, a source arriving from 90° to the side will generate a substantially different set of binaural cues for a young infant than for an older child or an adult. Thus, as the head size changes throughout development, the correspondence between location and directional cues has to undergo constant recalibration (Clifton et al. 1988).

Methods Used to Evaluate Binaural and/or Bilateral Inputs

The patient population being evaluated determines which methods are appropriate for perceptual testing. When evaluating binaural hearing we are often interested in the general category of spatial hearing abilities, which include three primary areas: (1) sensitivity to binaural cues, (2) sound localization, and (3) speech understanding in noise. For all three areas, the easiest population to test is adults with NH, because listening is intuitive to them, and instructions about what aspects of the sound they should pay attention to and report on are fairly straightforward. Older children with NH, for similar reasons to those stated for adults, are also generally easy to involve in testing. However, the population of interest here is children who are deaf and fitting with CIs.

Devising tests is rather challenging for this population because listening is not always intuitive, and perception in the spatial domain is an emerging ability that is likely to depend on experience in a more protracted time scale than the emergence of spatial hearing in NH children. Some of the anecdotes collected in the Binaural Hearing and Speech Lab at the Waisman Center (discussed by Litovsky 2013) are informative regarding the issues that most fundamentally affect children who are fitted with bilateral CIs; these children often report that they do not perceive sounds as arriving from particular locations. They appear to develop these skills with experience, in particular by matching what they hear to what they see. Although little is known about the mechanism through which auditory-visual inputs are integrated in these children, the studies discussed below highlight the use of behavioral testing utilizing visual markers that enable the children to indicate where the sound sources are perceived to be.

Methods to Measure Sensitivity to Binaural Cues

Much of the literature that is related to binaural and bilateral hearing focuses on questions related to the acuity of the auditory system: the extent to which listeners are sensitive to small differences between sound source locations, or between ITD/ILD values that are presented to listeners over headphones. On any given trial, the values of ITD or ILD are presented over several intervals, and the listeners' task is to determine whether the sound was perceived towards the right ear or left ear. ITD or ILD values are typically varied using a staircase procedure, whereby the values are decreased following a correct response and increased following an incorrect response. The goal in the experiments is to provide the child with enough information to compare on two stimulus intervals; one example is a stimulus that is presented from the right followed by the left, or the left followed by the right. On any given trial, the child is asked to report whether a sound source was perceived to move right-left or left-right. Feedback regarding correct responses helps the child learn what cues to focus on and achieve best performance. A schematic of the temporal sequence of stimuli is shown in Fig. 10.2. Panel A shows stimuli presented over headphones to NH listeners; these stimuli can vary in content but typically consist of brief tone bursts or noise bursts. The first set of bursts shows a stimulus that reaches the left ear before the right ear; hence if the child is able to extract binaural cues from the stimulus, s/he will perceive a sound on towards the left ear. The second set of bursts have the opposite temporal sequence, with the right ear leading the left ear, thus the child would perceive a stimulus near the right ear. In an experiment, this trial type and one in which the opposite sequence

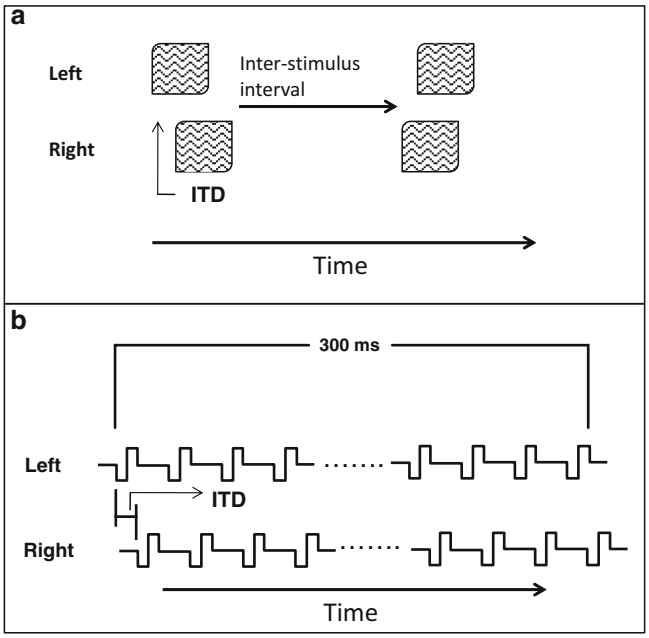

Fig. 10.2 (**a**) For stimuli presented over headphones schematics of the temporal sequence of binaural stimuli is shown. First, a pair of stimuli arrives at the left and right ear with an ITD favoring the left. Then, followed by a brief delay a second pair of stimuli arrive and the right and left ear with an ITD favoring the right. (**b**) Schematic of pulsatile stimuli presented to the cochlear implant arrays with binaural stimulation. In this example biphasic pulses are presented to the left ear followed by the right ear with a fixed ITD

occurs are presented in random order. Furthermore, the size of the ITD is varied, in order to find the smallest ITD for which the child can reliably hear the difference between left-right and right-left. In order to produce similar effects with ILDs (not shown), the levels of the stimuli in the two ears are adjusted to create perceptual images that are near the right or left ear, and a similar sequence of stimuli is presented.

Figure 10.1b shows examples of electrically pulsed signals that are used to make similar measurements, with deaf individuals who are fitted with bilateral CIs. This schematic (Litovsky et al. 2010) shows biphasic pulses, presented to select pairs of electrodes in the right and left ears, after extensive testing shows that the patient perceives the stimuli to produce similar pitch percepts, and that when the electrodes in the two ears are activated simultaneously, a fused auditory image is perceived (rather than independent sounds at the two ears). In order to establish precise levels of acuity, subject attention and motivation has to be very high, and thus little is known about binaural sensitivity in young listeners, particularly children with hearing loss.

A somewhat easier behavioral method that can be used to measure spatial hearing acuity in very young infants and children is typically done in the free field, using loudspeakers, but the same ideas as described above for ITD/ILD apply. Figure 10.3 shows a schematic diagram of loudspeakers

placed in a room at 10° increments; the locations of the loudspeakers can be set so they are flexible, in order to allow presentation of sounds at smaller intervals, for children who show sensitivity better than 10°. The aim of this behavioral test is to find the smallest change in the location of a sound that the infant or child can reliably discriminate. In the schematic diagram, the illustration is for sounds that are emitted from 0° (front), followed by presentation to the right or left. The size of angle is determined by the child's performance. Typically, larger angles are used first, and once it is established that a child can discriminate changes from the front to the right vs. left at larger angles, smaller angles are used. Testing is most efficient if conducted using an adaptive staircase procedure (see Litovsky 1997; Grieco-Calub and Litovsky 2012), and finding the angle at which performance is above chance (>70.9 %; Levitt 1971). Estimates of spatial hearing can also be obtained by fixing the loudspeaker locations for sets of 20 trials at a time, and obtaining data at numerous angles, then fitting the data to a psychometric function and finding the angle at which performance reached >70.9 % correct (Litovsky et al. 2006a). There are pros and cons to each of these methods, particularly for children with CIs for whom location information can be difficult to extract and who may need to first learn the task with the fixed-angle method before proceeding to the adaptive angle method. It is noteworthy that the adaptive staircase procedure has been used with NH infants as young as 6 months of age (e.g., Ashmead et al. 1991), and both methods have been used with toddlers who are either NH or who use CIs, age 2.5 years (Grieco-Calub and Litovsky 2012). The ultimate goal in this task is to assess auditory location acuity, known as the minimum audible angle (MAA), which is defined as the smallest change in a sound source location that the listener can discriminate accurately and reliably (e.g., Mills 1958; Litovsky and Macmillan 1994; Litovsky 1997). A more complex task, described below, is one in which children are tested on their spatial mapping ability, that is, on how well they know where a sound is coming from, rather than only discriminating its location based on hemifields. A significant issue to note here is that a child's ability to discriminate right vs. left might not automatically provide the necessary cues for a map of space and for accurate knowledge about where sounds are coming from (Hartmann and Rakerd 1989; Grieco-Calub and Litovsky 2010).

Methods Used to Measure Sound Location Discrimination and Sound Localization

In everyday environments, the listener is typically interested in finding a source of importance (such as the voice of a parent or teacher, a musical instrument or a toy), and the subsequent task is to be able to direct attention to the source, extract meaning from its content and respond to the content. These abilities are essential for achieving successful communication. To aid in this process, the ability to quickly identify the location of a source can be quite useful. In order to achieve this task, the listener needs to have a well-developed map of auditory space that organizes locations of sounds in the world relative the listener's head and relative to other sources in space. To date, research has produced a plethora of information about this ability in NH listeners, who have been tested through methods that incorporate verbal reports of locations (e.g., Wightman and Kistler 1989), eye gaze (Populin 2008), pointing towards the source location with the finger or head (see Middlebrooks and Green 1991), or pointing to a location on a proxy for space such as a spherical model of auditory space (Good and Gilkey 1996). The cognitive load required for these tasks might be high and training can take numerous hours before the data are repeatable. Thus, simplified versions of these tasks have been implemented in children. Nonetheless, the error rates observed in these experiments suggest that adults can generally localize sounds with a resolution ranging from a few degrees to ~10°.

When developing tests for young children, we have focused on utilizing ecologically valid methods that attract the children's attention, provide motivation, and provide results that are replicable. With children ages 4–5 years and older, interactive computerized testing platforms have been successful in that the children find the task intuitive and the response method is learned relatively quickly. The child typically sits in a room facing an array of loudspeakers, similar to the distribution shown in Fig. 10.3. A computer monitor placed under the loudspeaker in the front position displays the array, with icons corresponding to each location. On each trial a sound is emitted from one of the loudspeakers and the child uses a computer mouse to indicate on the computer monitor which loudspeaker emits the sound (Grieco-Calub and Litovsky 2010; Litovsky and Godar 2010).

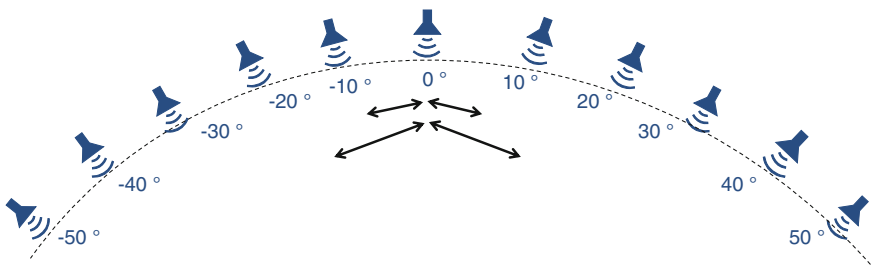

Fig. 10.3 Schematic diagram of loudspeakers placed in a room at 10° increments

Testing toddlers is, as indicated above, more challenging than testing older children, because instructions are more difficult to give, attention spans are much shorter, and redirection of attention away from distractions and to the task at hand can be more challenging. Towards that end, a novel method for assessing spatial hearing skills in toddlers was recently developed (Litovsky et al. 2013), whereby the child reaches for a sounding object that is hidden behind a curtain. The child faces an array of loudspeakers that are hidden behind an acoustically transparent curtain, and is shown a toy that will be the object of interest and attention. The toy is hidden behind the curtain at a location corresponding to one of the loudspeakers, and at the onset of the trial, the child hears a prerecorded voice from one of the locations, inviting them to find the object; by reaching through the correct space in the curtain the child can obtain the toy and is reinforced for a "correct" response. This Reaching For Sound (RFS) methodology lends itself to testing over dozens of trials with great interest on the part of most young children, and the method is successful with toddlers as young as 18 months of age. The RFS method is robust beyond sound localization measures and has been implemented in recent studies on speech perception and discrimination of toddlers with CIs (Hess CL. Speech discrimination and spatial hearing in toddlers with bilateral cochlear implants. Unpublished PhD Dissertation and University of Wisconsin-Madison 2013).

Methods Used to Measure Speech Understanding in Noise and Related Phenomena

The ability of a child to segregate speech from noise in complex auditory environments has been studied primarily in NH children, with a growing interest lately in understanding also how this ability emerges in children who are deaf and use CIs. There are some clear similarities between the two populations, under conditions that maximize spatial cues that both populations of children are able to hear. The goal of controlled experiments on speech intelligibility in noise is to measure the ability of children to identify the content of speech sounds that they know; rather than testing vocabulary, these tests only utilize stimuli that the children have been familiarized with, and are known to the children. A second goal is to create scenarios that mimic everyday listening situations, such as when a voice of interest is facing the child in front (target speech), and other voices (maskers) occur from locations that are either co-located with the target speech or spatially separated from the target speech.

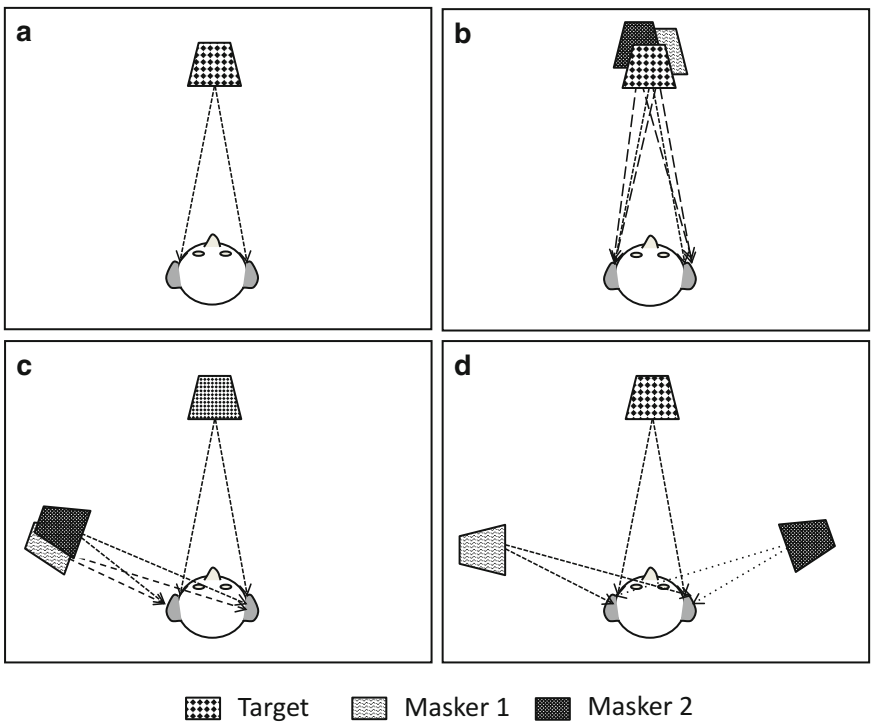

Fig. 10.4 (**a**) Target speech is presented from the front, in quiet. (**b**) Target speech is presented from the front and two maskers are presented from the front as well. (**c**) Here, there are two maskers and both are displaced towards the left ear. There are six signals in total, three at each ear from each source. However, the directional cues provided by the target are different from those of the maskers. In addition, as will be described below, the fact that the target speech reaches the right ear with a favorable signal-to-noise ratio (SNR) means that the "head shadow" effect creates a favorable listening condition. (**d**) In this symmetrial configuration the target speech is in front, and the two maskers are presented from the right and left, creating a situation in which there is no 'better ear' and the listener must use binaural cues to spatially segregate the speech from maskers

Figure 10.4 shows the four scenarios that are most informative about the ability of children to use spatial cues to segregate speech from background interferers, or maskers. Panel A shows the simplest case, in which the target speech is presented from the front, in quiet, and the stimulus reaches each ear. Panel B depicts an example of a masking condition with two maskers added to the front location. Each source emits sounds that reach both ears, creating a complex array of signals that need to be pulled apart in order for the child to extract meaning from the target speech. The benefit for speech intelligibility typically occurs when spatial cues are made available, in particular those shown in Fig. 10.4c; here, there are two maskers and both are displaced towards the left ear. There are six signals in total, three at each ear from each source. However, the directional cues provided by the target are different from those of the maskers. In addition, as will be described below, the fact that the target speech reaches the right ear with a favorable signal-to-noise ratio (SNR) means that the "head shadow" effect creates a favorable listening condition.

For many children, the condition shown in Fig. 10.4c results in an effect known as spatial release from masking (SRM), whereby performance is better compared with the condition in which maskers are co-located with the target (Litovsky 2005; Misurelli and Litovsky 2012). Performance is typically measured by obtaining the speech reception thresholds (SRTs) in quiet, as well as the co-located condition and the spatially separated conditions. SRM is thus quantified as the difference in SRTs between the co-located and separated conditions. In SRT terms, higher values indicate poorer performance, i.e., that a larger SNR was required in order for the child to correctly identify the target words. Thus, if SRTs are higher in the co-located than separated conditions, SRM would be positive, indicating that the child experiences a benefit when target/maskers are spatially separated. In other words, the child is able to take advantage of location cues in order to extract the meaning of the target words in the presence of the maskers.

Outcomes in Children Fitted with Bilateral Cochlear Implants

The following section described results from studies on binaural and spatial hearing that are relevant to pediatric bilateral CI users. As the data are considered, some of the limitations that occur in CI users will be discussed. These are summarized in Table 10.1.

Sensitivity to Binaural Cues

In the field of implantable auditory prostheses, the "gold standard" for testing exquisite levels of sensitivity to stimulation involves the use of direct electrical stimulation. This is unlike the free field, where the microphone picks up the signal and transmits it to the speech processor, which then acts on the signal in numerous additional ways. Instead, the microphone and speech processor are bypassed. Electrical stimulation is presented to the patient through research processors which allow the experimenter to stimulate electrodes along the cochlear array in a selective manner, and to tightly control the stimuli in each ear, at each electrode. In the case of binaural hearing this is particularly important, because the CI processors in the two ears are not temporally coordinated, which creates problems with the level to which ITDs are preserved with fidelity. In addition, the CI speech processor stimulates all electrodes at fixed-rate stimulation that is typically higher than the frequencies at which ITDs are easily encoded. In order to study binaural sensitivity one must therefore simplify the stimuli and maximize the possibility that patients will be able to extract information from the electrical pulses.

Figure 10.2b shows the type of stimuli that can be used, with simple trains of biphasic pulses, presented to select pairs of electrodes in the two ears. An ITD or ILD can then be imposed on these train pulses, to study the extent to which patients are sensitive to these cues. One key factor to keep in

Table 10.1 Limitations that occur in pediatric bilateral CI users

Site of limitation	Problem or limitation
Cochlear implant speech processor	Signal processing compromises acoustic cues: • Lack of temporal fine structure • Fixed-rate stimulation may not be ideal for capturing spatial hearing cues
Microphone	Compression distorts ILD cues
Cochlea	Spread of excitation along the basilar membrane leads to interaction amongst nearby electrodes
	Limited number of channels
	Poor specificity of stimulation on a frequency basis
Between the cochleae in the two ears	Potential mis-match in insertion depth, leading to mis-matched frequency inputs for electrodes that are anatomically matched in the two ears
Cochlea, auditory nerve	Neural degeneration; asymmetrical across the ears?
Binaural pathways	Degeneration of binaural circuitry due to lack of binaural inputs during development

mind is that binaural hearing in the acoustic system depends on the intrinsic wiring of inputs from the two ears whereby frequency-matched inputs are received at the level of the brainstem where ITD and ILD information is further processed. The studies on this topic in adults, in the past two decades, have shown that it is important to be able to stimulate electrodes that are matched by perceived pitch, because that indicates areas of the cochlea that stimulate auditory nerve fibers with the same frequency sensitivity (van Hoesel 2004; Litovsky et al. 2010). In fact, deliberate mis-matching of stimulation leads to the perception of binaural inputs diffuse or unfused, and those stimuli are poorly lateralized compared with pitch-matched inputs (Kan et al. 2013).

This background is critical towards our understanding of the issues that should be considered with young children who are bilaterally implanted, because at the clinical level the frequency allocation of information sent to the two ears is not deliberately matched by place of stimulation. Although there may be some matching across the ears by electrode number, if the two electrode arrays are not inserted with the identical insertion depth, a mis-match in frequency allocation across the ears is likely to occur. The extent to which children adapt to the potentially mis-matched inputs is not known. Further, little is understood regarding the extent to which children, whose neural pathways are stimulated at a time when neural plasticity is in place, are better than adults at compensating for this problem. Initial investigations on this topic suggest that children with bilateral CIs are able to use ILD cues to perceive sounds as occurring from the right or left; however, their ability to use ITD cues is poor. In contrast, when NH children are presented with a similar task using acoustic stimuli, they can reliably use either ITDs or ILDs to perform the same task (Salloum et al. 2010). This is not thought to be a developmental issue because ITD sensitivity on a right-left discrimination task is fairly well developed in NH children by age 4 (Van Deun et al. 2009): thresholds are reported to be, on average, 40 μs for 4-year-olds, 20–35 μs for 5–9-year-olds, and 12.5 μs for adults. The concern is that bilateral CI users are not receiving binaural inputs with fidelity during their everyday listening through their speech processors. Thus, when presented with these cues on a controlled experimental task, their auditory system may not be able to process the information in a useful manner. In contrast, ILD cues are received by the CI processors with greater fidelity, and all bilateral CI users seem to have sensitivity to those cues (for recent review, see Kan and Litovsky 2014). More recent and extensive studies in both NH children and in pediatric bilateral CI users are under way in the Litovsky lab at the University of Wisconsin-Madison. Results suggest that, similar to the Salloum et al. study, ILD sensitivity is easier to induce than ITD sensitivity. Moreover, children with onset of deafness after age 3 (postlingual) seem to have some access to ITD cues and perform better than children with congenital deafness (Ehlers et al. 2013; Litovsky 2011a, b; Kan and Litovsky 2014). The former are most likely able to rely on the fact that their auditory system was able to code that information prior to onset of deafness, and the cues that are provided during the experiments are stimulating pathways in the binaural system that had established ITD coding during development. This topic is of great interest in terms of future treatment of bilateral CI users, most of whom are congenitally deaf, and consideration should be given to advantages that might be gained through the development of CI processors that capture and present ITD cues.

Sound Location Discrimination and Sound Localization

In order to understand spatial hearing in children who are fitted with CIs, it is important to consider how the natural progression of spatial hearing emerges in NH infants and children. Thus, the standard to which CI users are compared can be considered in the context of expectations and rehabilitation. In NH infants, head orientation towards sound sources begins at birth as a reflexive response to an environmental stimulus. Newborns respond to sounds presented from the right vs. left in a reliable manner, although this head-orienting behavior is not conditioned and will only be observed for a limited number of trials (Muir et al. 1989). The head-orienting behavior is refined during the first 6 months of life and becomes an easily conditioned behavior through visual reinforcement (Moore et al. 1975); hence this has become a standard method of assessing auditory sensitivity in clinical audiology.

Using the head-orienting measure, studies with young infants have shown that the ability of infants to discriminate sounds to the right vs. left undergoes a steep maturational progression early in life. Summary of data from experiments described below is shown in Fig. 10.5a. MAA thresholds are near 25° at 2–4 months of age, decrease to approximately 10° by 6 months, and are as small as ~5° by 18 months of age (see Litovsky 1997). While MAA thresholds continue to mature into childhood, reaching 1° by 5 years of age, the 5° thresholds at 18 months suggest that young toddlers have a well-developed skill regarding discrimination of spatial cues at a prelingual stage in development. Studies described thus far used fixed-level stimuli, and it is possible that monaural level cues were available to the children. Thus, more recent studies have tried to minimize or eliminate overall level cues at each ear by roving the levels; thus the listener could solve the task by comparing the level cues at the two ears. Grieco-Calub et al. (2008) reported MAA thresholds near 10° for 2.5-year-old toddlers, and obtained slightly higher thresholds averaging 14.5° in a later study (Grieco-Calub and

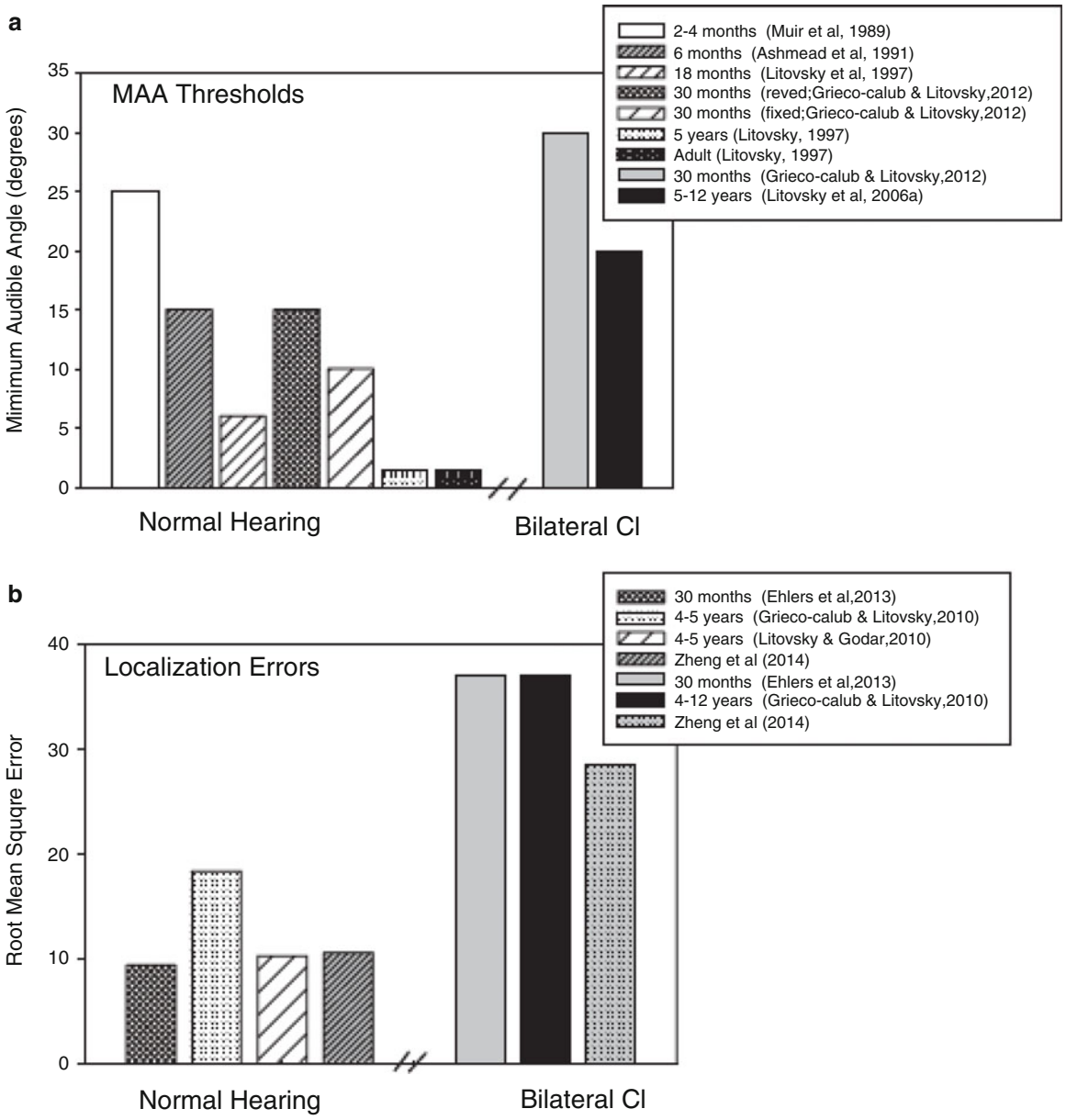

Fig. 10.5 (**a**) Summary data of minimum audible angle (MAA) thresholds are shown from a number of studies. (**b**) Summary data of root-mean-square (RMS) errors from localization measures are shown from a number of studies. With kind permission from Springer Science and Business Media (Litovsky 2011b)

Litovsky 2012). Thus, in young children with NH, localization acuity undergoes considerable maturation during the first 5 years of life, and the acuity of performance depends on the task and stimuli. During this time of life when the auditory system undergoes considerable maturation, there is an important interplay between the auditory inputs that are available to the children, the integrity with which the auditory system can process the information, and the ability of the listener to utilize those cues on everyday listening tasks.

In children who are deaf and who receive bilateral CIs, the ability to extract information regarding source locations to the right vs. left is complicated by the fact that they are typically not implanted in both ears until about 1 year of age; some children receive both CIs before a year of age, while other children are several years old at the time of the second implantation. The clinical practice regarding this issue varies and is beyond the scope of this chapter. Needless to say, there are many complications that are involved in determining the success of bilaterally implanted children, and some of the limitations known to us to date are included in Table 10.1. In some ways, it is quite remarkable that bilaterally implanted children are able to localize at all, and that

some of the children perform at levels that are within the performance levels observed in the normal hearing population. Summary of the data from bilaterally implanted children is shown in Fig. 10.5a, alongside the summary of results from NH children. Grieco-Calub and Litovsky (2012) tested 27 toddlers with an average age of 2.5 years, who received their second CI by 18 months of age. The MAA thresholds ranged from 5.7 to 69.6° (mean 31°). Unilaterally implanted toddlers were unable to perform the task, and the bilateral group was unable to perform the task if one of the CIs was removed, providing evidence for the use of a second CI when children discriminate sounds that are presented from the right vs. left. From an ecological standpoint, an average of 31° discrimination would provide these children with ample cues to know whether a sound of interest (voice, vehicle, etc.) is on their right or left. From a neuroscience perspective, the issue is more to do with the acuity of the neural mechanisms involved, and here there is a clear gap between the NH and bilateral CI groups. It is quite interesting that 5/27 toddlers tested had MAA thresholds within the range observed for the NH group, and all had more than 12 months of bilateral listening experience. Thus the role of auditory experience in the bilateral CI group might be an important factor in considering emergence of spatial hearing skills.

The head-orienting task, used for studies described thus far, has one potential flaw in relation to toddlers: a lack of ecologically interesting testing engagement. The reinforcement provided is at times boring and thus potentially questionable regarding the children's interest in the task. The variability observed within study and across studies may be due to this issue. More recently Litovsky and colleagues have developed and implemented a more ecologically interesting task for toddlers, whereby the task is to reach for a sounding object that is hidden behind a curtain (see earlier in chapter for description). The Reaching for Sound (RFS) method has proven to be fruitful with both NH children and subjects who are implanted with CIs. The RFS method was inspired by studies on "reaching in the dark" with NH infants, showing that sound location can be identified as early as 6 months of age based on auditory cues alone (Perris and Clifton 1988). In addition, at 6 months of age NH infants use their reaching behavior to indicate that they can discriminate sound source distance, and that they are not using intensity cues to solve the problem (Litovsky and Clifton 1992). This work is reviewed in more detail by Litovsky (2011a, b). In the CI population, the reaching behavior was motivated by testing in the light, for hidden objects that the child is motivated to find. Litovsky et al. (2013) tested bilaterally implanted toddlers with source locations at ±60°, ±45°, ±30°, or ±15°. First, discrimination was conducted for each of these location pairs, when listening bilaterally or with a single CI. As shown in Fig. 10.6, all toddlers were able to perform the task when using both CIs, and unilateral CI use was poorer. These results suggest that the RFS method is quite useful for yielding good performance from all toddlers tested, and that as reported above, bilateral CI use produces better results than unilateral CI use.

The MAA studies with bilaterally implanted children actually began prior to the toddler studies. Litovsky et al. (2006a) studied children ages 3.5–6 years and found that compared with unilateral listening conditions, bilateral listening provided an advantage for right vs. left discrimination. With both CIs activated, 9/13 children tested were able to perform the MAA task above chance, and the majority of the children demonstrated MAAs that were at least as good as 20°. Thus, the best-performing children demonstrated thresholds in the range of those observed with infants or toddlers with NH, who had similar hearing ages to the CI users. Also notable is the finding that of the nine children who had good MAAs, eight showed performance that was superior to the performance observed with one of the CIs turned off. The other 4/13 could not perform the right vs. left discrimination task; and "appeared to have little understanding of the fact that sounds can carry information regarding spatial location" (Litovsky et al. 2006a). The factors listed in Table 10.1 are considered to be highly relevant here in terms of the limitations contributing to the poor performance observed in these children. Notably, these children were older than the toddlers discussed above, both when they were tested and when they were bilaterally activated; the latter is most likely to be a cause of limitation. In a follow-up study, Godar and Litovsky (2010) focused on examining how MAA thresholds change over time, for children who are unilaterally implanted and transition to using bilateral CIs. Results were compared for intervals at the unilateral use stage, then at 3 months and 12 months following bilateral activation. For most children, MAA thresholds improved after transitioning to bilateral CI use, at 3 months, and even more so at 12 months after bilateral activation. More important, for these children, MAA thresholds remained poor, although that could have been due to the fact that they no longer received listening experience with a single CI on a daily basis.

Compared with sound location discrimination, sound localization taps an additional level of auditory perceptual processing, whereby auditory spatial mapping is involved, and localization perception is much more accurate than just hemifields discrimination. In addition, because localization involves the identification of the location of a sound source from amongst many options, this is a more difficult task than a 2-alternative forced-choice task used for the MAA measure. Initial studies with bilateral CI users were conducted with the same children who had been implanted with the second CI at relatively older ages (4–12 years of age). Comparison with NH children are quite important because the baseline needs to be well established as far as what the

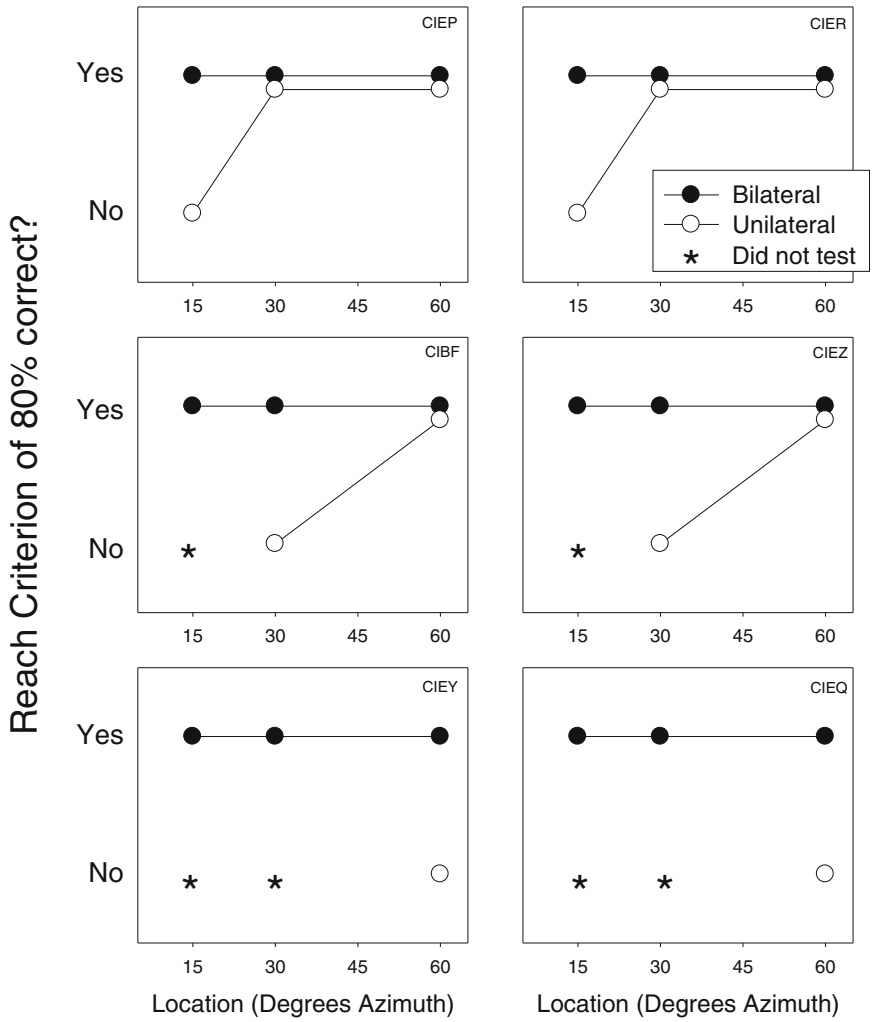

Fig. 10.6 Results from a study on spatial hearing using the "reaching for sound" paradigm are shown. Each panel shows data from a single child. The *x*-axis shows the locations used and the *y*-axis depicts whether the children passed a criterion of 80 % correct or not ("yes" or "no"). *Filled* and *open symbols* show results form bilateral and unilateral testing conditions, respectively. With permission from Litovsky et al. (2013)

expectation might be for emergence of spatial hearing in CI users. NH children ages 4–10 years show average error rates ranging from <5° to >30°. Root-mean-square (RMS) errors reported by Grieco-Calub and Litovsky (2010) were 9–29° (average of 18.3° ± 6.9° SD) in NH children ages 4–6 years old; note that <10° is within the range observed in NH adults. Two other studies reported smaller RMS errors of 1.4–38° (avg 10.2° ± 10.72° SD; Litovsky and Godar 2010) and 4–10° (Van Deun et al. 2009). These values overlapped with the RMS errors measured in NH adults. The RFS method described above was recently also adapted to measure sound location identification in toddlers, with a task requiring them to select one of nine locations as the perceived location of the sound source. Most of these toddlers were able to identify locations correctly on >95 % of trials (RMS errors <10°), and a small group of 2.5-year-olds selected the incorrect locations more frequently (RMS errors near 30°). Figure 10.5b shows average RMS from this and numerous other studies, for NH and bilateral CI users.

In bilateral CI users, sound localization studies were initially conducted with children who had very little experience listening with their CIs, and who were ages 4–12 at the time of activation of bilateral hearing. Litovsky et al. (2004) reported that RMS errors were near chance (~55°) after 3 months of bilateral CI users, suggesting poorly developed spatial hearing skills. Later studies investigated children with greater amount of listening experience, with notable improvement for some children. Grieco-Calub and Litovsky (2010) reported RMS values of 19–56° for spondaic speech stimuli; these values fell into a similar range of RMS errors (13–63°) reported by Van Deun et al. (2010) who used a broadband bell ring as the stimulus. Interestingly, using the RFS methodology, Ehlers et al.'s (2013) preliminary findings with toddlers show average RMS errors of 37° (range 11–52°), which is well within the range observed with the older children. The difference might be due to the difference in number of loudspeakers (9 for toddlers and 15 for older children); however that is unlikely to be the primary expla-

nation, because even with a 7-loudspeaker array some of the older children did not perform well on the localization task (Grieco-Calub and Litovsky 2010). Another possibility has to do with the exposure to bilateral hearing during early stages in development: the toddlers had been bilaterally implanted at a younger age than the children, and had more of an opportunity to become used to the bilateral cues and to use them on a sound localization

Speech Understanding in Noise and Related Phenomena

One of the overarching goals of providing bilateral CIs to young children is to enhance their ability to understand speech in everyday noisy listening situations. The question as to how to study the benefits from bilateral CIs compared with the use of a single CI led us to utilize the spatial release from masking (SRM) measure to evaluate sound source segregation abilities in these children. The key comparison in these studies is between conditions in which the target and masker(s) are co-located, and conditions in which they are spatially separated. Any improvement on the separated condition relative to the performance observed in the co-located condition is denoted as positive SRM; negative SRM refers to a disadvantage from spatial cues, which is seen at times in patients who use hearing aids or CIs. In NH adult listeners SRM can be as high as 12 dB improvement in the signal-to-noise ratio required to correctly identify the target speech; large SRM typically occurs when binaural cues are available, and when the target/maskers are similar or confusable (similar voices; Durlach et al. 2003; Jones and Litovsky 2008, 2011). The magnitude of SRM is also thought to be divided into both monaural and binaural and components (Hawley et al. 2004). Bilateral CI users are typically able to benefit from monaural-driven SRM, but have little access to the binaural cues that provide additional benefits for source segregation based on binaural cues.

Studies on NH children began about a decade ago. Litovsky (2005) first demonstrated SRM in NH children ages 4–7, using target stimuli consisting of spondaic words, and maskers that were either temporally modulated speech shaped noise or sentences spoken by a different-sex talker from the targets. Targets were presented from the front at 0°, and maskers were presented from locations that were either co-located with the target or spatially separated from the target. Using a novel 4-alternative forced-choice (4AFC) task, children indicated which target word they heard. Litovsky (2005) reported SRM values of 5–7 dB. In fact, SRM values were higher with two maskers (7.4 dB) than with a single masker (5.2 dB), indicating that the more complex auditory environments promote larger benefit from spatially separating potentially interfering sounds from the source of interest. Two further studies demonstrated that SRM is well developed at young ages. Garadat and Litovsky (2007) pursued this line of investigation in 3–4-year-old children, and reported similar, or slightly higher SRM values for that population, suggesting that the ability to use spatial cues to segregate target speech from maskers is developed by 3 years of age. Most recently, Hess CL. Speech discrimination and spatial hearing in toddlers with bilateral cochlear implants. Unpublished PhD Dissertation and University of Wisconsin-Madison (2013) measured SRM in toddlers, and found that the effect was fairly mature by 2.5 years of age. In those two studies, SRM was only evaluated for the conditions with maskers displaced asymmetrically around the head (see Fig. 10.4c); thus the "head shadow" might have been a highly dependable cue, and the extent to which binaural cues were used was not clear.

The first study with bilaterally implanted children was by Litovsky et al. (2006b) who used a similar design and stimuli as described thus far. The masker locations however were varied so that they were towards the side of either the first CI or the second CI. Results were compared with those from a group of children who used bimodal hearing (a CI in one ear and hearing aid in the other ear). For many of the children, the fact that both ears received input meant that there was an advantage to hearing the target speech at lower levels (lower SRTs) than those obtained in the unilateral listening condition. However, there was large inter-subject variability for this effect. For spatially separated conditions, the bimodal children, on average, did not have SRM; rather they had a "binaural disruption" effect, such that SRTs were higher for the separated than for the co-located conditions. This might indicate that the bimodal users lacked the ability to integrate information from the two ears in a way that benefited their source segregation. Other studies on similar measures with bimodal fitted children have not reported a similar disruptive effect (e.g., Ching et al. 2005, 2006). The differences, which should be further explored, might be due to variation in amplification approaches, different amounts of residual hearing in the unimplanted ear of the Ching et al. studies. In contrast with the bimodally fitted children, the bilateral CI users, on average, showed SRM that fell into the range observed in NH children. However, the effect was larger when the maskers were near the second CI than when they were near the first CI.

A more systematic evaluation of SRM was conducted by Misurelli and Litovsky (2012) who tested children ages 4–6 and 7–9 on similar tasks, with the added condition shown in Fig. 10.4d, whereby the maskers were symmetrically placed to the right and left, minimizing or eliminating the better-ear "head shadow" cue. In the NH groups, children were still able to demonstrate SRM in the symmetrical condition, although the values were smaller than with the asymmetrical condition. In the bilateral CI groups, SRM was achieved in

both age groups with asymmetrical maskers, but was very difficult to achieve with symmetrical maskers. Here again the contribution of monaural head shadow to spatial separation of target speech from maskers seems to be an important contributing factor.

Conclusions

Young children are fitted with hearing aids and/or CIs so that language acquisition and verbal communication can be developed, ideally at age-appropriate levels. CIs were designed to provide the signal processing necessary for stimulating the auditory nerve so that patients could hear speech, in quiet and in noise. For children, the goal was to provide each individual with the skills needed to function in a mainstreamed auditory environment. Bilateral CIs were not designed in a way that mimics the binaural system's ability to compute source locations and to squelch noise or reverberation based on interaural comparisons. Thus, to the extent that children who are bilaterally implanted show benefits from two CIs reflects the ability to their brain to interpret the signals from the two ears using rudimentary processing of binaural information. The studies that were reviewed here primarily focus on work conducted by Litovsky and colleagues, where parallel work is conducted in children with NH and with CIs. It is clear that, on average, bilaterally implanted children have a gap in performance relative to their NH peers. However, in many cases, the bilateral CI users' performance falls within the range of performance observed in the NH groups. That does not mean that the CI users are "the same" as the NH children, but it does mean that they are capable of resolving complex information about source location on the tasks that were described here. Many practitioners are concerned with being able to identify the age at which bilateral implantation will result in maximal recovery of function and minimal loss of auditory system integrity. The answer depends on numerous factors that can vary across individuals. Many of these factors were highlighted in Table 10.1. Future work will ideally focus on providing better understanding of how auditory system degeneration can be overcome, both peripherally and centrally. Because central mechanisms are thought to be more amenable to change following stimulation, stimulus-dependent learning and training can play an important role in habilitation.

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Language and Literacy Skills in Children with Cochlear Implants: Past and Present Findings

11

Susan Nittrouer and Amanda Caldwell-Tarr

Introduction

The topic of language acquisition in deaf children is one that can easily evoke visceral responses from clinicians and researchers in the field when it arises, a situation that might surprise anyone who does not have regular interaction with deaf individuals. To laypeople, hearing loss is seen as a problem of just that—hearing. But for those of us who work in deafness-related fields, it is more closely characterized as a problem of communication. Where children are concerned—especially since cochlear implants arrived on the scene—the problem of hearing loss is viewed primarily as a load placed on language learning, rather than as a problem of auditory sensitivity. In fact, the major challenge faced by scientists and clinicians is finding ways to facilitate language learning so that deaf children can progress through childhood unscathed by the deleterious consequences that can result from hearing loss. (Throughout this chapter the terms *deaf children* and *children with severe-to-profound hearing loss* are used more or less interchangeably to refer to children with average auditory thresholds no better than 70 dB hearing level, and generally much worse.)

Regardless of one's particular view concerning what language deaf children should be learning (the omnipresent question of whether it should be spoken or signed language) or how that learning should be facilitated, most professionals would agree that two key ingredients need to be provided in order for language to blossom: clear sensory input and adequate experience. Both of these ingredients are in short supply for deaf children, regardless of which language they are being encouraged to learn. (Throughout this chapter, terms referring to the *teaching* of language are avoided because of the philosophical perspective taken by the authors that language is not taught, for the most part. Rather, language emerges, or blossoms within children as part of the natural developmental process, facilitated by appropriate nurturing.)

If the decision is made that a child born with severe-to-profound hearing loss should be brought up learning a spoken language, strong constraints are imposed on the sensory inputs available because of that hearing loss. Although vision provides some access to the signals generated in the course of spoken language production, that information is limited because many articulatory gestures are not observable visually. Acoustic signals serve as the primary vehicles of transmission for sensory information generated during spoken language production. And even though cochlear implants have done a tremendous job of providing access to acoustic signals—effectively solving the problem of sensitivity—they nonetheless provide only degraded versions of natural speech. Spectral detail is greatly constrained by signal processing and delivery with cochlear implants, hampering access to many acoustic cues to phonemic categorization. Temporal structure arising from the actions of the larynx is largely absent, as is the harmonic structure generated by those actions. That factor accounts at least partly for the tremendous decrements in speech recognition observed when implant users must function in poor listening environments, such as noise or reverberation. And the problems associated with trying to listen in those tough environments serve to constrain the amount of language experience deaf children with cochlear implants (CIs) can obtain because many natural listening environments consist of some noise, arising from other speakers, the environment or reverberation. In the final analysis, both input and experience with spoken language are constrained for children with CIs.

Where sign language is concerned, it is true that deaf children have no sensory restriction on their access to the structure of that language: all components of sign language are available visually. Input is adequate for children who are

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deaf, but have no visual impairments, at least in principle. However, that point is only relevant for deaf children born to deaf/Deaf parents who are proficient signers themselves. Those children, who constitute about 5 % of all children born deaf, acquire sign language in a manner similar to how hearing children acquire spoken language (Lederberg et al. 2013). Generally speaking, though, deaf children are born to parents with normal hearing who have no proficiency in sign language. Even with the sincerest of intentions on the part of parents to facilitate their children's acquisition of sign language, these children will have highly constrained access to appropriate models of sign language. In the home, parents are unable to provide the kinds of rich language input that is usually provided to children. And it is extremely difficult to provide enough time in intervention to foster adequate exposure to sign language to make it a true first language.

This chapter is focused on the acquisition of spoken language by children with CIs; in particular, the acquisition of spoken English. Depending on the source of the estimates, something in the range of 93–96 % of children born with hearing loss are born to parents with normal hearing (Gallaudet Research Institute 2011). When those hearing losses are severe enough to warrant a cochlear implant, parents with normal hearing typically reach the decision to give their children implants for the specific purpose of facilitating the acquisition of spoken language, not sign language.

This chapter is divided into two sections. First, a review is provided of relevant data on language and literacy outcomes of children with severe-to-profound hearing loss who use CIs. Where language is concerned, both receptive and expressive language abilities are reviewed, providing a benchmark of how well children are faring with CIs. Where literacy is concerned, only reading is discussed in this chapter. Although of interest, there are not sufficient data on the writing abilities of deaf children to make conclusive statements at this time. A greater research effort needs to be dedicated to studying the writing skills of these children before we will have a collective account as comprehensive in nature as the one we have for their abilities in the areas of speech production and recognition, and reading.

Data of two sorts are discussed in this first section: (1) data from tests that are standardized in nature and meant to provide overall pictures of how children with CIs are performing with respect to language development, and (2) data from experimental protocols focusing on specific language-learning mechanisms. This review is largely restricted to school-age children.

In the second section of this chapter, a description is provided of data collected in our laboratory. These data come from a longitudinal study of children, both with hearing loss and with normal hearing. Specifically, performance levels will be described for children in this study at the age of 8 years. In this study, both standardized measures as well as experimental protocols have been incorporated in order to examine which specific language and cognitive mechanisms support the acquisition of spoken language and literacy.

Review of Research by Others

Anyone who is old enough to recall working with severely to profoundly deaf children before cochlear implants were available will readily recall the idiosyncratic language patterns of these children. There were, of course, the ubiquitous speech production errors that severely diminished intelligibility. Some of these speech production errors were problems with source support, such as breathy voice or deviant nasality. Other problems in production had to do with a failure to generate and coordinate the movements of the vocal tract, so omissions, epenthesis, and substitutions were frequent. But even when a listener could "hear through" those errors, the morphosyntactic and lexical constructions of the utterances were peculiar (e.g., Baumberger 1986; de Villiers et al. 1994; Quigley et al. 1976; Wilbur et al. 1976). Sentence structures were typically simple, with a lack of "sparkle" features such as adjectives beyond the ordinary or compound constructions of any kind. Inflectional morphemes (such as plural -s) were often missing. Contractions were rarely used. Function words were frequently absent. The pattern of language production and accompanying errors were recognizably unique to deaf children, a fact that might be attributable to the very formal approach taken to teaching these children language or to the late age at which most language skills were acquired. When tests standardized on hearing children could be implemented, deaf children of school age generally achieved age equivalency scores of roughly 3 or 4 years (Bishop 1983; Watson et al. 1982). When elicited productions of deaf and hearing children were analyzed, similarly low age equivalencies were observed for the deaf children (de Villiers 1988). These extremely delayed language abilities made the very information obtained by these assessment tools of little value for helping school-age deaf children acquire better language. Because they were cognitively well past the maturational levels of the children with whom they were matched in terms of language, typical language learning mechanisms could not be engaged. These qualities of the language of severely to profoundly deaf children (i.e., very low age equivalency scores and highly stylized structures) led to the development of evaluation instruments specifically designed for deaf children, such as the Scales of Early Language Communication Skills for Hearing-Impaired Children (Moog and Geers 1975). From these instruments teachers were able to get the kinds of data they needed to develop effective intervention programs for school-age deaf

children. However, outcomes of these specialized tools made it difficult to gauge the language performance of deaf children, relative to that of children with normal hearing.

Once CIs were introduced to the treatment arsenal for deaf children, outcomes immediately improved, which meant that assessment tools could be modified. Many of the speech and language qualities so recognizable in deaf children before CIs were available have all but disappeared, with perhaps the most salient change involving speech production. The numerous source problems previously heard in the speech of many deaf children are rarely found any longer. Expressive language has improved for deaf children, as well, to the point where it is commonplace to use assessment tools designed for children with normal hearing. With the goal of early intervention set as having these children ready to enter mainstream classrooms with normal-hearing children by the traditional start of school it is important that we have an idea of how proficient deaf children with CIs are in terms of language skills, relative to same-age peers with normal hearing. Assessment tools designed for children with normal hearing suit that purpose. Overall, the question now becomes whether deaf children with CIs have the language skills required to keep up in regular classrooms. To help answer that question, data were examined from recent studies evaluating the spoken language abilities of children with CIs.

Review of Research by Others: Standardized Measures

For this chapter, we searched the literature for published reports of studies that made use of standardized assessment instruments to evaluate the performance of children with CIs on five kinds of language skills.

Lexicality

Results for tests of both receptive and expressive vocabulary measures are reviewed. This skill refers to the number of individual words a child has in her lexicon. Although slightly different from semantics, the two skills—vocabulary and semantics—are closely related. Semantics refers to how a speaker is able to convey word meaning in connected discourse. Naturally, the larger one's lexicon is, the more precisely that word meaning can be conveyed. Standardized evaluations of vocabulary can take two forms. Tests of receptive vocabulary involve having children listen to words in isolation and identify the picture from a small set that represents each word. Tests of expressive vocabulary require children to look at a picture and provide a word to label it. An example of a receptive vocabulary measure is the Peabody Picture Vocabulary Test (PPVT) (Dunn and Dunn 2007). An example of an expressive vocabulary measure is the Expressive One-Word Picture Vocabulary Test (EOWPVT) (Brownell 2000).

Grammar (Morphosyntax)

Results of both receptive and expressive morphosyntactic abilities are reviewed. These skills refer to how well a child can use syntax to combine words into sentences, and appropriately incorporate morphological units into those words and sentences. Several standardized tests have been developed to quantify the language level of children in general, including the Test of Language Development (TOLD) (Hammill and Newcomer 2008), the Clinical Evaluation of Language Fundamentals (CELF) (Semel et al. 2013), and the Comprehensive Assessment of Spoken Language (CASL) (Carrow-Woolfolk 1999). These tests provide robust metrics of morphosyntactic abilities, both receptive and expressive.

Phonology

This level of structure refers to the actual sound patterns of the language; in particular, how phonemes are arranged. Having well-developed sensitivities to this level of linguistic structure is critical to a wide variety of language processes. For example, phonemic units form the substance used to store language in short-term memory buffers. Without strong sensitivity to the phonological level of linguistic structure, it is difficult to store sufficiently long sequences of language material to support the comprehension of sentences with complex syntax, which are often long. It is also important to have well developed sensitivity to phonological structure in order to acquire awareness of some morphological structures because morphemes can consist of single phonemes, such as the plural –s. And because words are stored in the lexicon according to phonemic structure, at least for adults (Luce and Pisoni 1998), refined sensitivity to this level of structure is a prerequisite for eventually developing large vocabularies. Finally, it is critical that a child develop adequate sensitivity to phonemic structure in order to learn to read because the symbols of our writing system largely represent individual phonemes. A commonly used standardized test of phonological sensitivity and abilities is the Comprehensive Test of Phonological Processing (CTOPP) (Wagner et al. 1999).

Reading

Two kinds of skills are often evaluated when it comes to reading: the ability to recognize isolated words and the ability to comprehend passages that are read. Reports concerning the emergence of both of these skills in deaf children were sought for this review. One other kind of skill is sometimes measured in regard to children's reading acquisition, and that has to do with fluency, which is measured by tallying the number of words a child can correctly read in a specified amount of time. That skill has not been examined extensively for children with CIs, but an earlier report from this laboratory showed no differences in fluency between children with CIs and those with normal hearing

(Nittrouer et al. 2012). Consequently, fluency was not considered in this review of research by others. However, it is examined in the second section of the chapter.

Several tests of word reading are available. For example, in our laboratory we frequently use the Word Reading subtest of the Wide Range Achievement Test (WRAT) (Wilkinson and Robertson 2006). In these assessments, the child is asked to read a sequence of unrelated words that become increasingly harder as the list progresses. When it comes to reading comprehension, this skill is usually assessed by asking children to read a passage, and then answer questions to assess comprehension of that passage. Generally speaking, several passages and associated questions are used, of increasing difficulty. In our laboratory, we have used the Qualitative Reading Inventory (QRI) (Leslie and Caldwell 2006), which contains several passages at each grade level and comprehension questions related to each passage. That structure is typical of reading assessments.

Working Memory for Speech

Finally, recent reports on working memory for speech (or phonological working memory, as it is also called) were sought. Although not exactly a language skill, this cognitive function so strongly underlies language performance that it was considered important to examine. Typically, the concept of working memory refers to how efficiently an individual can preserve a sequence of phonologically relevant items in a short-term memory buffer, although the additional ability of performing some sort of action on those items is often incorporated into the definition. In order to assess working memory by the first of these definitions (i.e., simple storage of verbal items), a child may be asked to repeat a sequence of digits in the order in which they were produced. The number of digits the child can correctly recall is used as the dependent variable, and is known as forward digit span. To assess the second description of working memory (i.e., storage and processing of verbal items), the child is asked to repeat a sequence of digits, but backwards. Thus, the process that must be performed on the digits is to reverse the order. This task is known as backwards digit span.

Criteria for Including Reports

In our search to find reports related to each of the five skills listed above, certain constraints were imposed. First, the report had to concern children with CIs who did not use sign language as a primary communication mode. There are intervention and educational programs that use signing systems, usually English based, to support the acquisition of spoken language. Studies including children in those sorts of programs were not excluded from the review because the goal of those programs is steadfastly to facilitate the development of spoken language in deaf children; it just happens to be the educational philosophy of the programs that a signed language can facilitate the acquisition of a spoken language in children who are unable to hear speech clearly. Nonetheless, all dependent measures used in the studies reported here had to involve spoken language. If a study granted children the option of responding in sign language, that study was not included in this review.

All studies had to involve school-age children, meaning they were in roughly grades kindergarten through high school. There was also the presumption that most, if not all, children in any study selected had been born with severe-to-profound hearing loss or lost their hearing very early in life. The children in the studies included in this review must have received their CIs relatively early in life, as well. In particular, most children in any single study must have received them before the age of 3 years. And only studies with English-learning children were included in this review. We also restricted the range of publication dates, from 2008 to 2013, a 5-year span.

The studies themselves had to adhere to specific procedures that underlie rigorous research. In particular, the study had to include at least 20 children with CIs in order to provide any reasonable degree of power. There needed to be some evidence of the validity of the assessment tools used, and the reliability of measurement procedures.

Outcomes for Literature Review

Table 11.1 lists the set of reports culled from the literature matching the selection criteria established for this review. A general conclusion that may be drawn from these studies is that, regardless of which language skill is examined, children with severe-to-profound hearing loss who use CIs are performing, on average, one standard deviation below the mean of age-matched children with normal hearing. That means that children with CIs obtain mean standardized scores of roughly 85 or mean scaled scores of 7. An irrepressible optimist might view these outcomes as clear evidence that CIs have changed the landscape completely for children with severe-to-profound hearing loss. These kinds of scores would not have been possible before CIs became available. This collective finding means that roughly half of the children with that degree of hearing loss are in what may be described as the normal range of language abilities for their age. That is a tremendous advance over performance levels of the past. The glass truly may be seen as half full.

The realist, on the other hand, looks at these scores and recognizes that children with severe-to-profound hearing loss still are not attaining the levels of language proficiency that they presumably would have attained had they not been born with those hearing losses. One standard deviation below the mean is the 16th percentile in terms of population ranking. This means that half of the children born with severe-to-profound hearing loss are displaying language abilities in the lowest 15th percentile rankings of children with normal

Table 11.1 Summary of outcomes of standardized testing with children with cochlear implants

Authors (year)	Numbers	Measures	Results
Lexicality (Vocabulary)			
Schorr et al. (2008)	39 CI and 37 NH matched for age and gender, ages 5–14	PPVT and EVT	CI means were ≈1 SD below control means PPVT: NH-112, CI-87 EVT: NH-106, CI-91
Geers et al. (2009)	153 CI, ages 5–7	PPVT, EOWPVT, EVT	CI means were ≈1 SD below normative mean PPVT: 86 EOWPVT/EVT: 91
Johnson and Goswami (2010)	39 CI (20 early implant, 19 late implant) and 19 NH matched for reading level, ages 5–15	EOWPVT	CI means in both groups were ≈2 SDs below control means EOWPVT: NH-108, CI-80 and 76
Conway et al. (2011)	23 CI and 26 NH matched for age, ages 5–10	PPVT	CI mean was ≈2 SDs below control mean PPVT: NH-114, CI-86
Fitzpatrick et al. (2012)	21 CI, age 10	PPVT	CI means were >1 SD below normative mean PPVT: 77
Grammar			
Schorr et al. (2008)	39 CI and 37 NH matched for age and gender, ages 5–14	TOLD	CI mean was >1 SD below control mean TOLD: NH-12.3, CI-8.4
Geers et al. (2009)	141 CI, ages 5–7	CELF	CI means were >1 SD below normative mean CELF: 79
Fitzpatrick et al. (2012)	21 CI, age 10	CELF	CI means were ≈2 SDs below normative mean CELF: 71
Tobey et al. (2013)	160 CI, ages 6–12	CASL	CI means were >1 SD below normative mean CASL: 76 and 78
Phonology			
Schorr et al. (2008)	39 CI and 37 NH matched for age and gender, ages 5–14	CTOPP	CI mean was ≈1 SD below control mean CTOPP: NH-12.3, CI-8.7
Geers and Hayes (2011)	112 CI, ages 15–18	CTOPP	CI mean was ≈1 SD below normative mean CTOPP: 6.9
Fitzpatrick et al. (2012)	21 CI, age 10	CTOPP	CI means were between 1 and 2 SDs below normative mean
Reading			
Spencer and Tomblin (2009)	29 CI and 29 NH matched on mother's education and word comprehension, ages 6–17	WRMT	CI means on both tasks were at least 1 SD below control mean WRMT-WA: NH-117, CI-101 WRMT-WC: NH-108, CI-93
Johnson and Goswami (2010)	39 CI (20 early implant, 19 late implant) and 19 NH matched for reading level, ages 5–15	NARA-R	CI means were ≈1 SD below control mean NARA: NH-99, CI-85 and 81
Geers and Hayes (2011)	112 CI, ages 15–18	PIAT	CI total mean was ≈1 SD below normative mean PIAT: 83
Working memory			
Pisoni et al. (2011)	108 CI ages 8–9 and 112 CI ages 15–16	WISC-III Digit Span	CI means were <1 SD below normative means WISC: 6.44, 6.38
Harris et al. (2013)	66 CI, ages 6–12	WISC-III Digit Span	CI means were 1 SD below normative means

Note: Numbers: shows numbers in CI group and control group, if applicable, and age range in years; *Measures*: *PPVT*, Peabody Picture Vocabulary Test (Dunn and Dunn 1997); *EVT*, Expressive Vocabulary Test (Williams 1997); *EOWPVT*, Expressive One Word Picture Vocabulary Test (Brownell 2000); *TOLD*, Test of Language Development (Hammill and Newcomer 1997); *CELF*, Clinical Evaluations of Language Fundamentals (Wiig et al. 2004); *CASL*, Comprehensive Assessment of Spoken Language (Carrow-Woolfolk 1999); *CTOPP*, Comprehensive Test of Phonological Processing (Wagner et al. 1999); *WRMT*, Woodcock Reading Mastery Tests (Woodcock 1987): Word Attack (WA) and Word Comprehension (WC); *NARA-R*, Neale Analysis of Reading Ability-Revised (Neale 1997); *PIAT*, Peabody Individual Achievement Test (Dunn and Markwardt 1989); *WISC-III*, Wechsler Intelligence Scale for Children, 3rd Ed. (Wechsler 1991); *Results*, given relative to means of normative sample or specific control group in study, and as standard or scaled scores when possible; standard scores have normative means of 100 with SDs of 15; scaled scores have normative means of 10 with SDs of 3

hearing. That kind of language proficiency makes it difficult to compete in the mainstream. We, as a profession, cannot be satisfied with these outcomes. The glass remains half empty. Strong research efforts need to continue in order to find ways to improve these outcomes.

Review of Research by Others: Nonstandard Measures

The clear conclusion to be drawn from the studies reviewed above is that in spite of the beneficial effects accrued by deaf children from CIs, they still are not performing as well as children with normal hearing in terms of their language abilities. The half of children with CIs whose standardized test scores are within the normal range—defined as better than one standard deviation below the mean of children with normal hearing—are in all likelihood not performing at the levels that they would attain, if they did not have severe-to-profound hearing loss. Even at that, comparing scores from standardized instruments to published norms underestimates the true magnitude of the average deficit for these children with CIs. Parents who choose to participate in research studies tend to be heavily involved in their children's upbringing, and are often well educated. Both these factors positively influence language development and are associated with high scores on standardized tests. Evidence of this claim is provided by the findings shown in Table 11.1. Means for children in the control groups of almost every study were above the normative means, often by as much as one standard deviation. That trend suggests that the children with CIs in these studies would have been performing comparably, if it were not for their hearing loss. The goal of current research efforts must be to move the language performance for children with severe-to-profound hearing loss who wear CIs to the levels they would achieve if it were not for the hearing loss.

In addition to studies that make use of standardized instruments are ones that investigate the mechanisms underlying each language skill considered above. These studies point us to the kinds of underlying skills that need to be measured and sharpened in order to improve the overall language performance of individual children with CIs. For these reasons, several experiments on the mechanisms that underlie the language skills discussed above are reviewed here.

For the selection of experiments to be discussed in this section, criteria were again imposed. Any study that is discussed had to focus on a specific mechanism, using nonstandard assessment methods. That meant that scores could not be standardized on a larger sample. Accordingly, each study had to include its own control group of children with normal hearing in order to be included in this review section. Again, participants in the studies selected for review had to be learning language primarily through an oral method of instruction, although sign support was permissible as long as there was a clear focus on spoken language. Dependent measures had to consist of spoken language. Not every study meeting these criteria could be included, but a representative sample was selected.

For this review, a restriction regarding date of publication was not imposed. It is reasonable to restrict date of publication when considering benchmarks of how well children with CIs are doing in order to ensure that performance with current devices and intervention procedures are being taken into account. However, when it comes to understanding the mechanisms that underlie the skills measured, those principles would not be expected to change over time or as a function of changes in treatment for a specific group of individuals.

Vocabulary Acquisition

Looking first at vocabulary skills, Table 11.1 reveals that both the receptive and expressive vocabularies of deaf children with CIs are smaller than the vocabularies of their age-matched peers with normal hearing. It appears that vocabulary growth is slowed. In our longitudinal study (e.g., Nittrouer 2010), we have found that vocabulary growth is roughly 2 years delayed for children with CIs. This factor can make it difficult to function in school settings and can hinder the acquisition of literacy because it is affected by lexical knowledge (Wise et al. 2007).

Because of these observed deficits in vocabulary, it is reasonable to ask how children with CIs learn words. Broadly speaking, learning a word to the point where it is a stable and readily accessible element in the lexicon involves three processes. The first stage of this learning is termed *fast-mapping*, which happens when a learner makes a connection between the sensory input and the referent (object, action, attribute, etc.). At first, these fast-mapped representations are not stable and not well specified in terms of meaning. At this point, the learner is able to pick the referent out of a closed set of pictures upon hearing the word, as is the protocol for receptive vocabulary tests, but likely could not retrieve the item from the lexicon in order to label the referent, according to the protocol for tests of expressive vocabulary.

The second step in word learning involves extending the word to other exemplars. Thus, the learner comes to recognize the group of referents that may be labeled with that word, as well as those that do not fit in the category. For example, many furry quadrupeds fit the category of *dog*, but not all of them. Discovering which ones are legitimate members of that category is the process of *extension*. Finally, the learner's experience hearing and producing the word must be sufficient so that the word is retained in the lexicon and can be retrieved at much later times. That process is termed *retention*.

There have been several studies looking at word learning in children with CIs, or children with severe-to-profound hearing loss more broadly (Lederberg and Spencer 2009; Tomblin et al. 2007; Walker and McGregor 2013). Two of these studies included children with normal hearing as control groups (Tomblin et al. 2007; Walker and McGregor 2013). In both cases, it was observed that children with CIs performed more poorly in terms of fast-mapping than age-matched peers with normal hearing. Walker and McGregor further observed poorer skills at extension and retention of new vocabulary items. These authors were able to show that the performance of children with CIs matched that of children with normal hearing who were roughly 14 months younger. Still another study traced the largest share of variance in fast-mapping abilities for children with CIs to their sensitivities to phonological structure in the speech signal, $r^2 = 0.72$ (Willstedt-Svensson et al. 2004). This finding can explain observed deficits in receptive and expressive vocabularies in children with CIs. If the word-learning process depends in large part on sensitivity to phonological structure, children with CIs could be expected to have difficulty because CIs do not provide a signal rich in the kinds of spectral and temporal detail that are thought to underlie phonemic representations. Consequently, their vocabularies suffer.

Sensitivity to Phonological Structure

The lack of sensitivity to phonological structure predicted for children with CIs likely poses problems for other kinds of language learning, as well. Beyond deaf children, this lack of sensitivity is often suggested as a critical deficit underlying problems in reading and working memory skills: In both cases, evidence shows that proficiency in these areas depends strongly on children's abilities to recover phonological structure (primarily phonemic) from the acoustic signal. In fact, one predominant view is that developmental dyslexia can be explained by a single (core) deficit in sensitivity to phonological structure (Snowling 1998; Stanovich 1986; Wagner and Torgesen 1987; but cf., Pennington 2006). Children with normal hearing who get diagnosed as having dyslexia have also been found to demonstrate poorer working memory skills than their typically reading peers (Brady et al. 1983; Nittrouer and Miller 1999; Savage et al. 2007), a finding that has similarly been traced to poor sensitivity to phonological structure (Mann and Liberman 1984; Shankweiler et al. 1979; Spring and Perry 1983). Thus, it is reasonable to propose that much of the deficit in reading and working memory observed for children with CIs may be explained by poor sensitivity to phonological structure in the acoustic speech stream, which in turn arises because of the highly degraded signal they receive through their CIs.

Evaluating children's sensitivity to phonological structure in the acoustic speech signal can be accomplished with a variety of tasks, each tapping into different sorts of phonological skills. The terms *phonological awareness* and *phonological processing* are generally used to refer to slightly different phenomena, although the exact phenomenon to which each refers can vary across reports. In reality, the two terms might be seen as anchoring two ends of a continuum, with the boundary between awareness and processing being somewhat fuzzy. Strictly speaking, phonological awareness refers to the ability to recognize phonological structure—inflectional, syllabic, onset/rimes, and individual phonemes—in the acoustic speech signal. The term *phonemic awareness* is also used to refer to awareness, but strictly of phonemic structure. Phonological awareness tasks usually consist of asking children to explicitly judge similarity or difference in the phonological structure of words, assess whether words rhyme or not, count elements of one type or another, or blend or remove elements from target words or syllables. Phonological processing refers to children's abilities to take structure and use it in further processing, such as in the storage of words in a short-term memory buffer or in repeating non-words. In principle, children may be able to recognize phonological units in the signal, without being able to bring that recognition to the level required for conscious inspection and manipulation known as meta-linguistic awareness. In examining phonological awareness and processing, investigators need to take care to ensure that any differences found between experimental and control groups are not actually due to differences in that meta-linguistic component of testing. One way to do that is to include a range of phonological awareness or processing tasks in the experimental protocol. Patterns of variability across tasks can help identify where any observed problems reside. In particular, if group differences are smaller for tasks with low processing demands, then concern is heightened that children in the poorer performing group have difficulties with meta-linguistic awareness.

Phonological awareness can be further grouped according to the level of structure in the signal being examined. Children acquire sensitivity to various levels of phonological structure at different times during development. Evidence of this maturational effect was first offered by Liberman et al. (1974). They showed that typically developing children were able to count the numbers of syllables within words with better than chance accuracy by kindergarten, but it took until second grade for them to be able to count the number of sounds (or phonemes) in those syllables. The developmental hierarchy of phonological skills was further explicated by Stanovich et al. (1984), who tested kindergarten children on ten separate phonological awareness tasks. By ranking tasks according to mean accuracy of responses they established a developmental hierarchy.

A fundamental point that is easily forgotten when thinking about phonological awareness and processing skills in children is that the acoustic signal of speech is not comprised of sequences of isolable phonemes. Clinical and

experimental protocols ask children to perform chores such as counting or matching phonemes, or removing one from a sequence. The implementation of these tasks can reinforce natural impressions that speech signals consist of strings of separate phonemes. But that is not the case. Figure 11.1 shows the sentence *Everybody knows the story of Winnie the Pooh*, and illustrates that it would be impossible to place markers on the *x* axis indicating where one phoneme ends and the next begins. (Spectrograms display time on the *x* axis and frequency on the *y* axis. Energy distribution across frequencies is represented by the darkness of the tracings.) That situation represents the attribute of speech known as a lack of segmentation. Figure 11.2 illustrates another relevant attribute of speech signals, known as a lack of acoustic invariance. This figure shows a single word, *bug*, spoken by a man and by a child. It is apparent in this figure that the acoustic structure affiliated with that production differs drastically for each talker. Thus, not only is it hard to identify individual phonemes in the continuous speech signal, but the acoustic structure affiliated with each phoneme differs depending on factors such as who the talker is. These attributes of speech signals emphasize the fact that speech perception involves more than just the harvesting of either phonemes or acoustic cues from the signal. Several separate processes must be undertaken and coordinated. The listener must know which components of the signal require attention for the perceptual task at hand: recovering phonemes or recognizing the speaker, for example. Those signal components need to be organized appropriately and interpreted within the current linguistic and social context. These considerations emphasize the fact that phonemic structure is highly encoded in the acoustic signal. Consequently, tasks of phonological awareness tap into processes much more complicated than simply recognizing phonemes in the signal. They require appropriate attentional and organizational strategies, as well.

Phonological awareness is an especially important mechanism to evaluate in children with CIs if we want to understand the underpinnings of their language and literacy skills. There is very good reason to suspect that children with CIs will have diminished sensitivity to phonological structure in the speech signal: the signal processing of CIs does not preserve the kind of spectro-temporal structure that strongly supports recognition of phonological structure. At the same time, phonological awareness has reliably been shown to underlie the development of many other language skills: in particular, working memory and reading. In turn, working memory plays a role in the acquisition of morphosyntactic abilities, especially those related to complex syntactic structures. Sentences with embedded clauses tend to be long, so it is important that a child can store long strings of linguistic material in order to discover clause structure in those sentences.

The CTOPP is very commonly used to evaluate phonological awareness and processing. As Table 11.1 shows, when a standardized measure of phonological awareness and processing is needed, the CTOPP is often the test of choice. Nonetheless, there have been some experiments

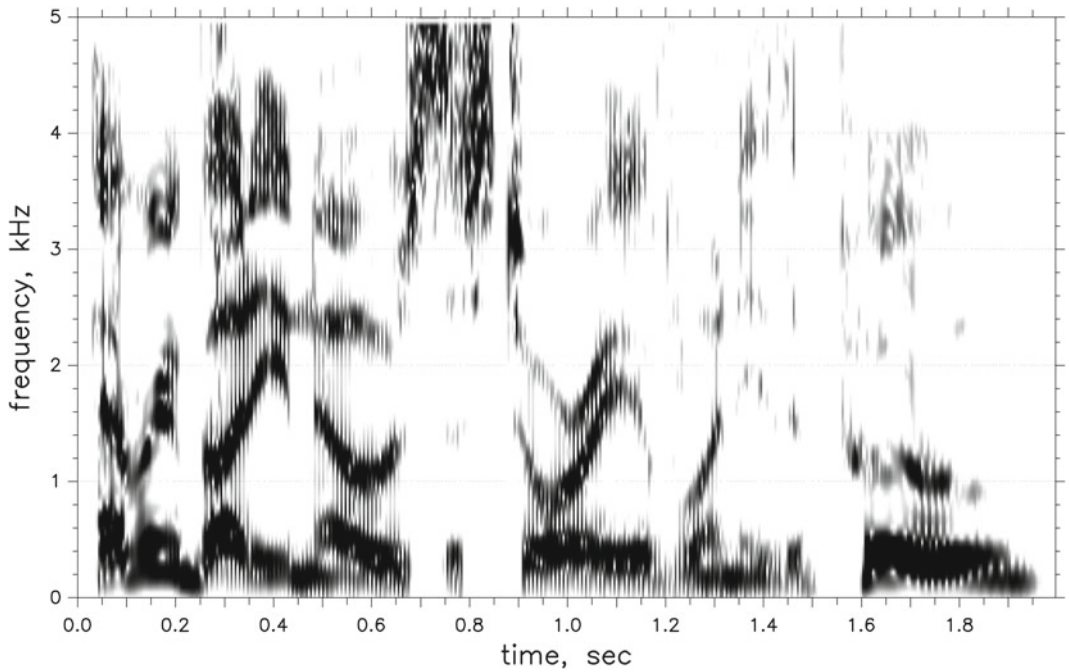

Fig. 11.1 A spectrogram of the sentence *Everybody knows the story of Winnie the Pooh* spoken by a man, illustrating the lack of clear segmental boundaries

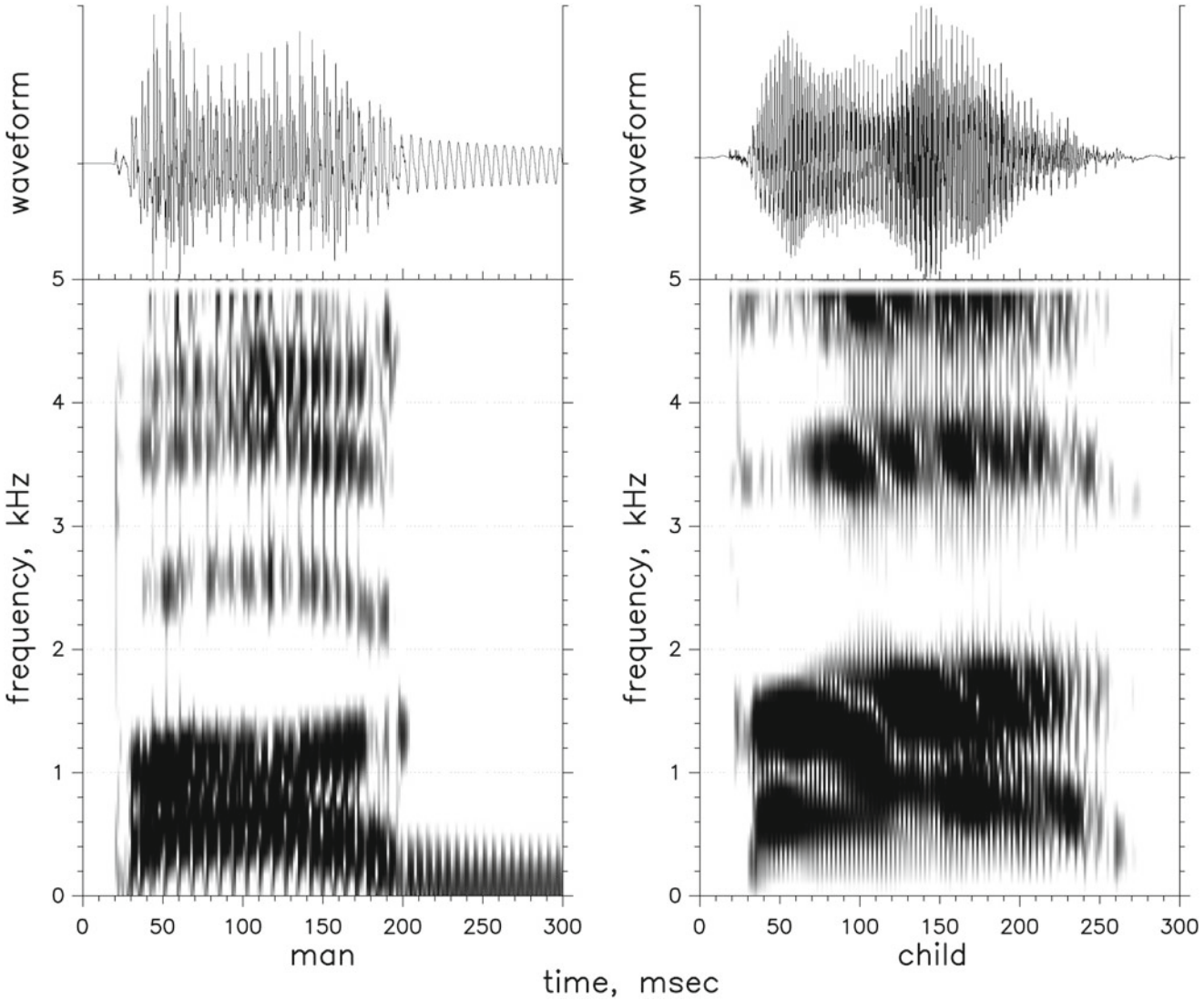

Fig. 11.2 The word *bug* spoken by a man (*left*) and a child (*right*), illustrating the lack of invariant acoustic structure

conducted that used nonstandardized measures of phonological awareness, and met the criteria for inclusion in this review. For example, James et al. (2009) examined phonological awareness in 19 eight-year-olds with CIs, 19 reading-level matched peers, and 19 chronological-age matched peers. These authors examined children's sensitivity to three kinds, or levels, of phonological structure: syllable, rhyme, and phoneme. The tasks were all visual, with pictures representing target words. The children with CIs performed as well as children in the two control groups on syllable awareness, but more poorly on rhyme and phonemic awareness. That finding would be predicted from the fact that syllable structure at the linguistic level is discernible from amplitude structure at the acoustic level. Recognizing phonemic structure, on the other hand, requires access to detailed spectro-temporal structure, precisely what is impoverished in cochlear implant processing strategies.

Grammar

Other studies have investigated the relationships among skills that are not strictly phonologically based. For example, Spencer et al. (2003) examined the relationship between reading comprehension and morphosyntactic skills for 16 nine-year-olds with CIs and 16 age-matched peers, for each group separately. To evaluate reading comprehension, the passage comprehension subtest of the Woodcock Reading Mastery Tests (WRMT) (Woodcock 1987) was used. To evaluate receptive and expressive language, the Concepts and Directions and Formulated Sentences subtests of the CELF were employed. Results demonstrated that the relationship between reading comprehension and oral language abilities was stronger for children with CIs than for the children with normal hearing: $r = 0.8$ vs. $r = 0.5$, respectively. A separate study by Connor and Zwolan (2004) replicated the general result. Taken together, those findings are important

because they suggest that the extent to which non-phonological language factors explain literacy acquisition may differ for children with normal hearing and those with CIs. Robust evidence supports the claim that children with normal hearing develop literacy skills largely through a phonological route. Any reading proficiency children with CIs manage to acquire may depend to a greater extent on language abilities not necessarily related to phonological knowledge. The reason for that difference in underlying mechanisms is likely the diminished access to acoustic structure that supports phonemic structure experienced by children with CIs. Unfortunately, it is generally agreed that in order to read much above a fourth grade level, sensitivity to phonemic structure is required (Goldin-Meadow and Mayberry 2001). Thus, the reliance on extra-phonological factors observed for the literacy skills of young school-age children with CIs might be a harbinger of limited proficiency to be found in their later literacy achievements.

There have been fewer investigations of the morphosyntactic skills of children with CIs using nonstandard measures than of other sorts of speech and language abilities. That may be due to the arduous nature of analyzing morphosyntax in language samples; it is much more efficient to use standardized test instruments. Nonetheless, one report using a measure that is not strictly standardized was conducted by Geers et al. (2003). It included 181 children with CIs, and 24 age-matched peers with normal hearing, all tested at 8- to 9-years of age. As the measure of morphosyntax, the Index of Productive Syntax (Scarborough 1990) was used. With this instrument, trained listeners review language samples from children. Occurrences of 56 syntactic and morphological forms are evaluated, providing scores in four categories: complexity of noun phrases, verb phrases, questions/negations, and sentence structures. When Geers et al. applied this index, the average score of children with CIs was 1.13 SDs below the mean of the control group. Thus, this study replicated the general finding that children with CIs are performing, on average, roughly one standard deviation below the means of children with normal hearing.

Summary

Results from several laboratories have been reviewed in this first section of the chapter, but it is far from an exhaustive set of studies on the topic of language and literacy in children with CIs. Over the past two decades there has been a well-focused effort on quantifying language outcomes in children with severe-to-profound hearing loss who use CIs, and measure how those outcomes have improved since CIs became available for children. Entering the search terms *language*, *cochlear implants*, and *children* together into the *PubMed* database produces more than a 1000 results. In this review section, we focused on a select few of those studies, using specific criteria. Nonetheless, the outcomes reported here generally match those of the numerous papers that we were unable to include. The data overwhelmingly indicate that CIs have allowed many children with severe-to-profound hearing loss to acquire language and literacy skills in the range of children with normal hearing, but the influences of that hearing loss have not yet been eliminated. The goal of future research and intervention efforts must be to find ways to more effectively ameliorate those influences. One approach that could facilitate that effort would be to construct a better model of the factors that underlie language acquisition for these children, and how those factors may differ for children with CIs and children with NH. That is what we are seeking to do in the research conducted in our laboratory.

Review of Outcomes at Second Grade from a Longitudinal Study of Children with CIs

In this next section, outcomes are reported for a sample of children in a longitudinal study conducted for 10 years in our laboratory. The project is titled Early Development of Children with Hearing Loss (EDCHL). In this chapter, language and literacy outcomes are reported for these children from data collected when they were all 8 years of age (mean age = 8 years, 6 months; SD = 5 months). They were tested during the summer following second grade in all cases. More detailed information about the original sample and procedures can be found elsewhere (e.g., Nittrouer 2010).

Participants

The children on the EDCHL project came from across the USA, and all had been tested as part of this project between four and eight times since their first birthdays. Forty-eight of these children had normal hearing (NH), and 50 of them had severe-to-profound hearing loss and wore one or two CIs. In order to participate in the study, children, their families, and their early intervention programs (in the case of the children with CIs) needed to meet specific criteria.

Criteria for Participation for Children

In order to participate, there could be no evidence of any physical, cognitive, or emotional deficit other than hearing loss (in the case of children with CIs) that could on its own be expected to impact development. While the first of these three requirements was easy to verify from clinical records, the last two were less transparent. Consequently,

assessments were made at each test time to confirm that none of these children had any disabling conditions other than hearing loss. The children in the NH group had their hearing screened each time they were tested, with octave frequencies between 0.25 and 8 kHz presented at 20 dB hearing level. All children were administered four subtests on the Leiter International Performance Scale—Revised (Roid and Miller 2002), which is a completely nonverbal test of cognitive functioning. The four subtests administered were Figure Ground, Form Completion, Sequential Order, and Repeated Patterns. From these four subtests an estimate of nonverbal intelligence can be computed that is labeled by the test authors as the Brief IQ. That metric is given as a standardized score, with a mean of 100 and a standard deviation of 15.

Emotional stability—defined here as the lack of emotional problems—was assessed using the Child Behavior Checklist (CBCL) by Achenbach and Rescorla (2001). This is an instrument completed by each parent separately and by the classroom teacher. Each of these three responders read 113 individual statements, such as *Argues a lot* and *Stubborn, sullen, or irritable* and had to rate how strongly the statement describes the child being assessed using a three-point scale (0–2). Weighted sums across items are computed to obtain two general indices, one of internalizing and one of externalizing tendencies. These weighted sums are given as standardized *T* scores, which have means of 50 and standard deviations of 10. Scores above 70 are considered to be in the clinical range. Internalizing problems refer to difficulties such as being emotionally reactive, withdrawn, or anxious. Externalizing problems refer to difficulties with rule breaking or aggressive behavior.

Finally, all children were screened with the Short Sensory Profile (SSP) by McIntosh et al. (1999), which is an abbreviated version of the Sensory Profile (Dunn 1999). This instrument is completed by parents, who rate according to a five-level scale how frequently 38 separate statements describe their children. Each statement concerns a specific kind of sensory processing, such as *Avoids going barefoot, especially in sand or grass* and *Responds negatively to unexpected or loud noises*. Impairments in the ability to process sensory inputs—such as defensiveness or over-responsivity—are widely reported for children with autism spectrum disorders (e.g., Kientz and Dunn 1997; Ornitz 1989; Osterling and Dawson 1994; Watling et al. 2001). By administering this instrument, we were able to screen the children in this study for tendencies that would place them on the autism spectrum. Scores on the SSP load on seven separate clusters: Tactile Sensitivity, Taste/Smell Sensitivity, Movement Sensitivity, Underresponsive/Seeks Sensation, Auditory Filtering, Low Energy/Weak, and Visual/Auditory Sensitivity. Results are not given as standardized scores. Instead, three ranges of scores are used that group children into three categories: Typical Performance, Probable Difference, and Definite Difference. Summing across the seven categories provides a total score that can be used, as well. Children on the autism spectrum have reliably been found to score lower than typical children in each category separately and on the total score, and reliably in the third category of Definite Difference (e.g., Tomcheck and Dunn 2007).

Table 11.2 shows means and standard deviations for pertinent scores from these screening measures. For the Brief IQ, it is clear that means for both groups were at the means for the normative sample, and standard deviations were similar, as well. A *t* test revealed no difference in scores across the groups.

For the CBCL internalizing and externalizing scores, two-way, repeated-measures ANOVAs were performed on each set of scores with respondent (mother, father, or teacher) as the repeated measure and group (NH or CI) as the between-subjects factors. Only for the externalizing scores was a significant effect found, and it was for respondents, $F(2, 162) = 3.86$, $p = 0.023$. That finding reflected the fact that mothers rated their children as having slightly fewer problematic externalizing behaviors than either fathers or teachers. But the differences were small: mean externalizing *T* scores were 46, 48, and 49 for mothers, fathers, and teachers, respectively. No Hearing Group × Respondent interaction was found. Consequently, means across the three responders were computed and are reported in Table 11.2, for both internalizing and externalizing behaviors.

Table 11.2 Group means and *standard deviations* for children with normal hearing ($N = 48$) and children with CIs ($N = 50$) for cognitive, emotional, and sensory processing measures

	Brief IQ		CBCL internalizing		CBCL externalizing		SSP auditory filtering		SSP total	
	M	*SD*	M	*SD*	M	*SD*	M	*SD*	M	*SD*
Normal hearing	105	*14*	48	*7*	47	*6*	24	*3*	168	*13*
Cochlear implants	100	*18*	47	*8*	48	*9*	22	*5*	166	*18*

Note: Brief IQ = standardized scores with a mean of 100 and *SD* of 15; CBCL (Child Behavior Checklist) = *T* scores with a mean of 50 and *SD* of 10; and SSP (Short Sensory Profile) = categorized into ranges describing performance. For SSP Auditory Filtering these are: Typical Performance (23–30); Probable Difference (20–22); and Definite Difference (6–19). For SSP Total Score these are Typical Performance (155–190), Probable Difference (142–154), and Definite Difference (38–141)

For the SSP, mean scores for both groups were in the Typical Performance range on all subtests and *t* tests revealed no differences between scores for children with NH and those with CIs, with one exception. Children with CIs scored significantly lower in the category of Auditory Filtering, $t(1,96) = 4.07$, $p = 0.046$, reflecting the fact that children with CIs did not attend to auditory input as well as children with NH. That difference could be predicted due to children in the CI group having hearing loss. Nonetheless, because of that difference, scores for this category are reported, as well as total scores.

In general it can be seen from Table 11.2 that mean scores for both groups were well within the average ranges on these screening instruments. That means that any group differences found for language and literacy measures can be fairly attributed to differences in hearing status, and the fact that children with CIs were learning language with a degraded signal.

For the children with hearing loss, further criteria had to be met in order for them to participate. There could be no evidence that the hearing loss was progressive in nature. As closely as could be determined, it needed to be present since birth. Better-ear pure-tone average thresholds for the frequencies of 0.5, 1, and 2 kHz (better-ear PTAs) needed to be poorer than 50 dB hearing level. The children needed to have been identified with hearing loss, received appropriate amplification, and started an intervention program by the time they were 2 years of age in order to be included in the study. For this group of children with CIs, mean age of identification was 6 months (SD = 7 months); mean age at which they received their first hearing aids was 8 months (SD = 6 months), and they began early intervention by a mean age of 9 months (SD = 7 months). Mean better-ear PTAs before receiving CIs was 100 dB hearing level (SD = 17 dB).

A few children are exceptions to the descriptions offered above, and they are the children who received their CIs late. Forty-three of the 50 children in this CI group received a first CI before 3 years of age, with a mean age of 16 months (SD = 5 months). Those children all had better-ear PTAs poorer than 80 dB hearing level, with a mean of 105 dB (SD = 13 dB). The seven children who received a first CI after 3 years of age (with a mean age of 58 months), all had better-ear PTAs better than 80 dB hearing level, with a mean of 71 dB. (SDs are not listed here because the group is so small.) These late-implanted children are also distinguished by the fact that they were identified with hearing loss and started intervention later than the early-implanted children: mean age of identification for the late-implanted children was 10 months and mean age of starting intervention was 12 months. These factors raise the specter that these late-implanted children form a distinct group. Because of that possibility, two-group *t* tests were performed on all 13 measures reported in this section of the chapter. Mean scores for the early- and late-implanted children were remarkably similar, and were not significant in any instance. Consequently, data at second grade from these seven late-implanted children are included with the larger group of children with CIs in this report.

Criteria for Parents and Early-Intervention Programs

In order for a child to participate in the EDCHL study, their parents and early intervention programs needed to meet certain requirements. All children had parents with normal hearing, and the language spoken in the home was predominantly English. In a few cases, grandparents visited who spoke a language other than English with each other and with the child's parents. However, in all cases parents spoke English with each other and with the children in this study. At every test session, parents were asked to reconfirm that it was their goal that their children would be fully mainstreamed in a regular educational setting by the start of traditional school age, without the need for a sign language interpreter. Some children, both with NH and with CIs, were exposed to a manual sign system from infancy through preschool. In all those cases, the stated purpose of using a sign system was to facilitate the acquisition of spoken language and/or to provide a means for the child to communicate wants and needs while learning to talk.

The early intervention programs in which children and their parents participated needed to provide services at least once per week during infancy and the toddler years. Those programs needed to be staffed by individuals with at least a Master's degree, and that educational background needed to be in a discipline related to communication and the needs of children with hearing loss. That typically meant that early intervention was provided by speech-language pathologists or teachers of the deaf. All of the children with CIs for whom data are reported in this section received early intervention services along with their parents at least once per week up to age 3 years, and then they attended preschool programs for children with hearing loss for an average of 16 h/week. They were generally mainstreamed into regular classrooms starting at kindergarten, but for a couple children, mainstreaming did not start until first grade.

Method

Children and one parent traveled to Columbus, Ohio for a day and a half of testing during the summer following second grade. Four to six children were tested during each of these "camps" in six sessions. Children had a minimum of 1 h between test sessions. In each session, several tasks were combined to make between 40 and 60 min of data collection. Undergraduate and graduate students were involved in data collection in each of the six sessions. These students were

thoroughly trained during the spring preceding the summer camps, and were required to practice procedures on at least 15 children with normal hearing whose data are not included in this report. Training emphasized testing details, such as how to provide verbal reinforcement for staying on task and working hard without providing reinforcement for giving correct answers. During the training of experimenters as well as during data collection itself, the program manager observed test sessions and reviewed video recordings to make sure that no experimenter strayed from standard protocol.

All procedures for stimulus presentation were made standard and automated. Any test instrument that is typically presented with live voice by a clinician or experimenter was presented on a computer monitor with audio presented on a high-fidelity speaker at 0° azimuth. All materials were presented at a 68 dB sound pressure level. Materials for these presentations were created by video recording a member of the laboratory staff producing test instructions and test items. High-quality audio was ensured on these videotapes by having the staff member wear a FM transmitter, and the signal from the receiver was fed into the video camera. With the exception of two tasks, children were videotaped as they were responding, and care given to recording the relevant dimension of the responses. When responses involved pointing, for example, the video camera was positioned so those responses could be seen on the video recording. When responding involved verbal responses, clear shots of the children's faces were obtained. Figure 11.3 shows the setup for data collection for the passage comprehension subtest of the CASL. In this case, the video camera recording children's responses was positioned behind them in order to capture the pointing responses. All tasks used in data collection were preceded by appropriate training. Scoring was done using the video records at a later time, with the stipulation that the staff member who collected a specific kind of data could not score responses for those data. All scoring was done by two independent staff members so that reliability could be checked.

The two tasks that were not video-recorded were the phonological awareness and the working memory tasks. In these cases, responses were entered directly into the computer by the software that controlled the experiments.

Outcomes are presented for the same set of language skills reviewed in the previous section: lexicality, grammar, phonological awareness, reading, and working memory. In sum, there were 13 measures that were examined.

Lexicality

Two measures were used to assess the size of children's lexicons and their abilities to use words in spoken language. Expressive vocabulary was assessed with the EOWPVT. This task requires the child to provide the words that label a series of pictured items shown one at a time on separate pages. Standardized scores were used as dependent measures.

Children's skill at using their lexical knowledge as part of spoken language (i.e., semantics) was evaluated by the number of different words they used in a 20-min narrative sample consisting of a story retelling. For this narrative, each child entered the sound booth and the experimenter explained that she had been called away for a few minutes. The equipment was set up for the child to view and hear a video of the book *The Day Jimmy's Boa Ate the Wash* (Noble 1980). This story was video-recorded with a narrator reading the printed material, but with separate staff members saying the material that appeared in quotes in the book. Full images of each face were

Fig. 11.3 Setup for testing

shown to ensure optimal opportunity for speech reading. Illustrations from the book were shown when appropriate. The experimenter explained that she hadn't seen the video story yet, and asked the child to watch carefully so it could be told to the experimenter when she returned. After the story was finished, the experimenter reentered the sound booth, and asked the child to tell her the story in as much detail as possible. If the story retelling did not take a full 20 min, the experimenter supplemented the time by asking questions about personal experiences the child had that paralleled some of those of the children in the story. Later the story retelling was transcribed by members of the laboratory staff. Those transcriptions were submitted to the Systematic Analysis of Language Transcripts (SALT) software (Miller and Iglesias 2010) for analyses of morphosyntactic structures, including the number of different words (NDW). For most SALT measures, including NDW, the first 100 utterances were used in the analysis. The NDW score indexes how well children use their vocabulary knowledge in their generative language.

Grammar

Children's abilities to understand morphosyntactic structure was assessed using the paragraph comprehension subtest of the CASL. In this task, children listen to progressively more complex stories, and have to answer comprehension questions by pointing to one of four choices on an easel. The stories and questions were video-recorded by a staff member. It is characterized by test authors as a measure of receptive syntax.

Generative grammar was assessed by three measures obtained with SALT analysis: mean length of utterance in morphemes (MLU), number of pronouns, and number of conjunctions. Although MLU is frequently criticized for being insensitive to language differences once children reach MLUs of roughly 5, we have found it continues to distinguish between syntactic capabilities for children with hearing loss and those with NH past that stage.

Phonological Awareness

Three measures of phonological awareness were used. Multiple measures are always used in our laboratory so that differences among groups will not be diminished by selecting a task that is either so easy that even children with phonological delays can perform it, or so hard that even children who are developing typically have difficulty. Using multiple tasks also provided an opportunity to evaluate whether children with CIs seem to have any special difficulties with meta-linguistic analysis. For these second graders, the three tasks used were the initial consonant choice, final consonant choice, and phoneme deletion tasks. Again, all test stimuli were video-recorded using laboratory staff members as talkers. In the first two tasks, children saw and heard a target word that they were required to repeat correctly. They were given three opportunities. If they could not repeat it correctly, that test item would not be included. However, all these children were able to understand the target words without difficulty. After repeating the target word, children saw and heard three word choices and had to select which of the three started or ended with the same sound, depending on which task was presented. In the phoneme deletion task, children saw and heard a non-word and had to repeat it correctly. Next they were asked to say the non-word without one of the segments. They needed to delete the correct segment and blend the others to create a real word. This task involved more phonological *processing* than the first two tasks, so required greater meta-linguistic awareness. In order to complete the phoneme deletion task, the child not only needed to be sensitive to phonological structure, but also needed to be able to manipulate segments within the non-word. By including this task, we were able to get an indication of whether any differences between groups would best be attributed to deficits in sensitivity to segmental structure, or to diminished capacities to engage meta-linguistic awareness. Each task had many items (i.e., 32 or 48), and all have been used extensively in this laboratory and others so they were known to be reliable (e.g., Nittrouer and Burton 2005; Nittrouer et al. 2012; Pennington et al. 1990; Stanovich et al. 1984). The percentage of items answered correctly was the dependent measure for each task.

Reading

The Qualitative Reading Inventory (QRI) was used to assess word reading, paragraph comprehension, and fluency. Although this last measure had not been found to distinguish children with NH and those with CIs when they were tested at kindergarten, it seemed worthwhile to examine it again because fluency is commonly used in educational settings to assess reading skill.

The QRI has both narrative and expository passages written at various levels of reading ability. In this study, children read each passage and were asked ten comprehension questions about each one. Three passages were used at each test age. One passage was a narrative written at one level below grade level, one was a narrative at grade level, and one was an expository at grade level. Children were video-recorded reading each story and responding to questions. Staff members scored the number of words read correctly and the number of questions answered correctly. Finally, the time required to read the passage was computed from the videotape, and the number of correct words read per minute was used as the metric of fluency.

Working Memory

On this task, children were asked to recall the order of strings of six monosyllabic nouns presented as auditory lists. In this case, video presentation was not used. A single set of words served as stimuli, and recognition was checked for each

child both prior to testing and after testing was completed. If a child had difficulty recognizing even a single word auditorily, testing would not have been conducted (if it happened during the pre-test) or data would have been removed from analysis (if it happened on post-test). However, all children readily recognized these simple nouns.

This test procedure has been used often to examine short-term memory (e.g., Brady et al. 1983; Spring and Perry 1983), and this particular task with these particular words has been shown to have good test-retest reliability (Nittrouer and Miller 1999). In this procedure, pictures of each noun are shown at the top of a touch screen monitor, and the words are played in random order at a rate of one per second. The child's task is to touch the pictures in the order that the words were heard. Ten lists are presented. The dependent measure is the percentage of words recalled in the correct order.

Results

Data for the 13 measures described above were screened for normal distribution and homogeneity of variance. Data on all measures met the criteria.

Overall Performance

Means and standard deviations for the measures described above were computed. Two-group *t* tests were performed, and Cohen's *d*s were obtained. These last values index effect sizes by representing group differences according to standard deviations. Thus, a Cohen's *d* of 1.00 represents a group difference of one standard deviation.

Looking first at lexicality, means and standard deviations of those measures are shown in the two left-most columns of Table 11.3, with statistical outcomes shown below. The EOWPVT scores indicate that the expressive vocabularies of children with CIs were not as large as those of children with NH. Based on NDW, it is clear that using those lexical items to represent semantic variation in spoken language was not a skill that was as well developed for children with CIs as it was for children with NH. However, the difference in performance between the two groups of children is not as great for NDW as for EOWPVT. Thus, although the lexicons of children with CIs were not as large as those of children with NH, their skills at using those vocabulary items were less delayed.

Looking next at grammar, results for those measures are shown in the right-most columns of Table 11.3. Here it is seen that children with CIs were not performing as well as children with NH, but none of these effect sizes are as large as that found for expressive vocabulary with the EOWPVT.

Scores for the measures of phonological awareness are shown in the left half of Table 11.4, with statistical outcomes shown below. These measures reveal some of the largest effect sizes observed in this study, with the final consonant choice task showing the single largest effect. Of pertinence is the finding that children with CIs performed better on the phoneme deletion than on the final consonant choice task, $t(47) = 3.232$, $p = 0.002$, whereas children with NH performed indistinguishably on the two tasks. That finding for children with CIs provides support for the suggestion that they do not have diminished capacities for meta-linguistic awareness or phonological processing because they were able to do relatively well on the phoneme deletion task, the more meta-linguistically challenging of the tasks. Rather, it is recovering phonological structure that remains a challenge for them. (These were the only measures that could not be collected for all 98 children. One child with CIs was not able to understand the instructions for phoneme deletion, and another child with CIs became ill part way through testing, and could not complete the initial consonant choice or phoneme deletion tasks.)

Reading scores are shown in the right half of Table 11.4, with statistical outcomes shown below. Of these, fluency shows the weakest effects, making it a less sensitive metric of group difference than the other two measures. Paragraph comprehension shows the greatest difference between children with NH and those with CIs.

Scores for working memory are not shown in the tables described above. However, mean recall was 56 and 43 % correct for children with NH and those with CIs, respectively (SD = 16 % for each group). The *t* test performed on these data showed a significant group effect, $t(1,96) = 3.97$, $p < 0.001$, with a Cohen's *d* of 0.81. Thus, children with CIs are poorer at retaining verbal material in short-term memory, which could interfere with their learning of syntactic structures.

In summary, all 13 of these measures revealed significantly poorer abilities for children with CIs than for those with NH. The magnitude of those differences was generally between three-quarters of a standard deviation and one standard deviation, matching effect sizes found in the data of other investigators and summarized in the first section of this chapter. Consequently it seems fair to conclude that even with early identification, good intervention, and CIs, children with severe-to-profound hearing loss still experience significant delays in language acquisition because of that hearing loss. The challenge facing clinicians and scientists is to enhance our understanding of the mechanisms underlying the acquisition of spoken language for children with CIs in order for us to appropriately modify intervention techniques so that these children may one day reach their full potential.

Computing Latent Variables

Analyses of sorts other than those that merely measure differences in abilities between children with NH and those with CIs were performed to help us understand how spoken language skills interact with each other for these

Table 11.3 Group means and *standard deviations* for children with normal hearing ($N=48$) and children with CIs ($N=50$) for measures of lexicality and grammar. Results of two-group *t* tests performed on the measures, along with Cohen's *d*s are shown below. Degrees of freedom were 1, 96 for all analyses

	Lexicality				Grammar							
	EOWPVT		NDW		CASL		MLU		Conjunctions		Pronouns	
	M	*SD*	M	*SD*	M	*SD*	M	*SD*	M	*SD*	M	*SD*
Normal hearing	110	*14*	292	*56*	112	*12*	6.27	*1.35*	106	*41*	231	*70*
Cochlear implants	95	*19*	248	*65*	99	*21*	5.43	*1.34*	79	*41*	177	*72*
t value	*4.56*		*3.61*		*3.61*		*3.11*		*3.17*		*3.70*	
p value	*<0.001*		*<0.001*		*<0.001*		*0.002*		*0.002*		*<0.001*	
Cohen's *d*	*0.90*		*0.73*		*0.76*		*0.62*		*0.66*		*0.76*	

Note: EOWPVT (Expressive One-Word Picture Vocabulary Test) and CASL (Comprehensive Assessment of Spoken Language) = standardized scores with means of 100 and *SD*s of 15; NDW (Number of Different Words), Conjunctions, and Pronouns = count of occurrence of each in 100-utterance sample; MLU (Mean Length of Utterance) = count across the language sample

Table 11.4 Group means and *standard deviations* for children with normal hearing ($N=48$) and children with CIs ($N=50$) for measures of reading and phonological awareness. Results of two-group *t* tests performed on the measures, along with Cohen's *d*s are shown below. Degrees of freedom were 1, 96 for all measures, except for Initial Consonant Choice (1, 95) and Phoneme Deletion (1, 94)

	Phonological awareness						Reading					
	Initial consonant		Final consonant		Phoneme del.		Comprehension		Word read		Fluency	
	M	*SD*	M	*SD*	M	M	M	*SD*	M	*SD*	M	*SD*
Normal hearing	87	*13*	70	*18*	71	*22*	21	*3*	200	*5*	122	*32*
Cochlear implants	65	*26*	36	*26*	50	*31*	16	*6*	191	*15*	104	*38*
t value	*5.45*		*7.34*		*3.84*		*4.16*		*4.05*		*2.62*	
p value	*<0.001*		*<0.001*		*<0.001*		*<0.001*		*<0.001*		*0.010*	
Cohen's *d*	*1.07*		*1.52*		*0.78*		*1.05*		*0.80*		*0.51*	

Note: Initial Consonant Choice, Final Consonant Choice, and Phoneme Deletion = percent of correct responses on each phonological awareness measure; Comprehension = number (out of 30) of comprehension questions answered correctly on the Qualitative Reading Inventory (QRI); Word Reading = number of words read correctly on the QRI; and Fluency = mean number of words read per minute on the QRI

children. In particular, factor analysis was performed to see if data across the 13 separate measures described above could be reduced to reveal a smaller set of latent variables. Specifically, factor analysis with varimax rotation was done on these measures. Although not strictly confirmatory in approach, this analysis was not exploratory, either. Predictions about how these measures might combine to create a few components could be made based on traditional models of linguistic structure. In particular, linguistic structure is generally viewed as having duality of patterning, in which a limited set of phonemic elements get combined to create almost an infinite variety of words and those words can be combined according to a finite set of rules to create sentences with almost infinite meanings (e.g., Hockett 1958; Studdert-Kennedy 2005). According to that model of duality, it was reasonable to expect prior to this analysis that the separate measures might reveal two latent variables based on these levels of structure: phonological and morphosyntactic. Thus the resulting variables would reflect language abilities associated with sensitivity to and processing of phonological structure or morphosyntactic structure.

Table 11.5 shows the standardized component matrix that resulted from the analysis, with significant effects indicated by bolded text. As expected, the variance in each of the individual measures is well explained by one of the components, but not the other. Furthermore, it seems appropriate, based on the measures associated with each component, to label the first of these a phonological component and label the second one a morphosyntactic (grammar) component. In this analysis, the number of participants relative to the number of measures was slightly less than optimal, but the strength of the components derived and the fact that those resolved components are conceptually sound militate against rejecting the outcomes because of that concern. Tabachnick and Fidell (1989) argue that in a situation such as this one (i.e., resolved components are strong and conceptually sound) five cases per measure is sufficient.

Table 11.5 The proportion of variance on each measure explained by the principal component

	Components	
	1	2
EOWPVT—expressive vocabulary	***0.814***	0.166
NDW—semantics	0.222	***0.875***
CASL—receptive syntax	***0.711***	0.346
MLU	0.235	***0.825***
Conjunctions	0.055	***0.890***
Pronouns	0.144	***0.761***
Initial consonant choice	***0.838***	0.104
Final consonant choice	***0.693***	0.153
Phoneme deletion	***0.823***	0.105
QRI—paragraph comprehension	***0.746***	0.383
QRI—word reading	***0.762***	0.071
QRI—fluency	***0.763***	0.221
Working memory	***0.622***	0.065

Note: *bolded text* indicates significant effects, with a $p < 0.05$

Scores for the two latent variables derived from this analysis were computed for each child, using the children with NH as the standard. Consequently, the mean for children with NH was 0.00 and the standard deviation was 1.00 on both the variables of phonological processing and of morphosyntax. It was found that means for the children with CIs were −1.86 (SD = 2.01) for the phonological processing variable and −0.87 (SD = 1.20) for the morphosyntactic variable. That means that the children with CIs in this study were trailing the children with NH in acquisition of both phonological and morphosyntactic skills, but they were further behind in phonological skills. That outcome could have been predicted from the fact that CIs provide signals that are highly degraded, allowing only limited access to the acoustic properties that underlie phonemic categories. Morphosyntactic structure can more readily be learned from how words are combined and knowing when to use each word. Even if the representations of those words are more global (i.e., less phonemically differentiated) for children with CIs than for children with NH, the rules for combining and using words may be learned.

Explaining Variance

Finally, Pearson product-moment correlation coefficients were computed for all pairwise combinations of the dependent measures examined, with one exception. Reading fluency was not included in this analysis because it was not found to be especially sensitive to differences between children with NH and those with CIs.

The primary motivation for this particular analysis was that by examining relationships among separate language measures, ideas should be derived concerning which skills would best be targeted in intervention. Table 11.6 shows correlation coefficients for each group separately: those for children with NH are on the top in each cell and those for children with CIs are on the bottom. Computing correlation coefficients separately for each group allowed us to examine whether the same pattern of relationships across skills could be observed for children in both groups. Again, that should help in designing interventions. A serious risk to the design of effective treatment options is encountered when strategies are based on patterns of language development found for children with NH because those patterns may or may not hold for children with CIs. In fact, examining Table 11.6 reveals that the most striking outcome is that many more of these correlations were significant for the children with CIs than for those with NH. Out of the 65 correlations performed, 51 were found to be significant for the children with CIs, while only 19 were significant for the children with NH. Fisher's z tests for the difference between correlation coefficients were performed on all pairs of coefficients in order to see if the strength of the relationship between individual pairs of measures were different for the two groups of children. In 28 cases, Fisher's z was significant, and in all those cases it was because the relationship was stronger for children with CIs than for those with NH. In Table 11.6, significant z scores are indicated by bolding. These outcomes indicate that language skills are less diversified for children with CIs than for children with NH.

Summary

This second section of the chapter reported data for second graders that come from an ongoing longitudinal study. All results are consistent with the pattern of outcomes reported in the first section of the chapter, from other studies. Even though the children in the longitudinal study have no risk

Table 11.6 Pearson product-moment correlation coefficients for pairs of dependent measures computed for children with NH (*top rows*) and children with CIs (*bottom rows*) separately

	E.V.	NDW	CASL	MLU	Conj.	Pro.	IC	FC	PD	Comp.	W.R.	W.M.
EOWPVT	1	–	–	–	–	–	–	–	–	–	–	–
NDW	0.***099*** ***0.456****	1	–	–	–	–	–	–	–	–	–	–
CASL	0.***437**** ***0.722****	***0.135*** ***0.483****	1	–	–	–	–	–	–	–	–	–
MLU	***–0.018*** ***0.433****	***0.541**** ***0.820****	0.349* 0.524**	1	–	–	–	–	–	–	–	–
Conjunctions	–0.179 0.189	0.706** 0.771**	–0.051 0.274	0.681** 0.680**	1	–	–	–	–	–	–	–
Pronouns	–0.014 0.265	***0.214*** ***0.857****	0.182 0.321*	***0.274*** ***0.693****	***0.283*** ***0.845****	1	–	–	–	–	–	–
Initial consonant	***0.171*** ***0.700****	0.089 0.205	***0.214*** ***0.568****	0.056 0.251	0.073 0.092	0.218 0.060	1	–	–	–	–	–
Final consonant	***0.144*** ***0.468****	***–0.137*** ***0.259***	0.141 0.439**	***–0.139*** ***0.343****	–0.092 0.156	0.071 0.200	0.559** 0.424**	1	–	–	–	–
Phoneme deletion	***0.232*** ***0.690****	***–0.284*** ***0.441****	0.309* 0.502**	***–0.142*** ***0.470****	***–0.304**** ***0.359****	–0.055 0.244	0.438** 0.667**	0.590** 0.531**	1	–	–	–
Reading comprehension	***0.496**** ***0.713****	0.350* 0.536**	***0.561**** ***0.788****	0.268 0.528**	0.143 0.330*	0.151 0.354*	***0.178*** ***0.637****	0.068 0.378**	***0.196*** ***0.644****	1	–	–
Word reading	***0.183*** ***0.574****	***–0.038*** ***0.327****	0.140 0.375**	***–0.053*** ***0.384****	–0.015 0.263	–0.039 0.224	0.543** 0.583**	0.401** 0.331*	0.508** 0.633**	***0.202*** ***0.527****	1	–
Working memory	0.193 0.473**	***–0.074*** ***0.321****	0.251 0.342*	***0.005*** ***0.362****	–0.025 0.197	0.023 0.165	0.351* 0.563**	0.311* 0.303*	0.299* 0.501**	0.253 0.315*	***0.124*** ***0.534****	1

*Individual correlation coefficient is significant at the 0.05 level
**Individual correlation coefficient is significant at the 0.01 level
Bolded text indicates that Fisher's *z* showed a significant difference between groups in correlation coefficients, at a minimum of the 0.05 level (one-tailed test), but in many cases significance levels were higher

factors for language delay other than hearing loss, and they all received appropriate and early treatment for that hearing loss, they trail behind their peers with NH by a substantial margin when it comes to language learning. Across the 13 measures reported in this section, the mean Cohen's *d* was 0.84. It was specifically found that children with CIs are further behind on skills requiring sensitivity to the phonological structure of the speech signal, rather than morphosyntactic abilities. That strong demonstration of phonological deficit surely reflects the fact that even with technological advances, CIs still provide degraded representations of spectro-temporal structure in the speech signal. Consequently these children have diminished access to some important cues to phonemic categories.

One piece of good news to come from these analyses is that morphosyntactic skills appear to be learned quasi-independently from the other language skills examined in this study, which are all strongly dependent on phonological structure. That outcome seems consistent with the proposal that young children with NH are less sensitive to word-internal segmental (phonemic) structure than are adults (Jusczyk and Derrah 1987; Locke 1988; Nittrouer 1992; Studdert-Kennedy 1981; Walley and Carrell 1983). For example, kindergartners have been found to judge similarity of non-word pairs based primarily on overall syllable shape, rather than on shared phonemes; by second grade, similarity is judged based on shared phonemes (Walley et al. 1986). As another example, the organization of the lexicon for 6-year-old children seems less clearly based on phonemic structure than are those of adults; instead children's lexicons seem based more on global acoustic patterns (Charles-Luce and Luce 1990). So although the rate of lexical acquisition is influenced by children's sensitivity to phonological structure (Willstedt-Svensson et al. 2004), children with normal hearing who are slightly younger than those for whom data are reported here acquire vocabulary items with less than adult-like sensitivities to phonological structure. It seems fair to suggest that the second graders with CIs in the EDCHL study may still be acquiring new vocabulary with those global representations. The independence of phonological and morphosyntactic skills revealed by the factor analysis reported in this section suggests that children with CIs can learn how to combine and when to use those lexical items in spite of having diminished sensitivity to phonological structure.

Finally, the results of correlational analysis reported in this section show that the language skills evaluated by the separate measures used in the EDCHL study are more interdependent for children with CIs than for those with NH. That finding emphasizes the need for enriching the language environments of children with CIs in a broad sense, rather than only working on separate language skills in piecemeal fashion, as might occur in pull-out intervention sessions.

Conclusions

This chapter has reviewed language and literacy outcomes for children with severe-to-profound hearing loss who receive CIs. Work by other investigators was reviewed, as well as work from an ongoing longitudinal study taking place in this laboratory. Outcomes were found to be consistent across studies, and reveal that children with CIs are performing on most language measures at roughly the 15th percentile of performance for children with NH. These findings suggest that language outcomes have not substantially improved for children with CIs since those devices first became available. Looking at the patterns of relationship across skills, it appears that morphosyntactic skills are not as affected by hearing loss and subsequent implantation as are skills dependent on sensitivity to phonological structure. Because the degraded nature of signals available through CIs likely diminishes access to the kinds of acoustic information needed to develop sensitivity to phonological structure, these language problems can be traced specifically to the nature of the signal children receive through their CIs. This situation means that ultimately solutions to the problems faced by children with CIs must involve the types of auditory prostheses we provide to them, but behavioral interventions should help, as well.

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12 Benefits of Cochlear Implantation on the Whole Child: Longitudinal Changes in Cognition, Behavior, Parenting, and Health-Related Quality of Life

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Using a dynamical systems approach, psychologists have demonstrated that there are interconnections across areas of development, and that deficits in one area can lead to cascading effects in others (Rubin et al. 2003; Smith and Thelen 1993; Thelen and Smith 1994). Thus, children with hearing loss, who have substantial delays in language development, are likely to experience deficits in learning and socioemotional development. Specifically, effects on selective attention, joint engagement, behavioral regulation, and social competence are well documented (Arnold and Tremblay 1979; Barker et al. 2009; Kennedy et al. 2006). To date, the majority of research on the effects of cochlear implantation in children has been focused on remediation of oral language skills, with less research into the broader impact of childhood deafness on cognition, behavior, and social functioning. Thus, an assessment and intervention process that takes into account the developmental progress of the "whole child" in relation to his/her family is warranted.

This chapter provides a broad review of the effects of cochlear implantation on young, deaf children's development and specifically includes data published from the largest, most nationally representative sample of children with cochlear implants in relation to their hearing peers (Childhood Development after Cochlear Implantation; CDaCI).

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Early Cognitive Development

Visual Attention

Evidence suggests that hearing and seeing are coupled systems in infancy (Bates and Dick 2000; Bates et al. 2003). Infants with normal hearing look in the direction of sound, track visual events whose temporal rhythms match what they hear, and more deeply process sights and sounds that are linked (Morrongiello 1994; Roberts 1994; Walker-Andrews and Lennon 1991). However, early childhood deafness disrupts this normal process, affecting their ability to direct, engage, and disengage visual attention (Quittner et al. 1994; Smith et al. 1998; Spencer 2000).

Studies of children with hearing aids and cochlear implants have consistently documented deficits in visual, selective attention (Dye and Hauser 2014; Quittner et al. 1994, 2007; Smith et al. 1998; Yucel and Derim 2008). These findings are counterintuitive since the visual system develops normally in this population and better visual attention would be adaptive for interpreting sign language, lip reading, or other forms of visual communication. This is likely due to a lack of integration of the visual and auditory systems early in brain development and the need for young deaf children to monitor their environment visually rather than auditorily (Markman et al. 2011). Although there is evidence that children and adults with hearing loss develop stronger visual skills in the periphery (Bavelier et al. 2006; Neville and Lawson 1987), selective attention requires focusing on targets in the center of the visual field.

Three key studies have demonstrated differences in visual attention skills in children with severe-to-profound sensorineural hearing loss (SNHL) who used hearing aids or cochlear implants (CIs) in comparison to age-matched hearing controls. Two studies used a computerized continuous performance task (CPT) that involved no sound

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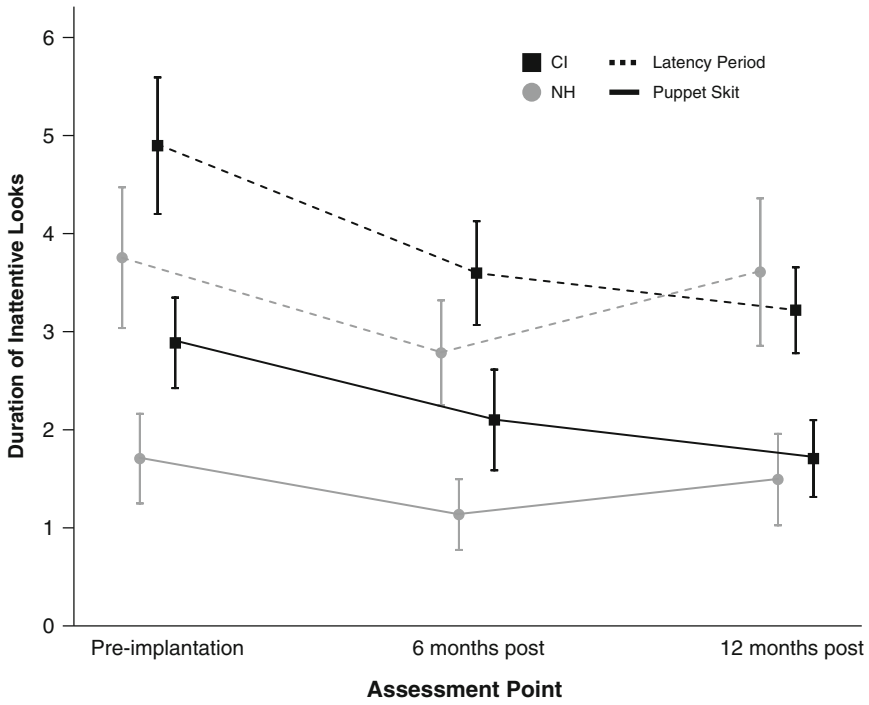

Fig. 12.1 Duration of inattentive looks averaged across the eight puppet skits and latency skits (error bars represent standard errors)

(Quittner et al. 1994; Smith et al. 1998). This task has been used to identify children with serious attentional deficits (Barkley 1988; Gordon 1986) and has well-established norms. In the first study to evaluate visual attention in children with SNHL using either hearing aids or CIs, both groups of children with hearing loss showed lower performance on visual attention tasks in comparison to hearing children. However, there was a significant interaction with age. At the younger age levels (6–8 years), 26 % of children with hearing aids and 52 % of children with CIs performed at or below the 5th percentile of the normative sample. At the older age levels (9–13 years), 39 % of children with hearing aids and 18 % of children with CIs fell in this range. Thus, older children in the CI group appeared to "catch up" and performed as well as their hearing peers (Quittner et al. 1994). In a recent study using the same task (Dye and Hauser 2014), both deficits in visual attention and similar age effects were found; the younger deaf children, ages 6–8, performed worse than the older children, ages 9–13. These authors speculated that the older children have developed better visual attention in the periphery but can inhibit this process when a task requires attention to the central visual field (Dye and Hauser 2014).

In the second longitudinal experiment with deaf children using hearing aids or cochlear implants and normal hearing controls, the CPT was completed twice over a period of 10 months. Results indicated that hearing children and older deaf children using CIs reached higher levels of performance than deaf children using hearing aids (Smith et al. 1998). Furthermore, significant correlations were found between visual attention scores and parental reports of the child's responsiveness to sound on the Meaningful Auditory Integration Scale (MAIS; Robbins 1991).

Finally, visual selective attention was measured using a 10-min videotaped Puppet Task (Ruff et al. 1990) in much younger deaf children (ages 5 months to 3 years), over 12 months postimplantation. At baseline, prior to implantation, deaf children demonstrated significantly longer and more frequent inattentive looks during the puppet skits than did hearing children. Longitudinal analyses revealed significant decreases in the frequency of inattentive looks for both groups, with a significant decrease in the duration of these looks for the CI group. The largest decrease in duration of off-task looks occurred at 6 months postimplantation, indicating that improvements occurred rapidly after restoration of auditory input, see Fig. 12.1 (Quittner et al. 2007). Notably these were the first observable changes in this large, nationally representative cohort.

Joint Engagement

Numerous studies have indicated that joint engagement between a parent and child develops within the first 3 years of life (Adamson et al. 2004). This developmental process lays the foundation for regulating attention and affect during interpersonal interactions, subsequently scaffolding representational skills and use of symbols in play (Beuker et al.

2013; Smith et al. 1988). As joint attention skills develop in the latter half of the first year, children begin to sustain these episodes of joint engagement, which facilitates the sharing of objects and events with a social partner, such as a parent (Adamson et al. 2004; Bakeman and Adamson 1984). As children begin to master language, by 18–30 months, there is a substantive transition to the use of symbols in play, which allows the parent and child to communicate about topics not outside of the current moment (e.g., dressing a doll and discussing favorite clothes). Thus, joint engagement is strongly linked to language development and is one mechanism by which children map language onto their environment (Tomasello and Farrar 1986; Wetherby and Prizant 2002).

Joint engagement has been conceptualized as a precursor to early communicative and linguistic development, is dyadic and interactional, and relies on coordinated exchanges between partners (Girolametto et al. 1994; Tasker and Schmidt 2008). Several joint engagement states have been identified, ranging from "disengaged" to "symbol-infused coordinated" joint engagement (see Table 12.1). The interaction must meet several criteria to be considered "symbol-infused coordinated joint engagement," which is considered the highest level of joint engagement. First, the dyad must be actively involved with the same object or event ("joint"). Second, the child must be using some form of symbolism ("symbol-infused"; e.g., using a block as a bed for a doll). Last, the child must coordinate his/her attention to the partner and shared object ("symbol-infused coordinated joint engagement") (Adamson et al. 2004).

Given that children with hearing loss have significant delays in oral language and difficulties with parent–child communication, the development of symbol-infused joint engagement is likely to be disrupted (Gale and Schick 2009; Prezbindowski et al. 1998; Quittner et al. 2013). Recently, in a cross-sectional study of 188 deaf (prior to implantation) and 97 matched hearing children, joint engagement was evaluated in a 10-min videotaped free play task (Cejas et al. 2014). Parent–child interactions were coded using Adamson's system (Adamson et al. 2004); percent time in each of the eight attentional states was calculated. Language was measured using both MacArthur-Bates Communicative Development Inventories (CDI; Fenson et al. 1993) and the Reynell Developmental Language Scales (RDLS; Reynell and Greuber 1990).

By the age of 18 months, hearing children were rapidly increasing the amount of time spent in symbol-infused states (i.e., child and parent taking turns to feed a doll) and by 36 months of age and older, the majority of their time was spent in this engagement state (93 %). In contrast, deaf children 18 months and older spent relatively little time in this state. At 18–36 months, they averaged only 8 % of their time in this state, which increased to 34 % at 36 months and older. In contrast to chronological age, analyses using language age revealed that the youngest deaf and hearing children spent

Table 12.1 Definitions of attention states

Engagement state	Definition	Example
Unengaged	There is no evidence that the child is actively involved with a person, object, or symbol.	Child is staring into space, staring at the floor or having a tantrum.
Onlooking	Child is watching another person, observing his/her activity, or the objects the person is manipulating; however, child is not engaged or involved with the activity.	Child watches while parent stacks the blocks to make a tower.
Object	Child is engaged with one or more objects, exploring or playing with objects him or herself.	Toddler is playing with his/her foot or playing with a car or truck.
Person	Child is actively engaged solely with a person (no objects are involved).	Parent tickles child and child reacts to the activity.
Supported Joint Engagement	Child is actively involved with an object or event with which the other person is also engaged, but the child does not acknowledge this involvement.	Parent and child take turns rolling a ball and child is only focused on the movement of the ball.
Coordinated Joint Engagement	Child coordinates his/her attention between person and objects. Child acknowledges the person's involvement.	Child holds a toy plane, shows it to mother, and then moves it through the air, while parent acknowledges their shared focus by laughing and saying "zoom".
Symbol-Infused Supported Joint Engagement	Child and parent are both focused on the same object or event and there is evidence that the child is actively attending to symbols (e.g., oral and sign language, symbolic gestures, symbolic play), but the child is not explicitly attending to the parent.	Parent assists the child as he/she focuses on pretending a block is a train.
Symbol-Infused Coordinated Joint Engagement	Child is coordinating his/her attention between the parent and a shared object or event, and the child is actively attending to symbols.	Parent and child are discussing a toy of mutual interest or they are taking turns feeding a doll.

similar amounts of time in symbol-infused states. However, delays in joint attention were still present in the older deaf group (18–36 months) when using language age. We plan to follow-up these children over 3 years of cochlear implantation to determine whether the implant facilitates the emergence of joint engagement. Interventions that promote a richer language environment and foster positive dyadic interactions may be helpful in increasing the use of symbols in play and thus, indirectly improve the growth of oral language.

Behavioral Development

Behavior problems in children have been shown to negatively impact a range of developmental, social, and educational outcomes (Masten et al. 2005; Pierce et al. 1999). Prior studies have documented strong links between language and behavior problems, with children diagnosed with language disorders showing a high incidence of behavior problems (Barker et al. 2009; Beitchman et al. 2001; Brownlie et al. 2004). Children with SNHL, in particular, exhibit higher rates of externalizing behavior problems (e.g., aggression, inattention, impulsivity), ranging from 30 to 38 % (Van Eldik et al. 2004; Vostanis et al. 1997) than normal hearing children (3–18 %; Hinshaw and Lee 2003). Some evidence suggests that visual attention is related to behavior problems for older children with hearing loss, but these findings need to be replicated and extended to younger children (Mitchell and Quittner 1996; Smith et al. 1998; Terwogt and Rieffe 2004). In particular, they had more difficulty than hearing children discriminating the target and inhibiting responses to nontarget information. Parent ratings on the Child Behavior Checklist (CBCL) indicated that one half of the hearing-impaired children had elevated levels of externalizing behavior problems, whereas teacher ratings indicated that one-third had significant behavioral problems.

Similar to externalizing problems, parents of deaf children report more internalizing problems (e.g., anxiety) than parents of hearing children (25–38 % vs. 2–17 %) (Albano et al. 2003; Hammen and Rudolph 2003; Van Eldik et al. 2004; Vostanis et al. 1997). A majority of these studies have been limited by a reliance on self- or parent–teacher report. In contrast, Barker and colleagues (2009) utilized both parent-report measures and videotaped observations to assess rates of behavior problems in deaf ($n = 116$) and hearing children ($n = 69$) ages 1.5–5 years. The Child Behavior Checklist (CBCL; Achenbach and Rescorla 2000) and three structured and unstructured videotaped play tasks (i.e., Free Play, Puzzle-Solving, Art Gallery) to assess Child Persistence and Child Negativity were used to make these comparisons (Quittner et al. 2004). Child Persistence was defined as the extent to which the child was actually involved in the task and Child Negativity was coded as the degree to which the child showed anger, dislike, or hostility toward the parent. These were the behavioral codes that we assessed during the parent–child interaction tasks.

Cross-sectional results indicated that across all measures, children with hearing loss evidenced more behavior problems than normal hearing children, with statistically significant differences on the CBCL Attention scale (i.e., hyperactive or inattentive behaviors), Internalizing Composite (anxiety or depressive symptoms), and videotaped Child Negativity (i.e., hostility toward parent during play). Surprisingly, the Externalizing Composite did not differ between the groups; however, a larger number of children in the deaf group scored within the at-risk range on the Aggressive Behavior Scale (Barker et al. 2009). A significant group difference was also found on duration of play during a solitary play activity, with hearing-impaired children engaging in less play than normal hearing children. In addition, children with hearing loss were also rated lower on Child Persistence than normal hearing children on both of the structured play tasks (Puzzle-Solving and Art Gallery). After accounting for language, there were no significant differences between the groups on sustained attention or amount of time children spent communicating with their parent.

One important but unanswered question is the direction of these effects. Do behavior problems lead to more significant delays in language or do deficits in language make it more difficult to regulate behavior? Recent longitudinal analyses in the same sample found that expressive language skills were strongly related to elevated externalizing behavior problems. Specifically, latent difference score modeling (McArdle 2009) suggested that for children implanted before age 2, receptive language and behavior problems shared a *reciprocal* influence. There was a bidirectional relationship, such that expressive language skills influenced rates of externalizing behaviors problems and vice versa. In contrast, for those implanted after age 2, the models suggested a *directional* relationship, with higher behavior problems leading to worse growth in receptive language (Romero et al. 2009, 2010). Receptive language skills did not influence the rates of behavior problems.

Social Functioning

Social competence is a broad construct that reflects a child's ability to interact effectively with those in the environment, such as peers, family members, and other adults (Waters and Sroufe 1983). It includes the ability to spontaneously utilize social skills in interactions in a flexible and adaptive manner (Lillvist et al. 2009). These social skills consist of reciprocity, perspective taking, complying with directions and rules, problem-solving, and responding to the actions of others (Cook and Oliver 2011; Spence 2003; Waters and

Sroufe 1983). It also encompasses the ability to express emotions appropriately and exhibit self-control (Gresham and Elliot 1990; Hogan et al. 1992). Thus, social competence is fundamental to the establishment and maintenance of positive relationships.

Social competence has a profound effect on several aspects of child development, facilitating family and peer relationships, emotion regulation, and academic achievement (Semrud-Clikeman 2007; Spinrad et al. 2006). It is also a strong predictor of important developmental outcomes, such as social anxiety, antisocial behavior, and later psychopathology (Hymel et al. 1990; Ladd and Troop-Gordon 2003). Children with hearing loss, who have delays in language and deficits in attention (Barker et al. 2009; Mitchell and Quittner 1996; A. L. Quittner et al. 1994; Smith et al. 1998), may be at increased risk for delays in social competence and related sequelae.

Recently, we measured social competence in preschoolers (mean age of 3 years) using teacher report on the Social Competence and Behavior Evaluation (SCBE; LaFreniere and Dumas 1995) and the Social Emotional Assessment Inventory (SEAI; Meadow-Orlans 1983). Our sample consisted of 89 children with hearing loss and 56 children with normal hearing enrolled in the CDaCI study who were 2 ½ years of age or older at baseline (prior to cochlear implantation). Results indicated that children with hearing loss scored lower on the Social Competence and General Adaptability scales than the comparison group. Age at diagnosis and age at amplification were significantly correlated with scores on the SCBE, with younger children having better social competence scores. Language age, measured by the CDI, RDLS, and the Comprehensive Assessment of Spoken Language (CASL; Carrow-Woolfolk 1999), predicted both Social Competence and General Adaptability. Interestingly, when the children with CIs were compared to the normative data (i.e., same-age children with hearing loss) on the deaf-specific SEAI, no delays in social competence were reported. Note however, that this measure was developed several decades ago, prior to implementation of newborn hearing screening and cochlear implant technology.

Measures of social competence completed later by children with and without cochlear implants, ages 8–12 years, also indicated significant deficits in social competence for those with hearing loss. On the Social Skills Rating Scale (SSRS; Gresham and Elliot 1990), children with CIs rated themselves as having significant delays at 96 months post-implantation. Similar results were found on parent report measures in the CI group. Post hoc analyses did not find any difference in social competence as a function of school setting or mode of communication. Specifically, no significant differences were found for children using sign language or for children enrolled in self-contained classrooms for children with hearing loss. This suggests that this effect was not driven by differences in school setting or mode of communication.

Impact on Family System

Parenting Stress

Raising a child with hearing loss is associated with significant parental stress due to substantial long-term challenges including communication, education, and health care (Barker et al. 2009; Lederberg and Golbach 2002; Marschark et al. 2011). Ninety percent of children with SNHL are born to hearing parents (Albertini 2010), thus, an immediate "mismatch" between the hearing status of the parent and child presents a significant barrier to communication (Quittner et al. 2004). In addition, previous studies have identified other factors that contribute to parenting stress, such as age at diagnosis, extent of hearing loss, mode of communication, and perceived social support (Asberg et al. 2008; Quittner et al. 2010; Pipp-Siegel et al. 2002).

The earliest study of parenting stress used both general and context-specific measures in 96 parents of young, deaf children ages 2–5 years and 118 age- and gender-matched children (Quittner et al. 1990). In this study, a disease-specific parenting stress measure was developed, the Family Stress Scale (Quittner et al. 1990, 2010). This measure consists of 16 items, 11 related to general family stressors and 5 specific to early childhood deafness. Items were rated on a 5-point Likert scale, ranging from 1 ("not at all stressful") to 5 ("extremely stressful"). This measure has yielded good internal consistency—Cronbach's alpha ranging from 0.76 to 0.89 (Quittner et al. 1990, 2010). Results in this early study found higher levels of stress on both the general (Parenting Stress Index; PSI; Abidin 1983) and context-specific measures of stress when compared to the matched hearing control group.

Several later studies of stress in parents of children with hearing loss have used primarily generic measures, such as the Parent Stress Index and have found contradictory results. Meadow-Orlans (1994) assessed stress in 20 parents of 9-month-old infants with mild-to-profound losses and 20 parents of similar-aged children without hearing losses. Her results indicated that there were no statistically significant differences in stress between the groups; however, 25 % of mothers in the deaf group compared with 5 % in the hearing group scored higher on this stress measure than the clinical cutoff. More recently, Pipps-Siegel and colleagues (2002) used the PSI in 184 hearing mothers of young children with mild-to-profound losses, ages 6 month to 5½ years and found no differences in stress levels between these parents and the

normative sample. Finally, Lederberg and Golbach (2002) evaluated stress in 23 mothers of deaf children and 23 mothers of normally hearing children 2 years of age using the PSI and the Questionnaire on Resources and Stress-Short Form (QRS-SF; Friedrich et al. 1983). Their results suggested that mothers of 2-year-old deaf infants reported similar levels of stress on the PSI compared to the mothers of children with normal hearing, but elevated stress on the QRS-SF. These elevations were not found when the children were 3 and 4 years of age. This suggests that generic measures of parenting stress (i.e., Parenting Stress Index) are not sensitive to the specific stressors of raising a child with hearing loss.

One possible reason for these conflicting results is the use of generic stress measures which do not capture the unique challenges experienced by hearing parents of children with hearing losses. In an effort to resolve this debate, we evaluated parenting stress in the CDaCI study with 181 parents of children with severe-to-profound hearing loss and 92 parents of children with normal hearing, mean age of 27 months. We used both the PSI, a generic measure, the Family Stress Scale, a deaf-specific measure, the Child Behavior Checklist (CBCL), a broad behavioral screener, and Child Negativity coded from videotaped parent–child interactions (Quittner et al. 2010). Child Negativity was defined as the degree to which the child shows anger, dislike, or hostility toward the parent. For example, a child rated as having high Negativity is repeatedly and overtly angry with the parent and forcefully rejects their ideas while resisting assistance. After controlling for maternal education and family income, parents of deaf children reported *more context-specific*, but not general parenting stress, than the comparison group. The top four stressors reported by parents of children with hearing loss were as follows: (1) communication, (2) finances, (3) educational concerns, and (4) child safety. In contrast, the top stressors for parents of hearing children were as follows: (1) finances, (2) discipline, (3) safety, and (4) following routines. Both the parent-reported and observed child behavior problems were higher in the deaf than in the hearing groups, which was associated with oral language delays. Furthermore, hearing status was related to parenting stress by way of language delays and child behavior problems. Thus, children who had more difficulties expressing themselves orally and exhibited more externalizing behaviors (e.g., impulsivity, inattention) had parents who endorsed greater stress in their parenting role. Other studies have also shown that high levels of parenting stress are associated with poor social and emotional development in the child (Crnic and Low 2002).

Maternal Sensitivity

Assessment of parenting behaviors is particularly important for children with severe-to-profound hearing loss. Early in development, parent–child interactions provide scaffolding for the normal growth of cognitive, behavioral, and communicative skills. Substantial disruptions in this process occur when the parent is hearing and the child has a hearing loss (Quittner et al. 2010). Observational studies have reported that hearing mothers of young deaf children have more controlling, directive, and intrusive interactions with their children; thus, displaying less warmth and positive affect with their deaf children than mothers of hearing children (Lederberg and Prezbindowski 2000; Meadow-Orlans and Steinberg 1993). The consequences of these dyadic interactions include less secure attachment, difficulties sustaining attention, and slower development of communicative competence (Barker et al. 2009; Bornstein 2000; Cruz et al. 2013).

To date, few studies have measured the quality of parent–child interactions and their effects on oral language; however, parental involvement has been shown to have a major impact on the growth of language in cochlear implant recipients. After accounting for maternal education, degree of hearing loss, and mode of communication, Pressman and colleagues (1999) found that greater maternal emotional availability (i.e., the ability to read child cues and respond appropriately, resolve parent–child conflict, and tolerate a wide range of affect while maintaining a positive tone) during videotaped free play sessions was a positive and unique predictor of oral language learning.

In the CDaCI study, we videotaped and coded parent–child interactions during 20 min of structured and unstructured play tasks. We used the National Institute of Child Health and Human Development's Early Childcare Study codes (NICHD 1999) for maternal sensitivity, which included a composite consisting of four scales: (1) Sensitivity/Responsivity, (2) Respect for Child Autonomy, (3) Positive Regard, and (4) Hostility (reverse-coded). For example, while completing a puzzle, a highly sensitive mother would encourage the child to complete the puzzle while providing praise and encouragement for effort. In contrast, a mother low on sensitivity would be quick to correct the child on errors, make negative comments about the child's effort, and fails to provide praise or support throughout the task. These interactions were coded on a 7-point scale from 1 (very low) to 7 (very high), with good interrater reliability (average intraclass correlation coefficient ranged from 0.79 to 0.93). We also rated cognitive and linguistic stimulation as predictors of oral language.

Longitudinal modeling at 3 and 4 years after implantation indicated that maternal sensitivity and cognitive stimulation predicted significant increases in the growth of oral language (Niparko et al. 2010; Quittner et al. 2013). Four years post-implantation, children of parents with higher maternal sensitivity had only a 1-year language delay compared with a 3-year delay in children of parents with low maternal sensitivity (see Fig. 12.2). Similar results were found for cognitive stimulation. Finally, linguistic stimulation was also related to improved language, but only in the context of

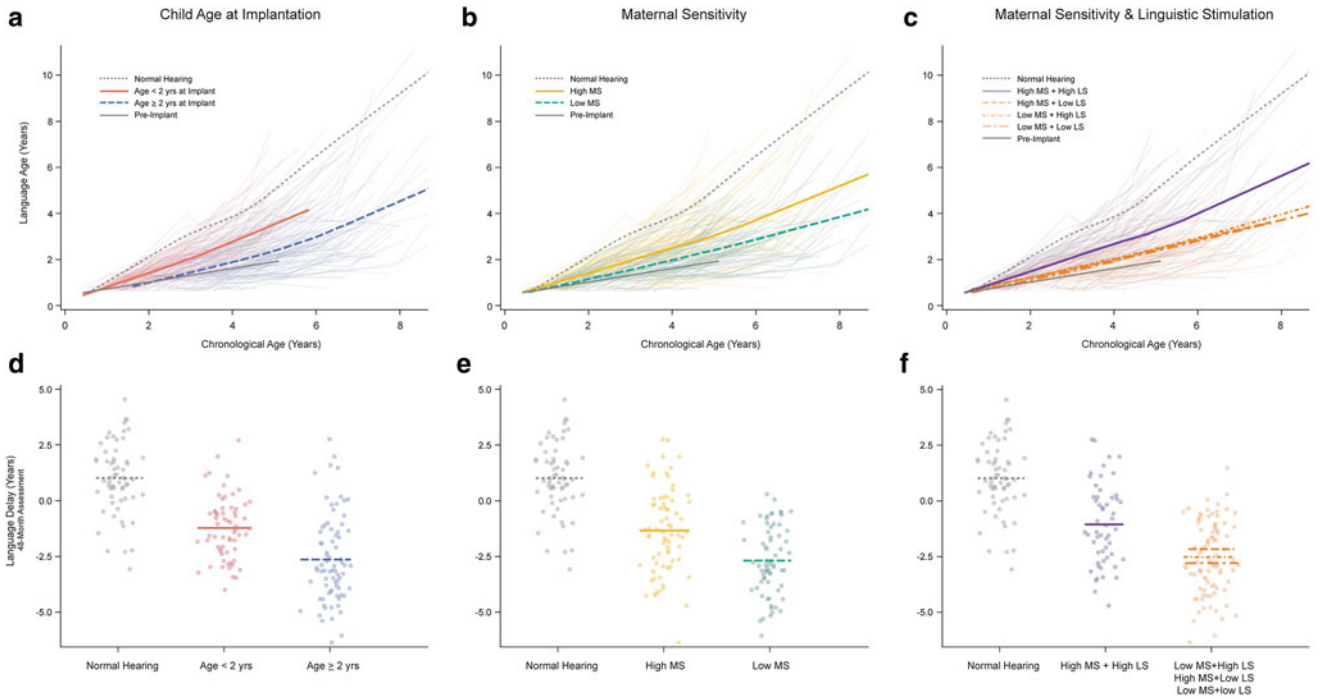

Fig. 12.2 Compares language development, measured by language age between (**a**) age at implantation, (**b**) high and low maternal sensitivity [MS], (**c**) the interaction between MS and Linguistic Stimulation [LS]. Compares normal hearing and cochlear implant users, (**d**) divided by age at implantation, (**e**) divided by high and low MS, (**f**) divided by the interactions between MS and LS, on language delay at 48 months postimplantation

highly sensitive parenting (Quittner et al. 2013). Importantly, our longitudinal analysis has indicated that the magnitude of the effects of maternal sensitivity on the growth of oral language emerged, over time, to be similar to those found for age at implantation, suggesting that parent behaviors are a critical target for intervention.

Facilitative Language Techniques

Specifically, the way parents' talk to their children has also been shown to affect oral language outcomes. Both quantitative linguistic input (e.g., number of different word types, mean length of utterance) and qualitative linguistic elements (e.g., open vs. closed-ended questions, labeling vs. expansion) have been associated with better receptive and expressive language development in children with cochlear implants (DesJardin and Eisenberg 2007; DesJardin 2009; Spencer 2004).

The first study evaluating facilitative language techniques (FLTs) was conducted in 32 children with cochlear implants, ages 2–7 years. Parent–child interactions were coded for use of FLTs during a Free Play and two storybook activities. The results suggested that the use of higher level FLTs (see Table 12.2) was positively associated with children's receptive and expressive language (DesJardin and Eisenberg 2007). In contrast, lower level techniques were negatively correlated with language outcomes. In an extension of this study, which examined FLTs in relation to children's phonological awareness and reading skills over 3 years, the authors found that higher level FLTs were significantly related to better awareness and reading ability.

A more recent study using the CDaCI study population measured FLTs in a Free Play and Art Gallery task in 93 deaf children implanted before age 2 (Cruz et al. 2013). During the unstructured Free Play task, the parent and child were directed to "play as you would at home." During the structured Art Gallery task, the parent–child dyad looked at and discussed a series of five art posters mounted on the walls of the playroom at different heights. The parent was instructed to discuss each picture with their child, while determining their favorite and least favorite. This task demands more communication between the parent and child. We examined both the frequency of FLTs, number of word types, and mean length of utterance by parents. Higher- vs. lower-level FLTs predicted growth in expressive language 3 years postimplantation. Number of different word types used by parents predicted growth in receptive language over time. In addition, parents increased the frequency with which they used higher level techniques from prior to implantation through 3 years of follow-up. Interestingly, the structured Art Gallery task elicited more and longer parental utterances

Table 12.2 Description and examples of FLTs

FLT	Description	Example
Lower level FLTs		
Linguistic mapping	Putting into words or interpreting the child's vocalization that is not recognizable as a word.	Child hands mother a toy cat and vocalizes—mother says, "kitty."
Comments	Statement or phrase that signals that a message has been received or an utterance to keep conversation going.	Mother says, "yeah!" or "thank you."
Imitation	Repeating verbatim the child's preceding vocalization without adding any new words.	Child says, "cup" and mother says, "yes cup."
Label	Stating the name for a toy, picture, or object.	Grandmother says, "There is a doggie."
Directive	Tells or directs child to do something.	Parent says, "Look!" or "You play with this cup."
Closed-ended question	Stating a question in which the child can only answer with a one-word response.	Father asks child, "Is that your favorite?" or "Do you like that picture?"
Higher level FLTs		
Parallel Talk	Parent talks aloud about what the child is directly doing, looking at, or referencing.	Child is looking directly at the picture of a bee and parent says, "The bumble-bee is flying over the flowers."
Open-ended question	Parent provides a phrase/question in which the child can answer using more than one word.	While looking at a picture, parent says, "What is happening in this the picture?"
Expansion	Parent repeats child's verbalization providing a more grammatical and complete language model without modifying the child's word order or intended meaning.	Child says, "baby cry" and the caregiver says, "The baby is crying."
Expatiation	Same as Expansion, but parent adds new information to the child's utterance.	While looking at the picture—child says, "swim water" and mother says, "Yes, we are going swimming at the beach. This summer we are going to the beach."
Recast	Caregiver restates the child's verbalization into a question format.	Child says, "puppy gone" and the caregiver says, "Is the puppy gone?"

and different word types than the unstructured Free Play task. Thus, this paradigm could easily be translated into clinical practice in a cochlear implant center.

Health-Related Quality of Life

Over the past two decades, tremendous progress has been made in defining and measuring health-related quality of life (HRQoL) and in recognizing its importance as a health outcome (Palermo et al. 2008; Quittner et al. 2009). More than 50 years ago, the World Health Organization proposed the first definition of HRQOL as "a state of complete physical, mental, and social well-being, and not merely the absence of disease or infirmity" (World Health Organization [WHO], 1947, p. 29). A consensus definition of HRQoL has now emerged, with agreement that it is multidimensional and includes four core domains: (1) disease state and physical symptoms, (2) functional status (e.g., performing daily activities), (3) emotional functioning, and (4) social functioning (Hays 2005; Rothman et al. 2007).

More recently, the Food and Drug Administration (FDA) has formally recognized the importance of patient-reported outcomes (PROs) and their relevance to the approval of new medications and treatments (FDA 2009). A PRO instrument is defined as any measure of a patient's health status that comes directly from the patient/parent proxy and assesses how a patient "feels or functions or survives with respect to his or her health condition" (p. 2 FDA Guidance). This may include observable behaviors or perceptions (e.g., ability to communicate, social functioning) or nonobservable outcomes known only to the child/proxy (e.g., anxiety). Seeking input from children and their parents represents a significant shift in health outcomes research and is particularly relevant for children with chronic conditions, in which a more collaborative, multidisciplinary model of care is required (Palermo et al. 2008; Quittner et al. 2008).

Severe-to-profound hearing loss is associated with measurable deficits in HRQoL, reflecting the broad effects of hearing loss and its concomitant effects on language learning, social and emotional functioning, and academic performance. Studies of cochlear implantation (CI) typically focus on clinical measures of efficacy related to communication (e.g., auditory skills, speech), which do not represent the device's effects on overall functioning. HRQoL measures provide a crucial assessment of the impact of CI's on everyday functioning (physical, emotional, social). To date, there are no CI-specific HRQoL measures for young children and their parents (Morretin et al. 2013).

A review of the literature on childhood deafness and cochlear implantation indicates that a majority of studies have used generic rather than condition-specific measures.

We found only one condition-specific HRQoL measure for school-age children using hearing aids, FM, or cochlear implants. This measure, the Hearing Environments and Reflection on Quality of Life (HEAR-QL; Umansky et al. 2011) was developed using existing literature and focus groups with children with mild-to-profound hearing losses either unilaterally or bilaterally and their parents. The developers did not use the process of instrument development advocated by the FDA and used an extremely heterogeneous sample of children in creating the items. Furthermore, they did not include the relevant domains of functioning that represent key HRQoL dimensions (e.g., physical functioning—balance problems; use of device—either hearing aids/CIs) (Morretin et al. 2013).

A recent study utilized parents' ratings of their children's developmental growth and HRQoL to compare the benefits of cochlear implantation in different age groups (Clark et al. 2012). Parents scored their child's general health, pain level, disposition, behavior, and ability to get along with others on a Visual Analog Scale (VAS). They also used the VAS-Development (VAS-D), adapted from the Infant/Toddler Child Health questionnaire (Landgraf 1994) to report on the child's expressive and communication skills, motor skills, and learning abilities. Parental ratings of children's development over the 4 years postimplantation showed improvement, especially for the youngest age group. These results were consistent with prior research which found greater rates of speech acquisition post-CI for children implanted prior to age 2 (Niparko et al. 2010).

The advantage of parental assessments over clinical objective measurements is that they take the child's everyday functioning and overall development, rather than their performance on speech and language measures at a single session. In addition, Clark et al. (2012) showed that, compared to parents of children implanted after 18 months, parents of children implanted before 18 months observed smaller developmental delays, and these results converged with measures of auditory skills and language. Although both groups made developmental gains over the 4 years, those children implanted later lagged behind. For children with longer periods of hearing loss, the use of hearing aids did not alleviate these deficits in development. This study highlighted the importance of early cochlear implantation.

Another metric of HRQoL is cost-utility ratings. Cost-utility assessments evaluate the costs associated with a medical intervention versus the gain in quality of life years given complete health. This ratio of monetary cost to quality-adjusted life years (i.e., QALY) is essentially a cost-effectiveness analysis of a healthcare intervention. QALY are the product of the years gained by the intervention and the quality of health defined on a scale of 0.0 (death) to 1.0 (perfect health). Younger age at implantation is associated with more gains in quality-adjusted life years (QALY). Semenov et al. showed that children receiving implants at <18 months gained, on average, 10.7 QALY. Cochlear implant recipients between 18 and 36 months gained 9.0 QALYs, and children older than 36 months gained 8.4 QALYs over their projected lifetimes (Semenov et al. 2013).

Although quantitative analyses have documented impressive benefits on QALYs, even more important is whether children with cochlear implants themselves, perceive this benefit. Using data from the CDaCI study, Meserole and colleagues (2014) showed that children 6 years postimplantation rated themselves as experiencing levels of HRQOL that were comparable to their hearing peers on the Child Health & Illness Profile-Child Edition (CHIP; Rebok et al. 2001) matched for child age, sex, maternal education, and family income. Children with CIs reported worse physical and emotional functioning, but comparable levels of well-being, self-esteem, and success in academics and peer relationships. In comparing parent reports of HRQOL for these 6- to 11-year-old children with CIs versus hearing peers, only academic achievement and social problem-solving were rated lower by the parents in the CI vs. hearing group (Meserole et al. 2014). In addition, higher parental stress, using the FSS, was associated with lower HRQoL scores. For every 1-point increase in parental stress, there was a 4-point decrease in Satisfaction and an 8-point decrease on the Global Score. Given these preliminary findings and its importance as a health outcome variable, a condition-specific HRQoL instrument is urgently needed.

Conclusions and Recommendations for Early Intervention

Numerous studies have documented the effects of childhood hearing loss on broad areas of development, including cognitive, behavioral, social, and daily functioning. However, recent advances in diagnosis and earlier use of cochlear implant technology have greatly improved the long-term developmental outcomes of this population. In general, implanting children before 18 months of age has greatly facilitated their development of oral language, attention, and behavioral regulation. Further, parental behaviors are now shown to exert a measurably strong influence on the progress children made in these areas. Both maternal sensitivity and the use of facilitative language techniques (FLTs) were significantly related to oral language skills after implantation and thus, should serve as critical targets for early intervention.

Our programmatic, longitudinal studies on the CDaCI cohort indicate that early implantation provides a unique opportunity for "catch-up" growth; however, in comparison to hearing peers, children with CIs continue to face challenges in domains such as joint engagement, internalizing

and externalizing behaviors, and social functioning (Cejas et al. 2014; Barker et al. 2009). Furthermore, parents of children with hearing loss continue to report high levels of stress in relation to their parenting role in areas such as communication, finances, safety, and education. This chapter has highlighted both areas of great improvement and areas in need of further remediation. To move the field forward and maximize the gains children with CI's can achieve, a more family-centered approach to intervention is needed.

The first step toward creating this type of approach is to proactively screen young, deaf children for delays in cognition, behavioral development, social–emotional functioning, and health-related quality of life. Currently, the focus of most pediatric implant teams is heavily weighted toward assessment of audiological functioning and language skills. In contrast, a family-centered approach would include evaluation of parental expectations for the implant, parenting stress, and key developmental milestones related to attention and behavioral regulation. This would assist in identifying children with comorbid diagnoses, such as attention-deficit disorder or autism, to provide appropriate treatment (Cruz et al. 2012). Additional expertise may be needed from other specialists, including psychologists, social workers, and developmental pediatricians. Based on the results of this broad-based screening, recommendations might include parent training in behavioral management, coaching of Facilitative Language Techniques (FLTs), and social skills training.

Future directions for research include development and testing of a parenting intervention for young, CI recipients and their families, focusing on increasing parental sensitivity, use of FLTs, and appropriate behavior management strategies. Finally, there is currently no deaf-specific HRQoL measure for children receiving cochlear implants. This is an important direction given that this type of instrument also serves as a broad screener of daily functioning. It could also be used to measure the efficacy of cochlear implants and early intervention programs.

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Part IV

Cochlear Implant Outcomes in Special Populations

Redefining Cochlear Implant Benefits to Appropriately Include Children with Additional Disabilities

13

Nancy M. Young, Constance Weil, and Elizabeth Tournis

May their vulnerability open our hearts so that they can find care and we can find compassion.

- Daniel Gottlieb, PhD, *Letters to Sam*

Abbreviations

AAC	Alternative augmentative communication
ADHD	Attention-deficit hyperactivity disorder
ASD	Autism spectrum disorder
CI(s)	Cochlear implant(s)
CMV	Cytomegalovirus
CP	Cerebral palsy
DQ	Developmental quotient
IQ	Intelligence quotient
LD	Learning disability
SNHL	Sensorineural hearing loss

Historically, most deaf children who received a cochlear implant (CI) have been those without other known co-occurring disabilities. However, large portions of the pool of children who meet audiological candidacy criteria for an implant have co-occurring conditions likely to impact their progress. In light of the emphasis on early identification and management of hearing loss and the importance of early intervention, increasing numbers of children are being evaluated for CIs at an age when it may not be possible for comorbid conditions to be definitively diagnosed or the severity of permanent disability to be known. The co-occurrence of complicating conditions with hearing loss, either sensorineural hearing loss (SNHL) or auditory neuropathy spectrum disorder, is not surprising. Many etiologies of hearing loss, in particular prematurity and congenital infection, may impact the developing brain, thereby increasing the likelihood of additional neurodevelopmental disabilities. A growing number of clinicians, including this chapter's authors, believe that these children can benefit from CIs. However, broader thinking about the benefits of implantation and what defines success, as well as the development of outcome measures beyond traditional measures of word recognition and spoken language, are necessary.

Although CIs have been available for children for nearly 30 years, only recently have children with co-occurring disabilities been considered appropriate candidates by some CI programs. The reason for lack of application to this population is in part explained by the early history of CI. The first CI system was developed by William House, MD, at a time when the CNS was viewed by most of the scientific community as lacking the neuroplasticity necessary for neurostimulation to be useful. In addition, the initial implant design consisting of a single intra-cochlear electrode was viewed with extreme skepticism. The initial controversy and lack of acceptance of CIs by the scientific community as well as deaf culturists (Fig. 13.1) influenced research in the field, pushing it toward demonstration of efficacy as defined by spoken language ability and mainstream placement in school. These studies typically excluded children with co-occurring disabilities to both enhance study population homogeneity and demonstrate that this level of efficacy could be achieved. The controversy had a

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Fig. 13.1 Anti-cochlear implant buttons purchased from deaf protestors during the 1994 International Cochlear Implant Speech and Hearing Symposium held in Melbourne, Australia

similar influence on clinical practice. Most implant programs did not view children unlikely to achieve spoken language as appropriate cochlear implant candidates.

Predicting the outcomes for children with cochlear implants (CIs) is a complex task, one that is considerably more challenging in children with additional disabilities. Such children are estimated to account for 40 % of children with hearing loss (Gallaudet Research Institute 2003). This estimate is consistent with reports from pediatric cochlear implant centers who do not exclude children with multiple disabilities (Wiley et al. 2005; Birman et al. 2012).

A broader definition of candidacy and successful outcomes in children has evolved over time as implant programs gained experience, technology advanced, and CIs became accepted as a standard treatment for childhood hearing loss. However, published literature on outcomes and clinical care of children with multiple disabilities, especially those with cognitive impairment, is sparse. There is no uniform consensus among CI programs that these children are candidates for cochlear implantation, but it is not in keeping with current thinking about improving the lives of individuals with disabilities to exclude such children from an accepted treatment for deafness.

Disabilities That Co-occur with Hearing Loss

Conditions that co-occur with hearing loss include learning disability (LD), intellectual (cognitive) disability, global developmental delay (diagnosis used for children under 5 years of age), language and communication disorders such as autism spectrum disorder (ASD), disorders of attention, blindness and low vision (defined as vision that cannot be corrected), cerebral palsy (CP), and emotional disturbance, as well as other medical, sensory, and motor problems (Gallaudet Research Institute 2003; Roush et al. 2004; Edwards 2007). The prevalence of most of these co-occurring disabilities in children with hearing loss is often beyond that in the general population (Table 13.1). For example, the general population prevalence of intellectual disability is approximately 1 %, with a 6 in 1000 prevalence for severe intellectual disability according to the Diagnostic and Statistical Manual of Mental Disorders, 5th Edition (DSM-5) (Edwards et al. 2006). Table 13.1 shows that the prevalence of intellectual disability in deaf children is 9.8 %. Increased prevalence is likely related to a combination of the impact of hearing loss on development as well as the presence of underlying etiologies of hearing loss that directly affect the developing brain.

Table 13.1 Prevalence of additional disabilities co-occurring with deafness (Gallaudet Research Institute 2003)

Additional disability	% of deaf children
No additional disability	60.1
Learning disability	10.7
Intellectual disability	9.8
Attention-deficit disorder	6.6
Blindness and low vision	3.9
Cerebral palsy	3.4
Emotional disturbance	1.7
Other conditions	12.1

Intellectual Disability

There has been much hesitancy to implant children diagnosed with or at significant risk for intellectual disability because of its association with reduced language abilities (Edwards et al. 2006). The DSM-5 defines intellectual disability as an intelligence quotient (IQ) of 70 or below, with concurrent deficits in adaptive behavior (American Psychiatric Association 2013). Individuals with low IQ who have developed adaptive behaviors that permit them to function independently are not considered to have intellectual disability. Adaptive behavior refers to daily living skills, including social and community engagement, and the ability to communicate one's needs, care for one's health and safety, manage one's home, self-care, and participate in leisure activities. Individuals with intellectual disability are characterized as learning and processing information more slowly than those without intellectual disability, having difficulty with abstract concepts such as money and time, and often having difficulty understanding the subtleties of social interactions.

Approximately 85 % of people with intellectual disabilities have "mild" intellectual disability. Many within this group can achieve some academic success, they usually meet elementary academic levels or beyond given sufficient support, and they are mostly self-sufficient. They can often live independently within their communities with a minimal level of additional supports such as assistance with life decisions. Additional time, instructions, and reminders may be needed for other life skills such as finances, nutrition, shopping, and transportation.

Individuals with "moderate" intellectual disability, about 10 % of those with intellectual disability, have adequate communication skills but complexity is more limited. Social cues, social judgment, and social decisions (particularly romantic decisions) regularly need support. Most self-care activities can be performed but may require extended instruction and support. Independent employment can be achieved in positions that require limited conceptual or social skills. However, additional supports may be required. Independent living may be achieved with moderate supports such as those available in group homes.

"Severe" intellectual disability describes 3–4 % of this population. Communication skills are very basic. Self-care activities require daily assistance. Many individuals in this category will require safety supervision and supportive assistance. Residence in supported housing is usually necessary. The final 1–2 % of those with intellectual disability has "profound" disability. They are dependent upon others for all aspects of daily care, usually requiring 24-h care and support. They have limited communication skills and usually have co-occurring sensory or physical limitation.

For young children with cognitive delays, intellectual disability is not used as a diagnosis as developmental progress in very young children can fluctuate for a variety of reasons. Therefore, instead of IQ, developmental assessments of children between birth and age 6 years provide a development quotient (DQ). DQ is determined by evaluating and placing a numerical value on how a child functions in multiple domains (cognitive, receptive, and expressive language, fine and gross motor functioning) in comparison to their typically developing peers of the same age. DQ can provide a useful baseline and a means to track progress and identify emerging problems that need to be addressed.

Since intellectual disability is not diagnosed in children less than 6 years of age, children with DQs below a certain level are considered developmentally delayed. However, measures used to determine DQ do correlate by varying degrees with IQ and intellectual disability (Baker et al. 1983; Ramsden et al. 2011; Deary et al. 2013). Thus, DQ is useful in identifying children at risk for permanent disability.

Children with developmental delay may have a diminished capacity for learning, reasoning, and understanding as well as diminished ability to function and cope within the environment. The term "developmental" is employed to convey that these deficits may improve, especially if intervention is provided. For example, infants with undiagnosed hearing loss of significance who are otherwise developing typically would initially be expected to have normal DQ. However, between 9 and 12 months, when early patterns of babbling and use of phonemes normally become more purposeful and responsive to others, their DQ will typically decline. The change in DQ provides a warning that an investigation including hearing evaluation is needed. Similarly, between 18 and 24 months of age, DQ of children with undiagnosed or inadequately managed hearing loss will decline if they lack the ability to perceive and understand language needed to follow directions. The decline in their DQ does not mean that they are destined to have lower IQ or intellectual disability.

Measuring DQ in deaf children who are typically language impoverished often requires a different approach that employs primarily nonverbal measures. DQ is impacted not only by hearing loss but also by other conditions such as visual and motoric problems. These latter conditions may render the use of nonverbal tests particularly challenging. Therefore DQ results may underestimate a child's cognitive potential, especially if comorbid conditions are present. Early intervention offers the opportunity to improve outcomes for some children by taking advantage of the developing brain's neuroplasticity.

Some etiologies of hearing loss are associated with an increased incidence of developmental delay and eventual diagnosis of intellectual disability. For example, Down syndrome is the most common chromosomal cause of mild-to-moderate intellectual disabilities, and children with Down syndrome have a higher prevalence of sensorineural hearing

loss in comparison to the general population (Park et al. 2012; Austeng et al. 2013).

Prenatal infections are also an important cause of sensorineural hearing loss and intellectual disability. A prime example is congenital cytomegalovirus (CMV), the leading cause of congenital infections worldwide, and the most common non-genetic cause of hearing loss (Manicklal et al. 2013). It is estimated to cause one-third of childhood SNHL. Symptoms at birth of congenital CMV infection include enlarged liver and spleen, microcephaly, seizures, retinitis, and intracranial calcifications. SNHL is the most common symptom of CMV among the 10–15 % of children with symptoms at birth (Dollard et al. 2007). The occurrence of other disabilities, including intellectual disability and visual impairment, is more frequent and severe in the symptomatic group than in those with asymptomatic infection who later develop sequelae. Normal cognition has been reported in one-third of children with symptomatic CMV at birth and the presence of microcephaly may be the most specific predictor of poor cognitive outcome (Noyola et al. 2001).

Postnatal bacterial meningitis remains an important cause of acquired deafness and is associated with cognitive impairment and learning disability in young children. Although its incidence has declined in the USA and other countries that have early childhood vaccination programs for *H. Influenzae* and pneumococcal disease, early identification of hearing loss and urgent implant evaluation are necessary because of labyrinthine ossification that may preclude optimal electrode insertion (Young and Tan 2010). Other conditions such as significant prematurity, low birth weight, anoxia, and hyperbilirubinemia are also risk factors for both hearing loss (auditory neuropathy spectrum disorder and SNHL) and intellectual disability.

In sum, there is a wide range of disabilities for which intellectual disability may co-occur, and the diagnosis of intellectual disability itself covers a wide range of functioning in both cognitive and adaptive realms. Increased ability to hear may have a measurable impact in a number of areas for this population. However, these types of improvements are usually not systematically assessed by cochlear implant programs. Therefore the range of benefits of cochlear implantation may not be recognized or appreciated by professionals focused primarily upon auditory and spoken language skills.

Visual Impairment

Children with congenital deafness and significant visual impairment are at high risk for developmental delay, learning problems, and intellectual impairment. Although dual-sensory deficits alone may increase the risk of learning and cognitive problems, the etiologies that result in both visual and hearing impairments are often ones that cause brain damage and thus many additional developmental problems. Congenital disorders that cause vision problems often impact the early stages of brain development, which results in more than an isolated visual problem.

Deafness associated with low vision or blindness also often arises because of insults including prenatal infection due to CMV and extreme prematurity (defined as age of less than 25 weeks at birth) which are known to affect overall brain development. Children with cortical visual impairment, a common neurological disorder in preterm infants, have bilateral visual impairment due to non-ocular brain disease (Good et al. 2001). This condition may not be diagnosed early in life, and it rarely occurs in isolation. The most common cause of cortical visual impairment is neonatal hypoxic-ischemic injury. In fact, at least 60 % of children with neonatal injury of this type have cortical visual impairment (Good et al. 2001). Its presence should be suspected in children in whom MRI of the brain detects periventricular leukomalacia, a characteristic type of brain injury that may be seen in preterm infants. Other causes of cortical visual impairment include head injury, bacterial meningitis, encephalitis, congenital toxoplasmosis, and complications of cardiac arrest and open heart surgery, all of which are also risk factors for childhood SNHL. Cortical visual impairment may improve, but most children will not regain normal vision (Huo et al. 1999).

Children with CHARGE syndrome are a significant subgroup of the visually and hearing impaired that has complicating conditions including cognitive, behavioral, and motor abnormalities. A very different group of children are those with Usher syndrome (US) type 1, the most common cause of deaf-blindness in the USA. In this disorder, progressive visual impairment due to retinitis pigmentosa begins later in childhood, subsequent to critical stages of early brain development.

Regardless of etiology and presence of complicating conditions, including impaired cognition, there are obvious advantages for deaf children with low vision or blindness not to be reliant upon visual or tactile communication methods alone and to have hearing to connect them to other people and to their environment.

Motor Impairment

Motoric deficits may impact cochlear implant candidates in a variety of ways. For some children, their degree of motor impairment means that sign language will not be an effective means of expressive communication. For others, motor difficulties may be limited to oral motor dysfunction and its impact on swallowing, spoken language, and articulation. Children in whom expressive communication via sign or

spoken language is limited by motor dysfunction may benefit from augmentative communication devices (Davis et al. 2010; Lee et al. 2013).

Cerebral palsy gives rise to a spectrum of neurologically based movement and posture disorders arising from nonprogressive damage to the developing brain. Its prevalence is estimated at 3.6 cases per 1000 (Yeargin-Allsopp et al. 2008). The types and severity of motor abnormalities vary, as does the cause of CP. Prenatal causes including brain malformations, genetic abnormalities, and intrauterine infections are estimated to account for 44 % of cases. Perinatal asphyxia and complications at delivery may also result in CP. About 25 % of cases are of unknown etiology. Many children with CP have additional findings which include epilepsy, cognitive impairment, and visual impairment. Sensorineural hearing loss of varying degrees is reported to occur in 12–25 % of children with CP (Ashwal et al. 2004; Odding et al. 2006). Although the injuries underlying CP are nonprogressive, the child's functional ability may vary over time (Rosenbaum et al. 2002; Shevell et al. 2009). There are three main forms of CP—spastic, dyskinetic, and ataxic. Spastic CP results in stiffness and difficulty moving, dyskinetic is characterized by excessive uncontrolled involuntary movements, and ataxic by poor balance, lack of proprioception, and poor coordination. In the most severe cases, motor function may be so limited that the child is "locked in." For some of these children, communication of a reliable positive versus negative response may be very difficult to obtain, making evaluation of learning and cognitive potential as well as auditory responsiveness pre- and post-implant very challenging.

Autism Spectrum Disorder

Autism spectrum disorder (ASD) is characterized by early onset and persistent deficits in social communication and social interaction and restricted, repetitive patterns of behavior, interests, or activities. By definition, these symptoms cause clinically significant impairment in social, occupational, or other important areas of functioning. The term "spectrum" refers to the wide range of symptoms, skills, and impairment that children with ASD may exhibit. Some children are mildly impaired while others are severely disabled. The prevalence of SNHL has been reported to be higher than that seen in the general population, with profound loss having a significantly higher likelihood of being comorbid with ASD than lesser degrees of loss (Rosenhall et al. 1999; Szymanski et al. 2012).

Most children with ASD are not diagnosed until after age 4 years, although it is possible to reliably diagnose this condition in children as young as 2 years (Lord et al. 2006; Kleinman et al. 2008). Therefore, diagnosis of ASD subsequent to CI would be expected in many children with profound hearing loss. For this reason, co-occurrence of ASD in implanted children who do not make expected progress should be considered.

Disorders of Learning

Learning disorders are the most common co-occurring conditions with hearing loss (Table 13.1). They can be caused by the brain's inability to receive, process, store, respond to, or communicate information. The DSM–5 defines a learning disorder as a "neurodevelopmental disorder of biological origin manifested in learning difficulty and problems in acquiring academic skills markedly below age level and manifested in the early school years, lasting for at least 6 months; not attributed to intellectual disabilities, developmental disorders, or neurological or motor disorders." LDs are a group of disorders that may affect development of spoken language, literacy, and/or mathematical ability. They are not usually identified until a child is of school age and is not making appropriate progress. The presence of LD is not reflective of IQ, and the causes of learning disabilities are not fully understood. Multiple factors that impact brain structure and development have been implicated, including hereditary causes, exposure to toxins such as alcohol and lead, and poor nutrition.

One of the best predictors of growth in children's vocabulary early in life is the number of words heard per unit of time from their mothers (Huttenlocher et al. 1991, 2010; Hart and Risley 2003). In regard to reading disabilities in normal-hearing children, many have a deficit in phonological processing skills, skills important in word recognition (Torgesen et al. 1994; Shaywitz 1996). With these studies in mind, it is not surprising that auditory deprivation secondary to congenital hearing loss alone is likely to limit consistent exposure to spoken language, thereby increasing the risk of impaired language learning and literacy. When hearing loss co-occurs in children with learning disorders, learning problems may be compounded.

As most children are implanted prior to school age, the presence of LDs is not usually a consideration in CI candidacy. However, LDs may impede post-implant auditory skill development and language acquisition as well as academic progress (Isaacson et al. 1996). Appropriate diagnosis and academic support are important. In the authors' experience, children with learning disabilities and a cochlear implant, including those with good word recognition ability and oral communication, are often transferred from the mainstream to a total communication program without first providing them with the support for their specific LD that a hearing child would typically receive. Although visual support of learning through sign language may be the best approach for some children, thoughtful educational management based upon understanding the child's individualized needs is preferred and in keeping with the concept of inclusion.

Attention Disorders

Disorders of attention, including attention-deficit hyperactivity disorder (ADHD) inattentive type, hyperactive type, and combined type, are common brain disorders in children and adolescents. They usually become evident in preschool or early elementary school. Symptoms include inattention, distractibility, impulsivity, and hyperactivity. Although these behaviors are normal for all children at times, for children with ADHD these behaviors are more severe and occur more often. To be diagnosed with an attention disorder according to DSM-5, a child must have symptoms for at least 6 months and to a degree greater than other children of the same age, with symptoms present before age 7 (American Psychiatric Association 2013). These disorders are often associated with poor academic performance, behavioral issues, and social problems. Co-occurring conditions may include LDs, anxiety/depression, oppositional defiant conduct, and bipolar disorders. The exact cause of ADHD is unknown, although genetics are thought to play a significant role. Environmental factors such as lead exposure, prenatal drug exposure, and extreme prematurity and low birth weight are also risk factors. A wide range of prevalence rates of ADHD have been reported in children with hearing loss, some considerably higher than the 6.6 % shown in Table 13.1. Because many of the symptoms overlap with those seen in children with hearing loss alone, accurate diagnosis may be challenging, especially in children who are language impoverished. The median age at diagnosis of ADHD is 7 years (Kessler et al. 2005). Therefore, most CI children will be diagnosed with attention disorders after implantation. Appropriate referral for careful evaluation, diagnosis, and management is important to optimize the academic progress of children in whom hearing loss and attention disorders co-occur. Proper management of ADHD can also improve the chances that the child will make progress in auditory skills and language acquisition with a cochlear implant (Pundir et al. 2007).

Reported Cochlear Implant Outcomes in Children with Additional Disabilities

Study of children with additional conditions is complicated by differing opinions about what qualifies as an additional disability as well as what defines each type of disability. Further complicating matters is that these conditions range in severity. Some conditions such as LD, intellectual disability, and ADHD cannot be diagnosed in very young children and may be more challenging to diagnose in children with limited language skills. Compounding of one disability by another is an important consideration. Improved hearing through a CI may provide these children with means of interacting with others and the physical environment, as well as enabling them to develop language learning and reasoning that might not otherwise be achievable in the face of co-occurring disabilities.

There have been many reports in the literature, primarily retrospective, describing performance by children with additional disabilities who did receive a cochlear implant, indicating that some programs have not ruled out candidacy based on presence of multiple disabilities. These reports vary widely in both number of subjects and the nature and severity of the additional disabilities.

A few authors focused specifically on the prevalence of additional disabilities among children receiving implants in their programs. Birman et al. reported that of 88 children implanted in a 12-month period, 33 % had additional disabilities (Birman et al. 2012). They noted that additional disabilities were most often found in children with syndromes and chromosomal abnormalities, jaundice, prematurity, CMV, and inner ear abnormalities. Wiley et al. retrospectively investigated the presence of additional disabilities among children with and without GJB2 mutations who received a cochlear implant (Wiley et al. 2006). Of 46 children evaluated, 16 had GJB2 mutations, 12 were negative for these mutations, and 17 were untested but had no other identifiable risk factor or etiology for hearing loss. In the GJB2-positive group, 44 % had additional disabilities as did 33 % in the GJB2-negative group and 41 % who did not receive GJB2 testing. In one study, 38 % of the program's 175 implanted children had additional disabilities, with 58 % of those having just one complicating condition, and 22 %, 9 %, 7.5 %, and 3 % having 2, 3, 4, and 5 additional disabilities, respectively (Nikolopoulos et al. 2008). Other reports in the literature show the specific additional disability group being studied to be anywhere from 19 to 34 % of the investigators' CI populations (Edwards et al. 2006; Holt and Kirk 2005; Berrettini et al. 2008; Oghalai et al. 2012). However, few of the reports reviewed indicate the degree to which the programs have accepted children with additional disabilities or their criteria for acceptance.

Studies of efficacy of CIs in children with additional disabilities have used a wide variety of measures to evaluate outcomes such as auditory skills, speech intelligibility, language development, communication, psychological benefits, and parent perception of benefit. Methods include traditional measures of speech perception, speech intelligibility, and language development as well as parent questionnaires, structured parent interviews, and video assessment. Some studies included children with a wide range of additional conditions while some focused on specific disabilities such as developmental disabilities, mild cognitive delay with no other disabilities, mild or moderate mental retardation, and deaf-blindness. Table 13.2 provides a listing of relevant studies, showing what additional disabilities the subjects had, the number of subjects, and the skills assessed. Some of the findings are summarized in the following sections.

Table 13.2 Reported studies of cochlear implant outcomes in children with additional disabilities

Author	Additional disabilities	*n*	Skills assessed
Isaacson et al. (1996)	Learning	5	Speech perception, environmental sound recognition
Saeed et al. (1998)	Vision	2	Speech perception
Hamzavi et al. (2000)	Learning, cognitive, sensory-motor integration, attention, motor	10	Speech perception
Waltzman et al. (2000)	Motor, language, developmental, learning, cognition, autism, attention, vision, cerebral palsy, sensory integration	29	Speech perception
Bauer et al. (2002)	CHARGE syndrome	5	Speech perception
Loundon et al. (2003)	Usher syndrome	13	Speech perception, speech production
Donaldson et al. (2004)	Autism spectrum disorder	6	Speech perception, expressive and receptive language, parent questionnaire: quality of life
Ramirez Inscoe et al. (2004)	CMV (autism spectrum disorder and other unspecified disabilities)	16	Speech perception, speech production
Holt and Kirk (2005)	Mild cognitive delay	19	Speech perception, receptive and expressive language
Lee et al. (2005)	Congenital CMV (vision, motor, cognitive)	6	Speech perception
Wiley et al. (2005)	Cerebral palsy, motor, learning, cognitive, language, vision	16	Parent structured interview: communication skills
Edwards et al. (2006)	Developmental delay	11	Speech intelligibility, speech perception
Berrettini et al. (2008)	Cognitive, cerebral palsy, autism spectrum, attention, language and learning, epilepsy	23	Speech perception, parent questionnaire: communication, quality of life
Dammeyer (2009)	Visual	5	Video observation: communication, attention, emotional response
Nikolopoulos et al. (2008)	Autism spectrum, behavioral, cognitive, language/communication, orofacial, physical, visual, vocal tract	67	Speech intelligibility
Wiley et al. (2008)	Motor, vision, cognitive, autism spectrum, developmental delay	14	Auditory skills
Lee et al. (2010)	Cognitive	15	Speech perception, speech intelligibility, language development
Meinzen-Derr et al. (2010)	CHARGE, cognitive, motor, vision, autism, cerebral palsy	20	Receptive and expressive language
Southwell et al. (2010)	CHARGE syndrome	3	Case reviews: speech perception
Steven et al. (2011)	Cerebral palsy, cognitive	36	Speech perception
Birman et al. (2012)	Developmental delay, cerebral palsy, visual, autism, attention	29	Auditory performance
Oghalai et al. (2012)	Developmental delay	12	Cognition, adaptive behavior, familial stress, communication
Wiley et al. (2013)	Visual (deaf-blind)	91	Developmental age, receptive and expressive language
Ahn and Lee (2013)	CHARGE syndrome	6	Speech perception, speech intelligibility
Byun et al. (2013)	Cerebral palsy	8	Speech perception, speech production, receptive and expressive language
Jatana et al. (2013)	Usher syndrome	26	Auditory performance, communication method

n = number of children with a CI and additional disability studied. Some reports also included children without additional disabilities, control groups, adults, or pre-implant studies on larger numbers of children. These subjects are not included in the "n" shown

Hearing

In the Birman et al. study, auditory performance, as measured by the Categories of Auditory Performance score at 12 months, was significantly lower in those with compared to those without additional disabilities (Birman et al. 2012). However, most studies do report improvement in speech perception performance post-implantation in many children with additional disabilities, even though the rate of improvement may be slower than for typically developing deaf children with an implant (Isaacson et al. 1996; Holt and Kirk 2005; Saeed et al. 1998; Hamzavi et al. 2000; Waltzman et al. 2000; Ramirez Inscoe and Nikolopoulos 2004; Lee et al. 2005, 2010; Wiley et al. 2008). For example, Waltzman

et al. found that children with a wide range of additional conditions likely to affect their progress and a minimum of 1 year of implant experience demonstrated significant improvement post-implant in the ability to perceive phonemes, words, and sentences using audition alone, although their rate of skill growth was slower than for children without co-occurring conditions (Waltzman et al. 2000). Wiley et al. also reported on children with a variety of additional disabilities, finding progress post-implantation in their auditory skills, with rate of change similar to those without additional disabilities (Wiley et al. 2008). However, those with additional conditions started at a lower level and, therefore, remained at a lower level when compared at similar follow-up times to the typical CI children. Holt and Kirk found that although children with mild cognitive delays had significantly lower scores than typically developing children with a CI, they all showed improvement in their speech reception and language over time on all of the traditional measures of speech and language development (Holt and Kirk 2005).

Several small retrospective series describe a range of speech perception and spoken language skills post-CI in children with CHARGE syndrome (Bauer et al. 2002; Lanson et al. 2007; Southwell et al. 2010; Ahn and Lee 2013). The majority of children were reported to have improvement in awareness of environmental sounds, and, in some cases, improved speech perception was measurable. Improved sound awareness was reported to improve quality of life, although this outcome was not formally evaluated. Young, Tournis, and Crimmins found that one child of their six with CHARGE syndrome developed measurable open-set speech discrimination after 3 years of CI use while two others showed closed-set discrimination abilities after longer periods of device use than typically developing children with a CI (unpublished data, 2012). Most authors commented upon lack of appropriate tools to measure the positive impact of a CI in this population. Development of spoken language as the primary mode of communication was the exception and occurred primarily in children in whom there was no evidence of intellectual disability.

To date, the literature on outcomes of CI children with CP is sparse and retrospective. A report of eight children implanted prior to age 3 years found that four children with less severe CP and better cognitive ability performed as well as age-matched children without co-occurring disabilities on standard measures of auditory skills and spoken language. The four with severe CP also had evidence of poorer cognition and performed poorly on these same measures (Byun et al. 2013). Another study categorized 36 children with CP by degree of physical impairment and by cognitive ability (Steven et al. 2011). Higher cognitive function was the most important prognostic indicator of speech reception category achieved. The authors did note that cognition was challenging to measure in this population and for this reason it was not done in the six children under age 2 years. They noted that 34 of 36 children are full-time CI users and commented that the impact of improved hearing on quality of life should not be underestimated.

Based on their findings, Hamzavi et al. concluded that the majority of the children they studied, who had disabilities including hemiparesis, sensory integration disturbance, and intellectual deficits, were successful implant users because of the benefits of hearing from a safety perspective as well as the psychosocial benefits perceived by the families (Hamzavi et al. 2000).

Speech Production

Several studies assessed speech intelligibility in children with multiple disabilities who received a CI. Nikolopoulos et al. found that 30 % of those with additional disabilities developed no intelligible speech compared to a rate of only 3.7 % in those without additional diagnoses or suspected disabilities (Nikolopoulos et al. 2008). The number of additional disorders was the factor most related to speech intelligibility, with language and communication disorders the most important individually contributing conditions. On the other hand, 70 % of the multiply disabled children did develop at least some intelligible speech. Another study suggested that CI outcomes in children with mild mental retardation, including speech perception, speech intelligibility, and oral language, were equivalent to those of children without an additional disability (Lee et al. 2010). The children with moderate mental retardation were implanted at an older age and had lower performance post-implant. The authors suggest that earlier implantation for children with moderate mental retardation might be beneficial.

Language/Communication Skills

Studies of language development and communication skills generally show that children with multiple disabilities are able to improve their skills over time with implant use (Wiley et al. 2005; Holt and Kirk 2005; Berrettini et al. 2008; Dammeyer 2009; Meinzen-Derr et al. 2010). "Communication skills," versus language and speech intelligibility per se, were often evaluated using parent questionnaires, video observation, or other tools not typically employed in standard CI studies (Wiley et al. 2005; Oghalai et al. 2012; Dammeyer 2009; Palmieri et al. 2012). Wiley et al. used a structured interview of parent-perceived CI benefit adapted to address special considerations within the multi-handicapped population (Wiley et al. 2005). All parents reported progress in their child's communication post-implant and indicated that the children wore their devices

consistently and exhibited improved awareness of environmental sounds and interest in their environment. The study by Palmieri et al. also used a questionnaire to assess global benefits of CI in children with additional disabilities (Palmieri et al. 2012). This tool, the Deafness and Additional Disabilities questionnaire, was designed to measure changes in behavior during everyday activities that require specific neuropsychological and perceptual skills. Improvement occurred in the majority of children, encompassing a wide spectrum of abilities including cognitive, social, relational, hearing, and linguistic skills needed for everyday activities.

Dammeyer used video observation of communication in deaf-blind subjects with and without their CIs and parent interviews to assess perceived benefits. Improvements in communication included attention, emotional response, and overall quality of communication when the CI was in use (Dammeyer 2009). Parents perceived their child's attention and communication as the most important CI benefits.

Oghalai et al. evaluated intelligence and adaptive behavior in children with developmental delay, comparing changes after implantation to a group of children implanted without developmental delay (Oghalai et al. 2012). They found that although children with developmental delay started out with a lower level of adaptive behavior than the group without developmental delay, after CI they improved at the same rate. However, they continued to have a lower rate of intellectual development than those without developmental delay. Because the delayed children were implanted at an older age, and age at implant was significantly related to rate of change in cognitive development, the authors speculate that such children might have improved cognitive development if implanted earlier. They consider appropriate parental expectations of CI in a child with developmental delay to be "improved sound awareness and slightly better interactions with their surrounding environment but not necessarily the development of oral communication skills."

One of the few studies to evaluate outcomes in autistic spectrum disorder reported on seven children, three diagnosed before and four subsequent to implantation (Donaldson et al. 2004). The mean age at CI was 4.7 years and average implant use was 25 months. Two children had measurable speech perception. Communication was typically multimodal: In addition to natural gestures used by six children, one used spoken words, four used word approximations, and three used sign language (two in conjunction with word approximations). All parents reported worse than expected performance if their child was diagnosed after implantation in comparison to a more positive view of performance if implanted subsequent to diagnosis of ASD. Five of six parents reported that they would recommend a CI to another family with a similar child.

Wiley et al. evaluated 91 primarily deaf-blind children after CI; 27.5 % of the children had CHARGE syndrome (Wiley et al. 2013). With the implant, only 22 % could follow directions related to functional use of objects and only 12 % could communicate with complex sentences using spoken language. Developmental ability, not age at implant, was the largest contributor to the language outcomes using oral communication. The authors pointed out that current measures used with implanted children do not assess the impact on quality of life that may occur in individuals with significant dual-sensory deficits. The effectiveness of cochlear implantation in children with Usher syndrome has been documented in several studies (Loundon et al. 2003; Jatana et al. 2013). Jatana et al. reported that 3.7 % of their very large series of implanted children had US (Jatana et al. 2013). The majority of these children developed significant open-set speech perception and oral communication skills, with 69.2 % using oral or primarily oral communication by the time of last follow-up.

Assessment of Cognitive, Emotional, Behavioral, and Adaptive Functioning

Early cochlear implant research was focused on demonstrating effectiveness of this auditory prosthesis particularly for development of speech perception and spoken language. More than two decades later, progress after implantation continues to be determined primarily by measures of word and sentence recognition, language level, and speech intelligibility. Pisoni points out that for the first 20 years of CI research, little beyond demographic factors such as age at CI and enrollment in oral education have been identified as influencing outcome variability in typically developing deaf implanted children (Pisoni 2000). Further understanding as to why some of these children are able to achieve skill levels almost commensurate with their hearing peers while others do not has remained a mystery. This lack of knowledge limits preoperative counseling and impedes development of individualized habilitation strategies.

Over the past decade, there has been increased interest in studying the cognitive factors that affect speech perception performance of cochlear implant recipients. Understanding the interrelated underlying processes that affect CI outcomes is also central to understanding the impact of CIs on children with multiple disabilities and their families. However, an even broader approach is needed that considers the impact of a CI on the child's neurocognitive, communication, and psychosocial development. Test measures beyond current standard speech perception and spoken language evaluations are needed that include cognition, social engagement, behavioral and emotional functioning, daily living skills, and quality of life, as suggested by a number of authors who have attempted to study these populations (Wiley et al. 2005; Palmieri et al. 2012; Johnson et al. 2008).

Comprehensive evaluation by a medical psychologist is of particular value in children at risk for some type of developmental delay. Such an evaluation serves multiple purposes. It provides a valuable snapshot of the child's current functioning, provides a baseline against which progress may be measured, and helps to guide management toward the aspects of the child's development in need of intervention. Evaluation typically includes assessment of (1) cognitive functioning, including IQ; (2) social, emotional, and behavioral functioning; (3) adaptive (daily living) skills; and (4) achievement level. A variety of instruments are available to assess each of these areas in children of varying age (see List).

Possible Assessment Tools by Category

Cognitive Functioning/Developmental Level (Ages 0–5)
- Bayley Scales of Infant and Toddler Development: Third Edition
- Wechsler Preschool and Primary Scale of Intelligence: Fourth Edition
- Differential Ability Scales; Second Edition
- Mullen Scales of Early Learning

Intelligence (Ages 6–16)
- Wechsler Intelligence Scale for Children: Fourth Edition

Nonverbal Intelligence
- Leiter International Performance Scale: Third Edition
- Test of Nonverbal Intelligence: Fourth Edition
- Hawaii Early Learning Profile: interview and activity/observation based assessment which includes domains for Cognitive, Language, Gross Motor, Fine Motor, Social-Emotional, and Self-Help

Academic Achievement
- Woodcock-Johnson Tests of Achievement: Third Edition
- Wechsler Individual Achievement Test: Third Edition
- Bracken Basic Concept Scale Revised

Adaptive Functioning
- Vineland Adaptive Behavior Scales: Second Edition
- Adaptive Behavior Assessment System: Second Edition
- Woodcock-Johnson Scales of Independent Behavior, Revised

Quality of Life
- PedsQL: Pediatric Quality of Life Inventory; parent and child reports for children ages 8–12
- Childhood Illness-Related Parenting Stress: The Pediatric Inventory for Parents
- Youth Quality of Life-Deaf and Hard of hearing (YQOL-DHH) Module—ages 11–18

Behavioral/Emotional Adjustment
- Achenbach Child Behavior Checklist, parent and teacher rating scales
- Behavior Assessment System for Children: Second Edition, parent, teacher, self-report forms
- The Strengths and Difficulties Questionnaire (SDQ); (Goodman 1997)—Ages 3–16, parent and youth forms
- A structured diagnostic interview

Evaluation of social, emotional, and behavioral functioning includes assessment of variables such as the quality and frequency with which the child engages with others, mood (for example, anxiety and depression/withdrawal), emotional regulation, oppositional behavior, attention and hyperactivity, and family functioning. Evaluation of adaptive functioning provides information about a child's daily living skills both at home and in community settings. Achievement level refers to the child's level of acquired knowledge. In a younger child, this could mean knowledge of body parts, colors, and counting skills, while for older children, academic achievement including reading, writing, and mathematics is assessed. Cognitive evaluation may include assessments of visual reasoning and visual spatial skills, attention and memory, processing speed, receptive and expressive language, and motor skills. The evaluation process often includes questionnaires for parents and professionals to obtain their observations about the child's capabilities. The child's medical history and known diagnoses may direct the evaluator to be especially vigilant to look for known associated comorbidities.

Notably, nonverbal tests of reasoning are frequently used to evaluate cognition and IQ of deaf children because of limited language ability. However, the majority of nonverbal tests important to the evaluation process rely upon the child having the necessary visual and motor skills to respond to the assessment tasks, making the assessment of deaf children with visual and motor impairments very challenging and limited. These language and physical impairment limitations in testing may result in an underestimation of cognitive potential in some children.

A comprehensive assessment can help assure that a child's caregivers have the best possible understanding of the child's functioning. This is beneficial for any deaf child receiving a cochlear implant, but is especially important to maximize outcomes for those with multiple disabilities in whom the benefits of implantation may be in areas other than speech perception and language and may be more difficult to ascertain.

Discussion

As cochlear implantation has become the standard treatment for severe-to-profound hearing loss, candidacy criteria have broadened, and intentional implantation of children with more severe disabilities has increased (Nikolopoulos et al. 2008; Berrettini et al. 2008; Hamzavi et al. 2000; Waltzman et al. 2000; Wiley et al. 2008; Meinzen-Derr et al. 2010; Donaldson et al. 2004; Filipo et al. 2004; Daneshi and Hassanzadeh 2007; Bacciu et al. 2009; Gérard et al. 2010). However, the published research on outcomes remains limited and largely retrospective. How to define and measure benefit in this very diverse population is rapidly becoming an important question. Most authors evaluate children with additional disabilities using standard hearing, word recognition, and spoken language measures. Few evaluate language in sign as an outcome measure despite research demonstrating that a CI doubles the rate of growth of both spoken and sign language for children in oral and total communication educational programs, respectively (Robbins et al. 1997). As relatively little has been published about outcomes and how to best serve these children after implantation, information is lacking for parents and implant team members who evaluate and serve these children. The current situation is ripe for meaningful translational research.

Language and cognition feed upon each other. An internal language process is needed to translate experience. Therefore, better language helps to build reasoning and cognition, which in turn enables language growth. The sooner we can intervene to improve language through hearing or enrichment of nonverbal learning, the better. Without such intervention, the problems may compound, and the gaps in both language and cognition may widen and potentially become irreversible. In addition, the role of nonspeech sound in daily life is often overlooked when defining the benefits of hearing (Berliner 1975). Individuals with hearing often take for granted that this sense is constantly providing an important connection to the world, even during sleep and from all directions simultaneously. Hearing enables us to determine our location and distance from other living and inanimate objects in our common environment. Even without visual input, our hearing provides us with a sense of security that we may be warned of unseen danger by permitting continuous contact with the environment.

Because early implantation can potentially impact the development of both language and cognitive skills and provide important contact with the environment, excluding certain children as candidates for cochlear implantation may deny them an intervention that might be uniquely suited to enhance communication and cognitive functioning, as well as adaptive functioning (daily living skills), and social and emotional well-being.

Based on several decades of cochlear implant experience delivered in the setting of a tertiary care pediatric hospital, it is the belief of these authors that children with multiple disabilities and their families often derive meaningful benefit from implantation. In terms of candidacy, there are few young children we would exclude as implant candidates based just on developmental delay and risk for cognitive impairment, if parents are engaged and willing to support their child's needs. We are most likely to advise against implantation in two situations. First, if despite a significant evaluation period which includes supportive counseling and guidance, the family is not committed or unable to be highly involved; and second, if the child clearly lacks evidence of responses to any sensory stimulation and shows no social responsiveness despite receiving therapeutic intervention. After implantation, measuring benefit in children with additional disabilities is often challenging. Standard speech perception and spoken language tests may not prove fruitful, especially within the time frame for which progress is expected by neurotypical deaf children. Expanding the ways in which we measure outcomes of CIs will help identify other benefits of improved hearing in this population, such as improvements in the quality and frequency of social engagement, emotional well-being, and adaptive capabilities that are not captured by the standard measures of progress.

In sum, post-implant assessments may serve to discourage the families and professionals serving children with comorbidities if only speech perception and spoken language are measured, and if the longer time often necessary for skills to emerge is not taken into consideration. This situation may result in less consistent implant use, poorer follow-up, and premature withdrawal of therapy services necessary for auditory skill development. A more comprehensive assessment and understanding of all of a child's abilities across multiple areas of functioning will enable both families and professionals to develop appropriate goals for the children with additional disabilities and appropriately assess progress toward these goals.

There are both philosophical and practical reasons why CI programs may not consider children with additional disabilities to be candidates and/or be unable to meet their post-implantation needs. These reasons include concerns about appropriateness of CI if spoken language is not a likely outcome, cost-effectiveness of CI in this population, additional time and resources needed to appropriately serve these children and their families, and the ethics of this intervention in children who may derive limited benefit based on standard measures. Not surprisingly, access to this technology remains problematic for many of these children. This situation is compounded when implant centers choosing not to implant this population do not inform parents that candidacy criteria

vary among CI programs and that their child might be considered a candidate elsewhere.

There are many challenges when working with children with multiple disabilities, especially those more severely affected. Delays in obtaining a recognizable and reliable conditioned response may render evaluation of hearing and optimal CI programming difficult. Therefore, the audiologist may need to modify testing methods to take advantage of whatever type of response the child may be able to consistently and reliably demonstrate. Behavioral observation audiometric techniques, such as watching for a consistent eye shift or tightening of the body in response to sound, may be useful for some children. For those children who have experience with an alternative augmentative communication (AAC) device, its use in the sound booth may enable or enhance the evaluation process. It may be hard to locate providers of aural habilitation who are experienced or even willing to work with multiply disabled children. In addition, much work may need to be done to ensure school placements that address the child's other disabilities while supporting implant use, auditory skill development, and sign language and other communication methods, including AAC devices. Further, the families of multiply disabled children are often under tremendous social, emotional, and economic stress that may interfere with consistent follow through. The availability of resources such as a social worker to support parental needs is invaluable. These families also often need assistance with coordination of care with multiple medical specialists, as well as assistance to obtain therapies from early interventionists and schools, and to secure an appropriate school placement.

A Model of Service for CI Programs Serving Children with Multiple Disabilities

Achieving the best outcomes for children with multiple disabilities is best accomplished by a multidisciplinary approach involving professionals beyond the core team members: CI audiologist, aural habilitation/speech and language therapist, and implant surgeon. Drawing upon the expertise of a developmental pediatrician and/or medical psychologist, social worker, and educator to understand the complex needs of these children and to provide support to families is invaluable. This type of collaborative multidisciplinary approach also provides a rich source of knowledge and support to the CI team members and community-based professionals who may be unfamiliar with the child's co-occurring conditions. A shared philosophy and vision are also important to successful collaboration. Ewing and Jones recommend that the following shared assumptions guide multidisciplinary collaborations of this nature: (1) Every child, including those with multiple and severe disabilities, is capable of learning. (2) Social relations and peer acceptance are important for all children. (3) Involvement of families and communities is critical. and (4) Commitment to a multidisciplinary model of service delivery with sharing of knowledge, resources, and coordination of care is a must (Ewing and Jones 2003).

Conclusion

Deaf children derive a broader range of benefits from cochlear implantation beyond spoken language. Those with comorbidities such as cognitive, dual sensory, and/or motor impairment may have lesser potential to develop age-appropriate language and the ability to talk, and yet may still derive significant benefits. Much research is needed to develop and implement appropriate measures of progress, early intervention strategies, and school-based learning approaches for this complex population. Providing hearing to these children through a CI, especially at a young age when neuroplasticity is greatest, provides an opportunity for CI programs to help such children with additional disabilities to develop to their full potential. The challenges faced by implant centers providing services to this population are significant, although the results are often well worth the effort. These children are best served when CI teams implement a multidisciplinary approach that recognizes the potential limitations of children with severe disabilities while emphasizing their potential to learn.

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Cochlear Nerve Deficiency

14

Claire Iseli, Oliver Adunka, and Craig Buchman

Definition

Cochlear nerve deficiency (CND) is clinically defined as a small or absent cochlear nerve as determined by high-resolution imaging (Buchman et al. 2006, 2011). It was first described by Casselman et al. in 1997, based on parasagittal MRI findings (Roland et al. 2012; Casselman et al. 1997). The definition can be expanded to include both the nerve and its osseous conduit. The latter consists of the internal auditory canal (IAC) and the bony cochlear nerve canal (BCNC), the bony pathway through which the cochlear nerve travels in order to reach the inner ear structures. The BCNC region is also referred to as the cochlear aperture and the cochlear fossette (He et al. 2012; Wilkins et al. 2012). The term "deficiency" is used in preference to aplasia or hypoplasia whether due to agenesis or degeneration, as it does not imply causality (Adunka et al. 2007).

The bony structures through which the seventh (facial) and eighth (cochleovestibular) cranial nerves traverse the temporal bone, including the cochlear nerve bundle as it enters the inner ear, are best seen on high-resolution computer tomography (HRCT). CND is presumptively diagnosed by the presence of a stenotic or absent IAC (Buchman et al. 2006). The diameter of the IAC considered stenotic varies between 2 and 5 mm (Buchman et al. 2006; Kang et al. 2010; Pagarkar et al. 2011; Valero et al. 2012; Walton et al. 2008). The diameter of the BCNC as measured on CT is characterized as normal when greater than 1.8 mm in diameter, mildly stenotic between 1.0 and 1.8 mm, and severely stenotic if less than 1.0 mm (Adunka et al. 2007; Valero et al. 2012) (Fig. 14.1).

Nerves are directly imaged by magnetic resolution imaging (MRI). The cochlear nerve is defined as "absent" if it is not visible on axial, coronal, or reconstructed parasagittal oblique views of the IAC (Buchman et al. 2006; Walton et al. 2008). It is defined as "small" if it is present but smaller than the other nerves in the IAC (superior and inferior vestibular nerve bundles and facial nerve) or than the cochlear nerve on the contralateral side (Buchman et al. 2006; Adunka et al. 2007). Most commonly its diameter is compared to the facial nerve in the midportion of the IAC (Casselman et al. 1997; Valero et al. 2012; Walton et al. 2008; Warren et al. 2010). At this point the cochleovestibular nerve has separated into its cochlear, superior vestibular, and inferior nerve bundles. The cochlear nerve is defined as "small" if it is present but smaller than the facial nerve at the midpoint of the IAC, "rudimentary" if just a single unbranching vestibulocochlear nerve is present, or "absent" if no cochleovestibular nerve is visible (Walton et al. 2008) (Fig. 14.2). Within the cerebellopontine angle (CPA), before entering the IAC, the cochleovestibular nerve diameter should be twice as large as the facial nerve (Warren et al. 2010).

CND can be further classified by the associated labyrinthine abnormality (Casselman et al. 1997; Govaerts et al. 2003) into

Type I—Absent cochleovestibular nerve with normal or abnormal labyrinth.
Type IIa—Present common vestibulocochlear nerve with no cochlear branch and labyrinthine dysplasia.
Type IIb—Present common vestibulocochlear nerve with no cochlear branch and normal labyrinth.
Type III—Present common vestibulocochlear nerve with no vestibular branch (not proven to exist).

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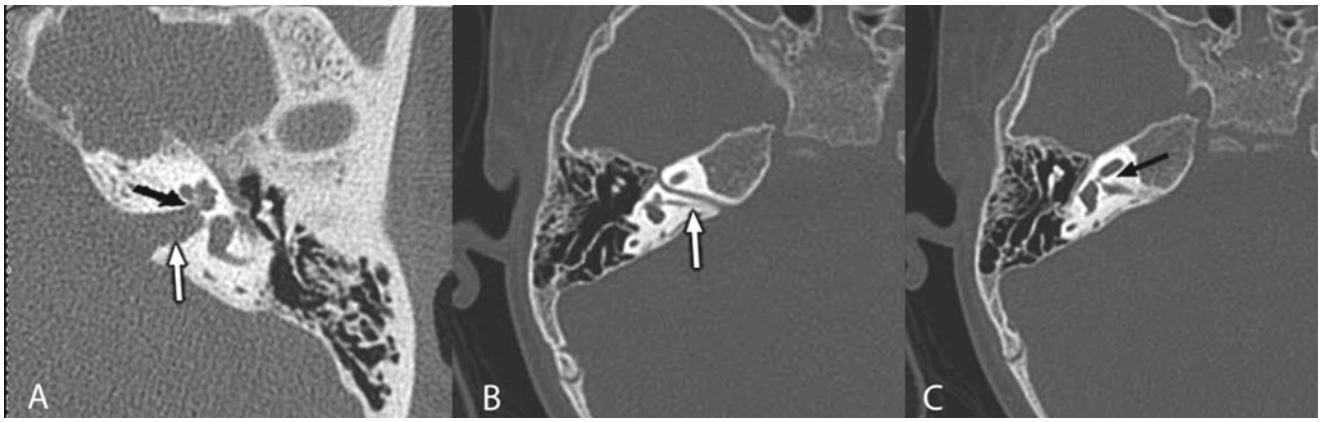

Fig. 14.1 Axial section high-resolution CT of the temporal bones demonstrating (**a**) left ear with internal auditory canal (IAC) of normal caliber (*white arrow*) and patent bony cochlear nerve canal (BCNC) (*black arrow*); right ear with (**b**) narrow IAC (*white arrow*); and (**c**) absent BCNC (*black arrow*)

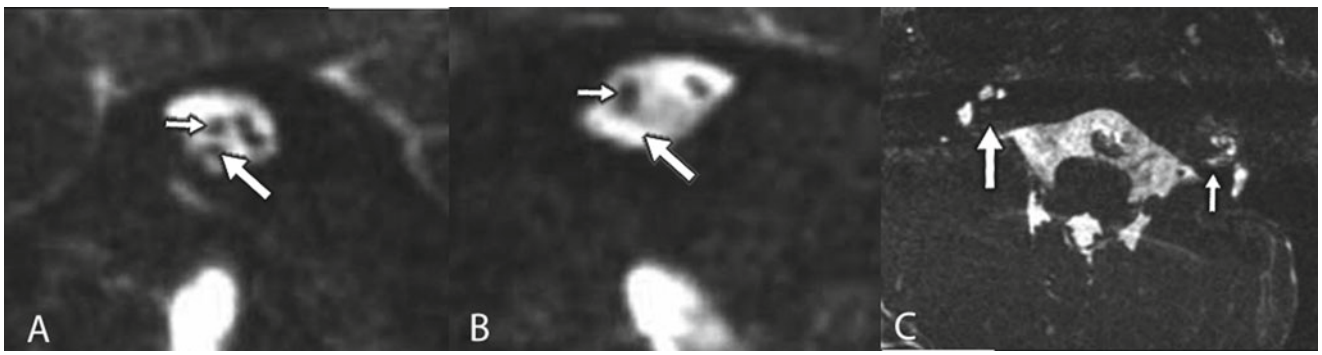

Fig. 14.2 Magnetic resonance imaging of cochlea and internal auditory canal (IAC). (**a**) Sagittal section demonstrating normal appearance of four nerve bundles within the lateral aspect of the IAC, including the cochlear (*large arrow*) and facial nerve (*small arrow*). (**b**) Sagittal section demonstrating normal caliber IAC, facial nerve (*small arrow*), and absent cochlear nerve (*large arrow* points to expected position of cochlear nerve). (**c**) Axial section demonstrating absent right IAC (*large arrow*) and narrow left IAC (*small arrow*)

Epidemiology

It is estimated that CND occurs in approximately 1 % of children who have bilateral sensorineural hearing loss (SNHL) (Roland et al. 2012; Wu et al. 2009; Parry et al. 2005; Adunka et al. 2006). CND has been reported to occur in 2.5–11 % of those children who received a cochlear implant (Buchman et al. 2011; Kang et al. 2010; Valero et al. 2012). The reported incidence of bilateral versus unilateral CND varies between one-third and 100 %, a range that may not reflect the true incidence of this pathology, but rather the patients selected for imaging (Adunka et al. 2006, 2007; Walton et al. 2008; Teagle et al. 2010). In individuals with bilateral CND the severity of nerve deficiency (small vs. undetectable cochlear nerve) of each ear is variable (Walton et al. 2008). Most often the cochlear nerve bundle is undetectable (84–90 %) rather than small on high-resolution imaging (Adunka et al. 2006, 2007). The incidence of CND in children with unilateral SNHL is not well understood as MRI of the labyrinth and internal auditory canals has not routinely been done in this population.

CND has been identified in a subset of children diagnosed with auditory neuropathy spectrum disorder (ANSD). ANSD may be present in 10–14 % of children with severe-to-profound bilateral SNHL, though its incidence may be underestimated as diagnosis requires an appropriate ABR protocol as well as an experienced clinician to interpret the waveforms (Buchman et al. 2006; Valero et al. 2012; Walton et al. 2008; Teagle et al. 2010). The incidence of CND confirmed by appropriate imaging in children with ANSD varies widely between 6 and 75 % (Buchman et al. 2006; Valero et al. 2012; Walton et al. 2008; Teagle et al. 2010; Roche et al. 2010).

Embryology

In animal models, it has been demonstrated that the otic vesicle (precursor to the labyrinth) has a trophic effect on the cochlear neurons. Without the otic vesicle these neurons do not stabilize or survive. In contrast, the cochlear itself can develop completely independently of nervous excitation (Casselman et al. 1997; Adunka et al. 2006). Temporal bone studies of patients with CND support that the same is true in humans. These studies have demonstrated the presence of a normal organ of Corti in the absence of spiral ganglion cells (Buchman et al. 2006; Nelson and Hinojosa 2001). Other supportive evidence comes for electrophysiological studies. There is a subpopulation of individuals with CND whose auditory brainstem response (ABR) recordings demonstrate the presence of a cochlear microphonic (CM) which suggests the presence of outer hair cells (Adunka et al. 2006). These findings suggest that the cochlear nerve is not required for cochlear labyrinthine development. In contrast, the formation of the IAC, which occurs in week 9 of gestation, is dependent on the presence of the vestibulocochlear nerve (Walton et al. 2008; Miyanohara et al. 2011). The nerve either fails to develop completely, in which case there will be no IAC, or the nerve can develop only partially (hypoplastic), or undergo post-developmental degeneration. In the latter scenarios the IAC will be present (Walton et al. 2008; Miyanohara et al. 2011). Secondary degeneration may occur due to vascular injury, trauma, or compression of the nerve early in gestation (Miyanohara et al. 2011). In most cases a vascular insult seems unlikely as the facial nerve shares a common vascular supply with the absent cochlear nerve and is typically present (Buchman et al. 2006).

Associations

There is no known association with specific perinatal complications, preterm delivery, perinatal infections, hyperbilirubinemia, or family history of hearing loss (Adunka et al. 2006). Over half of those with CND have an additional significant comorbidity, such as a central nervous system disorder or developmental delay (Walton et al. 2008). CND is associated with congenital syndromes, labyrinthine anomalies, ANSD, and intellectual disability (Buchman et al. 2011). Syndromes have been identified in 30–50 % of individuals with CND (Buchman et al. 2011; Walton et al. 2008; Adunka et al. 2006). Associated disorders include the following syndromes: CHARGE (*c*oloboma, *h*eart, *a*tresia, *r*etardation, *g*onadal, *e*sophageal), VATER (*v*ertebrae, *a*nus, *t*rachea, *e*sophagus, and *r*enal), Down, Duane, Moebius, oculo-oto-radial, Goldenhar, and branchio-oto-renal, as well as congenital facial paralysis and congenital hydrocephalus (Buchman et al. 2011; Adunka et al. 2006, 2007; Pagarkar et al. 2011; Valero et al. 2012; Walton et al. 2008). CHARGE is by far the most common associated syndrome. One review of children with CHARGE reported that CND was present based upon MRI of 13 of 14 profoundly deaf ears (Holcomb et al. 2013).

Bony labyrinthine abnormalities have been identified in the vast majority of ears with CND, although a small number of cases in which inner ear structures appear normal have been reported (Buchman et al. 2006, 2007, 2011; Kang et al. 2010; Pagarkar et al. 2011; Walton et al. 2008; Warren et al. 2010; Adunka et al. 2006; Carner et al. 2009). The associated labyrinthine abnormalities include common cavity, incomplete partitions I/II/Mondini, large vestibular aqueduct, and varying degrees of vestibular hypoplasia (Carner et al. 2009; Giesemann et al. 2012). The frequency of labyrinthine abnormality is higher in ears with a smaller than normal diameter IAC (Pagarkar et al. 2011; Giesemann et al. 2012). Common cavity deformity and complete labyrinthine aplasia are most commonly associated with CND (Giesemann et al. 2012).

Children with ANSD are an important subgroup of children with CND. ANSD is definitively diagnosed by the presence of the cochlear microphonic when appropriate ABR recording is performed. The wave forms used for auditory threshold determination are absent or have very poor morphology. Otoacoustic emissions are typically present during infancy but may disappear over time, especially if amplification is not used. ANSD is a heterogeneous disorder in terms of its cause, clinical presentation, management, and response to cochlear implantation.

Clinical Picture

Most children with CND have behavioral auditory thresholds in the severe-to-profound range (Buchman et al. 2006; Walton et al. 2008; Adunka et al. 2006). However, there are reported cases of CND in which behavioral thresholds are in the moderate range, or even normal at certain frequencies (Pagarkar et al. 2011; Adunka et al. 2006; Miyanohara et al. 2011). As with ANSD, when hearing is present, patients with CND usually have worse than expected speech understanding than would be expected based upon the pure tone thresholds (Walton et al. 2008). The vast majority of children with CND will be identified as in need of further evaluation by newborn hearing screening regardless of screening methodology. However, the subpopulations of CND children with ANSD typically do have measurable OAEs in the affected ear(s). As otoacoustic emissions are very commonly used to screen hearing in well baby nurseries, these children may not be identified by universal newborn hearing screening (Buchman et al. 2006, 2011; Adunka et al. 2007). CND may occasionally present as progressive or sudden loss in a

child who had passed newborn hearing screening ABR evaluation (Buchman et al. 2006). Therefore a high index of suspicion must be maintained and ABR technology must be employed to identify these children if CND presenting as ANSD is suspected.

Assessment

Audiology

Both transient evoked OAEs and distortion product OAEs have been found in approximately 10 % of those with CND (Buchman et al. 2006; Pagarkar et al. 2011; Oker et al. 2009). ABR typically demonstrates no response or a cochlear microphonic consistent with the diagnosis of ANSD (Buchman et al. 2006; Adunka et al. 2006, 2007; Kang et al. 2010; Carner et al. 2009). The cochlear microphonic (CM) typically occurs between 0 and 2 ms after stimulus onset. When rarefaction and condensation click stimuli, which have opposite polarity, are presented, ears with ANSD generate mirror image responses to each type of stimulus (Fig. 14.3). This mirror inversion of response is due to the abnormal synchrony that underlies ANSD. This phenomenon of mirror inversion is also used to help identify the cochlear microphonic which might otherwise be difficult to distinguish from electrical artifact. It is therefore important to use a protocol to diagnose ANSD that includes both rarefaction and condensation click stimuli. The possibility of electrical artifact being mistaken for the cochlear microphonic may also be eliminated by disconnecting the sound tubing delivering the stimulus to the ear. In this situation the cochlear microphonic will disappear, whereas electrical artifact will not. A distinguishing feature of the cochlear microphonic from a neural response is the relationship between latency of the wave form and intensity of the stimulus. The latency of the cochlear microphonic should remain the same with change in the stimulus level, as compared to a neural response latency which will change with the stimulus level (Buchman et al. 2006; Teagle et al. 2010).

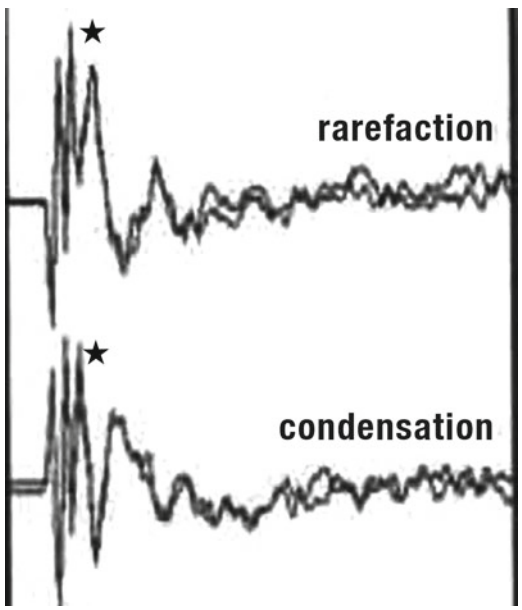

Fig. 14.3 Auditory brainstem-evoked response recording in an ear with auditory neuropathy spectrum disorder (ANSD). The presence of the cochlear microphonic is first identifiable (*star*) by the mirror image recordings seen in response to rarefaction and condensation click stimuli

Children diagnosed with ANSD or CND should be tested by behavioral audiometry by age 6 months. Behavioral tests are particularly important if the child will be fitted with amplification, as ABR should not be relied upon for determining amplification levels in this population (Roland et al. 2012; Teagle et al. 2010; Young et al. 2012). The presence of unaided or aided sound detection in an ear with CND is a positive sign that the ear may have improved auditory thresholds should a cochlear implant (CI) be provided (Warren et al. 2010; Young et al. 2012).

Auditory Steady State Response (ASSR), which is helpful in defining threshold in children with severe SNHL, is not useful if ANSD is present. ASSR may produce artifact in ears with ANSD that may be incorrectly interpreted as threshold (Kang et al. 2010; Warren et al. 2010; Buchman et al. 2007).

Imaging

High-resolution three-dimensional (3D) MRI is the most sensitive standard imaging technique available to identify CND. As CND may be present when the bony IAC appears normal, MRI is a much more sensitive diagnostic modality than high-resolution computer tomography (HRCT) (Parry et al. 2005; Pakdaman et al. 2012). However, HRCT provides more detailed information about the labyrinth and the BCNC.

HRCT slice thickness should be less than 1 mm to achieve maximum accuracy (Carner et al. 2009). It is particularly good at assessing the bony conduit for the cochlear nerve from the cerebello pontine angle to the cochlea, identifying labyrinthine abnormalities including vestibular aqueduct enlargement (Buchman et al. 2006; Pagarkar et al. 2011; Trimble et al. 2007), other temporal bone pathology (Roche et al. 2010), and external and middle ear anatomy (Buchman et al. 2007). In particular, HRCT is far superior in detecting modiolar deficiencies than MRI (Parry et al. 2005; Trimble et al. 2007). HRCT is also superior at providing the surgeon with information about the position of the fallopian canal, which contains the facial nerve as it courses through the middle ear and mastoid (Parry et al. 2005). This landmark is

particularly pertinent in cases of semicircular canal dysplasia, where the pathway of the facial nerve is more likely to be aberrant (Roche et al. 2010). The presence of stenosis of the IAC is best determined by HRCT. However, the presence of a bony IAC of normal caliber does not eliminate the possibility of CND, with 50–70 % of those with CND having a normal IAC (Buchman et al. 2006; Adunka et al. 2006, 2007; Pagarkar et al. 2011; Walton et al. 2008; Miyanohara et al. 2011; Carner et al. 2009; Giesemann et al. 2012).

HRCT is also the best way to image the BCNC, through which the cochlear branch of the eighth nerve enters the inner ear. The presence of a stenotic or non-patent BCNC is concerning for CND (Adunka et al. 2007; Pagarkar et al. 2011). However, both narrow IAC and small BCNC have been observed in temporal bones in which a normal cochlear nerve was imaged by high-resolution 3D MRI (Adunka et al. 2006; Ahn et al. 2012). Therefore definitive radiological diagnosis of CND relies on an appropriately performed MRI.

MRI of the inner ear and IACs should be performed with submillimeter T1- and T2-weighted 3D imaging techniques, such as constructive interference in steady state (CISS, Siemens) or fast imaging employing steady-state acquisition (FIESTA, GE) sequences. Axial images and oblique sagittal acquisitions perpendicular to the course of the nerves through the IAC are helpful in distinguishing the nerve bundles within the IAC (Buchman et al. 2006; Casselman et al. 1997; Adunka et al. 2006, 2007; Valero et al. 2012; Carner et al. 2009). MRI not only allows direct measurement of the nerve diameter but also has the added benefit of permitting assessment of the brain (Pakdaman et al. 2012). Brain imaging is useful as up to 40 % of pediatric patients undergoing cochlear implantation have been reported to have cerebral abnormalities (Trimble et al. 2007). In particular those with CND often have significant differences in MRI characteristics of the lateral lemniscus and inferior colliculus both ipsilaterally and contralaterally on diffusion tension MRI images compared to controls (Wu et al. 2009).

The major limitation of MRI is the difficulty in distinguishing absence of the cochlear nerve from a hypoplastic nerve whose course may be indistinguishable from other nerves, particularly in the setting of a narrow IAC (Adunka et al. 2007). Examples have been reported of a single nerve seen on MRI in the setting of a small IAC but clear clinical evidence of both auditory and facial nerve function (Adunka et al. 2006; Young et al. 2012).

Given the high rate of associated labyrinthine abnormalities in those with CND, there is benefit to performing both MRI and HRCT in this population (Buchman et al. 2006; Parry et al. 2005; Adunka et al. 2006; Oker et al. 2009; Trimble et al. 2007).

MRI and HRCT findings suggestive of CND must be augmented with electrical and behavioral methods of hearing assessment. These additional clinical evaluations are necessary because it is possible that nerve fibers or bony passageways exist that are too small to be detected on imaging. It is also possible that cochlear nerve fibers may be intertwined with those of other nerves bundles in the IAC, as demonstrated by examples of significant auditory perception with amplification or CI despite seemingly absent nerves on imaging (Adunka et al. 2007; Pagarkar et al. 2011; Valero et al. 2012; Walton et al. 2008; Young et al. 2012; O'Leary and Gibson 1999; Thai-Van et al. 2000). One case has been reported of an undetectable BCNC but good function with hearing aids (Pagarkar et al. 2011) and another with bilateral narrow BCNC and normal hearing (Ahn et al. 2012). This limitation may be overcome in the future with functional MRI, which may be able to assess the cortical response to sound (Thai-Van et al. 2000).

Electrical Testing/Other

The role of other functional tests to improve preoperative and intraoperative prediction of benefit with cochlear implantation is still evolving. Of particular importance are preoperative cortical auditory evoked potentials (CAEP) because these recordings are noninvasive and require no observations of behavior to determine central reception of sound. These responses can be detected down to 10 dB SL above hearing thresholds (Roland et al. 2012). They require less synchrony than ABR to be recorded and so are seen more often in ANSD. The latency of the P1 wave is age specific and can also be used to assess maturation of the auditory system, as the latency of this wave should reduce with age if the central auditory system is exposed to sufficient sound (Roland et al. 2012; He et al. 2012). The P1 CAEP is being used by some centers as part of the CI candidacy evaluation, although the ability of these responses to predict CI outcomes in individual patients requires further study (Roland et al. 2012). CAEP could potentially be used to aid in timing of auditory brainstem implant (ABI) surgery, as early intervention is important to outcomes (Roland et al. 2012). A limited number of studies of CI-evoked CAEPs have been done. One study reported seven children having poor speech outcomes, despite three having P1 wave forms (He et al. 2012). These findings may indicate poor correlation between CI-evoked CAEPS and outcomes.

Preoperative electrocochleography and promontory stimulation with electrophysiological recording have been used to study ears in which absence of the eighth nerve is suspected. These tests are also under investigation as to whether they will be helpful in predicting outcomes of cochlear implantation in patients without CND (Adunka et al. 2006; O'Leary and Gibson 1999). A detectable response from

these types of tests would suggest the presence of a partially functional auditory system (Oker et al. 2009). Post-insertion electrical compound action potentials (ECAP) and electrical ABR (eABR) may be recorded once the CI is placed. Interestingly ECAP may be more predictive of CI outcomes in inner ears with malformations and CND than those without these abnormalities. It has been reported that 0–27 % of ears with absent or abnormal ECAP achieved some open-set speech perception, compared to 81 % with robust ECAP responses (Buchman et al. 2011; Kang et al. 2010; Valero et al. 2012). Like CAEPs, eABR may help to monitor central auditory maturation, as wave V peak latencies should decrease with cochlear implant use and age (Valero et al. 2012). The accuracy of eABR is uncertain, as reported results of correlation to outcomes have been highly variable (Valero et al. 2012; Walton et al. 2008).

Treatment Options

Overall, CND poses multiple management challenges. In general, small rather than absent nerves, and behavioral or electrical evidence of auditory function, increase the likelihood of responses to amplification, CI, or ABI. The challenge to the clinician is to offer the greatest chance of functional hearing without taking on unnecessary risk to the child. It is important to approach each child individually rather than trying to apply a preformed treatment algorithm. Ideally the treatment chosen should be implemented as early as possible to minimize auditory deprivation and/or to allow maximum integration of non-oral communication methods into the child's home environment to ensure that communication with family is maximized.

Noninvasive

In the past, evidence of CND was seen as a contraindication to amplification and CI as no connection between the cochlear and the brainstem was thought to be present (Buchman et al. 2006; Valero et al. 2012; Pakdaman et al. 2012). Early attempts in the 1980s and 1990s with CI showed poor behavioral outcomes (Valero et al. 2012), so nonverbal forms of communication (such as sign language) were encouraged exclusively.

There is little to be lost, beyond cost, in providing early amplification to children with CND. The presence of auditory responses with amplification may provide clinically useful information about each ear. However, if the child does not make good progress in their auditory skill development, they should be rapidly transitioned to other potentially more effective treatments to prevent long periods of auditory deprivation (Teagle et al. 2010). In light of the possibility of limited speech recognition despite amplification or future surgical interventions, use of other habilitation methods such as cued speech and sign language should be encouraged simultaneously with efforts to improve audition.

In unilateral disease it is important to appropriately amplify the contralateral ear and in the setting of asymmetrical hearing losses it is critical to monitor the better hearing ear closely to watch for delayed neuronal loss (Buchman et al. 2006).

Cochlear Implantation

As demonstrated by auditory responses to electrophysiological tests in ears with no eighth nerve imaged with high-resolution 3D MRI, it is possible that a small number of nerve fibers may function to convey acoustic information to the brainstem and beyond (Valero et al. 2012). For this reason, CI has recently regained favor when performed in carefully select patients with CND. Some centers view implantation as contraindicated if there is no evidence of a cochlear nerve either on imaging or functional testing (Buchman et al. 2006; Walton et al. 2008). In patients in whom CND is present bilaterally, it is usually advisable to implant the ear with the larger nerve (Kutz et al. 2011).

Although there are a minority of cases of open-set speech perception enabling oral communication by children with CND (Young et al. 2012), overall the results with cochlear implantation are significantly poorer than children with normal anatomy implant at a similar age (Valero et al. 2012; Teagle et al. 2010; Kutz et al. 2011). There is also a higher rate of non-auditory sensations (Valero et al. 2012), including facial nerve stimulation requiring electrode deactivation (Kutz et al. 2011; Sennaroglu et al. 2009). The increased frequency of non-auditory sensations is likely related to the higher charge per unit phase required to achieve sound perception when CND is present (Valero et al. 2012). In addition, longer refractory periods after nervous excitation are often present in these children. This situation may require use of slower rates of stimulation. Mapping can be further confounded by development delay and medical comorbidities that limit the use of behavioral responses during programming sessions. It is often advisable to start with low stimulation levels and modify these as the child becomes more familiar with the device, so as to avoid painful stimuli (Buchman et al. 2011). It often takes 3–6 months to achieve a stable map (Buchman et al. 2011). In some children the sound perception results do not appear to improve with time but instead plateau after an initial gain (Valero et al. 2012). However some children may make significant progress after prolonged consistent use of their CI (Young et al. 2012). Though few children achieve open-set speech, most will gain some benefit from implantation and have increased

environmental awareness (Buchman et al. 2011; Kang et al. 2010; Valero et al. 2012; Govaerts et al. 2003; Teagle et al. 2010; Oker et al. 2009; Young et al. 2012; Kutz et al. 2011). Speech intelligibility in this group is also often poor (Kang et al. 2010). Yet in one large study, approximately one-third of patients with CND were able to attend mainstream school (Buchman et al. 2011). Results of recent trials are presented in Table 14.1. It is difficult to truly gauge the best possible outcomes with CI in this group as few of those reported upon were implanted under 2 years of age (Kutz et al. 2011).

Unlike minimal harm in attempting hearing aid use, the clinician should always be cognizant of the potential risks of cochlear implantation, including meningitis and facial nerve injury. These risks are somewhat increased in ears with labyrinthine abnormalities which are more frequently present in ears with CND (Oker et al. 2009). Furthermore in those with concurrent syndromes or medical comorbidities there may be an increased risk of anesthetic complications.

It is critical that preoperatively parents are given realistic expectations of more limited CI outcomes, including the possibility that no sound perception or rejection of the device by the child may occur. During the habilitation process, it is important that the family and professionals working with the child be made aware that responses to only loud sounds may be due to non-auditory percepts and not auditory awareness of sound (Buchman et al. 2006, 2011). Children with CND who receive a CI are likely to benefit from early introduction of non-oral communication augmentation (Buchman et al. 2011).

Auditory Brainstem Implantation

A small number of auditory brainstem implants (ABIs) have been performed in children with CND. The majority of children implanted with ABI first underwent cochlear implantation and received no benefit (Colletti and Zoccante 2008). All had ABIs placed via the retrosigmoid approach (Sennaroglu et al. 2009; Colletti and Zoccante 2008; Colletti et al. 2001, 2002, 2004). Intraoperative and postoperative eABR or neural response telemetry (NRT) and postoperative HRCT have been used to evaluate the positioning of the ABI electrodes relative to the cochlear nucleus (Colletti et al. 2001). The results of ABI have so far been variable. The majority of children have benefitted from increased awareness of environmental sounds with some children achieving varying degrees of speech detection and word recognition. Evaluation of benefit has been confounded by the presence of other conditions such as ADHD or severe developmental delay in some of the recipients (Sennaroglu et al. 2009). One study noted a significant improvement in cognitive function in all 26 of the children implanted (Colletti and Zoccante 2008). Most ABI-implanted children have some non-auditory stimulation which may present as facial stimulation, gag or swallow reflex stimulation, nystagmus, or vertigo (Sennaroglu et al. 2009; Colletti and Zoccante 2008; Colletti et al. 2001, 2002, 2004). If this occurs without any sound perception then the electrode responsible is simply turned off. If non-auditory stimulation is concurrent with auditory stimulation but primarily occurs in response to high levels of stimulation, programming of the responsible electrode to reduce stimulation levels may resolve the problem (Sennaroglu et al. 2009). Despite the proximity to important central anatomy, only one intracranial complication, an intra-cerebellar clot requiring surgical evacuation, has been reported (Colletti and Zoccante 2008). Though ABI surgery is an intracranial procedure for which there is a small risk of significant lower cranial nerve and central nervous system complications as well as bacterial meningitis, it has so far been accomplished with a good record of safety in the hands of experienced neurosurgery/neurotology teams.

Conclusion

High-resolution three-dimensional MR has led to an appreciation that CND is more common than had been previously recognized. CND may be present in individuals with either SNHL or ANSD. Responses to amplification and to CI are typically much poorer than seen in individuals without CND. However, variability in outcomes has been reported that is likely related to both limitations of MR resolution and individual ability to make use of degraded auditory input. The issue as to whether ABI is the preferred treatment modality for this population is being explored.

Table 14.1 Published speech outcomes with cochlear implantation in cochlear nerve deficiency

Author	Case No. size CN	Controls	Age at surgery	Associated abnormalities	Follow-up	Environmental sound detection (SD)	Closed-set speech	Open-set speech
Valero et al. (2012)	19 small	19 age matched	Mean 4 years (1–12)	CND: 12 with CHARGE/other inner ear abnormality. 2 with other comorbidities	1–14 years	All improved SD. Early plateau of benefit	12/19	8/19 (limited)
Young et al. (2012)	10 7 absent 3 small	Nil	Mean 2.6 years (1.1–5.2)	7/10 syndromes (2 CHARGE, 2 Goldenhar, 1 Clouston, 1 Down, 1 BOR). 9/10 associated inner ear malformations	1–6 years	7/10 improved SD, 3 no response	4/10	3/10
Buchman et al. (2011)	22	54 non-CND inner ear malformation	Mean 4 years (1.3–18)	82% associated inner ear malformation. 77% suspected syndrome	1–10 years	19/22 improved SD, 3/22 no response	16/22	4/22 (limited)
Kutz et al. (2011)	7 absent 2 small	Nil	1.5–8.3 years	1 pt-microcephaly, 1 CHARGE. All some inner ear malformation	1.1–7.1 years	All improved SD	3/9	Nil
Kang et al. (2010)	6 absent	15 age matched	1–3 years	1 pt abnormal cochlear, 3 pts some vestibular abnormality	12–36 months	5/6 improved SD, 1/6 rejected device. Early plateau of benefit	5/6	1/6
Song et al. (2010)	13 narrow IAC	Nil	2–13 years	9/13 had associated inner ear malformations, 10/13 had associated medical comorbidities/syndromes	1–6 years	10/13 improved SD, 3 had no sound perception	7/13	Nil
Warren et al. (2010)	3 absent	Nil	0.5–4 years	2 pts—CHARGE, 1-pt hemivertebrae + other. All had cochlear hypoplasia and absent SCC	5–9 months	All improved SD	1/3	Nil
Chadha et al. (2009)	2 small	8 non-CND inner ear malformation	1–2 years	1 had associated inner ear malformation	1–2 years	All improved SD	2/2	2/2
Oker et al. (2009)	2 small bilateral	Nil	1.5–3 year	2 pts—some vestibular abnormality. 1 minor CHARGE	2–5 years	Better SD with bilat than unilat	1/4 ears	1/4 ears
Bradley et al. (2008)	6 small	Nil	1.5–6 years old	3 had significant medical comorbidities/ syndromes	2–6 years	All improved SD	1/6	Nil
Walton et al. (2008)	15 ANSD + CND	39 ANSD without CND	Mean 4 years	CND: 93% inner ear abnormality and 67% medical comorbidity	1–2 years	All improved SD	Nil	Nil
Govaerts et al. (2003)	3 absent 1 small	Nil	2–11 years	3/4 had associated cochlear abnormality	1–6 years	2/4 rejected device. 2 improved SD	1/4	Nil
Acker et al. (2001)	1 small	Nil	4.5 years	Nil	3 months	Improved SD	1/1	Nil
Ito et al. (1999)	1 small	Nil	4 years	Common cavity deformity	4 months	Improved SD	1/1	Nil

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The Neuroscience of the Pediatric Auditory Brainstem Implant

15

Robert Shannon, Lilliana Colletti, and Vittorio Colletti

Introduction

In the early 1700s philosophers were debating whether the senses were innate or learned. William Molyneux, an Irish philosopher, posed a question that has become known as "Molyneux's problem." Essentially, this thought experiment asked whether a person, born blind, but familiar with object shape from touch, could identify an object by sight alone if they had their vision restored. Molyneux and his friend, English philosopher John Locke, both argued that the blind person would not be able to tell a ball from a cube using sight alone. They both believed that the brain did not have an intrinsic template for sensory information—that it had to be learned. They further assumed that even if this shape information was learned from touch it would not transfer to vision without visual experience. Since that time, there have been cases where people, congenitally blind from cataracts, regain sight from surgery to remove or replace the lens of the eye. In most cases they could technically see, but could not recognize objects because their brains had not been trained on visual input. Even after many months of experience most such people functioned as if they were still blind (Gregory and Wallace 1963). More recent data shows some limited ability to learn basic visual pattern recognition skills in adulthood after congenital blindness (Kalia et al. 2014).

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It appears that at least two factors influence the development of the brain's ability to process sensory information: receiving information during the critical period of brain development in childhood, and the quality of the sensory information. The brain is a complex and dynamic organ—some aspects of brain development are highly choreographed by biological stages of plasticity. We now know that complex sensory development must occur quite early—before about 5 years of age, or the window of plasticity closes (e.g., Ruben and Rapin 1980). If no sensory experience is obtained prior to age 5, then later restoration of that sense is difficult if not impossible. Other aspects of brain plasticity remain after age 5, but the level of organization needed to process a complete new sensory modality appears to be not available after 5 years of age.

In our lab at the House Ear Institute (Shannon 2015) we had direct experience with a man in his early 40s who had been profoundly deaf since birth from Usher's syndrome. He was working as an electrical engineer but was losing his sight from retinitis pigmentosa. Although he had never heard sound he was confident that he could learn to recognize sounds because of his training as an engineer—he knew mathematically what a sine wave was and what an auditory filter was. He understood the physics of speech sounds and their spectrograms. He felt that with this knowledge he could learn to differentiate and recognize sounds from a cochlear implant. Following cochlear implantation, CI researcher Qian-Jie Fu developed a computerized training program for him to practice simple auditory distinctions: loud vs. soft, high vs. low pitch, one sound vs. two sounds, etc. (This software is available for free at http://www.emilyshannonfu-foundation.org/esff_software.html.) Another training program simply presented spoken words that identified everyday objects: ear, nose, eye, fingers, etc. He worked diligently at this task but never made much progress. The sound from the implant always evoked an emotional feeling that included some degree of sadness, probably because his unused auditory cortex had been repurposed to accommodate emotional information. Electric stimulation of the

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auditory system was now triggering emotions instead of, or in addition to, sound sensations. Although he knew the sounds mathematically he was not able to learn even simple patterns acoustically. After 2 years of dedicated work and practice he gave up. This outcome is consistent with the Molyneux's conjecture and with the prior experience with congenital blindness.

What is the relationship between brain development and sensory information? What are the critical periods for sensory brain development? How much sensory information is necessary for brain development? The answers to these questions are now better understood thanks to restoration of hearing with cochlear implants (CIs) and auditory brainstem implants (ABIs) in children. This chapter briefly reviews the findings on CIs and ABIs and discusses the results in the context of neuroscience and brain development.

Cochlear Implants

In Molyneux's time it was rare for a person born with a sensory deficit—blindness or deafness—to recover the sense, so there was no way to resolve his conjecture. However, the thought experiments of these early philosophers can now be tested in scientific detail with the advent of cochlear implants and auditory brainstem implants for the congenitally deaf. Early in the application of CIs it was apparent that providing CIs to congenitally deaf adults was of little value. Just like cataract surgeries in the congenitally blind, these CIs resulted in only rudimentary auditory capability for adults, even after many years of experience.

However modern cochlear implants can restore good functional hearing to children born deaf. The results are now completely clear, as this volume attests, that cochlear implants provide sufficient auditory information for children to develop functional hearing. Most children can identify sounds, recognize speech, and produce speech well enough to interact in a mostly normal fashion with the hearing world. However, we also know that congenitally deaf children don't typically achieve these results if implanted after the age of 8. The plasticity of the brain in adapting to new sensory information appears to diminish after the age of 5 years (Niparko et al. 2010). Although cochlear implant outcomes are best when the child is younger, some neural plasticity remains so that older children can still benefit from a CI, but on average their outcome is not expected to be as good as children implanted early. Some evidence suggests that if a brain region is not used by its natural sense, it becomes "colonized" by some other function (Lee et al. 2001; Shepherd et al. 1997). Once hearing is restored by a cochlear implant it is difficult to dislodge the "interloper" after age 5 years. Of course, children who have even limited early auditory experience can use the CI information more effectively because their brains have received some input from the auditory system and their brain will have developed some abilities to interpret auditory information.

Lee et al. (2001; Giraud and Lee 2007) showed PET images of congenitally deaf children implanted with CIs at different ages. The area of auditory cortex that responded to the CI diminished as the duration of deafness increased. It appeared that the auditory cortex in these children now responded to sign language or other things and not as strongly to acoustic sound. The auditory performance of these children was proportional to the area of the cortex that responded to acoustic sound. Children who had a long period of deafness showed poor speech recognition with the cochlear implant and strong responses in auditory cortex to sign language. Children with a short period of deafness showed a larger area of the cortex that responded to sound and had excellent speech recognition with the CI.

Children implanted at early ages have shown dramatic hearing abilities with cochlear implants (Govaerts et al. 2002; Rubinstein 2002; Manrique et al. 2004; Robbins et al. 2004; Svirsky et al. 2000, 2004; Dettman et al. 2007; Niparko et al. 2010, Colletti et al. 2014). Measures of speech recognition have shown an almost normal trajectory of speech recognition development as long as the CI was provided prior to 3 years of age. Recent research has shown improved performance with even earlier implantation, so that cochlear implantation prior to 12 months of age is now the norm. There is now evidence that implantation prior to 12 months of age provides improved long-term auditory performance compared to those implanted after 12 months (Colletti et al. 2011; Waltzman and Roland 2005).

The brain develops remarkable pattern recognition abilities, and this development begins even prior to birth. In a normally hearing child, hearing begins at least 1 month prior to normal term birth. At birth newborns can already distinguish their mother's heartbeat and voice from others (Smith et al. 2007; Kisilevsky et al. 2003, Kisilevsky and Hains 2011) and can recognize music that was played in utero (Lecanuet et al. 2000). This shows that the brain's auditory pattern recognition is already in action prior to birth. And so children with congenital deafness have a disadvantage already at birth because their brain's processing of auditory sensation was not "jump-started" in utero.

Even if prosthetic stimulation is available during the time of critical brain plasticity, the quality of the sensory information will have a large effect on the ability of the brain to use it. If the quality of sensory information is poor, then even the developing adaptive brain may not have sufficient information to work with. The pattern of neural activity provided by normal acoustic stimulation and the pattern produced by electric stimulation are highly different, with the acoustic pattern being much richer in cues than the electric. Psychophysical studies in persons with CIs have shown

almost normal perceptual abilities in timing, e.g., gap detection, forward masking, and modulation detection (e.g., Shannon 1990). But CI listeners have reduced abilities in the perception of intensity (Zeng and Shannon 1994) and frequency resolution (Nelson et al. 2008). But not all of those cues are equally important for speech—some cues can be dropped with little consequence for speech perception while other cues are essential. So which aspects of the pattern of sensory information are more important for sensory development and which are less important? Fu and Shannon (1998) showed that distortions in amplitude mapping only have a small effect on speech recognition, while Shannon et al. (2004) showed that the number of spectral channels necessary for recognition depends on the difficulty of the listening situation. We know that the information provided by a cochlear implant is sufficient for understanding speech. Congenitally deaf children implanted with a CI are mostly able to understand speech and develop spoken language. In spite of the many differences between the patterns of acoustic and electric activity, the brain is able to make optimal use of the information. Congenitally deaf children with CIs reach the same level of performance as adults with CIs who had prior normal-hearing experience. Even though the implant activation has highly abnormal timing and a smeared representation of tonotopic place, the brain is able to extract the information fully from the signal. This observation suggests that the limiting factor in modern cochlear implants is in the signal processing. The same high level of performance is achieved by both adults who have heard prior to deafness and by children who are learning to hear with only the signal provided by the CI. It is now clear that the most serious limitation of CI signal processing is the limited number of spectral channels. Even though modern CIs contain up to 20 electrodes, the electrical field interactions and neural interactions limit the effective number of spectral channels to 8–10 (Friesen et al. 2001). If we can improve the tonotopic specificity we should be able to improve the speech recognition abilities of patients with CIs.

Auditory Brainstem Implants

The auditory brainstem implant was developed for patients without an intact auditory nerve, making them unable to benefit from a CI. The ABI is similar to a CI except that the electrode is placed on the cochlear nucleus in the brainstem instead of in the cochlea. The cochlear nucleus is the first auditory relay station beyond the cochlea in the pathway of auditory information to the brain. An ABI provides a case of more extreme distortion of the sensory pattern of information than a CI. The ABI electrodes rest on the surface of the cochlear nucleus, a complex structure with three main divisions (anteroventral: AVCN, posteroventral: PVCN, and dorsal: DCN). Each of these divisions has unique cell types and physiological response properties. Each division has at least one tonotopic axis. The ABI electrode is 8 mm in length, so that it likely stimulates both PVCN and DCN. However, the tonotopic axes in these divisions are not well represented on the surface. So the surface electrode ABI probably stimulates a mix of cell types, mostly low-pitch tonotopic regions, and at least two major functional divisions. Previous experience with adults shows that the pitch map of electrodes is complex and non-monotonic (Otto et al. 2002). We know little about the temporal pattern of neural activity evoked by ABI firing. Based on what is known from stimulation of the auditory nerve by a CI we can assume that the ABI produces extreme, nonnatural phase locking to electric pulses.

Even though it was known that CIs could provide sufficient information for speech recognition in adults and children, little attention was paid to auditory brainstem implants because ABI results in adults were poor compared to CIs (Otto et al. 2002). However, Colletti et al. (2004) showed excellent open-set speech recognition in adults with ABIs. In the USA, most recipients of an ABI were individuals with neurofibromatosis type 2 (NF2). In contrast, ABI recipients implanted by Colletti in Italy lost their auditory nerve from trauma or severe ossification (Colletti et al. 2004). The fact that the ABI could support open-set speech recognition in non-NF2 patients showed that the poor ABI results in NF2 patients were not due to the device or to electrode placement; rather, it suggested that the poor performance was related to the damage to the auditory pathway as a result of NF2.

In a CI the electrodes typically change systematically in pitch sensation from one end of the array to the other. The pitch representation may be shifted relative to normal if the electrode is placed too shallowly in the cochlea, but CI patients usually adjust in a few months to this shifted pitch representation. In contrast, the ABI electrode is placed on the surface of the cochlear nucleus in the brainstem. The ABI electrodes do not produce a monotonic change in pitch because the cochlear nucleus has multiple tonotopic maps and these are not accessible from the surface of the nucleus (Moore and Osen 1979; Moore et al. 1994). In the first few months of ABI use, adults find the sound quality strange and unhelpful. They typically require 3–6 months of experience to make sense of the sounds. Among adult ABI users who were deafened due to NF2, only a few out of hundreds were able to obtain open-set speech understanding (Otto et al. 2002). Among adult ABI user who did not have NF2, about 30 % were able to understand speech with an ABI well enough to have a conversation on the telephone (Colletti and Shannon 2005; Behr et al. 2007; Matthies et al. 2013, 2014). A few in this latter group achieved speech recognition levels comparable to the best CI outcomes: 100 % recognition of simple sentences presented in quiet, and 50 % recogni-

tion of speech presented in noise at signal-to-noise ratios of 3–4 dB. These high-performing ABI-implanted adults were able to successfully map this scrambled tonotopic information onto the speech pattern recognition that already existed in their brain. Such cases are interesting for neuroscience, because it shows that an adult brain, trained in speech pattern recognition with acoustic sound, can map that pattern recognition onto a new pattern generated through electrical stimulation that has little in common with the original pattern—different temporal properties (phase locking, etc.) and tonotopic organization that is highly scrambled compared to the acoustic representation. In this case the brain can "morph" the new pattern into the previously established speech recognition system in the brain.

Recent outcomes show that even some people with NF2 can achieve speech recognition with an ABI. Two surgeons (Behr et al. 2007; Matthies et al. 2013, 2014) have shown that about 30 % of their NF2 ABI patients were able to understand speech well enough to converse on the telephone. These surgeons use a different surgical approach that may cause less neural and vascular damage to the cochlear nucleus region. The fact that they are able to show improved speech recognition suggests that even subtle damage to the cochlear nucleus can have large effects on ABI performance.

ABI in Children

Colletti et al. (2014) also implanted the ABI in children with no auditory nerve. This was controversial because at that time most outcomes with the ABI were poor compared with CIs. Many people thought that it was not ethical to incur the risks of ABI surgery for a limited outcome, especially in children. However, Colletti had seen excellent CI-like outcomes in non-NF2 adults (Colletti and Shannon 2005) and so implanted the ABI in non-NF2 children. Colletti et al. (2010) demonstrated that the complication rate of ABI surgery in children implanted at his medical center was similar to that seen in a CI, so the surgical risk did not appear to be unreasonable. He selected children with congenital anomalies who had no auditory nerve based upon magnetic resonance imaging (MRI) or those who had failed to show any progress with a cochlear implant. Some of these children had severe ossification following meningitis and some had temporal bone fractures that severed the auditory nerve. After a few months, he started to see CI-like outcomes in some of the children. Figure 15.1 shows the outcomes over 7 years from 64 children implanted in Verona with ABIs (data from Table 1 of Colletti et al. 2014). The results show median scores over time on the CAP, a categorical evaluation of auditory performance (Archbold et al. 1995). On the CAP a category of 0 indicates no detection of sound, 4 represents identification of

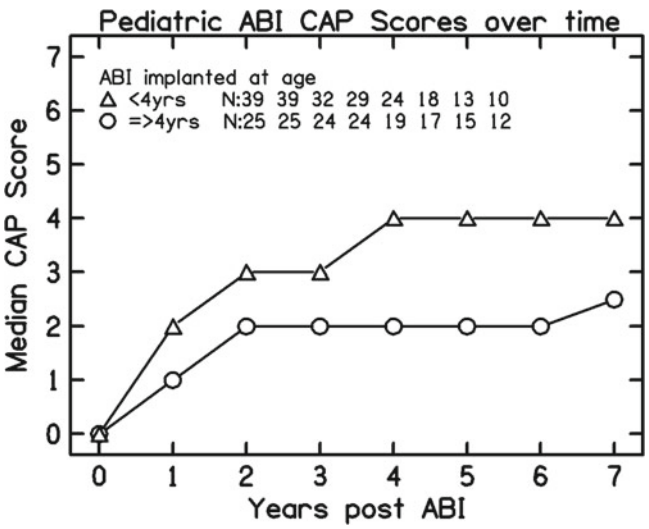

Fig. 15.1 Auditory performance as measured by the CAP test as a function of time after ABI surgery. The two groups or results show scores over time for children implanted before or after 4 years of age. Note that the number of children represented by each data point decreases over time: the *numbers in the legend* show the *N* for each successive point for the two groups

words from a closed set of alternatives, and 7 indicates the ability to converse on the telephone with a familiar person. Results in Fig. 15.1 are divided into groups that differ in the age at the time of ABI surgery and the numbers in the figure show the number of children at each time point. Note that the group that received the ABI before 4 years of age ($N=39$) had better performance, with the median score reaching high closed-set (CAP level 4) performance. These are children with diverse etiologies, but all had no auditory nerve visible by MRI, mostly from genetic problems and a few from disease or trauma. Lower median CAP scores were observed in the children who received the ABI after age 4 ($N=25$). These children showed an increase in CAP level but the median level only reached a level of 2, which represents only being able to make simple discriminations between sounds. Overall, of the 64 children, 11 (17 %) reached CAP level 7, which means that they are able to converse on the telephone with a family member, and 20 children (31 %) were able to achieve open-set speech understanding (CAP scores of 5–7). Twenty-two of the 64 children had previously received a CI and showed no response. These results show that the scrambled spatiotemporal pattern of activity from the ABI is sufficient for the brain to learn to hear even with no prior experience. This result shows the potential for the ABI to restore a significant amount of hearing function to children who cannot benefit from a CI. Good speech recognition is now being obtained in other centers with ABIs in children (Sennaroglu et al. 2009, 2011; Eisenberg et al. 2008, 2012). The implication of this observation for neuroscience is addressed in the next section.

The Role of Ear and Brain

Now let us return to Molyneux's problem. If a person does not receive any sensory input from birth and then that sensory input is restored as an adult, the person will not be able to use it very well, if at all. There are two issues to consider: At what age is the sensory information provided, and what is the quality of the restored sensory information? Cases of visual cataracts and cases of cochlear implants both show that sensory information provided as an adult to a person with congenital absence of that input is not effective. We assume that this is due to the plasticity of the brain not being as fully available in adulthood as in infancy. Learning complex pattern recognition from sensory input apparently requires a high level of brain plasticity, which is not available to adults. Outcomes from CIs and ABIs in children show better outcomes the earlier the device is implanted. This result suggests that the necessary neural plasticity is most available right after birth.

A newborn brain must learn about the universe from its sensory experience. Studies have shown how babies learn to distinguish subtle differences in sensory patterns by computing the probabilities from their sensory experience (Saffran 2002, 2003; Saffran et al. 1996; Jusczyk and Aslin 1995). Babies can be presented with sequences of nonsense sounds of only 2 min in duration and they will remember phonetic aspects of the sequences even 1 week later. When the babies' response to sounds has a desired effect on the mother's behavior, the distinction is quickly learned. But even in the case of normal hearing it takes many years to learn the complex patterns of speech. Hart and Risley (1995) estimated that children are exposed to more than 30 million words of direct interaction with their mother before the age of 5 years. This observation has given rise to the 30 Million Word project in Chicago to train parents on the importance of early word and language exposure in their children (Leffel and Suskind 2013). The learning of sensory probabilities is also well studied in vision, showing that people learn the detailed probabilities of the visual world over many millions of experiences (D'Antona et al. 2013). This kind of intensive experience has been popularized as the "10,000-h" rule, suggesting that mastery of any complex task takes 10,000 h of dedicated practice. Early development of sensory systems may require millions of repetitions to achieve mastery of complex pattern recognition.

So even if the sensory information is intact it can take thousands of hours and millions of repetitions to gain "fluency." What happens when the sensory information is degraded and distorted, as in the case of CI and ABI auditory input? If the sensory experience is dramatically reduced in quality, then the level of sensory development will be limited by this poor signal. Is it possible to learn patterns of sensory information that are dramatically different from the normal pattern? The pattern of CI stimulation is different from the pattern of acoustic stimulation—the activation by CI electrodes is compressed and probably shifted along the cochlea compared to the normal acoustic pattern. In the ABI neural activation patterns are much more scrambled in tonotopic order than with a CI. Will children still be able to learn the probabilities, the regularities in the sensory patterns when they are so distorted?

First let us consider cochlear implants. The CI activates neurons with electric currents. Research has shown that there are large differences in the spatial and temporal patterns of neural responses to electric stimulation compared to acoustic stimulation. How well will the brain be able to use such a neural pattern for hearing? Early single-channel CI devices presented an analog version of the speech wave form directly to a single intracochlear electrode—almost like just removing the wire from your stereo speaker and connecting it to a wire in the cochlea. Such stimulation did not use multiple electrodes to access the normal tonotopic distribution of information in the cochlea. However, even with this signal some children were able to achieve limited open-set speech understanding—a level of performance that was unexpected and still not fully understood (Berliner et al. 1989).

In multichannel CIs the signal is split into different frequency bands and each band is presented to a different electrode, to take advantage of the normal tonotopic distribution of information in the cochlea. But even if we present many "channels" or electrodes of electric stimulation we know that not all of those channels are used effectively. Studies have shown that CI patients only receive 8–10 channels of effective information even if we present 16–22 channels of input (Friesen et al. 2001). The loss of information occurs because of interference between electrodes, either electrically or at the neural level. In a CI the information is also presented to a limited region of the cochlea, and is probably shifted in tonotopic space from the normal acoustic locations due to insertion in the basal cochlear region. Congenitally deaf children never learned the "normal" tonotopic map, so their developing brains simply take the pattern of implant stimulation as the "new normal." Speech recognition results show that most children with a CI can rapidly adapt to the degraded and distorted pattern of sensory information and achieve a high level of speech recognition and production. They are mostly able to reach the same level of speech recognition as adult CI users who have had normal acoustic hearing prior to deafness. This suggests that the brain is able to use the full information in this implant signal, in spite of the distortion compared to the normal acoustic pattern.

But not all children achieve this level of speech recognition. About 10 % of children implanted with a CI show minimal auditory capabilities even over a long period of time (Niparko et al. 2010). These children may have deficiencies in their residual auditory nerve or central processing

limitations that limit their performance (Govaerts et al. 2003; Casselman et al. 2008; Carner et al. 2009; Buchman et al. 2011; Young et al. 2012). In these cases speech recognition is not limited by the signal processing of the device but by the status of their auditory nerve. Some of these children may be candidates for an ABI. Colletti's results (Colletti et al. 2014) indicate that ABI outcomes are poorer in children with multiple disabilities compared to children without additional disabilities. Clinical trials are now under way in several centers to refine the selection criterion for children to receive an ABI.

In spite of the ABI distortions in time and spatial activation patterns, some children are achieving high levels of open-set speech recognition with the ABI (Colletti et al. 2014; Sennaroglu et al. 2009, 2011). This result demonstrates the flexibility of the developing brain when presented with degraded and tonotopically scrambled auditory information. The fact that some children can achieve high-level CI-like performance shows that even in the ABI case the brain is capable of getting the maximum information out of the implant signal. One unsolved problem with ABIs is why the rate of children obtaining good speech recognition is lower than for CIs. It is possible that the genetic problems that cause the loss of their VIII nerve also affect development of the cochlear nucleus and other central auditory nuclei. It is also possible that some children have more complex central problems in development that limit their ability to use the information provided by an ABI.

Cognitive Development

One understudied aspect of auditory implants is the effect of prosthetic sensory experience on cognitive development. Work with deaf children and with CIs has shown reductions in executive function, including visual working memory (Pisoni 2000; Pisoni and Cleary 2004; Khan et al. 2005; Edwards et al. 2006; Shin et al. 2007; Le Maner-Idrissi et al. 2008; Fagan and Pisoni 2009; Kronenberger et al. 2014). At first glance this is a puzzling finding. Why would a sensory deficit in hearing affect visual memory? One possible answer comes from the consideration of how memory depends on the senses. It has long been known that people with synesthesia have exceptional memories (Rothen et al. 2012). Synesthesia is a rare phenomenon where activation of one sense, e.g., vision, is accompanied by linked sensations in another sense, like hearing or taste. For example, some persons with synesthesia always see a specific color associated with a number, or sense a specific color to be associated with a particular pitch; all eights may also be perceived as blue, or middle C is always accompanied by the sensation of green. It is thought that the extra sensory experience provides the brain with an extra dimension of information about objects and events. This extra dimension of sensory experience provides more cues for the storage and retrieval of memories. It is possible that the deficit in memory (and other cognitive deficits) in deaf children is due to the loss of dimensionality of sensory experience (Fagan and Pisoni 2009). If only two sensory dimensions (e.g., vision and smell) provide cues to a memory event, the memory may be weaker than if three sensory dimensions (e.g., add hearing) contribute to the memory. The early development of executive function may also be delayed by the reduced dimensionality of sensory experience. So having one less sensory dimension (deafness) may degrade memory and cognitive development.

If CIs and ABIs restore the auditory dimension do they also restore deficits in memory and executive function? Is the restoration of memory proportional to the restoration of sensory function? We do not yet have answers to these questions, but research is under way to quantify such possible effects. One study (Colletti and Zoccante 2008) in nine children with an ABI showed that even the sensory information provided by an ABI provided significant improvements in selective visual-spatial attention and fluid reasoning (i.e., reasoning or the ability to solve problems in novel situations). It appears that adding another dimension of sensory experience, even a limited dimension, can provide sufficient additional information to improve cognitive development.

Issues for the Future

There is still much research to be done to fully understand the role of prosthetic hearing on speech, language, and cognitive development. It is clear that it is of critical importance to provide the best sensory information at as early an age as possible. Brain development is moving fast in the years 0–3 and every month lost is a lost opportunity. If a child with an implant is not making progress we need to determine if this is due to a poor adjustment of the processor, a partial failure of the device, or a problem with their neurons due to the etiology of hearing loss. Once this is determined we can improve the adjustment, replace the device, or, in the case of the CI implanted child, contemplate moving to the ABI. But whatever solution is correct, the brain plasticity clock is ticking. Figure 15.2 presents a schematic of the normal fitting cycle for a CI or ABI in a child. Initially the child cannot provide feedback on the qualities of the hearing sensation because they have no experience with sound. The initial maps are often fit based on the levels of evoked auditory brainstem electrophysiological responses (ABR) and avoiding any non-auditory side effects (NASE), especially for the ABI. Trained pediatric audiologists can manage the child's attentional state and observe behavioral responses. Once an initial map setting is achieved, the child must adapt to it. This adaptation may take several weeks or even months

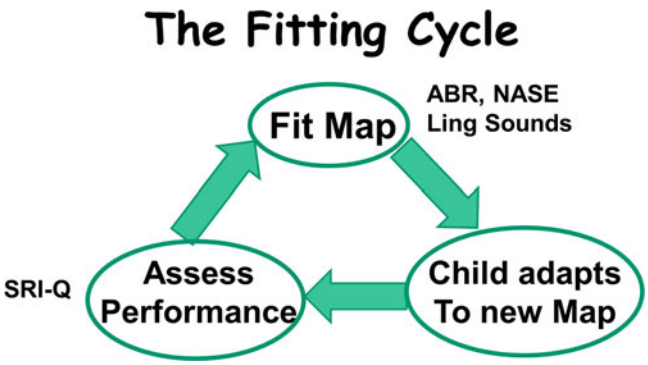

Fig. 15.2 Schematic representation of the fitting cycle for an ABI. Initial test measures indicate the ability of the child to detect and discriminate sounds with the new map, but additional tests are made after a period of adjustment where the child learns to use the new information provided. The cycle must be short to achieve an optimal mapping as soon as possible to take advantage of brain plasticity, but long enough to allow the child to accommodate to the new information

before any assessment can be made of the efficacy of the fitting. This is an area where better methods and tools are needed. The sooner we can determine if a device setting is inadequate the sooner we can adjust the map in an attempt to fix the inadequacy. This cycle can't occur too quickly because it takes some time for the child to adapt to the changes before we can reasonably assess their use of acoustic cues. However, this fitting cycle should occur early in the process so that the final map can take advantage of the plasticity of the brain.

New brain imaging methods may hold promise in this process. Cortical potentials and optical imaging may be able to show new patterns of cortical activity before any behavioral manifestations (Friesen et al. 2009; Aslin 2012, Martinez et al. 2013). We assume that a strong physiological response at the auditory cortex is a sign that the information from the implant is activating the brain in the desired fashion. Even if the individual child has not yet learned how to use this information, normal developmental plasticity gives us some confidence that the information will be used if it is present. Behavioral techniques, such as the rate of babbling development, may offer a metric for assessing early auditory progress (Oller and Eilers 1988; Kishon-Rabin et al. 2005; Schauwers et al. 2008; Ertmer and Goffman 2011). At the present time these tools are not fully developed to allow this kind of assessment, but there is hope that, individually and in combination, they will be useful in the near future.

Conclusion

New outcomes with ABIs in children show the power of brain plasticity. A complex, novel, and distorted pattern of neural activity presented by the prosthesis to the brainstem is capable of providing high levels of open-set speech recognition in some pediatric recipients. This observation is not only greatly satisfying to clinicians, but also important for neuroscience. Children without an auditory nerve cannot receive hearing from a CI. But some of these children can receive CI-like hearing from an ABI. Questions remain about the best etiology for the ABI and why some children show only sound awareness and simple discrimination with the ABI. More research is necessary to further develop the ABI device, fitting, and evaluation. But the results now show that an ABI can provide useful hearing to children who cannot benefit from a CI.

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Cochlear Implants as Treatment of Single-Sided Deafness in Children

16

David R. Friedmann, J. Thomas Roland Jr., and Susan B. Waltzman

Introduction

Since the implementation of newborn screening programs, detection of hearing loss has improved drastically but early detection of unilateral hearing loss (UNHL) in children remains a challenge. The prevalence of unilateral sensorineural hearing loss is estimated at 13 per 1000 newborns and 3 % of school-aged children (Niskar et al. 1998). Unilateral hearing loss can be progressive and often go undetected due to the presence of a normal hearing ear. Therefore, the recorded prevalence of UNHL in children actually increases with age. Unilateral auditory neuropathy spectrum disorder and cochlear nerve deficiency may also contribute to the diagnostic difficulty. Most of these children have isolated hearing loss, although in some cases the hearing loss may be associated with cognitive impairment.

The nature of the hearing loss may vary from a transient mild asymmetric loss as a result of a middle ear effusion to single-sided deafness (SSD). SSD refers to an asymmetric condition in which a patient has one ear with severe-to-profound sensorineural hearing loss with normal hearing in the contralateral ear.

Hearing loss may be congenital or acquired and is most commonly of genetic origin. Other causes include perinatal infections including meningitis, complications of prematurity, and head trauma. The most common finding in unilateral sensorineural hearing loss is related to temporal bone anomalies including enlarged vestibular aqueduct, cochlear dysplasia, or cochlear nerve deficiency. Nearly one-third of patients with UNHL have such temporal bone anomalies related to the inner ear structures (Laury et al. 2009; McClay et al. 2008; Simons et al. 2006; Song et al. 2009) though rates as high as 65 % have also been reported (Masuda et al. 2013). While many cases are congenital, progressive hearing loss has also been documented with a higher prevalence in older children (Song et al. 2009) and as such children presenting even with a mild unilateral hearing loss should be followed carefully and frequently.

Conductive hearing loss can also contribute to UNHL as a result of otitis media with effusion, tympanic membrane perforation, ossicular chain abnormalities, and microtia/aural atresia. Some of these conditions are temporary and/or may be corrected surgically or medically; although such conditions can have significant effects, the focus of this chapter is unilateral sensorineural hearing loss.

Impact of Unilateral Hearing Loss and the Binaural Advantage

Though it was once felt that unilateral sensorineural loss had minimal impact on communication development, since the 1990s studies have documented speech and language delays related to such a hearing loss (Bess et al. 1998; Bess and Tharpe 1988; Bovo et al. 1988). Lieu et al. conducted a longitudinal 3-year study that followed children aged 6–12 years with a longitudinal hearing loss to determine the natural history of this type of hearing impairment. They documented that elementary school-aged children with UNHL have significantly worse language comprehension, oral expression, and oral composite scores than their siblings with normal hearing (Lieu et al. 2010). They also found that 25 % of these children were identified as having poor academic performance (Lieu et al. 2012). This functional impact may be explained by increased listening effort by the single good ear in complex listening environments leading to fatigue and low self-esteem (Bess et al. 1998) and may lead to social embarrassment and isolation and impact on quality of life.

Delays in treating UNHL may contribute to the negative impact on communication development and may affect the potential benefits intervention has to offer. For example, studies

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evaluating whether a congenitally deafened ear is affected by time to introduction of auditory stimulation suggest a sensitive period during which one should intervene to prevent abnormal development of central pathways through cross-modal plasticity that may re-wire the auditory cortex for other sensory input. Using P1 latency of the cortical auditory evoked potential as an index of maturation of the auditory pathway, Sharma et al. demonstrated different waveform morphology in those implanted at an older age than those implanted before 3.5 years of age (Sharma et al. 2005). In addition to asymmetry in the auditory brainstem, there are cortical effects from USNHL as well. There is evidence from functional imaging studies that those with a unilateral profound hearing loss suffer sensory deprivation and resultant cortical reorganization from monaural stimulation that may inhibit development of contralateral pathways (Rouger et al. 2012) and changes in electrophysiologic parameters may not be reversed after a prolonged delay before treatment (Gordon et al. 2013). This may have implications regarding whether an ear should receive a cochlear implant (CI).

An understanding of the advantages afforded by bilateral auditory inputs provides a framework for understanding the available treatment options. The binaural gain is multifactorial and includes the *head shadow effect* which dampens the sound wave to the ear away from the sound source, resulting in a better signal-to-noise ratio on the "unshadowed" side while sound is attenuated on the opposite side, *binaural summation* which is the added benefit of two hearing ears rather than simply one, and the *binaural squelch effect* or "release from masking" which suppresses competing sounds by recognizing auditory signals stemming from different acoustic sources responsible for the so-called cocktail party effect. Lastly, amplitude and latency differences between the two receiving ears result in psychoacoustic specific interaural time difference (ITD) and interaural level difference (ILD) that aid in *sound localization*.

Treatment Options

Perhaps the least understood aspect of unilateral pediatric hearing loss is when treatment is indicated. Experience suggests that some children benefit from noninvasive interventions such as preferential seating in school or some form of assisted listening or sensory aid technology; however, determining optimal treatment and timing for a given patient remains a challenge. Treatment efficacy may be difficult to measure directly and some children overcome such deficits without intervention. While improvement in academic performance is an important measure, it may not be directly attributable to treatment, nor is it necessarily sufficient evidence that the child does not still have a functional impairment relating to their hearing loss that would benefit from treatment.

FM Units and Hearing Amplification

Amplification with conventional hearing aids is only likely to benefit children with mild-to-moderately severe hearing loss and not those with true SSD.

Preferential seating and the use of FM units in the classroom for school-aged children are beneficial because FM technology increases the signal-to-noise ratio for the child in the classroom setting. However, FM technology cannot restore hearing in the poorer ear or improve "hearing" in all listening environments. As this is the least invasive modality, it is often one of the first interventions taken with school-age children.

Contralateral Routing of Sound and Bone Conduction Implant Hearing Systems

Contralateral routing of sound (CROS) amplification devices are a viable option for patients with SSD. With CROS, a microphone is placed on the poorer hearing ear and transmits the signal to a receiver on the better hearing ear. Bone conduction devices also route sound to the contralateral ear of individuals with SSD. They provide transcranial amplification by transferring sound through bone conduction. Sound arriving to the poorer hearing ear is converted to vibrations by a sound processor and then transmitted through the skull to the better hearing cochlea of the opposite ear.

Bone conduction implant systems are most commonly used to address SSD, although most recipients have been adults. They consist of an internal implant that is surgically attached to the skull and an externally worn sound processor. The first bone conduction systems had a percutaneous abutment that connected the implant with the sound processor (Baha Connect System, Cochlear LTD, Australia, and Ponto System, Oticon Medical, Sweden). More recently transcutaneous systems have become available that rely on internal and external magnets to couple the implant to the sound processor (Baha Attract System, Cochlear LTD, Australia, and Sophono System, Medtronic, Ireland). Surgical implantation of these implants is approved by the Food and Drug Administration at age 5 years and older. For younger children, the sound processor may be worn with a headband ("softband"). A nonsurgical bone conduction hearing system specifically designed to address SSD was commercially available between 2011 and 2015 (SoundBite Hearing System, Sonitus Medical, California). This system consisted of a hearing device placed in the mouth against the teeth and an externally worn microphone. One significant advantage of bone conduction devices is that there is no risk of occlusion of the good ear as with CROS amplification. Percutaneous bone conduction implant systems that couple by an abutment have been shown to be effective in improving hearing in difficult listening situations in pediatric populations (Christensen et al. 2010).

Both the CROS and bone conduction hearing systems may have undesirable effects in certain listening situations including hearing in noise. For example, when noise is present to the ear with SSD, it may be routed to the better hearing ear, worsening the signal-to-noise ratio and making listening more difficult. This problem is likely part of the reason why there has not been widespread use of either CROS or bone conduction amplification in the pediatric population. More recently some manufacturers of bone conduction systems and CROS systems have incorporated advanced noise suppression features to mitigate this problem. For this reason, there is renewed interest in the use of CROS in the pediatric population with SSD. Further study of newer CROS and bone conduction systems is needed to measure and compare the benefit of these devices in both adults and children.

Although both CROS and bone conduction devices offer amplification, they lack the advantages of binaural hearing that require sound to arrive at each ear independently for the processing of timing and pitch differences to be integrated by the brain. They do not restore hearing to the affected ear and hence do not offer the benefits of binaural hearing. Though some clinicians have speculated that bone conduction systems might allow for perceived differences in sound quality reaching the cochlea to be perceived differentially, this has not been borne out. Recent studies have failed to show objective improvement in localization abilities and have demonstrated only modest improvements in difficult listening situations (Battista et al. 2013; Wazen et al. 2005). This is likely a result of auditory input from all locations being transferred to the single functioning cochlea. Still, quality-of-life assessments in children with SNHL using a percutaneous bone conduction implant system do suggest a perceived benefit (Christensen et al. 2010; Doshi et al. 2013).

Cochlear Implants

Cochlear implantation is the only treatment that has the potential to restore bilateral auditory input to patients with SSD. CI technology is far from restoring normal hearing. Not all the benefits of binaural hearing are realized in those with CIs for reasons that may include a frequency mismatch between electrical stimulation to electrode array inserted into the basal portion of the cochlea and acoustic stimulation in the contralateral ear. Still, a CI is the only available treatment option to address stimulating the deafened inner ear rather than routing the sound to the hearing cochlea. Because there is limited published information on cochlear implantation in the pediatric SSD population, to date adult data provide the rationale for pediatric cochlear implantation in this population.

Some of the earliest literature is from Van de Heyning et al. who first reported on the use of cochlear implantation in SSD as a tinnitus suppressant in patients with debilitating and intractable tinnitus (Van de Heyning et al. 2008). A follow-up publication focused on the subjective improvement of restored binaural hearing (Vermeire and Van de Heyning 2009). Arndt et al. (2011) compared a group of adults with SSD treated with what the authors termed "pseudo-binaural" hearing with CROS and bone conduction amplification compared with "real" binaural hearing afforded to CI users. Preoperatively, all patients were tested in an unaided condition and then, in a random order, given trial periods of CROS and softband bone conduction sound processor use. After testing in each condition, all patients received a CI. This study showed a demonstrable benefit in sentence comprehension in the CI group at the 6-month postoperative interval compared with the other treatment groups (Arndt et al. 2011). Additionally, localization ability in sound field was significantly better with CIs compared to performance in the other three conditions.

Hansen et al. (2013) reported on the results of cochlear implantation in patients with Meniere's disease who progressed to profound sensorineural hearing loss with one ear. They reported significant improvement in word and sentence scores, though the ability to localize sound in this cohort showed much more modest improvement (Hansen et al. 2013). Clinical trials are being performed to further study the efficacy of CIs in SSD patients. In summary, though limited, the current literature seems to support the concept that stimulation from a CI on one side can be integrated with acoustic stimulation from the contralateral side to enable auditory input to both ears in patients with SSD.

SSD Evaluation

Clinical evaluation of a child for cochlear implantation begins with a comprehensive audiologic evaluation. These findings as well as the identification of specific genetic causes from the family's history may dictate the specific medical work up to be undertaken (e.g., electrocardiogram for suspected Jervell and Lange-Nielsen syndrome). A full history to account for possible etiologies and risk factors for hearing loss should be investigated to help guide further diagnostic testing and assessment. This includes birth history, etiology of hearing loss if known, onset of severe hearing loss (be it congenital, sudden, or progressive), prior or current amplification use, and presence of other symptomology such as vestibular symptoms.

All CI candidates require temporal bone imaging by computed tomography (CT) or magnetic resonance imaging (MRI) of the temporal bone. High-resolutions studies using either modality identify abnormalities of the cochlear labyrinth. MRI is particularly valuable as it is the most sensitive modality to identify cochlear nerve deficiency, a very

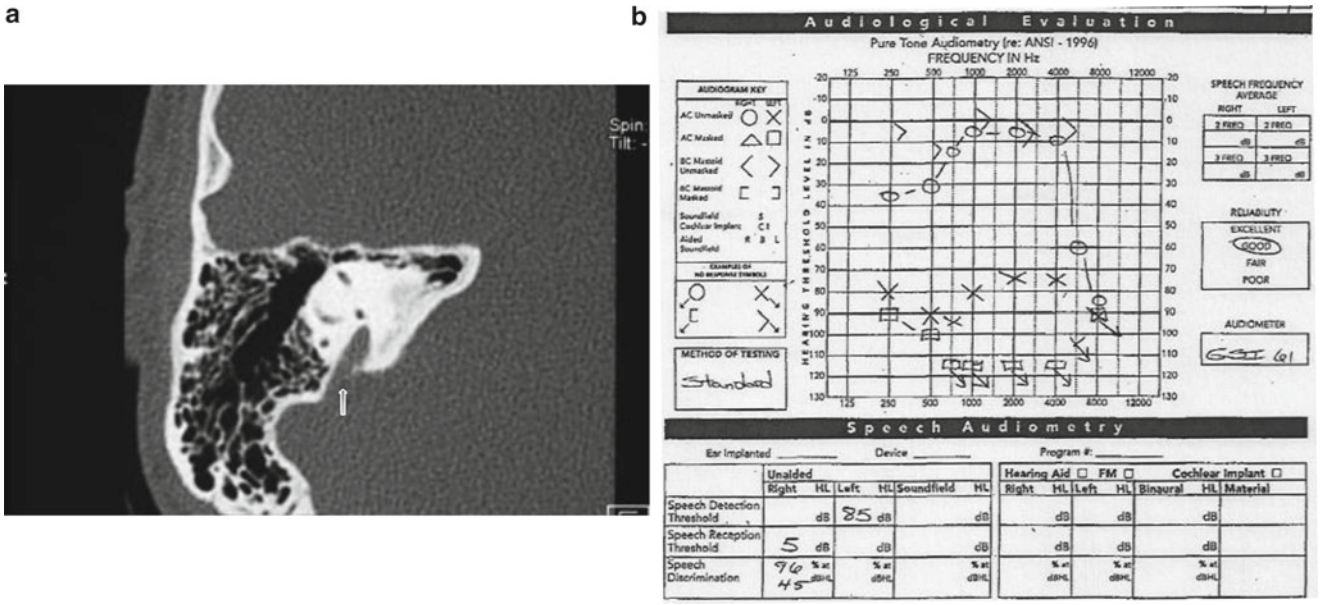

Fig. 16.1 (**a**) CT temporal bone axial section of right ear demonstrating wide vestibular aqueduct (*arrow*). This finding was present in both ears of this 10-year-old with a history of progressive unilateral hearing loss. (**b**) Audiogram demonstrating a left-sided profound hearing loss as part of her cochlear implant evaluation for treatment of single-sided deafness

unfavorable finding that may be fairly common in pediatric SSD. MRI is also preferable to CT for patients in whom labyrinthitis ossificans is suspected because it is more sensitive in determining cochlear patency. Imaging may also suggest the cause of progressive hearing loss by identifying congenital inner ear malformations, such as enlarged vestibular aqueduct. Implantation may be favored in these cases because of concern that patients will suffer progressive bilateral loss later in life (Fig. 16.1).

Although standard indications for cochlear implantation require that the patient not receive substantial benefit from amplification, the criteria for pediatric SSD candidates need to be approached differently. While candidacy criteria for cochlear implantation in this population remain to be defined, certain conditions may exclude a patient from further consideration. These include contraindications to surgery such as previously mentioned anatomic anomalies that preclude insertion of an electrode array. Unrealistic expectations of the patients and their families would be another critical consideration to address before proceeding.

Just as in any family with children undergoing evaluation for a CI, an important part of the preoperative counseling includes ensuring that patients and their families understand the range of possible outcomes as well as the considerable time and effort required for optimal performance with the device. Additionally, particular consideration should include discussion about subjective performance and progress over time, in addition to objective testing. An assessment of functional impairments may be more important than objective audiologic testing, most of which may be relatively normal with one hearing ear. These factors may be more important than the degree of hearing on the contralateral side. For those children who are of school age, one should inquire of the family whether they have noted difficulty in particular listening conditions, in social interactions, or in reports from teachers.

Another consideration is the very young child with SSD. With acknowledgement that some children with SSD grow up to be well-functioning adults and adapt well, these outcomes are difficult to predict. From the above literature, we have learned that the developing brain is at maximal neuroplasticity at a young age; a prolonged period of auditory deprivation may compromise ultimate auditory performance following treatment. At this point, our ability to predict individual benefits from implantation is limited. By analogy to adults, there are some adults who have lived with SSD without perceived difficulty, while others have found it challenging and no factors have yet been identified to know which patients fall into which group. It is important that the family understands all of these considerations when making the decision with the CI team.

Our institutional pediatric SSD protocol is listed in Table 16.1 and is adapted from our adult protocol. Pure tone air and bone conduction thresholds are performed using insert phones. Immittance measures including tympanometry and acoustic reflexes are performed along with otoacoustic emissions. Speech reception thresholds and speech discrimination are also determined using age-appropriate

Table 16.1 Institutional protocol for cochlear implantation in pediatric SSD patients

– Pure tone air and bone conduction thresholds
– Immittance measures including tympanometry and acoustic reflexes and otoacoustic emissions
– Speech reception thresholds and speech discrimination where age appropriate (CNC, HINT)
– Adaptive HINT is also done with sound field using CROS amplification
– Localization testing is done using a manufacturer-specific "direct connect" system
– Vertigo and tinnitus questionnaires are included in the evaluation

speech perception measures. The preoperative evaluation is performed using either a CROS hearing aid or the bone conduction sound processor worn with a headband that allows the child to be tested without undergoing surgery. In addition, localization testing post-implantation is done using a manufacturer-specific "direct connect" system. Direct connect testing allows direct delivery of sound to the sound processor. It allows for isolated testing of hearing in the implanted ear without providing any acoustic stimulation to the better hearing ear. It also permits sophisticated testing of sound localization outside of a sound booth specifically designed for this purpose. "Direct connect" assessment is expanding our abilities to specifically test for binaural advantages expected with cochlear implantation.

When related to tinnitus and vertigo, questionnaires are given to the parents. We have adapted our adult evaluation based upon the child's age with the unique considerations addressed above. The decision to proceed with implantation is ultimately a parental choice based on extensive discussions with the CI team. It is incumbent on the implant team to present the parents with all test results, treatment possibilities, and outcomes and to take into account educational, linguistic, and social development.

Cochlear Implant Outcomes in Children with UNHL

Because data on children with SSD who have received CIs are limited, it may be worthwhile studying children with asymmetric losses but not true SSD. One recent study detailed five implanted adolescents with asymmetric hearing loss (Cadieux et al. 2013). While the worse hearing ear was in the severe-profound deaf range, the better hearing ear (word scores 40–80 %) continued to use amplification and was not in the normal hearing range. Three of the five patients in this study had open-set speech recognition and significant bimodal improvement for speech recognition and localization compared to either modality alone following implantation. The other two subjects did not show bilateral speech recognition scores above those in the contralateral hearing aid-alone condition. Both of these patients had prolonged durations of deafness (12 and 15 years) and no hearing aid use. The impact of this duration of deafness may be a consideration unique to the pediatric population; to date, most published reports on adults with post-lingual SSD have included patients implanted after a shorter interval.

A recent publication focusing on three pediatric patients (ages 4, 10, and 11 years of age) with SSD who underwent cochlear implantation suggests a benefit in binaural integration and processing the combined electric and acoustic stimulation. These patients underwent open-set speech testing in background noise and localization tests 6 and 12 months after cochlear implantation. Additionally, subjective parental evaluation of the speech, spatial, and qualities scale (SSQ) was utilized (Hassepass et al. 2013). Enhanced sound localization abilities were demonstrated by reduced deviations in angle of error in identifying the sound source in the bilateral condition compared with the unilateral listening condition pre-implant. The children were still using their device consistently 1 year post-implantation.

There is one report of a patient with posttraumatic SSD in which expeditious implantation was done due to imminent fibrosis and ossification. Speech perception tests at 6 months demonstrated 90 % monosyllabic words and decreased angle-detection error suggesting improved sound localization (Plontke et al. 2013).

Similar results were observed in our own institutional experience which to date consists of three pediatric patients. S1 was implanted at the age of 10, is now 14 years old, and has enlarged vestibular aqueduct as depicted in Fig. 16.1. Preoperatively, S1 obtained 0 % on CNC words. Her score at the 1-year post-op interval was 18 % but dropped to 6 % by post-op year 3. Concurrently, there has been a progressive decline in performance in the non-implanted ear from 98 to 80 % at her most recent evaluation. S1 wears the device regularly and reports subjective benefit despite limited objective improvement. Evidence from our experience with SSD patients after cochlear implantation is that while the quality of the auditory percept may not be acceptable initially, as they lose hearing in the non-implanted hearing ear (as expected in cases of enlarged vestibular aqueduct), they begin to better integrate and interpret the CI signal.

S2 was implanted at the age of 6 and had PBK-word scores of 20 %; HINT-Q was 74 % and HINT-N was 46 % in implant-only condition at 3 months post-stimulation. Bimodal scores were 100 % showing that the signal was not being degraded by the addition of an electrical stimulus to the normal hearing ear. Interestingly, despite the apparent increase in performance, S2 only wears the CI in school and sometimes complains that it "bothers" the good ear.

S3 was 3 years old at the time of implantation. As of the 3-month postoperative evaluation, PBK-word scores were

96 % with the non-implanted ear alone and 32 % with the implant alone. In the bimodal condition the word score was 96 % attributable to the ceiling effect from having one normal hearing ear. Importantly, the combined signal did not cause a decrement in performance. On the sentence test, with noise-front S3 scored 100 % in the non-implanted ear, 70 % with the CI alone, and 100 % in the bimodal condition. The father reports that S3 no longer asks where sound is coming from and responds better to sound in general.

Overall, the children demonstrated varying degrees of open-set speech perception in the implanted ear and bilateral improvement in the presence of background noise. The parents and the schools report increased attention, higher grades (where present), a decrease in asking "what?", and importantly the children are no longer fearful of social situations such as playing with friends.

Discussion

To date, there are few published reports on the results of CI as a treatment for single-sided deafness in the pediatric population. While adults have achieved high levels of performance and appear to be satisfied with their choice, results with the small population of children who have been implanted are varied. Thus, issues remain regarding the recommendation of widespread use of implants as a treatment for SSD. Like many issues in medicine, it is the unknowns that preclude definitive recommendations. It is unknown in a given young child what the extent of his or her disability from SSD is likely to be during the developmental years or later in life. Similarly, as with standard CI patients, results are often unpredictable and variable. Unfortunately, attempting to clarify these unknowns introduces a paradox. Waiting until a child gets older may allow a better determination of the impact of the hearing loss on functioning and learning, but this wait introduces a longer duration of deafness, a negative relationship in predicting CI outcomes.

Issues remain. First, given the fact that many adults and older children with unilateral deafness have not received any treatment and are doing well, some parents are justifiably reluctant to have their children go through the implant process and be labeled as "special needs" when the child may do well without receiving any treatment. As mentioned earlier in the chapter, there are known benefits from binaural stimulation but it appears that some adults and children can overcome the deficits of unilateral stimulation. They develop normal language, do well in school and professionally, and have appropriate social interactions. To date, we are not able to predict who will perform well nor have we identified the factors that account for the differences. The lack of data often influences parental decision making and support for CI as a treatment for UNHL in children.

Secondly, as outlined earlier in this chapter, in addition to doing nothing, other surgical and nonsurgical treatment options exist including CROS amplification and bone conduction hearing options. Despite their differences, these devices all have one thing in common: they do not restore hearing to the deaf ear; rather, they route the signal to the good ear. While this may be helpful in certain listening situations, it does not offer the benefits of binaural hearing.

Another issue revolves around the amount of the hearing loss. While this chapter focuses on SSD, questions regarding degree of hearing asymmetry are relevant. What options are best for a given degree of unilateral hearing loss and when should treatment be considered? Most would agree that implanting infants for SSD is not supported by current data. Still, available CI data in pediatric patients suggests that developing neural pathways and the degree of auditory plasticity are affected by early stimulation; children diagnosed with SSD should be followed closely and evaluated for treatment as dictated by their behavior. For instance, is the child unable to focus or learn? Is he/she having behavioral issues? Parents and professionals will have to evaluate the consequences of an impoverished auditory system and decide how to proceed. A concern in school-aged children in particular relates to use or nonuse of sensory aids. It's important to determine at what ages a treatment is more likely to be met with opposition. Often younger children will be more accepting, so there is a need to predict which children need treatment and which intervention is most appropriate.

We know from CI studies of children and adults that certain patient factors will favor improved outcomes (e.g., shorter duration of deafness, age, cognitive issues, and patient motivation) and these should certainly be considered as part of the comprehensive evaluation. Advances in implant technology and programming methods leading to better outcomes may also influence the decision.

While criteria continue to be defined, CI candidacy for SSD is favored in patients with progressive conditions such as enlarged vestibular aqueduct, genetic conditions, autoimmune inner ear disease, ototoxicity, and certain metabolic diseases. Because the good ear is likely to decline eventually, reestablishing hearing in the poorer ear avoids the untoward sequelae of long duration of deafness and total auditory deprivation.

Despite the fact that these children might be mainstreamed in school, they may still be at a real disadvantage and struggling in academic, social, and developmental spheres. As such, a CI may allow them to integrate more easily and learn more effectively. With improvements in the accuracy and comprehensiveness of newborn screening programs, families of children with unilateral hearing loss will be more regularly confronted with questions related to when to seek treatment and which treatment to seek. The role of cochlear implantation for such patients is not yet clearly defined but,

to date, does offer these children the best opportunity for binaural hearing and should be considered an option. Studies are ongoing which will ultimately help to further clarify and define the role of cochlear implantation for children with unilateral hearing impairment.

Conclusion

Unilateral hearing loss in children can have a significant impact on linguistic and social development and academic performance. An informed discussion to include all available therapies and their respective advantages and disadvantages with the family and CI team is essential to the decision-making process. Early experience with pediatric CI recipients with UNHL suggests that cochlear implantation, with appropriate preoperative assessment and counseling and postoperative management, may offer these children the best opportunity to realize the benefits of binaural hearing.

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Part V

Maximizing Cochlear Implant Outcomes Learning

17 Elementary Cognitive Processes Underlying Verbal Working Memory in Pre-lingually Deaf Children with Cochlear Implants

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Despite advances in cochlear implant (CI) technology and early identification and implantation, unexplained variance is still observed in pediatric CI users' performance on a variety of speech, language, and higher level cognitive outcome measures (Geers et al. 2011). Based on research spanning more than a decade, we have identified verbal working memory as a basic, elementary cognitive process that appears promising as one of the core mechanisms of action underlying a portion of the variability observed in the speech and language outcomes in these children. Working memory is the ability to temporarily encode, activate, store, and maintain information for current and future information processing activities (Cowan 2001). Working memory serves as the "interface" or mental workbench between the sensory input and prior knowledge and experiences stored in long-term memory (Cowan 2001, 2008). As such, the efficient use of working memory is highly dependent on the quality of both the incoming sensory input and the rapid automatic access to long-term memory representations, such as linguistic knowledge.

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A major controversy in the field of working memory is whether information from all sensory modalities (e.g. visual and auditory) are stored together in a single, multimodal memory system (Barrouillet et al. 2007; Cowan 2005; Oberauer and Bialkova 2009), or whether information is stored in separate domain-specific storage components called the Phonological Loop and the Visuo-Spatial Sketchpad which respectively store auditory/verbal and visual/spatial information (Baddeley 1986; Baddeley et al. 1984). Our goal in this chapter is not to defend one class of models or the other, but rather to argue that working memory is a basic, core cognitive process necessary for the maintenance and manipulation of all forms of information in memory and is an inseparable component of all behavior assessments used to measure outcome and benefits following cochlear implantation. Moreover, we propose that information which is verbally coded holds a special status within working memory, regardless of whether one adheres to a multimodal or domain-specific framework (Morey et al. 2013). The special status of verbally coded material in working memory is a result of the extensive language experiences of typically developing, normal hearing listeners who also tend to be the subjects of interest in much of the basic working memory research literature. Pediatric CI users, who consistently demonstrate poor speech and language outcomes have less experience with language than otherwise healthy, normal-hearing peers (Geers et al. 2011). Thus, understanding how normal hearing children develop highly efficient verbally mediated working memory strategies is crucial to identifying specific working memory component processes that may be delayed or disturbed in many pediatric CI users.

Working memory involves first encoding sensory information from the environment, then maintaining representations of this information in a short-term memory store, and finally retrieving stable memory codes from long-term memory for use in a wide range of information processing tasks. In the remainder of this chapter, we consider *short-term memory* to refer specifically to the passive storage of information in immediate memory, while

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working memory includes short-term memory along with any active cognitive processing operations, such as recoding, chunking, or verbal rehearsal, which make use of the initial precategorical sensory information (see Cowan 2008 for a similar use of terminology). Our use of the term *working memory* differs slightly from the other researchers who reserve the construct of working memory for conscious, deliberate attentional control processes (Engle et al. 1999) or the involvement of the central executive (Baddeley and Hitch 1974). However, it is often unclear precisely when a task imposes additional processing demands on an observer. In normal hearing children, developmental changes have been observed in the relative contributions of the phonological short-term store, domain-general working memory processing, and lexical knowledge on a number of memory span and language processing tasks (Archibald 2013). Archibald (2013) found that in older children, variability in auditory digit span tasks was primarily accounted for by phonological short-term storage, whereas in younger children both phonological short-term storage and language processing contributed to digit span scores. We might similarly expect that auditory digit span, a task often considered to be a simple passive short-term memory task for older, typically developing, normal hearing children, would impose an additional processing load for deaf children who use CIs (Kronenberger et al. 2013). Thus, we will be less conservative here in discussing precisely what constitutes a working memory task than may be generally expected from the mainstream of research in human memory and cognition (Cowan 2008; Engle et al. 1999).

Working memory capacity, broadly defined, has been strongly linked to a wide variety of higher-level cognitive abilities in typically developing, normal hearing children; individual differences in working memory capacity contribute to the variance on measures of general intelligence and academic achievement (Engle et al. 1999), reading compression (Daneman and Carpenter 1980), and vocabulary development (Gathercole and Baddeley 1993). Moreover, both phonological short-term memory and additional working memory processing operations have been identified as core underlying factors which contribute to performance on a wide range of standardized outcome measures commonly used to assess language ability (Archibald 2013). Before discussing the three elementary cognitive processes of working memory—encoding, maintenance, and retrieval—in detail, we will first broadly describe verbal working memory delays observed in deaf children with CIs. We will also briefly summarize our current understanding of the relationship between verbal working memory and language development in normal-hearing children.

Working Memory in Normal-Hearing and Deaf Children

Our current research program has been focused on identifying the elementary foundational processing components of working memory that may be compromised in deaf children with CIs. We have argued in earlier papers that the deficits and delays that have been observed in CI users across a variety of working memory tasks are not elusive or idiopathic (Burkholder and Pisoni 2006; Conway et al. 2011). Instead, these delays and impairments are consistent with a large body of earlier theoretical and empirical research in the field of Cognitive Psychology and Cognitive Science and are natural byproducts of speech and language systems which developed under atypical circumstances. Identifying the locus of the sources of individual differences that contribute to speech perception and language by studying working memory dynamics, will not only allow for the development of novel targeted interventions for specific fundamental information processing tasks, but will also lead to better informed evidence-based assessment of the efficacy of cochlear implants and other sensory aids for hearing impaired children.

Working memory and its component processes contribute to the individual differences observed in children with CIs on a range of clinical speech and language outcome measures, as well as other neurocognitive measures such as the executive function abilities (Kronenberger et al. 2013). In a seminal study on auditory memory span in a group of 43 children who had received cochlear implants before 3 years of age, Pisoni and Geers (1998) found that Wechsler Intelligence Scale for Children (WISC) forward digit span, which requires verbatim reproduction of both item and order information from a spoken list of digits, was strongly correlated with several conventional endpoint speech and language outcome measures, including measures of open-set speech perception, speech intelligibility, and reading; these initial findings suggested a common underlying source of shared variance and a possible process-based neurocognitive mechanism of action linking these diverse behavioral measures together. Pisoni and Geers (1998) also found that the simple bivariate correlations with WISC forward digit span remained strong and significant even after removing variance associated with audibility as indexed by an independent measure of speech feature discrimination.

In a larger more extensive follow-up study, Pisoni and Cleary (2003) reported working memory delays and other disturbances in a group of 176 school-age CI users. These children were administered both the forward digit span and backward digit span (repetition of a number list in the reverse order of its original presentation) subsections of the WISC. Examining both the forward and backward digit

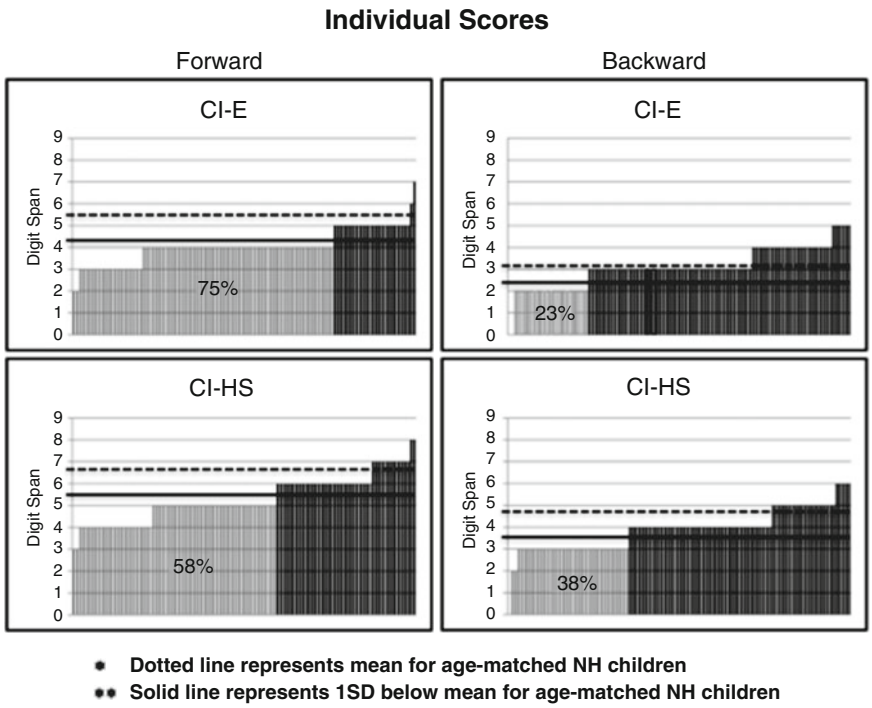

Fig. 17.1 Individual Forward and Backward Digit Spans in elementary school and 8 years later. Individual longest digit span scores at CI-E (age 8;0 to 9;11) and CI-HS (age 15;0 to 18;6) rank ordered from lowest to highest. The *top two panels* show the scores at CI-E; the *bottom panels* show the scores at CI-HS. The panels on the *left* show the forward span scores; the panels on the *right* show the backward span scores. The *horizontal dashed lines* within each panel show the mean obtained from the WISC III norms for typical-developing age-peers. The *horizontal solid lines* represent the scores that are 1 SD from the mean of the norm sample. The percentages shown in each panel represent the number of children who fell more than 1 SD from the mean of the norm sample (Pisoni et al. 2011)

spans, Pisoni and Cleary (2003) found that these deaf children with CIs recalled fewer digits than expected based on the published norms; moreover, they also recalled fewer digits in both forward and backward conditions than a comparison group of 45 age- and gender-matched normal hearing children. When tested 8 years later in high school, the absolute digit span scores of all CI users increased (Pisoni et al. 2011). However, once age differences were accounted for by comparison to age-based norms, the difference in forward digit span between CI users and their normal-hearing peers remained constant, and only 9 of the 112 CI users achieved forward digit span scores at or above the normed mean, despite 8 additional years of CI use (Fig. 17.1; Pisoni et al. 2011). Differences in backward digit span between the two groups actually increased (Pisoni et al. 2011). Performance on the backward digit span in the group of CI users suggested that these children were not developing the same cognitive control strategies during late childhood and adolescence as their normal-hearing peers.

Such failures to develop highly efficient cognitive and verbal working memory strategies implemented by normal-hearing children can have significant downstream effects on CI users' future speech and language learning. Pisoni et al. (2011) also found that measures of forward and backward digit span obtained during elementary school predicted high school performance on speech perception in noise assessed using Bamford-Kowal-Bench (BKB) sentences, receptive vocabulary measured with the Peabody Picture Vocabulary Test (PPVT), and receptive and expressive language measured using the Clinical Evaluation of Language Fundamentals (CELF). These findings are consistent with a large body of previous work on normal-hearing children, showing that the ability to rapidly encode, maintain, and retrieve robust phonological representations of spoken words in working memory is strongly linked to the development of more complex speech and language skills including vocabulary acquisition, syntax development, speech production, and reading comprehension (Baddeley et al. 1998; Gathercole and Baddeley 1993).

Working Memory and Novel Word Learning

One important role of verbal working memory in language acquisition is novel word learning. The phonological short-term memory store in Baddeley's (1986) multicomponent model of working memory is hypothesized to support the simultaneous maintenance of unfamiliar phonological repre-

sentations of novel unfamiliar words. Processing operations in working memory are used to decompose, reassemble, and combine individual phonemes into meaningful lexical units. The process of combining or *chunking* phonemes into robust stable lexical representations in long-term memory is assumed to be the foundation of early word learning (Baddeley et al. 1998). Using a longitudinal, crossed-lag correlational design, Gathercole and Baddeley (1993) were able to infer a causal relationship between working memory and language skills. Nonword repetition—an information processing task thought to isolate capacity of the phonological store unaided by long-term memory or other working memory processing—and receptive vocabulary were initially assessed starting at 4 years of age and then continued every year until 7 years of age. While vocabulary at age 4 was unrelated to vocabulary at age 5, nonword repetition skills at age 4 was found to predict unique variance in vocabulary at age 5, indicating that phonological short-term memory—not lexical knowledge—predicts vocabulary acquisition during early childhood. We have observed similar robust correlations between nonword repetition scores and speech and language skills in deaf children with CIs (Table 17.1; Carter et al. 2002; Cleary et al. 2002; Dillon et al. 2004a, b). More recently, we found that nonword repetition scores of these deaf children with CIs obtained at 8 years of age predicted a wide range of speech and language measures, including open-set word recognition, sentence recognition, speech intelligibility, and receptive vocabulary, at 16 years of age (Casserly and Pisoni 2013). Thus, both children with CIs and typically developing children with normal hearing rely on accurate and highly efficient rapid phonological encoding and storage during word learning and language development.

The primary difference in the findings obtained by Casserly and Pisoni (2013) and the earlier results reported by Gathercole and Baddeley (1993) is that in the CI users, nonword repetition predicted receptive vocabulary scores even 8 years later. However, once normal-hearing children reached 5 years of age, nonword repetition no longer predicted later gains in vocabulary knowledge. Instead, vocabulary predicted nonword repetition at the following test session. Gathercole and Baddeley's (1993) findings are consistent with the general claim that the relative contributions of phonological short-term memory and long-term lexical knowledge change throughout development (Archibald 2013). This body of work will be discussed more fully in a later section. For now, it is clear that in normal-hearing children, robust and highly efficient phonological processing and lexical encoding into short-term memory appears essential for early language learning. Increases in lexical knowledge also reinforces otherwise fragile, incomplete, and confusable phonological memory representations of spoken words and supports additional verbal working memory maintenance strategies which in turn lead to increases in short-term and working memory capacity.

Little is currently known about how a period of early auditory deprivation followed by experience with degraded and underspecified auditory input from a cochlear implant affects the developmental trajectory of working memory, particularly the reciprocal trade-offs observed between lexical knowledge and verbal working memory capacity. Our early findings suggest that CI users rely primarily on phonological short-term memory throughout a longer period of development (Casserly and Pisoni 2013). Initially, an early period of auditory sensory deprivation followed by degraded auditory input experienced by most CI users prevents the encoding of highly detailed phonological information into working memory, impairing early word learning skills. Because CI users lack the highly detailed phonological and lexical representations of their normal-hearing peers, they build lexicons with different organizational structures (Beckage et al. 2011; Kenett et al. 2013; Schwartz et al. 2013). Consequently, any working memory strategies which rely on efficient access to lexical knowledge would be compromised and may be atypical relative to the strategies that normal-hearing children routinely develop with fully specified input signals.

Table 17.1 Correlations between nonword repetition scores (for consonants correct, vowels correct, and subjective ratings from NH listeners) and several speech and language outcome measures (controlling for performance IQ, age at onset of deafness, and communication mode) in a group of cochlear implant patients

	Consonants ($N=76$)	Vowels ($N=76$)	Accuracy ratings ($N=76$)
LNT easy words	0.83***	0.78***	0.76***
LNT hard words	0.85***	0.71***	0.70***
MLNT	0.77***	0.74***	0.77***
Forward digit span	0.60**	0.62**	0.76***
Speech intelligibility	0.91***	0.88***	0.87***
Speaking rate	−0.84***	−0.81***	−0.85***
Word attach (reading)	0.75***	0.72***	0.78***
Rhyme errors (reading)	−0.63**	−0.68**	−0.54*

Data taken from Carter et al. 2002, Cleary et al. 2002, and Dillon et al. 2004a, b
*$p<0.05$, **$p<0.01$, ***$p<0.001$

Verbal Working Memory Strategies

Verbal working memory strategies can be used at any stage of information processing within the working memory system. During encoding, visual-spatial stimuli can be optionally recoded into stable phonological forms in immediate and long-term memory (Conrad and Hull 1964; Ford and Silber 1994). During maintenance, two language-mediated strategies—chunking and covert verbal rehearsal—can be implemented to create more robust representations: chunking occurs when prior long-term lexical associations are used to organize smaller linguistic units into larger units (Miller 1956); covert verbal rehearsal, the silent repetition of verbally coded information can be used to reactivate otherwise fragile and decaying phonological codes (Baddeley et al. 1975). During verbal recall tasks, the speaker must reconstruct the phonological code and articulate the semantic label before information is lost because of decay (Cowan et al. 1994, 1998). Although reconstruction and articulation themselves are not verbal working memory strategies, rapid and efficient response output processes are related to increases in verbal working memory capacities. Prelingually deaf, long-term CI users may take significantly longer to accumulate the necessary language experience to efficiently and consistently implement automatized verbal working memory strategies for encoding, maintenance, and retrieval of phonological information. Importantly, CI users' verbal working memory deficits are not specific to auditory input; CI users also show significant impairments when verbal information is presented visually, suggesting that their working memory system has been compromised beyond the extra processing load which might be encountered for recognizing degraded speech signals because of differences in audibility (AuBuchon et al. 2015a; Kronenberger et al. 2013).

We consider each of these component processes (phonological encoding and recoding, maintenance via covert verbal rehearsal and chunking strategies, and retrieval and reconstruction of phonological representations) one at-a-time. In addition to briefly describing the basic literature illustrating the use of these component processes in typically developing, normal-hearing children, we will also summarize relevant findings from our lab illustrating when deaf children with cochlear implants fail to efficiently utilize these verbal working memory strategies. We will also consider possible explanations of why CI users consistently display atypical working memory processes in a range of conventional behavioral tasks used to assess speech and language outcomes following cochlear implantation.

Stimulus Encoding and Phonological Recoding in Verbal Working Memory

Audibility is, of course, a very important consideration in early encoding of speech and spoken language. Hearing loss, background noise, and degraded, sparsely coded input signals from a cochlear implant all limit the amount and precision of the sensory information that can be encoded and maintained in active working memory during any given information processing task. It seems obvious that if an auditory signal cannot be reliably detected, it will not be reliably encoded, maintained, or retrieved by a listener. However, we wish to move beyond this obvious prediction and describe three possible sources of variability related to stimulus encoding which are particularly relevant to cochlear implant users, even under reliable encoding conditions in the quiet. Of the three sources of variability to be discussed in this section, the first—variability due to the perceptual similarity of the to-be-remembered items—is the factor most directly related to audibility and processing of degraded inputs from a cochlear implant. Even when a set of test signals can be reliably discriminated from one another, acoustically or visually similar items are simply more difficult to encode and maintain in short-term memory than perceptually dissimilar items. CI users are at a marked disadvantage compared to normal-hearing listeners in having access to highly detailed, perceptually distinctive episodic representations of these test signals, whether from current incoming sensory input or generated from their own lexical representations residing in long-term memory. The second source of variability comes from developmental and individual differences in phonological recoding operations. During phonological recoding, non-auditory signals are converted into verbal/phonological forms in working memory. The third source of variability reflects individual and developmental differences in the speed of stimulus encoding in memory which are present even during ideal encoding conditions.

Perceptual Similarity of To-Be-Remembered Items

The perceptual similarity of to-be-remembered items imposes variability into memory performance. We will begin by summarizing the literature which describes how normal-hearing listeners' memory performance changes as stimuli are manipulated to be more or less similar to other to-be-remembered items. Then we will elaborate our proposal that using a cochlear implant artificially increases the perceptual similarity among items, even when those items can be reliability discriminated from one another. Variability in the listener's ability to rapidly encode the fine acoustic-phonetic details of an auditory event and maintain highly detailed episodic representations of the early input in sensory memory or retrieve the fine acoustic-phonetic details of auditory stimuli from long-term memory should have downstream effects on memory performance.

Once encoded, some representations in working memory are more fragile than others because they are minimally specified and sparely coded. For example, the limitations of immediate memory for pure tones presented in isolation is well documented. When presented with a pure tone in isolation and asked to identify it from a small set of tones, perfect

performance can be accomplished up to a stimulus set size of about four to six tones for judgments based on pitch or loudness (Miller 1956). As the set of tones increases beyond this limit, identification performance asymptotes. Similarly, identification of unidimensional visual stimuli, such as colored squares, is very limited for judgments based solely on hue or brightness (Miller 1956). The limitations in absolute identification of unidimensional stimuli appear especially striking when compared to the fine-tuned sensitivity of the auditory system in detecting and discriminating between two tones based on frequency or intensity or the visual system in discriminating differences in hue or brightness (Miller 1956; Pollack 1953). High levels of discrimination performance suggest that items have been successfully encoded into working memory as echoic (for tones) or iconic (for squares) sensory representations; instead the limitation on absolute identification of unidimensional stimuli comes in perceptual processing during later maintenance and/or retrieval of those representations (Pollack and Ficks 1954).

Unidimensional representations of simple signals are very fragile because they are highly similar to one another, and are less perceptually distinctive and more easily confusable with each other (Miller 1956; Pollack 1953). Identification performance is much better for more complex, multidimensional stimuli which differ on several orthogonal perceptual dimensions (e.g. pitch and loudness) rather that when the stimuli differ by only a single set of perceptual attributes (e.g. pitch). Just as simple tones vary in perceptual similarity, speech sounds also vary in perceptual similarity. Using a four-alternative forced-choice reproduction task, Cleary (1997) demonstrated that sequence memory is poorer for perceptually similar isolated vowels than for dissimilar vowels. Cleary compared memory for sequences of isolated vowels that were near each another in the F1 × F2 perceptual vowel space ([ɪ], [ʊ], [ɛ], [ʌ]) to sequences of isolated vowels that were more dispersed in perceptual space ([i], [u], [æ], [a]). The relative locations of the vowels in perceptual space are shown in Fig. 17.2. So that no errors would be introduced in the reproduction task, subjects' responses were made by pressing one of four buttons. Prior to each block of testing, participants were presented with the four vowels in the set being tested in that block and were trained to map each vowel sound to a specific response button. Thus, participants could reliably discriminate among all the test sounds, indicating that individual vowels were successfully recognized and reliably encoded in both conditions. Nonetheless, when the vowels were presented in sequence, participants accurately reproduced longer sequences of dissimilar (i.e. far) vowels than sequences of similar (i.e. near) vowels. Cleary's findings on sequence memory for isolated vowels demonstrate that the negative effects of item similarity on working memory are not due to perceptual confusions during the early encoding process itself, but rather are due to the perceptual similarity of the representations of the vowels once they have been encoded and are being maintained in active working memory.

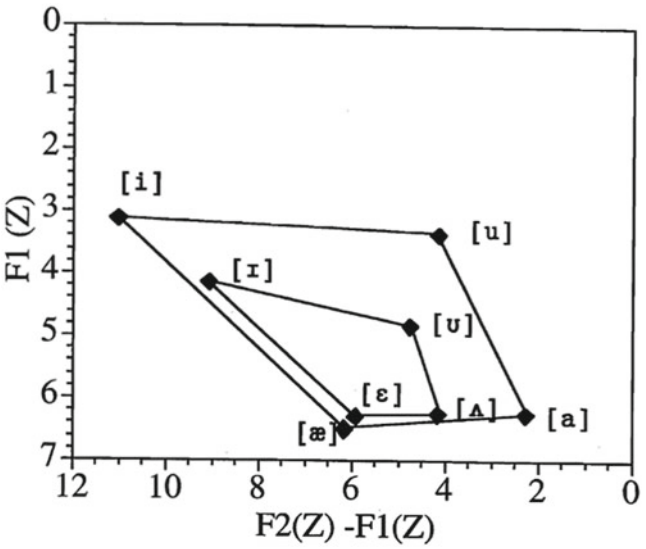

Fig. 17.2 Perceptual spaces for the vowel stimuli used by Cleary (1997)

Even when auditory signals are highly discriminable to CI users, the representational specificity as well as the fine acoustic-phonetic and indexical details of the auditory input typically heard by normal-hearing listeners may not be consistently encoded by CI users. CI users can recognize spoken words reliably in quiet testing conditions based on "coarse-coding" of the speech signal using broad phonetic categories, but their encoding of very fine sublexical details of speech and the indexical attributes of the vocal sound source are often compromised significantly compared to normal-hearing listeners.

Phonological Recoding

In normal-hearing listeners, item similarity is often exploited to assess the modality of internal representations and determine whether that representation's form differs from the form of the original stimulus. In all of the examples previously discussed, stimuli appear to be encoded into working memory in the same form that they were presented (e.g. a tone is initially encoded into an echoic sensory representation; a colored square is encoded into an iconic representation). However, as previously mentioned, the working memory system can also be used to rapidly *recode* input signals from their original forms into new, more stable memory codes. Phonological recoding occurs whenever a visual stimulus, such as a printed word, is recoded into a verbal or phonological code, as might occur during reading or when labeling a picture or object (Conrad and Hull 1964).

The Phonological Similarity Effect in Normal-Hearing Listeners

One of the classic demonstrations of phonological recoding in the study of human memory is the *phonological similarity effect*, a common finding in which lists of phonologically similar items (e.g. B, G, T, P) are remembered more poorly than lists of phonologically dissimilar items (e.g. F, H, Q, Y). Cleary's findings on the reproduction of vowel sequences was a good demonstration of the phonological similarity effect for auditorily presented isolated vowels. Importantly, phonological similarity effects for letters or words are consistently observed in adults even when the stimuli are presented via the visual, rather than auditory sensory modality (Conrad and Hull 1964). In this situation, the conventional interpretation of the phonological similarity effect is that visually presented input signals are rapidly and automatically recoded and represented in verbal short-term memory in terms of their phonological forms, which is a robust and highly efficient encoding strategy when the phonological representations are unique, but phonological recoding becomes less useful when those representations are more perceptually similar.

Auditory-verbal-linguistic input is automatically encoded into stable phonological representations in verbal short-term memory whereas visual input can be retained either as an iconic sensory-based visual representation or optionally recoded into phonological representations (Salamé and Baddeley 1982). The speed and efficiency with which someone can utilize automatized phonological recoding strategies is influenced by both developmental experience and other task demands. For example, when shown pictures of common objects, young children 3–5 years of age are able to remember more pictures if they have been instructed to name the pictures aloud, whereas older children 5–11 years of age did not benefit from the same explicit overt verbal labeling instructions (Ford and Silber 1994). In the labeling condition, the auditory form from the spoken verbal label is automatically encoded in memory as a phonological representation. However, in the silent condition, recoding from visual to phonological representations is optional. Because the older children performed equally well in both the overt and silent naming conditions, they likely utilized an automatic obligatory subvocal phonological recoding strategy even when they were instructed not to do so. However, Ford and Silber (1994) observed phonological similarity effects across all age groups, even when labeling was not instructed. Although the effect was larger in the older children, even children as young as 3 years of age recalled more pictures when the names of the objects were phonologically dissimilar (*dog*, *socks*, *ball*, *milk*, *fork*, *egg*, *bus*, *tree*) than when the names were phonologically similar (*car*, *cup cow*, *cat*, *key*, *clock*, *cot*, *cake*). Hulme and Tordoff (1989) reported similar phonological similarity effects in 4, 7, and 10 year-olds—although the effect increased with age.

Requiring the subject to engage in *articulatory suppression*, the continuous repetition of an irrelevant word such as "*the*" has been shown to interfere with routine phonological recoding strategies (Baddeley et al. 1984). When words or letters are presented visually, however, the phonological similarity effect disappears if the participant is also required to engage in articulatory suppression during stimulus presentation (Murray 1968). However, when stimuli are presented auditorily, the phonological similarity effect persists despite overt articulatory suppression because of the automatic phonological encoding of auditory-verbal input. Ford and Silber (1994) also reported that all of the age groups in their study performed more poorly when articulatory suppression was required, preventing phonological recoding. As with phonological similarity, articulatory suppression disproportionately harmed the oldest children. The findings that phonological similarity and articulatory suppression impair verbal recall in 3–5-year-olds—but not to the extent these manipulations impaired older children and adults—suggest that even the young children attempted to recode pictures of familiar concrete objects into stable phonological and lexical representations in memory but did so less efficiently than older children and adults.

The Phonological Similarity Effect in Deaf Children

Phonological similarity effects for visually presented materials have also been observed in deaf adolescents who use manual language (Conrad 1979), suggesting that some severely deaf adolescents who used sign language are able to optionally engage in phonological recoding, but with varying degrees of efficiency. In a large scale study of prelingually deaf 15–16.5 year-old adolescents who attended either a residential school for the deaf or a special program within a typical school, Conrad (1979) reported that both degree of deafness and speech intelligibility were important factors in predicting which deaf adolescents displayed these phonological similarity effects. Conrad (1979) found that a majority of adolescents with the most residual hearing (i.e. hearing thresholds below 85 dB) used phonological recoding strategies, although their normal-hearing peers were still more likely to utilize phonological recoding strategies. Moreover, those deaf adolescents with residual hearing who did use phonological encoding did so just as efficiently as their normal-hearing peers. The pattern changed for adolescents who had greater degrees of hearing loss; less than half of the adolescents who had hearing thresholds above 85 dB showed any evidence of phonological recoding strategies, and for those adolescents who did attempt phonological recoding, their efficiency decreased with increasing levels of hearing loss (Conrad 1979).

Speech intelligibility was also found to be predictive of which deaf adolescents used phonological recoding strategies (Conrad 1979). Phonological similarity effects were

observed in deaf adolescents who were classified by their teachers as having intelligible speech but not in deaf adolescents with the same degree of hearing loss who were classified as unintelligible. Conrad's (1979) findings can be interpreted in two different ways. It may be that pre-lingually deaf adolescents who have poor speech intelligibility also have poor, easily confusable inner speech. According to this account, these adolescents might attempt phonological recoding of visual material, but their internal representations are fragile, less distinct, and more perceptually confusable—even for those items classified as phonologically dissimilar under normal perceptual circumstances with hearing listeners. Another interpretation is that for some deaf adolescents, phonological recoding is a very difficult and inefficient processing strategy. These deaf adolescents may simply abandon any attempt at recoding visual input into a phonological form. Conrad's (1979) findings may also reflect a combination of both factors.

We have recently examined the use of covert phonological recoding (i.e. private speech) during completion of a variety of executive function tasks in a group of older children, adolescents, and young deaf adults who have used their CIs for at least 7 years as well as a group of age and nonverbal IQ matched hearing controls (AuBuchon et al. 2015b). The executive function tasks for our study were specifically chosen to minimize reliance on auditory input and audibility; thus almost all tasks used visual or visual-spatial presentation formats. For the normal-hearing sample, estimates of covert phonological recoding were found to be significantly related to almost all of the executive function tasks, suggesting that despite the visual presentation of the stimulus materials, typically developing, normal-hearing children, adolescents, and young adults automatically recoded the visual input into verbal-phonological forms in order to complete these tasks. However, for the cochlear implant groups, only the tasks which required a verbal output response (i.e., the Visual Digit Span test and the Retrieval Fluency subtest of the Woodcock Johnson) were related to estimates of covert phonological recoding. We suggested that, as a group, CI users are less likely to automatically implement phonological recoding strategies in these neurocognitive tasks. Instead, many of these CI users may have opted to maintain the visual-spatial representations of the test stimuli in order to complete the executive function tasks.

Perceptual Encoding and Phonological Recoding Speed

Typically developing, normal-hearing children consistently show individual differences and developmental variation in the speed of stimulus encoding, and these early encoding processes have been found to be closely linked to working memory capacity in normal-hearing children. In their well-known study, Case et al. (1982) estimated encoding speed in 3–6-year-old children by measuring their delay before beginning repetition of a single word heard from an audio recording. Older children were faster to initiate a spoken response suggesting that they had greater efficiency of early perceptual encoding and basic input-output operations. Importantly, the estimate of encoding speed obtained by Case et al. (1982) at each age was also related to a second, unspeeded task that was used to measure verbal working memory capacity. Greater encoding efficiency was found to be associated with larger estimates of verbal working memory capacity. In this study, individual differences in encoding speed were not a result of differences in audibility because Case et al. (1982) assessed only normal-hearing children.

To further remove any possible residual effects of audibility, Rapid Automatized Naming (RAN) tasks have been used extensively in the developmental literature to estimate encoding speed. In a RAN task, subjects are required to quickly and accurately name visually presented test items from a single category (i.e. digits, letter, colors, or objects). RAN tasks have been particularly useful in estimating phonological encoding speed in clinical populations. Results from RAN tasks have been shown to reliably distinguish between children with and without dyslexia (Denckla and Rudel 1974; Wolf et al. 2000). Children with dyslexia, who have difficulty with phonological encoding and recoding processes, are also slower on RAN tasks than typically developing controls (Denckla and Rudel 1974).

Recently, we obtained measures of perceptual encoding speed from a large group of long-term cochlear implant users ($N=57$) who completed two RAN tasks: (1) the *Digit Naming* task from the control condition of the Counting Interference Test (Hummer et al. 2011) and, (2) the *Color Naming* task from the control condition of the Stroop Color-Naming Test (Golden et al. 2003). When compared to a group of normal-hearing listeners matched 1:1 for age and nonverbal IQ, the CI users named nearly 20 % fewer digits in the Digit Naming task (83.40 named by the CI users and 99.6 named by the normal-hearing controls; $p<0.001$) and 14 % fewer colors in the Color Naming task (65.19 named by the CI users and 76.07 named by the normal-hearing controls; $p<0.01$). These new findings on perceptual encoding speed suggest that long-term CI users are less efficient than their normal hearing peers in encoding even highly familiar visual stimuli in a simple naming task with no other concurrent processing demands. The CI users' performance on Digit Naming, which is closely tied to phonological recoding efficiency (Christopher et al. 2012), suggests that when CI users carry out phonological recoding, they do so less efficiently than their normal hearing peers.

In addition to the group differences in phonological encoding efficiency observed between CI users and normal hearing controls in these two RAN tasks, we also found that for both the CI users and the normal hearing

Table 17.2 Correlations of perceptual encoding speed measures with speech, language, and executive function composite measures reported in AuBuchon et al. (2015b)

	Digit naming		Color naming	
	CI	NH	CI	NH
Speech perception composite	0.13	–	0.02	–
Language composite	−0.31*	0.002	0.19	0.09
Verbal WM composite	0.59***	0.55***	0.62***	0.61***
Inhibition-concentration composite	−0.64***	−0.76***	−0.67***	−0.82***
Fluency-speed composite	0.63***	0.76***	0.74***	0.77***

Note: *$p<0.05$, **$p<0.01$, ***$p<0.001$

controls, digit naming speed, and color naming speed were strongly related to forward auditory digit span as well as forward visual digit span, replicating Case et al.'s (1982) earlier results. Digit Naming and Color Naming were also strongly related to composite scores obtained from two other domains of executive functioning—Fluency-Speed and Inhibition-Concentration—in both groups of listeners (Table 17.2). Furthermore, for the long-term CI users, Digit Naming—but not Color Naming—was related to language skills as measured by the PPVT and CELF, a finding that is consistent with Christopher et al.'s (2012) proposal that digit naming, in particular, is sensitive to phonological recoding efficiency. These new findings on perceptual encoding speed in long-term CI users suggest that individual differences in encoding efficiency are an additional source of the variance in a variety of speech and language outcome measures routinely obtained from CI users.

Maintenance Strategies: Covert Verbal Rehearsal and Chunking

Short-term storage in verbal working memory is both time-sensitive and capacity-limited (Cowan 2001). Time limits refer to the length of time that items remain activated in immediate memory before their memory representations decay; capacity limits refer to the number of items that can be held in mind (Cowan 2001). Time and capacity limitations of verbal working memory can be reduced significantly by utilizing maintenance strategies such as covert verbal rehearsal and chunking. Covert verbal rehearsal is the silent repetition and reactivation of the phonological code of the to-be-remembered information in working memory. In contrast, chunking occurs when a subject utilizes associations in long-term memory to combine and integrate multiple individual cues and features into larger meaningful units (Miller 1956). Although other rehearsal strategies exist for maintaining visual and visual-spatial information in immediate memory (Logie 1995), verbal rehearsal is highly specialized for maintaining phonologically coded linguistic information in working memory and appears to be a more efficient than visual-spatial rehearsal strategies because it is more likely to be used in conjunction with other more sophisticated chunking strategies which are possible when long-term linguistic knowledge and prior experience is automatically activated in long-term memory (Morey et al. 2013).

Verbal Rehearsal Process

During verbal rehearsal, it is generally assumed that the phonological representations being maintained in verbal short-term memory are "reactivated" before they can decay, extending the time which they can be actively held in the phonological short-term store (Baddeley 1986). Verbal rehearsal is particularly useful for normal hearing young adults, and once initiated, can continue on with very little general attentional resources (Morey et al. 2013). Morey and colleagues (2013) reported a series of dual-task experiments in which participants encoded a set of visual items (colored squares) followed by a set of verbal items (auditory digits) or vice versa. Participants were then cued as to which set of stimuli—the squares, the digits, or either set—would be tested. Morey et al. (2013) found that memory for visually presented items was disrupted by encoding a verbal memory set, even when the cue indicated that only the visual set would be tested. Conversely, memory for verbally presented items was unaffected by subsequently encoding and maintaining a visual memory load, regardless of the later cue. This asymmetrical pattern of interference held up even when participants were disproportionally rewarded for correct responses in the visual task. One explanation Morey et al. (2013) provided for this dissociation was that initiating either verbal or visual rehearsal requires controlled attentional resources; however, once verbal rehearsal is initiated, verbal/phonological representations are likely to activate long-term memory traces and receive substantial downstream semantic support, making verbal rehearsal a more efficient and automatic maintenance strategy than visual rehearsal.

The asymmetrical advantage for verbal material described by Morey et al. (2013) has been observed repeatedly in studies of typically developing, normal hearing adults. However, automatic adult levels of verbal rehearsal do not emerge during development in an all-or-none fashion.

Instead, verbal rehearsal processes become faster and more efficient throughout development with increasing experience and knowledge of language (Gathercole and Adams 1994; Gathercole and Baddeley 1993; Jarrold and Hall 2013). Because covert verbal rehearsal is thought to occur at the same rate as overt speech production, temporal measures of articulation rate have been routinely used as a proxy to estimate verbal rehearsal speed (Baddeley et al. 1975). Two well-documented empirical findings in the human memory literature demonstrate the relation between articulation rate and verbal memory capacity.

Articulation Rate and Verbal Working Memory Capacity in Normal-Hearing Listeners

The first finding is the *word length effect* in which serial recall of visually or auditorily presented verbal items is better for lists of short words (e.g. fish, car, egg) compared to lists of long words (e.g. helicopter, kangaroo, umbrella; Baddeley et al. 1975; Hulme and Tordoff 1989). Similar to other traditional word span tasks, Baddeley et al. (1975) presented subjects with spoken word lists containing four to eight words. Following the final word in the list, subjects were asked to repeat the words in their serial order. Subjects could remember more sequences at each list length when short words—rather than long words—were used in the lists (Baddeley et al. 1975). This effect has been replicated using a wide range of verbal materials (Baddeley et al. 1975; Cowan et al. 1992; Hulme et al. 1986; Hulme and Tordoff 1989), and is consistently observed in children as young as 4-years-old (Gathercole and Adams 1994; Hulme et al. 1986). The explanation for the word length effect in immediate recall is that short words (i.e. words with faster overt articulation rates) will also be rehearsed more quickly, which results in the reactivation of more words before decay occurs (see however Cowan 1992; Lewandowsky et al. 2004).

The second well-documented empirical finding linking articulation rate to verbal working memory span was the discovery that individual differences in articulation rate predict verbal working memory capacity. Hulme and Tordoff (1989) estimated verbal rehearsal speed in 4-, 7-, and 10-year-olds by measuring overt articulation rate during a speeded word repetition task in which the child was taught a three word sequence of either short, medium, or long words and was instructed to repeat the sequence aloud ten times as quickly as possible. The articulation rate was calculated as the number of words articulated per second for each word length (i.e. short, medium, long). Memory span was determined by the mean number of words recalled, in order, from four-item lists (for 4-year olds) or six-item lists (for 7- and 10-year olds) constructed entirely with words of a given length (short, medium, or long). Overall, 4-year-olds had the slowest articulation rates and the smallest memory spans while 10-year olds had the fastest articulation rates and largest memory spans (Hulme and Tordoff 1989); moreover, for each age group, articulation rate slowed and memory span decreased as the words comprising the list got longer.

Hulme and Tordoff's (1989) seminal findings suggest that even children as young as 4-years-old utilize covert verbal rehearsal during verbal memory span tasks and that their slower rehearsal rate, as estimated by overt articulation, leads to poorer performance on memory span tasks. However, the relation between articulation rate and memory span is not consistently reported in the literature, especially in children younger than 7-years-old (Jarrold and Hall 2013). For example, Gathercole and Adams (1994) failed to observe such a relation in a group of 5-year-olds. In this study, children's speeded articulation rates were obtained for the set of nine digits as well as sets of one- and three-syllable words by having the children repeat a two item sequence from each stimulus set as quickly as possible. Gathercole and Adams (1994) only found a relation between articulation rate and memory span for the 5-year-olds when digits were used as the stimuli in both tasks (Table 17.3). Speeded repetition and memory span for one- or three-syllable words were not related in their 5-year-olds. Furthermore, the 5-year-olds also recalled more digits than one- or three-syllable words, suggesting that these children probably employed a specialized rehearsal strategy with the highly familiar digits that they did not use with spoken words. Gathercole and Adams (1994) proposed that children at this age were more practiced with repeating the nine digits together than they were with the particular sets of words. Thus, prior experience and processing activities with the specific stimulus materials used in the task may be necessary in order to reach levels of automaticity that make verbal rehearsal an efficient and effective maintenance strategy.

Articulation Rate and Verbal Working Memory Capacity in Children with CIs

In our initial studies of digit spans in children with CIs, we hypothesized that if covert verbal rehearsal speed contributes to the underlying source of individual differences in verbal working memory spans of normal hearing children, then it should also be an important factor in deaf children with cochlear implants (Pisoni and Geers 1998). Measures of perceptual encoding speed, such as speeded digit or speeded word repetition, were not obtained in this initial study. However, we did have access to each child's audio recordings of spoken sentences from the McGarr Sentence Intelligibility Test (McGarr 1981), a task routinely administered as part of the clinical CI test battery to assess speech intelligibility. Articulation rate was measured by calculating each child's average spoken duration of the seven-syllable McGarr sentences. As predicted, CI users had shorter digit spans and slower articulation rates than their normal hearing age-matched controls, indicating that the CI users had developed less-efficient verbal rehearsal strategies. Additionally,

Table 17.3 Correlations between measures of verbal rehearsal speed (VRS) and working memory span tasks for studies in which individual variation was reported

Group tested	*N*	Authors	Measure of VRS	Memory span task	*r* value
Deaf children with CIs					
8–9 years old	37	Burkholder and Pisoni (2003)	7-syllable McGarr sentence durations	WISC-III forward digit span	−0.52**
				WISC-III backward digit span	−0.63**
8–9 years old	176	Pisoni and Cleary (2003)	7-syllable McGarr sentence durations	WISC-III forward digit span	−0.55***
				WISC-III backward digit span	−0.42***
16 years old	112	Pisoni et al. (2011)	7-syllable McGarr sentence durations	WISC-III forward digit span	−0.42***
				WISC-III backward digit span	−0.14
7–25 years; >7 years CI use	57	AuBuchon et al. (2015b)	7-syllable McGarr sentence durations	WISC-III backward digit span	−0.14
				WISC-IV visual digit span	−0.27*
Normal hearing children					
5 years old	70	Gathercole and Adams (1994)	Rate to pronounce a pair of digits five times (digits/s)	Auditory Digit Span	0.30*
			Rate to pronounce a pair of 1-syllable words five times (words/s)	Auditory Word Span (1-syllable words)	0.05
			Rate to pronounce a pair of 3-syllable words five times (words/s)	Auditory Word Span (3-syllable words)	0.16
4.5 years old	16	Cowan et al. (1994)	Rate to pronounce a pair of words 10 times (words/s)	Cumulative memory span for word lists	−0.59*
8–9 years old	23				0.44*
8–9 years old	36	Burkholder and Pisoni (2003)	7-syllable McGarr sentence durations	WISC-III forward digit span	−0.37*
				WISC-III backward digit span	−0.04
7–25 years old	57	AuBuchon et al. (2015b)	7-syllable McGarr sentence durations	WISC-III backward digit span	−0.19
				WISC-IV visual digit span	−0.23a

Note: *$p<0.05$, **$p<0.01$, ***$p<0.001$. When rate is used as a proxy for VRS, faster articulation results in larger values. When duration is used as a proxy for VRS, faster articulation results in smaller values. Thus the signs on the correlations are expected to be reversed across these measures

articulation rate, a proxy for verbal rehearsal speed was also found to be strongly correlated with digit span in these young CI users (Table 17.3; Burkholder and Pisoni 2003; Pisoni and Geers 1998; Pisoni and Cleary 2003). Even after partial correlations were carried out to remove variance from word identification, language comprehension, and speech production, the partial *r* values remained significant, ranging from −0.29 to −0.49. These analyses provide support for the proposal that the relation observed between verbal rehearsal speed and working memory span in CI users is not due to limitations of audibility but reflect internal neurocognitive processes that are used to actively maintain verbal information in working memory (see Burkholder and Pisoni 2003).

In a follow-up study, Pisoni et al. (2011) found that the relation between verbal rehearsal speed and working memory span remained stable from elementary school to high school. Eight years after the first session of testing, 112 of the original 180 children described in Pisoni and Cleary (2003) returned for additional testing. Almost half of CI users tested in high school still displayed sentence durations that were on average longer than a group of age-matched NH controls, and as expected, high school measures of McGarr sentence duration and digit span were also strongly correlated (Table 17.3). More importantly, the McGarr sentence duration measured during elementary school ages was found to be a strong predictor of memory span 8 years later. McGarr sentence duration was strongly correlated with both high school digit span ($r=-0.50$) as well as improvement in digit span performance from elementary to high school ($r=-0.20$). Those CI children with faster verbal rehearsal speeds in elementary school showed greater improvements in digit span 8 years later (Pisoni et al. 2011).

More recently, we examined articulation rate, as estimated by McGarr sentence duration, and two digit span measures (backward auditory digit span and forward visual digit span) in another group of early-implanted long-term CI users, as well as a sample of normal hearing matched controls. Some of the CI users in this new study were among the first to receive multichannel CIs and are now young adults, making our study the first systematic study of these implant recipients after many years of experience using their implants. McGarr sentence duration was not related to auditory backward digit span in either group (Table 17.3). This finding is not surprising considering the processing requirement to reverse the order of the digits during recall which imposes an additional cognitive load, and may prevent or interfere with active rehearsal in both groups. In contrast, the relation between verbal rehearsal speed and forward visual digit span reached significance in the CI users and approached significance in the normal-hearing control group (Table 17.3).

The finding that verbal rehearsal speed is related to visual digit span in deaf children with CIs provides further converging evidence that the relation between articulation rate and digit span is not a spurious finding arising solely from the contribution of individual differences in audibility that influences both speech production and memory span. The results reflect limitations on how verbal information is processed and maintained in active working memory that is independent of input modality.

Chunking and Long-Term Memory Contributions

Just as covert verbal rehearsal can be utilized to extend the time limits of verbal working memory, chunking strategies can be utilized to extend its capacity limits. During the chunking process, multiple individual representations are combined into larger meaningful linguistic units by making use of existing associations in long-term memory (Miller 1956). Although a chunk's representation can exist in any form (e.g. visual, spatial, phonological), we suggest that special considerations should be given to verbal, or linguistically mediated, chunking strategies when studying the development of working memory span in children with CIs.

In his classic paper on the limitations of human information processing, Miller (1956) concluded that some experience is helpful, but that strong, automatic associations between long-term memory representations are essential to efficient chunking. Miller (1956) highlights the importance of experience and long-term memory in his original illustration of telegraph operators who routinely create larger chunks from simple patterns of *dits* and *dahs*. At first, telegraph operators can only recode patterns into single letters, but, with experience and practice receiving and sending Morse code, strings of *dits* and *dahs* can be recoded into meaningful words or even short sentences. Miller (1956) goes on to describe an experiment by his colleague, Sidney Smith, in which participants were taught to recode binary digits (e.g. randomly generated lists 0's and 1's) to the octal system. When participants trained for 10 min, only very slight increases in memory span for binary digits were observed using this chunking strategy. However, when Smith trained himself to recode from binary to octal digits to the point of automaticity, his own memory span increased proportionally to the recoding scheme used (e.g. in the 4:1 scheme, 4 binary digits *0010* would be recoded to a single octal digit, *2*; Miller 1956).

Miller's (1956) early conclusions have been supported by numerous studies of expertise in a variety of special populations. Expertise in a number of domains (e.g. chess, music, computer processing) improves performance on working memory tasks when the tasks use stimuli selected from those domains (Simon and Chase 1973). However, experience in any domain appears to selectively benefit only memory for stimuli in that domain. When asked to recreate the locations of pieces on a chess board, chess masters could correctly reproduce more pieces than even the Class A chess players who are considered the most highly skilled amateur players (Simon and Chase 1973). Importantly, however, the memory advantage of chess masters was only observed when the chess pieces were placed in a way to resemble a possible game of chess. The memory advantage of chess masters over players at other skill levels completely disappeared when the pieces were placed randomly on the board (Simon and Chase 1973). After thousands of hours of training and practice, experts use their long-term memory and knowledge of a specific domain to supplement the limits of working memory (Ericsson and Delaney 1999). Experts benefit during the early encoding stage of working memory because their familiarity with the specific content helps them to identify meaningful associations between items as well as detect subtle cues that will facilitate later retrieval (Ericsson and Delaney 1999).

Linguistic knowledge, like other forms of expertise stored in long-term memory, supports chunking in working memory. Participants in verbal working memory experiments are typically not viewed as "language experts," especially in tasks involving highly familiar materials such as digits or common words (but for an exception, see Ericsson and Kintsch 1995). Yet, participants' prior experience and lexical knowledge of a stimulus set has been shown to have strong effects on memory span (Hulme et al. 1991). Hulme et al. (1991) reported that adult native speakers of English had larger verbal memory spans for English words than for Italian words, presumably because English speakers already have stable robust long-term memory representations of the English words but not the Italian words. After learning English translations for the Italian words, memory span for the Italian words improved although it still was not at the same level as the English words; conversely, learning Italian translations for English words had no effect on memory span for English words. The English speakers' previous experience with the English words provided access to rich and highly organized lexical networks in long-term memory that were used to support the verbal memory tasks (Hulme et al. 1991). Learning the English translations of the Italian words created new long-term memory representations to support otherwise fragile and sparsely coded phonological memory traces, but these limited experiences did not result in the rich lexical networks needed to reach the level of efficiency obtained with familiar English words.

For typically developing, normal hearing children, linguistic experience and exposure to robust models of spoken language in their immediate home environment provides a foundation for the development of organizational and maintenance strategies in phonological memory from a young age. In a study using nonwords similar to the one discussed earlier, Gathercole (1995) assessed nonword repetition skills

in children at age 4 and again at age 5. At both time points, children heard nonwords that were all phonotactically plausible, but varied in adults' ratings of *wordlikeness*, or the degree to which they could pass for real words. At both testing sessions, children were more accurate at repeating nonwords with higher rating of wordlikeness, suggesting that even children as young as 4 years of age are able to use their limited linguistic knowledge of words they know in their native language to supplement phonological short-term and working memory for novel nonwords.

CI users also utilize linguistic knowledge in long-term memory, specifically, lexical knowledge and organization of the sound patterns of spoken words in lexical memory. When asked to repeat isolated spoken words CI users identify lexically "easy" words more often than lexically "hard" words (Kirk et al. 1995). Spoken word recognition, like nonword repetition, is a complex phonological memory task because it requires sensory encoding of the sound pattern, maintaining the phonological representation of that pattern in short-term memory, and then retrieving the lexical representation for a spoken response. However, CI users do not display the typical developmental trajectory in acquiring linguistic knowledge, so it should follow that they also do not follow a typical trajectory for utilizing that knowledge during working memory tasks like word recognition and nonword repetition (see Kirk et al. 1995; Casserly and Pisoni 2013).

Scanning and Retrieval Processes

A critical component of any working memory task is the retrieval and output stage. One retrieval process is *memory scanning*, the rapid serial search of a set of to-be-remembered-items in short-term memory during which the correct item for the current serial position is identified (Cowan 1992; Cowan et al. 1998). Short-term memory scanning, which can be measured by the pause durations between successively recalled items, has been found to be related to working memory span (Cowan 1992; Cowan et al. 1998). Because scanning time is very short—too short to involve explicit verbal rehearsal of all remaining items—scanning is assumed to be free of any phonological information (Cowan 1992). This interpretation is supported by findings showing that duration of memory scanning does not vary with word length; rather, short-term memory scanning time increases as the number of items on the to be remembered list increases (Cowan 1992; Cowan et al. 1994, 1998). However, in the case of verbal recall, after the subject scans and identifies an item in short-term memory, the subject must refresh the identified item's decaying phonetic code and construct an articulatory motor plan for recall and response output, both of these component processes rely on access to knowledge of the phonological structure of spoken words in long-term memory (Hulme et al. 1999). In the case of verbal recall, individual differences in scanning and phonetic implementation have been shown to contribute to the variability observed in working memory span (Cowan 1992; Cowan et al. 1994, 1998).

Pause Duration and Retrieval from Short-Term Memory

Cowan (1992) reported that 4-year-olds' verbal working memory spans were negatively correlated with their short-term memory scanning times, but only when scan times were obtained from a particular fixed list-length (e.g. three-word lists). He reasoned that the 4-year-olds who have faster, more efficient, scanning would also have larger spans than their slower-scanning peers. However, when scanning times were obtained from each child's maximum word span, the average interword pause lengths were roughly equal for all children. At their maximum span lengths, children with larger memory spans would be scanning more items between responses but would scan each item faster than children with smaller maximum span lengths who have fewer items to scan. Hence, children with different scanning rates will have similar interword pauses at span-length lists. Cowan et al. (1998) found that scanning time contributes to both individual and developmental differences in working memory span, indicating that this process also becomes more efficient over time.

In a seminal study on short-term memory scanning in deaf children with CIs, Burkholder and Pisoni (2003) found that scanning and retrieval speeds were much slower for 8–9 year olds with CIs than a group of normal hearing age-matched controls (Table 17.4). Using digital recordings of the verbal responses obtained during a standard WISC forward digit span task, Burkholder and Pisoni (2003) compared CI users to NH controls using three acoustic response

Table 17.4 Mean (SD in parentheses) interword pauses, individual articulations, and response latencies (s) of forward digit span reported by Burkholder and Pisoni (2003)

		Speech timing measure		
Hearing ability	List	Articulation duration	Response latencies	Interword pauses
Normal hearing	Three digit lists	0.56 (0.14)	0.63 (0.30)	0.16 (0.15)
	List limit	0.56 (0.18)	0.92 (0.61)	0.18 (0.16)
CI users	Three digit lists	0.53 (0.09)	0.77 (0.30)	0.43 (0.20)
	List limit	0.59 (0.13)	1.06 (0.57)	0.49 (0.28)

time measurements: (1) the response latency, time between the experimenter's final utterance and the child's initial utterance; (2) articulation rate, the average time taken to say a digit; and (3) pause durations, the average pause time between spoken digits in a recalled list. The two groups did not differ in response latency or articulation rate of digits during the recall task but they did differ in pause durations between recalled digits.

Replicating Cowan's (1992) earlier work, Burkholder and Pisoni (2003) found that when list length was held constant, either at three-item lists or four-item lists, CI users' pause durations were three times longer (450 ms) than their NH peers' pause durations (150 ms), providing support for the claim that CI users may not necessarily differ from normal hearing peers in working memory capacity, but instead differ in the speed of processing and retrieval of information within the working memory system. Using pause durations obtained from each child's maximum span length, the group differences still remained significant. This finding with the children who use CIs differs from Cowan's (1992) results in which children with different working memory spans displayed similar pause durations at maximum span length. Since this was a verbal recall task, the pauses between items in recall included both scanning time and phonetic implementation processes used for output. Thus, Burkhold and Pisoni's (2003) findings suggest that, in addition to slower verbal rehearsal speeds discussed earlier, CI users are also slower and less efficient at retrieving phonological codes of spoken digits from short-term memory, possibly because the initial phonological representations in memory were compromised and underspecified and therefore required more downstream support for phonetic implementation and response organization before the verbal response could be produced.

Speech Production and Retrieval from Short-Term Memory

It is possible that poor speech production skills may also contribute to CI users' lower working memory performance. Slower articulation during verbal recall may interfere with maintenance of later, to-be-recalled items because overt articulation of the current item during recall prevents rehearsal or reactivation of remaining list items, allowing them to decay (Cowan et al. 1992). At least some of the word length effect can be accounted for by articulation rate during recall (Cowan 1992; Cowan et al. 1992). However, we believe that the working memory impairments observed in CI users go well beyond any localized effects of audibility or speech production because we have found poor performance even when the stimulus materials are presented visually and responses are made manually (AuBuchon et al., 2015a). For example, poor implicit sequence learning was observed in a group of young CI users between 5- and 10-years-old in a working memory paradigm which utilized visual input and manual output (Conway et al. 2011). In this modified reproduction memory span task, CI users and normal-hearing controls saw a sequence of two to four colored squares (e.g. *red*, *blue*, *green*) on a computer screen. After the final item of the list, all four colors were displayed and the participant was asked to reproduce the list by touching the corresponding squares in their original temporal order. The sequence span task was modified so that, unbeknownst to the participant, during the initial portion of testing the order of the colors was probabilistically determined based on an artificial grammar (Conway et al. 2011). During the final portion of testing, implicit sequence learning was assessed by comparing performance on novel trials generated by the same previously experienced artificial grammar to novel trials generated by a second unfamiliar artificial grammar. Normal hearing controls reproduced more lists generated from the old grammar than from the new unfamiliar grammar, demonstrating that they had implicitly learned the underlying artificial grammar from the initial phase of testing. In contrast, CI users did not show any advantage for novel sequences generated by the previously experienced old grammar. Conway et al.'s (2011) results cannot be attributed to difficulties in perception or verbalization because the stimuli were visually presented and the responses were manual button pushes; instead CI users had difficulty encoding and retrieving covert sequential representations for these temporal patterns.

Verbal rehearsal, during which reactivation occurs in an item-by-item manner, supports maintenance of sequential order (Brown et al. 2000; Burgess and Hitch 1999). It is unclear whether less-efficient verbal rehearsal directly results in poorer learning of implicit sequential patterns as observed by Conway et al. (2011) or whether both poor implicit sequential learning and poor verbal rehearsal result from impaired brain-based timing signal of a system which developed without the early experience of sequencing auditory input (Conway et al. 2011). Regardless of which account is correct, it is clear that deaf children who experience the auditory and visual world via a cochlear implant show delays and disturbances in a wide variety of verbal working memory tasks, even when audibility and speech production are removed as potential limiting factors on performance. Differences also emerge when the stimulus materials are presented visually, eliminating audibility as an explanatory factor.

Effects of Experience on Estimates of Working Memory Span

Experience with a particular stimulus set or information processing task leads to greater efficiency during recoding, covert verbal rehearsal, chunking, and retrieval from short-

and long-term memory. Greater efficiency, particularly in utilizing chunking and retrieval from long-term memory, can have dramatic impacts on estimates of verbal working memory capacity. Here, we make a subtle distinction between *actual* working memory capacity and *estimates* of working memory capacity. Working memory capacity is limited by the number of chunks of information that can be maintained in the short-term store and by the amount of time that can pass before those chunks decay; however, a chunk is not limited in the number of individual item representations and associations contained therein (Cowan 2001, 2005; Miller 1956). Increasing the size of each chunk might change the overall amount of information held in working memory, but the number of chunks remains roughly constant (Miller 1956). Clinical measures of working memory, such as the digit span, assume that each item (e.g. digit) of the working memory task equals a single chunk. However, chunks are defined by the strength of the individual items' association in the participant's long-term memory, so it is the participant with their unique developmental histories who ultimately defines the size of the chunk.[1] Similarly, faster encoding, covert verbal rehearsal, and retrieval also increase the overall amount of information held in working memory, but the rate of decay remains roughly constant (Baddeley 1986). This logic leads to clinical estimates of working memory which reflect working memory capacity *with* the help of any recoding and maintenance strategies, such as chunking and covert verbal rehearsal, used during the working memory task.

Subject S.F. is a classic illustration of an individual who is able to form large chunks of information to increase his working memory capacity (Ericsson et al. 1980). With 230 h of intense laboratory practice, S.F. improved his digit span score from 7 digits to almost 80 digits. Ericsson et al. point out that S.F.'s absolute working memory capacity did not actually increase; rather the number of digits within each chunk increased. S.F.'s extended practice with this specific set of stimuli allowed him to adopt explicit mnemonic and organizational strategies that capitalized on S.F.'s existing long-term memory to extend chunk size. For example, when presented the digits *3-4-2-9*, S.F. recoded the individual digits into the larger chunk, "3 minutes and 49 point 2 seconds, near world record mile time," (Ericsson et al. 1980).

According to traditional methods of calculating digit span, S.F. would be given a score of four for recalling *3-4-2-9*, even though he recoded the four digits into one larger chunk. A hypothetical participant who was unable to similarly recode digits would have only one digit per chunk. If S.F. and our hypothetical participant could maintain the same number of chunks, but S.F.'s chunks consistently contained 4—rather than 1—digit, then S.F.'s working memory capacity would be estimated to be four times larger. In verbal working memory tasks, linguistic knowledge contributes to the efficiency of both chunking and covert verbal rehearsal. Thus, understanding the effect of experience-dependent learning on encoding, maintenance, and retrieval is both theoretically and clinically important to correctly interpreting and understanding the differences consistently observed between CI users and normal hearing peers on a range of working memory tasks.

[1]The role of the participant (or patient) in optionally utilizing maintenance strategies such as chunking is part of the impetus for our own conservative use of which tasks constitute a working memory task.

Summary and Conclusion

Working memory is a core foundational neurocognitive process that is an inseparable component of all behaviorally based information processing tasks, including all of the conventional endpoint product-based speech and language outcome measures traditionally used in clinical settings to assess benefits and track progress following cochlear implantation. As basic research on working memory progresses in this clinical population, we will be able to learn more about the complex interactions among speech perception, working memory, and long-term linguistic knowledge within the constraints of what we already know about working memory development and functioning in typically developing, normal hearing children and adults. These findings can then be used to develop better interventions for CI users who may be doing poorly. It is very clear from the findings presented in this chapter that rapid verbal coding has a special status within working memory and that verbal working memory strategies provide robust improvements to the basic limitations on working memory performance. The three primary information processing components of the working memory system—encoding, maintenance, and retrieval—play a critical roles in language development and lexical acquisition and, in turn, influence the efficiency of speech and language processing. For CI users, these three component processes have been found to be disturbed and impaired and are they are often atypical and delayed when compared to performance of normal-hearing typically developing age-matched peers on a range of conventional short-term and working memory tasks are routinely used to assess information processing capacity and efficiency.

The limitations of CIs place users at an immediate disadvantage during encoding of auditory information. The effects of degraded auditory input conveyed by a CI extend well beyond early auditory encoding and audibility of the signal to effect the quality, distinctiveness, and representational specificity of phonological representations and memory codes for speech and verbal information—even when they have been accessed from long-term memory. Phonological

recoding, which does not start out as an automatic process for normal hearing children but does become faster and more efficient with practice, may be more difficult for CI users to implement. Because phonological recoding is an optional strategy for visually presented materials, some CI users may only implement verbal coding strategies when required by the task, such as the need to produce a verbal output response. Finally, CI users, just as their normal hearing peers, vary in encoding and recoding speed. This source of variability likely contributes to the individual differences observed not only in working memory performance but also on other conventional speech and language outcome measures.

Rich lexical coding and a strongly interconnected lexical network support the maintenance strategies of covert verbal rehearsal and chunking which, to some degree, can be used to circumvent the capacity and time limits of short-term storage within working memory. Deaf children with CIs, like normal hearing children, can—and do—utilize these maintenance strategies in working memory tasks, although not as quickly or efficiently as normal hearing peers. Thus, when compared to published norms or age-matched normal hearing controls, the relative "language expertise" of the normal hearing peers should be taken into consideration as a limiting factor. Although the contribution of language might be discounted in basic research studies of working memory, typically developing, normal hearing participants are rarely, if ever, compared to a unique group of children with significantly less language experience such as prelinguistically deaf CI users.

Finally, variability in short-term memory scanning and retrieval time—which includes linguistically based phonetic implementation and reconstruction strategies in verbal recall tasks—influences performance on working memory span tasks in both normal hearing and implanted children. Planning and executing verbal output processes also temporarily delays recall, allowing more items to decay between responses. However, eliminating verbal recall does not fully account for the working memory deficits observed in deaf children who use CIs. The research findings presented in this chapter suggest that verbal working memory and the efficiency of its component processes provide a unified theoretically motivated account of the core information processing mechanisms of action underlying the variability in speech, language, and executive function outcomes in deaf children with cochlear implants.

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Working Memory Training in Deaf Children with Cochlear Implants

18

William G. Kronenberger and David B. Pisoni

Cochlear Implants and the Auditory Neurocognitive Model

Cochlear implants (CIs) provide access to auditory experience for many deaf children, allowing the potential for development of speech perception and spoken language skills. Much of the focus on outcomes following cochlear implantation has emphasized gains made by children in speech perception and spoken language skills, although considerable variation is present (Niparko et al. 2010; Geers and Sedey 2011). Within the past decade, there has been increasing awareness and evidence that a period of deafness followed by the degraded, underspecified auditory input provided by a CI may have a significant impact on the development of neurocognitive skills extending well beyond spoken language. The brain is an integrated, self-organizing system that develops based on transactional experiences between neural activity and stimulation from the environment, including auditory stimulation. As a result, deprivation in auditory experience may affect broad areas of cognitive functioning extending well beyond the proximal spoken language skills that are dependent on auditory experience. For example, auditory experience provides temporal patterns to the developing brain, which may be important in the development of domain-general sequential processing abilities ranging from pattern detection and memory for serially presented items to sustained attention to a sequence over time (Conway et al. 2009). The sequential input provided by auditory sensation may also provide opportunities for the developing brain to organize and integrate serial temporal information such as tones and sequences of speech sounds into organized holistic patterns and products such as words and sentences.

Kronenberger et al. (2012) proposed an Auditory Neurocognitive Model (Fig. 18.1) to explain potential links between hearing impairment and degraded/underspecified auditory experience from a CI and domain-general neurocognitive outcomes, particularly involving executive functioning. According to this model, auditory deprivation increases risk of deficits in three key areas of cognitive functioning that are critically important in the development of executive functioning and cognitive control: sequential processing, internalized use of language, and underspecified phonological and lexical representations of spoken language. Sequential processing is impacted by reduced experience to temporal patterns from auditory sources (Conway et al. 2009); internalized use of language is affected by language delays associated with reduced access to auditory experiences (e.g., despite marked improvement in speech and spoken language skills following CI, children with CIs are delayed, on average, in spoken language skills relative to normal-hearing controls); and underspecified neurocognitive representations of spoken language result from auditory experience that is poorly coded/specified, leading to difficulty differentiating and fluently processing the internal representations of spoken language (Pisoni et al. 2011). Executive functioning, in turn, is influenced by all three affected areas of cognitive functioning.

While there is no universally agreed-upon definition of executive functioning (EF), the construct of EF is typically

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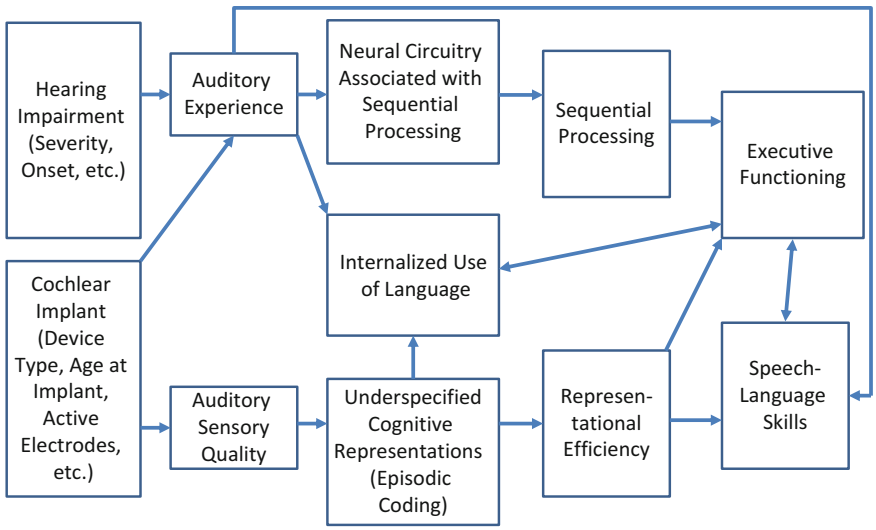

Fig. 18.1 Auditory neurocognitive model

depicted as the cognitive control and supervisory oversight processes necessary to carry out planned, purposeful, goal-directed activities. Components of EF therefore include controlled attention, working memory, self-monitoring, organization, goal direction (and resistance of distractions), and inhibition, all of which are necessary to evaluate, monitor, develop, and carry out plans and reach goals. Internalized language (Barkley 1997) and sequential processing (Conway et al. 2009) are core building blocks of EF because they support and promote control and goal direction over time. Cognitive efficiency, which is a combination of fluency/speed and information processing capacity based partly on robust cognitive representations that the individual can access rapidly, has also been shown to be a core component of EF (Cepeda et al. 2013). Thus, underspecified phonological and lexical representations resulting from auditory deprivation impact negatively on cognitive efficiency. Conversely, EF also influences performance on complex cognitive speed tasks because cognitive control (a central component of EF) is necessary for efficient processing of complex information that requires mental effort (Cepeda et al. 2013).

The results of several studies of children with hearing impairment and CIs provide support for the basic principles underlying the Auditory Neurocognitive Model. Figueras et al. (2008) report lower scores on several measures of executive functioning, including inhibition, planning, set-shifting, working memory, and attention, in children with CIs or hearing aids, relative to normal-hearing children. Parents reported more behavior problems related to executive functioning in children with CIs compared to norms (Beer et al. 2011). More recently, Kronenberger et al. (2013a) compared prelingually deaf, early-implanted, long-term CI users with a 1:1 age- and nonverbal IQ-matched sample of normal-hearing peers on measures of executive functioning in three areas: working memory, inhibition-concentration, and fluency-speed. CI users consistently scored lower than normal-hearing peers on measures of verbal working memory, inhibition-concentration, and fluency-speed, despite minimal demands on audibility or spoken language in the tasks used to measure these domains. Furthermore, differences were found on visual as well as verbally mediated measures of inhibition-concentration and fluency-speed, suggesting that differences in audibility are not responsible for the pattern of results.

In sum, EF is a multifaceted construct consisting of information processing domains ranging from attention to inhibition to working memory, which are used in the service of controlling cognition, affect, and behavior in order to carry out goal directed activity. The Auditory Neurocognitive Model hypothesizes that hearing impairment and degraded auditory experiences produce a downstream, domain-general effect on EF that is mediated in part by the direct negative effects of auditory deprivation on sequential processing, internalized use of language, and underspecified phonological and lexical representations. Empirical research supports the hypotheses of the Auditory Neurocognitive Model, demonstrating that children with CIs are at risk for domain-general (e.g., not solely dependent on audibility or auditory-verbal processing) delays in executive functioning in areas including working memory, inhibition-concentration, and fluency-speed. Hence, conditions affecting auditory experience such as deafness and exposure to the degraded auditory signals provided from a CI may put some children at risk for delays in general and specific areas of EF.

Working Memory: Definitions and Theories

Working memory is one of the core components of EF that may be impacted by hearing impairment followed by treatment with a cochlear implant, putting children with CIs at risk for delays and deficits. Working memory is the system

responsible for encoding, storing, and manipulating information during performance of other mental processing activities (Baddeley 2007). As a core component of EF, working memory is required for storing critical task-relevant information in order to maintain focus and response set so that optimal learning can occur and so that tasks can be completed. Working memory requires cognitive control of concurrent memory storage and information processing activities, in order to efficiently allocate limited cognitive resources to allow for both memory and information processing tasks to occur rapidly and efficiently in real-time.

According to one influential conceptualization of working memory, working memory consists of four components: a phonological loop (rote storage of auditory-verbal information), a visuospatial sketchpad (passive storage of visual information), a central executive (active, controlled attention and allocation of the resources of the working memory system), and an episodic buffer (component that accesses and integrates long-term memory with current information in working memory). The degree of involvement of each of the four components (and particularly the first three components) of working memory in a specific information processing task varies with the demands of the task: The visuospatial sketchpad and phonological loop are activated and accessed based on domain-specific demands in the visuospatial and auditory-verbal areas, respectively. Demands are greater on the supervisory central executive when cognitive load and simultaneous cognitive demands are increased.

Working memory is viewed as one of the elementary foundations of cognitive processing because it serves as a holding and processing area for information that is the focus of immediate attention and awareness (Baddeley 2007). The amount of information that can be held in working memory, and the individual's control over that information, form the basis of the cognitive content in immediate conscious awareness and attention that is available during information processing. Therefore, any deficits or disturbances in working memory may have significant downstream effects on cognitive, behavioral, and adaptive functioning.

Working Memory and Cochlear Implantation

According to the Auditory Neurocognitive Model, children with CIs are at risk for working memory delays because sequential processing deficits, language delays, and underspecified phonological and lexical representations affect the development of working memory as well as ongoing efforts to store and manipulate information in working memory. Working memory, particularly in the auditory-verbal-linguistic modality, requires processing of sequential speech and language patterns, both of which are adversely affected by a period of early auditory deprivation. Furthermore, processing of auditory-verbal-linguistic representations in working memory is less efficient as a result of the degraded, underspecified nature of those representations. Additionally, because the cognitive load involved in speech perception is much larger for children with a CI as a result of greater demands on controlled attention and mental effort to process the underspecified input from a CI, children with CIs have fewer cognitive resources (relative to normal-hearing children) to allocate for controlled attention and other information processing activities of the central executive during auditory-verbal working memory tasks.

A considerable amount of empirical research supports these hypotheses that deaf children with CIs are at-risk for working memory deficits. For example, children with CIs perform more poorly than normal-hearing peers on measures of learning and memory of implicit visual sequences (Pisoni et al. 2010). These implicit learning tasks require children to reproduce sequences of visual patterns, which follow a set of rules (grammars) that are not explicitly provided to the child. Children with CIs had shorter sequence memory spans than normal-hearing control subjects, and within the CI group, longer periods of auditory deprivation (e.g., later age at implant and longer duration of deafness) were related to poorer sequence memory spans. Other research has found that the visual memory spans and visual sequencing skills of children with CIs fall below average compared to normal-hearing controls (Conway et al. 2009; Pisoni and Cleary 2003).

Studies of auditory-verbal-linguistic working memory also demonstrate poorer performance in CI users relative to normal-hearing controls. CI users score below age norms on measures of auditory working memory such as digit span (Pisoni and Cleary 2003; Pisoni et al. 2008), which measures the child's ability to recall sequences of digits in either forward or backward order. Furthermore, the speed of retrieval of correctly recalled spoken digits by children with CIs is three times slower than that of normal-hearing children (Burkholder and Pisoni 2003). In recent studies of two separate samples of prelingually deaf children who received their CIs in early childhood (e.g., prior to ages 5–7 years) and who had used their CIs for extended periods of time (e.g., 7–10 years or longer), children with CIs scored well below normal-hearing controls on measures of working memory including digit span forward and digit span backward (Pisoni et al. 2011; Kronenberger et al. 2013a). Importantly, substantial differences in digit span forward remained between the groups even when the digits were presented in the visual, as opposed to auditory (spoken language), modality (Kronenberger et al. 2013a). Thus, differences in the audibility of test stimuli could not explain differences between the CI and normal-hearing samples.

In studies of the development of verbal working memory over time, Harris et al. (2013) found that mean age-based

growth curves of digit span forward and digit span backward in a large sample of children with CIs consistently lagged behind those of normal-hearing children throughout development up to age 16 years. While considerable differences were found in the individual growth curves over time, the mean growth curve of the CI sample lagged 0.5–1.0 standard deviations behind that of a normative sample. Kronenberger et al. (2013b) reported that 1/3 of their sample of children with CIs showed a very low baseline level of short-term verbal memory (based on digit span forward scores) and very slow growth of short-term verbal memory over time. A smaller, but still substantial, percentage of the CI sample (23 %) showed a similar very low baseline, low slope development of verbal working memory as measured by digit span backward scores. Thus, children with CIs are at risk not only for deficits in verbal working memory performance based on single time-point measures, but a substantial proportion of these children also show delayed growth in verbal working memory skills over time (Kronenberger et al. 2013b).

Working Memory and Language Skills in Children with Cochlear Implants

Working memory is an important component of executive functioning and has wide-ranging impact on individual learning and behavioral adjustment (Barkley 1997; Baddeley 2007). Additionally, working memory has particular importance for children with CIs because of its central role in language development, which is delayed (on average) in samples of children with CIs (Geers and Sedey 2011; Geers 2004). In normal-hearing and hearing-impaired children, working memory is also critical for the maintenance of phonological and lexical representations of spoken language during verbal information processing tasks. Working memory capacity is a critical determinant of the amount of auditory-verbal-linguistic information that the child can hold in mind during verbal processing and language-based learning. As a result, development of verbal working memory (both the phonological loop and the central executive) underlies and facilitates the growth of a broad set of spoken language skills including receptive and expressive vocabulary, comprehension, and speech production (Gathercole and Baddeley 1993).

Consistent with the importance of verbal working memory for language development, delays in verbal working memory are found in normal-hearing children with specific language impairment and related disorders of language and learning (Gathercole and Baddeley 1993). In samples of children with CIs, similar findings have been reported: Verbal working memory capacity is closely related to language development, even after accounting for conventional demographic and hearing history variables such as duration of deafness, age at implant, and number of active electrodes. Pisoni and colleagues (Pisoni et al. 2011; Pisoni and Cleary 2003) found significant correlations between verbal working memory measures using digit span and a wide variety of speech and language measures in children with cochlear implants. Speech perception, speech production, reading skills, verbal knowledge, and language skills were all strongly correlated with digit span, and significant correlations between digit span forward and speech perception remained even after controlling for demographic and hearing history variables (Pisoni and Cleary 2003). In a follow-up study using the same sample, Pisoni et al. (2011) found that the digit span forward scores of children with CIs obtained at about 8–9 years of age significantly predicted speech perception, vocabulary, and language performance after more than 10 years of CI use. Digit span backward scores predicted vocabulary and language scores 8 years later in the sample (Pisoni et al. 2011). Using a different sample, Harris et al. (2013) analyzed longitudinal data and found very similar results to the earlier findings reported by Pisoni et al. (2011): Early digit span forward scores predicted later speech perception and language scores, and early digit span backward scores predicted later language scores in children with CIs. Taken together, this body of research demonstrated not only cross-sectional, correlational relations between verbal working memory and speech-language skills in children with CIs, but also showed that early verbal working memory skills predict language performance longitudinally.

The prior studies showed that baseline verbal working memory skills measured at a single time-point in early childhood predict endpoint speech and language abilities in children with CIs several years later. Additional findings have also demonstrated that *changes* in verbal working memory over development (measured at multiple time points) predict both later endpoint speech and language performance in children with CIs and rate of development of speech and language skills over time. Pisoni et al. (2011) found significant correlations between improvement (i.e., differences) in digit span forward scores between ages 8 and 16 years and endpoint measures of speech perception, vocabulary, and complex language skills in children with CIs. Harris et al. (2013) replicated this finding, with a significant correlation between growth of digit span forward scores during development and endpoint scores on a measure of complex language skills. This relationship was present even after controlling for baseline digit span forward scores in the sample (Harris et al. 2013).

Kronenberger et al. (2013b) investigated the relationship between the developmental trajectories (as opposed to endpoint performance) of speech and language skills and verbal working memory. Different patterns of development of verbal working memory were related to the growth rate of speech perception, vocabulary, and language skills over time

(Kronenberger et al. 2013b). Hence, development of verbal working memory skills during childhood predicts concurrent development of speech and language skills as well as endpoint speech and language proficiency.

Executive Functioning, Working Memory, and Speech Language Skills: Summary

The Auditory Neurocognitive Model postulates that impoverished auditory experience causes proximal deficits in sequential processing and underspecified phonological and lexical representations that lead to distal weaknesses in executive functioning, speech, and language skills. Of the various components of executive functioning, working memory is particularly at risk because of its strong dependence on sequential processing and robust cognitive representations. Empirical research supports these hypotheses of the Auditory Neurocognitive Model, demonstrating working memory (particularly verbal working memory) deficits in children with CIs, which are independent of audibility of stimuli. These working memory deficits are significantly related to later development of speech and language skills in children with CIs, indicating that working memory is important not only in its own right but also as a mediator of speech and language development in the CI population. However, despite the potential importance of working memory for learning, adjustment, and speech-language development in children with CIs, few interventions have targeted working memory delays in this clinical population. Working memory therefore offers a highly novel venue for intervention to promote development in an at-risk area for children with CIs and to produce improvement in a heretofore understudied area underlying speech and language skills.

Working Memory Training Types and Programs

Working memory is of critical importance for broad domains of learning, development, and behavioral-emotional adjustment. Conversely, working memory deficits are central to disorders involving language acquisition/processing (Gathercole and Baddeley 1993) and executive functioning (Barkley 1997). As a result, working memory is a potentially fruitful target for novel interventions to address some of the core deficiencies of these disorders. In response to this potential, a variety of interventions have been investigated to assess their potential for improving working memory capacity and related executive functions in normal-hearing populations. These interventions range from physical and mental exercises to mindfulness/meditation to tutoring and educational interventions (Alloway 2011a, b; Diamond and Lee 2011).

One type of working memory training that has generated considerable enthusiasm and some scientific controversy is computer-based working memory training (WMT). WMT consists of video game-like memory exercises administered on a computer. The child must attend to stimuli on the computer, encode and store the information in memory, and accurately retrieve memory information in order to successfully complete the exercises (WMT programs also exist for adults, but the focus in this chapter will be on programs designed for children). WMT exercises for children typically use sequential spatial or verbal stimuli as targets for memory intervention and attempt to present the stimuli in entertaining fashion such as successive locations of a cartoon character or a code of numbers selected on a keypad. The task is for the child to hold some or all of the sequence information in working memory and produce a response demonstrating evidence of an accurate memory trace of the sequence.

Most WMT programs present stimuli using adaptive training algorithms, beginning with relatively easy tasks and progressing to harder tasks as easier tasks are successfully completed. Ideally, training difficulty is at the maximum information storage and processing capacity of the subject, increasing as the subject's capacity increases. If the child fails to complete tasks successfully, the difficulty level is adjusted and reduced. The number of tasks that must be passed or failed prior to modification of the difficulty of subsequent exercises varies from program to program and may be important for the efficacy of the WMT program (Gibson et al. 2013). WMT programs also vary in a number of other characteristics such as amount of required training time, type of stimuli (e.g., verbal vs. visual), type of working memory task (e.g., sequential memory vs. *n*-back), and demands for other concurrent cognitive processing during memory storage/recall (Gibson et al. 2012). Although many WMT programs consist of stand-alone training exercises, some are embedded within broader cognitive training programs that also attempt to improve attention, perception, processing speed, etc. (Rabiner et al. 2010). Examples of WMT programs include Cogmed, Brain Age, Posit Science Brain Fitness, Jungle Memory, WMPro, Lumosity, COGITO, Mindsparke, Brain Twister, and BrainTrain.

Two of the most widely used and researched WMT programs for children are Cogmed Working Memory Training (Klingberg et al. 2005) and *N*-Back Training (Jaeggi et al. 2011). The Cogmed Working Memory Training program is a 25-day program consisting of computer-based exercises that require the user to complete a series of memory tasks involving verbal, visual, or combined verbal and visual stimuli. Examples of exercises include reproducing a sequence of lights that illuminate one at a time on a 4×4 grid, remembering the order in which asteroids are lit as they move about on a computer screen, and remembering a series of single digit numbers in backward order. Because

the tasks are sequential and effortful, Cogmed exercises also demand the use of other executive functioning skills including sustained and focused attention, concentration, planning, and reasoning/problem-solving. Users complete Cogmed exercises at home for 30–40 min/day, 5 days/week, during a 5-week training period. On each training day, 8 WMT exercises are presented from among the 12 different Cogmed WMT exercises. The program uses an adaptive algorithm that presents users with problems of increasing difficulty at a level slightly higher than that at which they have recently achieved success. Cogmed exercises are completed on a computer at home or school, eliminating the need for frequent visits to a clinic during completion of the program. Users receive a phone call each week from a certified coach, who reviews program progress, assists with any problems, and encourages continued adherence to the program. Detailed information about daily performance on the program is sent to a secured internet site that can be accessed by the coach in order to monitor progress.

N-back WMT programs such as the one developed by Jaeggi et al. (2011) present stimuli in sequential order. Subjects are required to respond when a stimulus is the same as a stimulus presented *N* items earlier, where *N* typically varies from one to as many as ten items prior. For example, in a 2-back WMT program, subjects should respond (e.g., with a button press) when they see a stimulus that is the same as a stimulus presented two items prior in the sequence. Because subjects must keep *N* items in mind in WM in order to know whether to respond to each stimulus, larger values of *N* place greater demands on working memory. Furthermore, constant updating of memory is required, because items that were *N*+1 (or more) items before the currently presented item should be discarded from memory and replaced with the current test item. *N*-back WMT programs may present stimuli in more than one modality (e.g., visual and auditory), requiring subjects to simultaneously track two streams (modalities) of information, responding when a stimulus in either stream is the same as a stimulus presented *N* items earlier. These dual *n*-back WMT programs place even more demands on WM then the conventional (single modality/single stream) *n*-back WMT program.

Working Memory Training with Normal-Hearing Children

Efficacy Studies: Does Working Memory Training Work?

A growing body of research has investigated whether WMT programs can produce meaningful change in working memory capacity and related cognitive abilities. Studies in this area are heterogeneous, differing in the type of WMT evaluated, sample characteristics (age, cognitive delays, and clinical diagnosis), intensity of WMT, and outcome measures. Considerable enthusiasm has focused on WMT for children with working memory deficits, especially when these deficits are associated with clinical diagnoses such as Attention-Deficit/Hyperactivity Disorder (ADHD—a psychiatric disorder consisting of deficits in attention, concentration, organization, and in many cases behavioral self-control [impulsivity, hyperactivity]), learning disorders, and a range of speech, language, and communication disorders. Because deficits in working memory are a core feature of ADHD (Martinussen et al. 2005) and learning disorders (Gathercole and Pickering 2000), WMT has been suggested as a possible treatment to produce improvement in symptoms of these disorders.

Early WMT efficacy studies used small samples and often lacked appropriate control groups, as questions about feasibility and proof-of-concept were the primary goals of initial research. For example, Klingberg et al. (2002) assigned 14 children (age 7–15 years) with ADHD to either a treatment group who completed an adaptive WMT program or to a control group who completed the same exercises at a much easier level that never increased in difficulty (nonadaptive; e.g., remembering only two or three items at a time). The WMT group demonstrated significant improvement on measures of visuospatial WM (span board, a measure of memory for sequential spatial locations), fluid intelligence (Raven's Progressive Matrices), inhibition (Stroop Color-Word accuracy), and activity level (number of head movements) relative to the control group.

A benchmark WMT efficacy study was conducted by Klingberg et al. (2005) in order to address some of the limitations of prior studies. Using a multicenter, randomized, double-blind design, Klingberg et al. (2005) assigned 53 children with ADHD (44 of whom completed the study and were used in the analysis) to either Cogmed WMT or a nonadaptive placebo control condition with identical tasks presented at a very easy level (memory for two to three items) that likely did not present much challenge to WM. Outcome measures included near-transfer and far-transfer tasks. Near-transfer tasks are performance measures of WM that are not identical to the actual training exercises; far-transfer tasks are measures of abilities not consisting of WM performance but hypothesized to be related to WM. Near-transfer measures in the Klingberg et al. (2005) study included span board for visuospatial WM and digit span for verbal WM, whereas far-transfer measures included executive functioning tasks (Stroop Color-Word Test, Raven's Colored Progressive Matrices), a coded behavior task (number of head movements during a 15 min task, measured by an infrared camera), and behavior rating scales of everyday real-world behaviors completed by parents and teachers.

After the training period, subjects who received Cogmed WMT showed significantly greater improvement relative to the placebo group in span board, digit span, Stroop, Raven's, and parent ADHD rating (both inattention and hyperactivity/impulsivity) scores. No differences were observed between the groups on teacher ADHD ratings. Effect sizes for baseline-to-post-training change in the Cogmed group were in the medium to large range. The magnitude of improvement in the Cogmed group persisted at 3-month follow-up for most of these variables, although statistically significant differences between Cogmed and placebo were found only for the WM (near-transfer) and parent ADHD symptom ratings (far-transfer) at the follow-up visit.

The results of Klingberg et al.'s (2005) study have been partially replicated in several follow-up studies with the Cogmed WMT program, some of which used randomized, double-blind designs and some of which used unblinded/open-label (e.g., subjects were aware that they were receiving the intervention) designs (for comprehensive reviews of WMT studies, see (Melby-Lervåg and Hulme 2013; Epstein and Tsal 2010; Shipstead et al. 2012a, b; Morrison and Chein 2011; Klingberg 2010)). Holmes et al. (2009) investigated WMT outcomes in children with low WM in two schools, one of which was assigned to Cogmed WMT and one of which was assigned to a nonadaptive placebo control. Children completing Cogmed WMT showed greater improvement relative to controls on measures of visuospatial short-term memory ("short-term memory" was defined as tasks involving rote memory with little or no intervening additional cognitive processing required, as compared to "working memory" which involved memory concurrent with additional cognitive processing tasks), visuospatial WM, and verbal WM, as well as on a following directions task intended to be an analogue of an important classroom skill. No differences were found between groups for verbal STM, verbal intelligence, fluid/nonverbal intelligence, word reading, or math. Gains were largely maintained at 6-month follow-up, and an additional difference between the groups emerged in mathematics skills.

Green et al. (2012) found greater reduction in off-task behaviors in a classroom analogue task following Cogmed WMT compared to a control group in a randomized, double-blind, placebo-controlled design, although several other coded behaviors did not differ significantly between groups. The Cogmed WMT group also showed greater improvement on a verbal WM index but did not differ from the control group on parent-ratings of ADHD. Thorell et al. (2009) used a modified version of Cogmed WMT with preschoolers in a randomized, active placebo (playing video games), comparator (inhibition training with go/no-go, stop-signal, and flanker tasks), and wait-list-controlled study. Results showed significant gains on near-transfer tests of spatial and verbal WM and far-transfer tests of attention in the Cogmed group compared to the combined active placebo and wait-list groups (which did not differ from each other). Nutley et al. (2011) also studied Cogmed in a double-blind, placebo-controlled design with preschoolers (4 year olds) and found improvement in near-transfer WM skills but not in far-transfer reasoning skills. Other studies investigating Cogmed WMT using open-label designs have also reported improvements in near-transfer WM (Kronenberger et al. 2011; Beck et al. 2010).

Several studies of WMT using *n*-back training have also shown change in near-transfer and far-transfer skills following WMT. Jaeggi et al. (2008) conducted dual *n*-back WMT with a sample of young adults compared to a wait-list control and found greater improvements in WM and fluid intelligence in the WMT group compared to the control group. Furthermore, the gain in fluid intelligence was related to the time spent in WMT, suggesting a dose-response relationship. A subsequent study by Jaeggi et al. (2011) compared visuospatial *n*-back WMT to a knowledge training active placebo control task in a randomized design using a sample of typically developing (e.g., no diagnosis of ADHD or learning disability) children. Results showed no difference between groups at post-training on fluid intelligence, but children who showed the most improvement during the WMT task also showed the most improvement on fluid intelligence, relative to both the placebo and low-improvement WMT group. Other studies using *n*-back training have also reported improvement in near-transfer WM skills (Bäckman et al. 2011).

In contrast to the positive WMT results presented by the foregoing studies, some recent research has been less encouraging for WMT. Owen et al. (2010) conducted an entirely online study of over 11,000 volunteers who trained on two versions of a broad, internet-based cognitive training program that included (among other exercises) WMT, as well as a non-training, computer-use control group. Their results showed no evidence of near-transfer effects to reasoning, verbal short-term memory, spatial working memory, or paired-associates learning. However, Owen et al. had no in-person or phone contact with subjects to confirm or monitor adherence or outcome measurement, and the range of training sessions completed by subjects (1–188 training sessions) was very large. Redick et al. (2013) studied young adults (mostly college students) in a randomized, placebo-controlled trial of a dual *n*-back WMT program compared to an active placebo group who completed an adaptive visual search task and a no-contact control group. They found no significant differences among the groups on measures of fluid intelligence, multitasking, WM capacity, crystallized intelligence, or perceptual speed following training. The dual *n*-back training group self-reported more memory and intelligence changes than the other groups and were more likely to say that they changed the way that they carried out

their daily activities, but these self-reported changes were not reflected in the performance testing scores.

Redick et al.'s methodology differed from most other WMT methods in several ways (Redick et al. 2013): First, they emphasized aggregate scores for constructs such as WM and fluid intelligence, as opposed to individual test scores; their rationale for this approach was that aggregate scores better reflect the construct, which is the true target for improvement following WMT. Second, Redick et al. set their *p*-value for statistical significance at a more conservative $p < 0.01$, in order to reduce the chances of alpha error (inaccurately concluding that a difference exists between groups when such a difference does not exist); however, this increases the chances of beta error (inaccurately concluding that no difference exists between groups when such a difference does exist), particularly in small sample sizes. Third, Redick et al.'s two measures of WM emphasized memory under high concurrent cognitive load using either complex span (requiring a competing cognitive operation during the period in which the memory information is retained) or running span (requiring constant updating of working memory for a stream of information), consistent with findings reported by Engel et al. (1999) about the nature of WM. In contrast, many other WMT studies used measures of digit span or spatial span (span board) from classic clinical neuropsychology batteries.

Broad reviews that provide an overview of WMT efficacy research suggest that WMT typically has positive effects on the trained task and on near-transfer abilities, but as a result of limitations in the current body of research, there is less confidence about the effects (if any) of WMT on far-transfer outcomes. Melby-Lervag and Hulme (2013) conducted a meta-analysis of 23 WMT studies of participants 75 years or younger. They reported mean effect sizes across studies in the moderate to large range for near-transfer visuospatial and verbal WM skills immediately after WMT; this effect was diminished at long-term follow-up (an average of 5–9 months after post-testing) but still in the moderate range for visuospatial WM. The effect size for improvement in attention/inhibition following WMT fell in a small to moderate range immediately after training, while effect sizes for nonverbal reasoning, verbal skills, reading, and arithmetic were small. Long-term effects in these domains were also very small or negligible. Based on these findings, Melby-Lervag and Hulme concluded that WMT improved the trained tasks (e.g., the tasks that were completed in WMT) and near-transfer tasks (e.g., tasks involving WM, which were similar to those trained) but that there was little or no evidence for far-transfer effects to other tasks.

Epstein and Tsal (2010) reviewed a broader set of cognitive training programs and came to similar conclusions. They found clear gains on the trained tasks as well as on cognitive near-transfer tests, suggesting generalization to closely related cognitive functions. However, they noted mixed results about the efficacy of cognitive training on behavior as reflected in parent ratings, and no effect found for teacher ratings. Klingberg (2010), reviewing a smaller set of studies, found some evidence of near-transfer to WM in all reviewed studies, although about 1/3 of the studies found nonsignificant results for at least some WM measures. Evidence for far-transfer to reasoning skills (similar to fluid intelligence) was found in about half of the reviewed studies. Morrison and Chein (2011) concluded that near-transfer to WM abilities has been found in studies of Cogmed and *n*-back training, with more preliminary but promising findings for other cognitive abilities. They recommended caution regarding the endorsement of efficacy of WMT.

Shipstead et al. (2012a, b) have offered the most skeptical reviews of WMT efficacy, although their conclusions are broadly consistent with those of most other reviewers. They report studies showing consistent findings of improvement in tasks that are specifically trained as a part of WMT as well as frequent (but not universal) findings of near-transfer to WM tasks similar to those trained. However, they found little evidence for far-transfer effects to other cognitive abilities, reporting that findings for fluid intelligence are inconsistent at best. They also note that the evidence for improvement in attention following Cogmed WMT is stronger for sustained attention tasks than for selective or controlled attention (including controlling impulses) and that overall evidence for improvement in attention or ADHD symptoms is "sparse."

Neuroimaging Studies: Working Memory Training and the Brain

In addition to change in cognitive and behavioral outcomes following WMT, several studies have investigated changes in neurobiological functioning following WMT. Neuroimaging studies using functional magnetic resonance imaging (fMRI) have found changes in activity in regions of the prefrontal and parietal association cortex following WMT (Klingberg 2010; Dahlin et al. 2008; Olesen et al. 2004; Schneiders et al. 2011; Westerberg and Klingberg 2007). Interestingly, some studies report increases in prefrontal cortical activity after training, whereas others find decreases (Dahlin et al. 2008; Olesen et al. 2004; Westerberg and Klingberg 2007); this may be a result of interactions between different prefrontal regions (some of which increase in activity whereas others decrease) or may reflect an effect whereby WMT causes initial increases in activity during early or intense training, followed by decreases in activity associated with mastery. Positron emission tomography (PET) studies have shown changes in dopaminergic functioning in the prefrontal cor-

tex following WMT, although the specific dopamine receptors and systems involved have varied from study to study (Bäckman et al. 2011; McNab et al. 2009). Over all, the prefrontal and parietal areas affected by WMT overlap with those brain areas that are recruited during WM processing (Morrison and Chein 2011; Klingberg 2010).

Effects of Working Memory Training in Normal-Hearing Children: Summary

In summary, although empirical research, systematic reviews, and meta-analyses do not provide entirely consistent results about WMT efficacy, some converging trends are emerging. First, the most consistent finding is that individuals who undergo WMT show improvement on the trained tasks. While this finding indicates that some learning is taking place, it provides no information about whether this learning generalizes to other outcomes, which are the true goals of WMT. Second, there is fairly consistent evidence (with some exceptions) of near-transfer of WMT to untrained WM tasks administered immediately following WMT, suggesting that some improvement in performance on WM tasks is taking place (although there is disagreement about whether this actually reflects a change in WM ability). Third, some studies demonstrate generalization of WMT to far-transfer abilities, but other studies have found different results. Therefore, despite some promising results, there is disagreement among researchers about the generalization of WMT to far-transfer abilities such as fluid intelligence, attention/concentration, and academic skills. Finally, neuroimaging studies suggest that changes in brain functioning occur following WMT, in prefrontal and parietal areas that overlap with the brain regions recruited during completion of WM tasks; however, this research has not yet sufficiently investigated the long-term durability of these changes or their relationship with real-world behavioral outcomes.

In part, the limitations in conclusions that can be drawn about the far-transfer effects of WMT are a result of differing methodologies in WMT studies. WMT studies differ in the type of WMT used, the populations studied, and the methodology of outcome measurement. It seems unlikely, for example, that every type of WMT will have similar effects (Schneiders et al. 2011; Lucas et al. 2008), so there is some risk in aggregating studies using different types of WMT or in concluding based on one type of WMT that WMT in general is effective (or not) for far-transfer outcomes. Furthermore, there is no evidence that WMT should affect all populations (e.g., ages, diagnoses, cognitive ability characteristics) equally. As a result, the application of research using college students to research using preschoolers, children with ADHD, or deaf children with CIs is questionable. Additionally, there has been insufficient attention to subject factors that may be related to WMT efficacy, such as baseline WM skills, baseline executive functioning, and performance on the WM task. For example, while some studies report secondary analyses suggesting that baseline WM is unrelated to WMT efficacy (Jaeggi et al. 2008; Redick et al. 2013), others find more improvement in individuals with poor WM skills at baseline (Hunt et al. 2013). Finally, WMT studies use a wide range of outcome measures that vary from study to study; this complicates the comparison of results both within and across studies. This arises in part from the diversity of definitions of short-term and working memory, which is reflected in a diversity of measures of the WM construct that are sometimes only modestly correlated. Also, it is not uncommon to find significant improvement in some outcome measures and no difference in others within the same WMT study, despite the fact that the measures purport to assess the same underlying construct.

Working Memory Training for Children with Cochlear Implants: A Pilot Study

Rationale

Based on promising results of WMT research and the well-documented risks for poor working memory in children with CIs, our research team sought to investigate the feasibility and efficacy of WMT for children with CIs. The initial study emphasized feasibility and preliminary efficacy in an open-label design (e.g., not blinded, everyone receives WMT) with a small sample of children with CIs (see (Kronenberger et al. 2011) for study description and results, which are summarized in this section), in order to see if a more extensive randomized, blinded, clinical trial was justified. Although costly, double-blind, placebo-controlled studies with large samples are ideal, the use of pilot studies is a well-established first-step in the investigation of the efficacy of a novel treatment and can provide useful information about efficacy in the context of other studies (Gathercole et al. 2012).

Study Methods

Nine children aged 7–15 years were enrolled in the study (Kronenberger et al. 2011). Participants were required to have severe or profound bilateral hearing loss from birth and cochlear implantation prior to age 3 years. In order to document the need for improvement in working memory skills, participants' scores had to be at the 50th percentile (relative to norms) or worse on either a parent-completed behavior checklist of working memory (Behavior Rating Inventory of Executive Function [BRIEF]) (Gioia et al. 2000) or a measure of auditory digit span (Children's Memory Scale

Numbers subtest) (Cohen 1997). The 50th percentile was chosen as the cutoff (instead of a lower level of performance) because the demands of verbal working memory processing have been shown to be significant even for children with CIs who have average levels of cognitive ability (Pisoni et al. 2011). Participants scored in the upper end of the Average range on a measure of nonverbal reasoning (mean scaled score = 11.7, SD = 3.7, range 8–18; scaled scores have a population mean of 10 and population SD of 3).

There were four study periods and five assessment visits, with WMT occurring between the second and third visits: A waiting period of 2–5 weeks took place between an initial screening visit (Visit 1) and the pre-training baseline visit (Visit 2); then, a training period of 5 weeks occurred between the baseline visit (Visit 2) and the post-training visit (Visit 3); finally, a short-term follow-up period of 1 month was scheduled between the post-training visit (Visit 3) and the 1-month follow-up visit (Visit 4), and a long-term follow-up period of 5 additional months (for a total of 6 months after the post-training visit) took place between the 1-month follow-up visit (Visit 4) and the 6-month follow-up visit (Visit 5). At each study visit, participants and parents completed questionnaire and ability tests of program feasibility, working memory skills, and speech-language skills.

WMT was completed during the training period, which took place between Visit 2 (baseline visit) and Visit 3 (post-training visit). During this 5-week training period, subjects completed the standard version of the Cogmed WMT program, described earlier in this chapter, with weekly phone calls from trained coaches. A minimum of 20 completed Cogmed training days was required to remain in the study; all subjects were able to meet this requirement. During the other periods (waiting, 1-month follow-up, and 6-month follow-up periods), no WMT or other intervention was provided. Thus, the waiting period evaluated change in working memory during a period of no intervention, providing a contrast with the training period; administration of working memory tests at the beginning and end of the waiting period also allowed us to evaluate practice effects on the tests prior to initiating WMT. The follow-up periods allowed for the evaluation of the durability of WMT effects over a 1–6 month period following training.

Primary assessment measures for the study consisted of feasibility measures, working memory measures, and sentence repetition measures. Three types of feasibility measures were used: Cogmed performance information (uploaded daily from each participant's WMT sessions to the Cogmed internet site), a Program Feasibility Questionnaire (PFQ) completed by parents, and a Coach Rated Adherence and Motivation Scale (CRAMS). A performance improvement value was calculated to measure improvement in each of the 12 Cogmed WMT exercises; this value corresponded approximately to the increase in span length and difficulty from the second training session (allowing for adaptive increase to the child's maximum span at the start of training) to the end of training (highest score for the final five training sessions). Two aggregate performance improvement values were then obtained by taking the average performance improvement scores for the auditory-verbal exercises (exercises involving auditory-verbal memory information, either with or without a concurrent visual component) and the visual-spatial exercises (exercises involving visual-spatial memory information only). PFQ items requested information about challenges, problems, and satisfaction with the training program, rated on a 1 (strongly disagree) to 5 (strongly agree) scale with 3 = neutral; several PFQ items specifically targeted issues that might arise for a CI user, such as difficulty hearing or understanding the WMT exercises. CRAMS items (missing for two subjects) were rated on a 1–10 scale with 1 = not at all, 4 = somewhat/some of the time, 7 = much/much of the time, and 10 = very much/very much of the time.

Near-transfer verbal and visuospatial WM were measured with digit span tests (repeating sequences of single-digit numerals in either forward or backward order) and spatial span tests (reproducing sequential locations shown by the examiner on a span board, in either forward or backward order), respectively. In order to reduce training effects, different versions of the Digit Span and Spatial Span tasks were used at each of the first four visits (because there were only four versions of each of these tasks, the version from Visit 2 was reused at the 6-month follow-up visit). At the screening visit, subjects completed the normed versions of the digit span test (Numbers subtest from the Children's Memory Scale) and spatial span test (Spatial Span subtest from the Wechsler Intelligence Scale for Children, Fourth Edition, Integrated [WISC-IV-I]) (Wechsler et al. 2004) from well-established, existing test batteries. For subsequent visits, alternate versions of these memory span tasks were created using a random number generator, with the stipulation that each digit or location could occur only one time per item.

These three alternate versions of the span tasks were administered to subjects in a counterbalanced order at Visits 2–4, such that three subjects completed each version of the span task at each visit. Therefore, each subject completed all three versions of each span task during Visits 2–4, and the versions of the span tasks were evenly distributed across Visits 2–4 (e.g., each span task was completed by three subjects at Visit 2, at Visit 3, and at Visit 4). Repeated measures analyses of variance (ANOVAs) of the three alternate versions of the span tasks completed by subjects at Visits 2–4 showed no difference in performance, demonstrating that the alternate versions were equivalent in difficulty: digit span forward ($F(2, 16) = 0.76$, $p = 0.48$), digit span backward ($F(2,16) = 0.64$, $p = 0.54$), spatial span forward ($F(2,16) = 1.41$, $p = 0.27$), and spatial span backward ($F(2,16) = 0.52$, $p = 0.61$). In order to further test the equivalence of the digit span alternate versions, we also analyzed data from a group

of 53 healthy, nonreferred, normal-hearing adult males who were each randomly assigned to complete one of the digit span alternate forms ($N=15$, 19, and 19 completed each of the three alternate forms) at the first visit of a different study. Results of a between-groups ANOVA also showed no difference between groups on digit span forward ($F(2,50)=0.17$, $p=0.84$)) or digit span backward ($F(2,50)=0.72$, $p=0.49$).

To obtain measures of far transfer, parents completed the BRIEF for information about executive functioning behaviors in the everyday environment (the BRIEF Working Memory subscale was used as a measure of parent-rated working memory behavior at home), and participants completed the Sentence Memory subtest from the Wide Range Assessment of Memory and Learning, Second Edition (WRAML-2) (Adams and Sheslow 2003) as a measure of speech perception and sentence memory. Because there are no alternate versions of the BRIEF or WRAML-2, parents and subjects completed the same versions of these tests at each visit. The WRAML-2 Sentence Memory subtest was administered only at baseline, post-training, and 6-month follow-up sessions in order to reduce practice effects. Raw scores were used in analyses of all near-transfer and far-transfer measures.

Feasibility of WMT in the CI Sample

Almost all subjects (89–100 % of the sample) improved in every one of the specific Cogmed WMT exercises. All subjects showed positive performance improvement values on the global measures of auditory-verbal-linguistic exercises and visual-spatial exercises (Fig. 18.2), reflecting better WM performance on the Cogmed exercises at the end of training relative to the beginning of training. The improvement in auditory-verbal-linguistic exercises and visual-spatial exercises was significantly greater than 0 ($t(8)=6.05$ and 9.54, respectively, $p<0.001$), showing that improvement in the trained tasks took place.

Parents' responses to the PFQ showed that participants had no difficulty hearing or understanding the WMT exercises (Fig. 18.3). However, a majority of parents reported that considerable effort was required from both child and

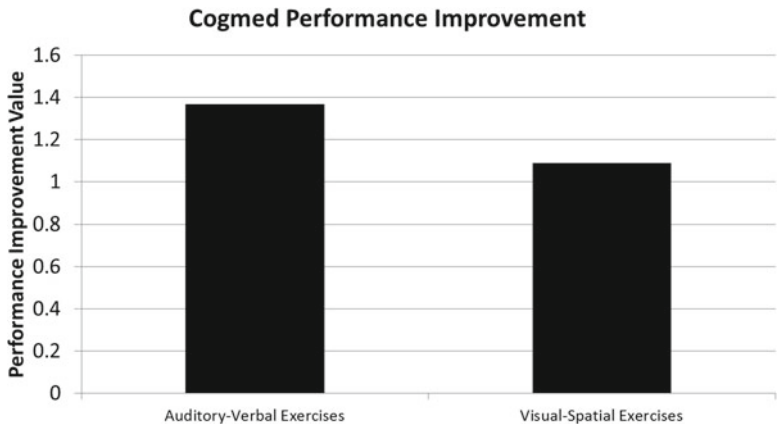

Fig. 18.2 Cogmed performance improvement during training

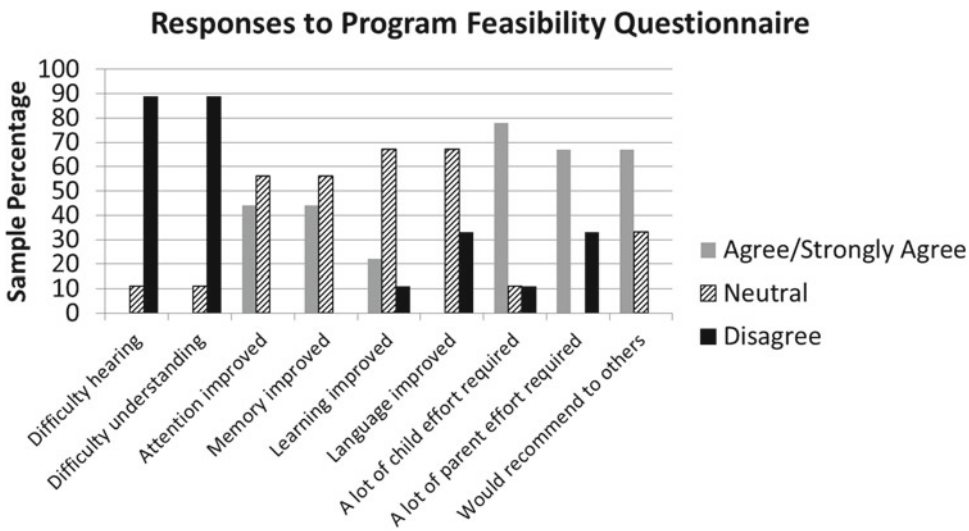

Fig. 18.3 Parent-report of working memory training feasibility. *Note*: Based on data from Kronenberger et al. (2011)

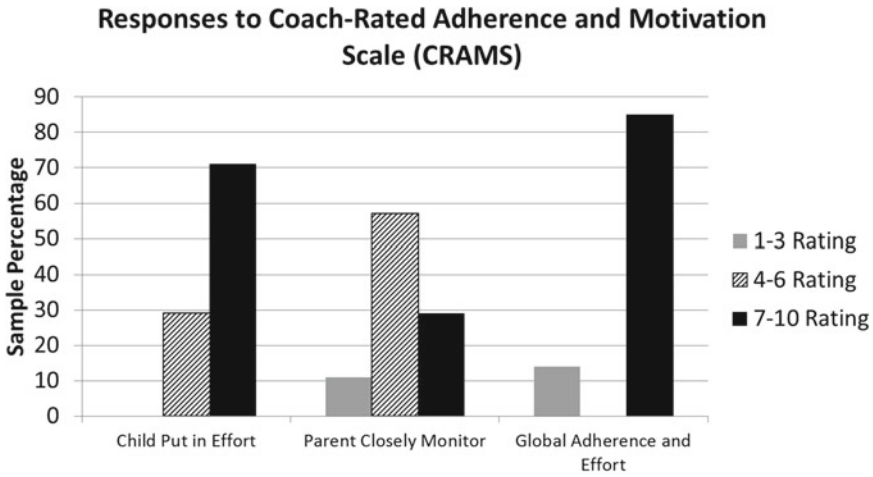

Fig. 18.4 Coach-report of adherence and motivation during working memory training. *Note*: 1 = not at all; 4 = somewhat/some of the time; 7 = much/much of the time; 10 = very much/very much of the time

parent to complete the full WMT program. Most parents were neutral when asked whether they believed that WMT improved their child's attention, memory, learning or language, but a large minority (over 40 %) of parents agreed with statements that their child's attention and memory improved following WMT. Parents were generally less convinced that WMT improved their child's learning or language, with 1/3 of parents disagreeing with the statement that their child's language improved following WMT. Overall opinion of the program was generally favorable, with 2/3 of the sample stating that they would recommend the program to other families and the others reporting a neutral opinion about recommendation to others; no parents disagreed with the statement that they would recommend the program to others (Fig. 18.3).

On the CRAMS, coaches reported that most participants put in adequate effort much or very much of the time, but most parents closely monitored the program only some of the time (Fig. 18.4). Nevertheless, coaches rated over 85 % of families as much or very much adherent to the program, consistent with findings that all subjects completed at least the required 20 training days.

Near-Transfer Effects of WMT

Changes during the waiting period, when no intervention took place, were minimal for Digit Span (forward and backward) as well as Spatial Span Backward, suggesting minimal practice effects from these tests (Fig. 18.5). Spatial Span Forward scores improved significantly during the waiting period.

Following the training period, scores for Digit Span Forward, Spatial Span Forward, and Spatial Span Backward improved significantly from baseline scores at the beginning of the training period, consistent with near transfer effects on these measures. Effect sizes using standardized change scores, which are an index of the number of standard deviations improved in each measure over each study period (obtained by subtracting the baseline score from the endpoint score for each period and then dividing by the standard deviation of the baseline score), were in the medium to large range (Cohen 1992). For Digit Span Backward, the standardized change value during the training period fell in the small to medium range and was not statistically significant (Fig. 18.5).

Standardized change scores for 1-month follow-up were calculated relative to the baseline prior to training; therefore, these scores reflected change from pre-WMT to 1 month following training. The 1-month follow-up change scores were similar to those immediately following WMT, although the slightly lower standardized change scores for the Spatial Span measures (at 1-month follow-up relative to the training period) were no longer statistically significant (Fig. 18.5). However, significant declines in standardized change scores were observed on all near-transfer measures by 6-month follow-up, suggesting that improvement in these measures was not long-lasting without continued working memory training. In sum, significant near-transfer effects were found for three of the four WM measures following training, but these effects declined by 6-month follow-up.

Far-Transfer Effects of WMT

BRIEF Working Memory subscale scores did not change during the waiting period but then declined significantly (indicating improvement in WM) following training, with a standardized change score in the medium range (Fig. 18.6). This improvement, however, declined by 1-month and 6-month follow-up periods and was no longer statistically significant. WRAML-2 Sentence Memory scores improved

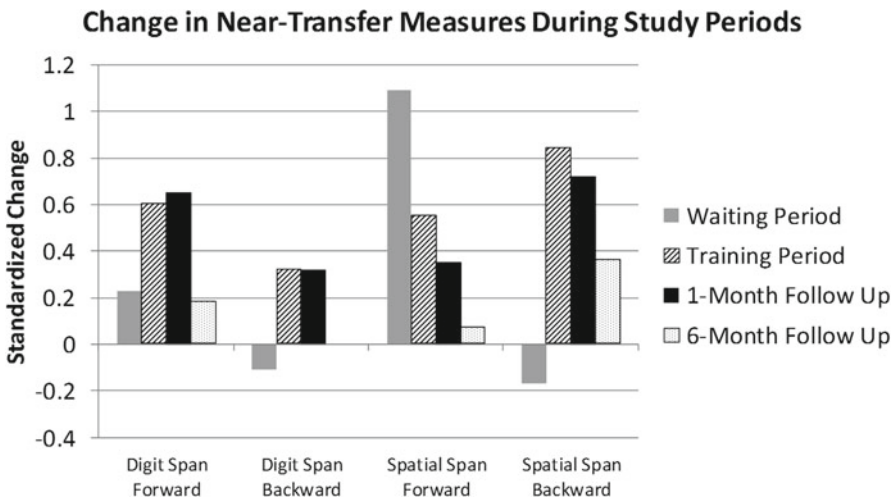

Fig. 18.5 Near-transfer change following working memory training. Standardized change scores for each period were derived by subtracting the baseline score from the endpoint score and dividing by the sample SD for the baseline scores. For the waiting and training periods, the score at the start of the period was used as the baseline score. For the follow-up periods, the score at the start of the training period was used as the baseline score. *p*-values are for paired *t*-test comparing endpoint and baseline scores (***p* < 0.01; **p* < 0.05). Based on data from Kronenberger et al. (2011)

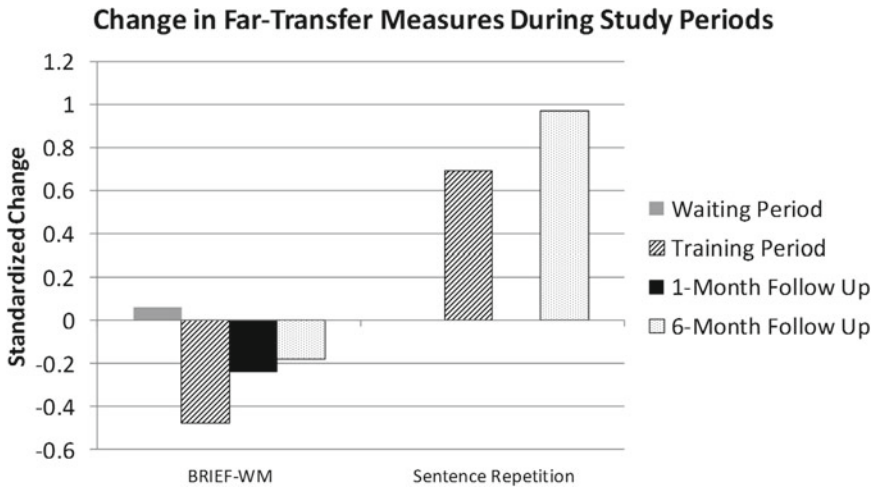

Fig. 18.6 Far-transfer change following working memory training. *BRIEF-WM* working memory subscale of the behavior rating inventory of executive function. Standardized change scores for each period were derived by subtracting the baseline score from the endpoint score and dividing by the sample SD for the baseline scores. For the waiting and training periods, the score at the start of the period was used as the baseline score. For the follow-up periods, the score at the start of the training period was used as the baseline score. Sentence repetition (Wide Range Assessment of Memory and Learning, Second Edition, Sentence Memory) scores were obtained only during the training period and 6-month follow-up period in order to minimize practice effects. *p*-values are for paired *t*-test comparing endpoint and baseline scores (****p* < 0.001; ***p* < 0.01; **p* < 0.05). Based on data from Kronenberger et al. (2011)

markedly during the training period, and this improvement persisted through the 6-month follow-up period (because of possible practice effects, the WRAML-2 Sentence Memory subtest was not administered at the beginning of the waiting period or at the end of the 1-month follow-up period). Thus, behavioral improvements reported by parents were significant after training but declined thereafter; substantial improvement in sentence repetition performance was observed after training and persisted through the 6-month follow-up.

Exploratory Analyses

Because deficits in fluency/speed and processing efficiency have been reported in samples of CI users (Kronenberger et al. 2013a; Burkholder and Pisoni 2003), we also conducted exploratory analyses with several measures of processing speed data obtained before and after each study period. The Coding subtest of the WISC-IV-I measures ability to rapidly reproduce a sequence of symbols based on a corresponding sequence of numerals; this subtest was admin-

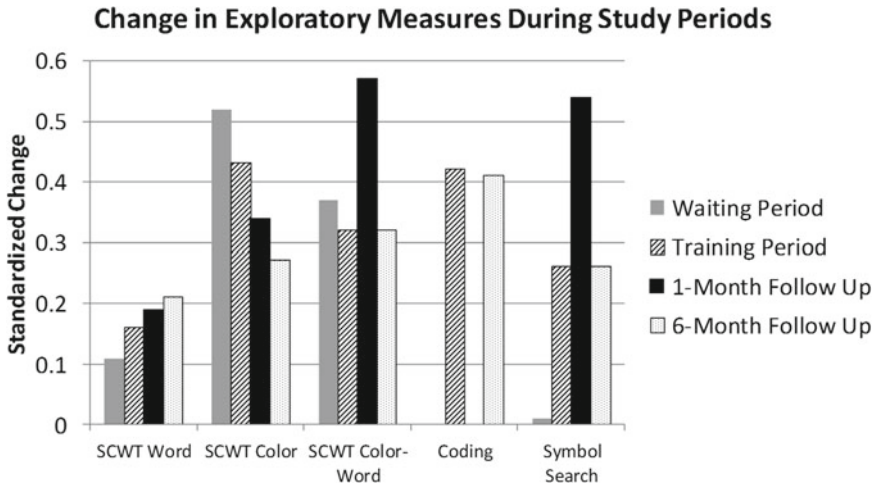

Fig. 18.7 Change in exploratory fluency-speed measures following working memory training. Standardized change scores for each period were derived by subtracting the baseline score from the endpoint score and dividing by the sample SD for the baseline scores. For the waiting and training periods, the score at the start of the period was used as the baseline score. For the follow-up periods, the score at the start of the training period was used as the baseline score. *p*-values are for paired *t*-test comparing endpoint and baseline scores (**$p<0.01$; *$p<0.05$; *a* $p<0.10$)

istered only for the training and 6-month follow-up periods in order to reduce practice effects. The Symbol Search subtest of the WISC-IV-I measures the ability to rapidly identify two identical symbols (or indicate if no symbols are identical) within a row of seven symbols; the pages in this subtest were administered in different orders at each visit (counterbalanced across participants) in order to reduce practice effects. The Stroop Color-Word Test (SCWT) requires subjects to rapidly read or name items in three conditions: word reading (read a series of color words [red, green, or blue]), color naming (name a series of ink colors used to print the stimulus "XXXX"), and color-word (name a series of ink colors used to print incongruent color words [e.g., "blue" written in red ink]); the first two conditions of the SCWT involve rapid automatized naming (RAN) processes, whereas the last condition requires inhibition of a more automatic/prepotent response (word reading) in favor of a more effortful response (color naming). In order to reduce practice effects, the standard (Golden and Freshwater 2002) version of the SCWT was administered at Visit 1, followed by three alternate versions of the SCWT created for the current study (administered in counterbalanced order across participants) at Visits 2, 3, and 4; at Visit 5, the version of the SCWT that had been administered at Visit 2 was used.

Finally, durations of pauses between spoken digits from the digit span test were obtained as measures of short-term memory scanning speed (Burkholder and Pisoni 2003), using digital waveforms from audiorecorded responses to the digit span test (see (Kronenberger et al. 2010) for a thorough description of methodology and results). These pause durations in the vocal responses are assumed to reflect the time needed to scan for verbal representations of the digits in short-term verbal memory. Faster scanning speeds (shorter digit span pause durations) are characteristic of individuals with better working memory skills (Cowan et al. 1998; Cowan 1999). Values for mean digit span forward pause duration and mean digit span backward pause duration were obtained for each visit by averaging all pause durations (in milliseconds, ms) for correctly repeated digit span sequences for each participant.

Results of the exploratory analyses produced standardized change values in the small to medium range for SCWT Color, Color-Word, Coding, and Symbol Search tests during the training and follow-up periods (Fig. 18.7). Although most of these standardized change values were not statistically significant, several approached significance with *p*-values <0.10. Large declines in average pause duration values for digit span backward were seen following the training and 1-month follow-up periods (pause duration values were not available for the 6-month follow-up), although, again, these did not reach statistical significance ($p<0.10$; Fig. 18.8); changes in pause durations were negligible for digit span backward during the waiting period and for digit span forward during any study period (Kronenberger et al. 2010).

In summary, exploratory analyses of processing speed and speech timing measures demonstrated numeric improvement on most measures following training, but typically at levels approaching but not reaching statistical significance. Because the study *N* was very small (and therefore the study might not have been sufficiently powered to detect a significant training difference), the tests of statistical significance of these processing speed measures should be interpreted with caution.

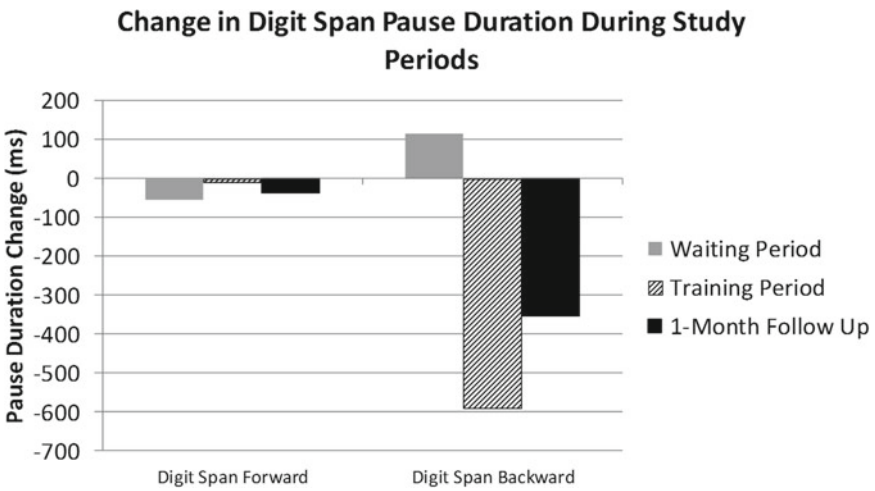

Fig. 18.8 Change in exploratory digit span pause duration measures following working memory training. Pause duration is the average length in time (in milliseconds, ms) between digits for correctly repeated sequences. Change scores for each period were derived by subtracting the baseline score from the endpoint score. For the waiting and training periods, the score at the start of the period was used as the baseline score. For the 1-month follow-up period, the score at the start of the training period was used as the baseline score. *p*-values are for paired *t*-test comparing endpoint and baseline scores ($a\ p < 0.10$)

Summary, Implications, and Limitations

Our pilot study of WMT in a small sample of prelingually deaf, early-implanted children and adolescents with CIs was a first attempt to evaluate the feasibility and potential effects of WMT for CI users with average to below average WM. Limitations of the study include the small sample size, lack of a placebo control group, and lack of subject blinding to condition. The small sample size limited study power, making the interpretation of nonsignificant results difficult, particularly for standardized change scores in the small to medium range. Small sample size also prevented us from investigating characteristics that might predict response to WMT, such as pre-training WM skills (all subjects were required to have evidence of average or poorer WM skills), pre-training attention problems, or performance on the WM exercises. Additionally, because subjects were not blinded to condition, changes in scores could be influenced by expectancy effects. Lack of a placebo control group makes it difficult to differentiate true change from practice or expectancy effects. Despite these study limitations, the inclusion of a waiting period and administration of most tests twice (at the beginning and end of the waiting period) prior to initiation of WMT likely accounted for some of the practice effects. Because the greatest practice benefits (e.g., steepest portion of the learning curve) are typically realized following the first repetitions of a task, change during the waiting period would presumably reflect the largest practice effects. However, only the Spatial Span Forward and SCWT Color subtests showed significant improvement during the waiting period; most other tests showed negligible or even negative change during the waiting period. Furthermore, alternate versions of several tests (Digit Span, Spatial Span, SCWT, Symbol Search) were used to further minimize practice effects. Nevertheless, the results of this study should be understood within their proper context as a preliminary, pilot study designed to assess the feasibility and efficacy of WMT in this clinical population.

With the caveats of the previous paragraph in mind, our pilot study of WMT in children and adolescents with CIs yielded several important findings:

1. In almost all cases, children with CIs had no difficulty with the mechanics of the Cogmed WMT exercises (e.g., difficulty hearing, understanding, or completing exercises).
2. Children with CIs improved on the trained Cogmed WMT tasks.
3. A considerable amount of effort from both parent and child was required to complete the Cogmed exercises, but coaches generally reported that parents and children put in adequate effort, with the assistance of the trained coaches. Therefore, close monitoring and coaching are likely to be essential elements of any WMT program for children with CIs.
4. Most parents were neutral or undecided about whether the WMT program produced an improvement in attention, memory, learning, or language, but a substantial minority (over 40 %) reported improvement in attention and memory.
5. Most parents expressed satisfaction with the WMT program in the form of agreeing that they would recommend it to others, although a substantial minority of the sample was neutral about recommendation.

6. Most near-transfer measures of working memory skills improved significantly following WMT, with medium to large effect sizes; however, these improvements declined by 6-month follow-up.
7. Far-transfer measures of sentence repetition skills and parent-reported WM behaviors at home improved following WMT, with medium to large effect sizes; sentence repetition improvements were maintained at 6-month follow-up.
8. Exploratory measures of processing speed improved numerically following WMT, with small to medium effect sizes; however, these improvements were, for the most part, not statistically significant, possibly because of the small sample size.

The results of this pilot study of WMT in children with CIs are encouraging, and they suggest the need for further research with larger samples and blinded, controlled designs. Aside from issues of methodology and design, the largest challenges appear to be with the amount of effort required to complete the program, robustness of the training effects (decline in gains at follow-up), variability in response from one child/family to the next (with about 40–70 % of families reporting improvement and recommending the program, but the remainder expressing neutrality about the program), and differences in abilities showing significant improvement (e.g., not all tested working memory or processing speed abilities showed improvement after WMT).

At this time, no study has yet investigated WMT efficacy in children with CIs using a blinded, controlled design, and almost no other research exists on WMT in CI users. Oba et al. (2013) evaluated the efficacy of working memory training using a visual digit span training task (recall of sequences of visually presented digits) with ten postlingually deafened adult CI users. Their results showed no improvement in auditory digit span, phoneme recognition, sentence recognition in noise, and digit recognition in noise, leading them to conclude that improvements observed in CI users after auditory training in earlier studies (Oba et al. 2011) are likely a result of improved auditory perception rather than improved attention or memory.

It is important to note, however, that the Oba et al. study differed markedly from our WMT study: First, all subjects in the Oba et al. sample were postlingually deafened, implying a fundamentally different influence of auditory deprivation on brain development and organization (e.g., the auditory neurocognitive model in Fig. 18.1 applies only to prelingually deafened children) than our prelingually deafened, early-implanted sample. Second, five of the subjects (50 % of the sample) in Oba et al. were over age 60 years, and two were over 70 years of age (including one subject aged 91 years). Thus, their results apply in large part to a geriatric sample, which may have a very different response to WMT than a pediatric sample, especially given marked differences in brain plasticity at young vs. old ages. Third, duration of CI use was highly variable in the Oba et al. sample, with two users having CIs for less than a year. Finally, the visual digit span task presented in Oba et al. is one very specific type of WMT; it is not clear whether other types of WMT might have different effects. Therefore, marked differences in our WMT study and the study of Oba et al. make it difficult to draw any general conclusions by integrating results across studies.

Summary and Future Directions

Deprivation in auditory experience as a result of early deafness and subsequent underspecified auditory input from a CI can produce downstream effects on a range of neurocognitive abilities extending well beyond speech perception and spoken language skills. Consistent with the Auditory Neurocognitive Model (Fig. 18.1), research has demonstrated that prelingually deaf children with cochlear implants are at risk for delays in fundamental areas of executive functioning, including inhibition/concentration, fluency/speed, and working memory dynamics. Working memory, especially verbal working memory, is significantly delayed in a large proportion of children with CIs. Because of the critical importance of working memory for language, learning, and daily functioning, these delays present a significant challenge to the cognitive development and quality of life of children with CIs and underlie some of the difficulties they experience in the development of spoken language skills.

Several computer-based WMT programs have been developed and evaluated in empirical research with populations of normal-hearing children, adolescents, and adults. Results of this research consistently show improvement in the WMT exercises and in untrained near-transfer tasks of WM that are conceptually similar to the WMT exercises, as well as changes in brain functioning following WMT. Results are less conclusive regarding far-transfer effects of WM to abilities such as fluid intelligence, attention/concentration, and academic skills; some placebo-controlled studies have found improvements in these areas, while others have not.

We conducted a pilot study of the feasibility and efficacy of a widely used WMT program (Cogmed) in a small sample of prelingually deaf, early implanted children and adolescents with CIs and average to below average WM skills. Results of our pilot study indicated that the WMT program was feasible and acceptable in the sample. Statistically significant improvement with medium to large effect sizes were found on most measures of near-transfer WM skills following training, as well as on far-transfer measures of parent-ratings of WM behaviors at home and sentence repetition skills. Exploratory measures of fluency/speed showed

numerical improvement following WMT, but most did not reach statistical significance. Almost all improvement (with the notable exception of sentence repetition and memory skills) declined during a follow-up period of no treatment, particularly 6 months following the completion of training. The study results provided preliminary evidence of the feasibility and potential efficacy of WMT for children with CIs, but additional research is needed with a large sample of children using a randomized, controlled clinical trial.

Clearly, a pressing need exists for a randomized, controlled clinical trial of WMT in children with CIs in a design powered to detect small to medium effect sizes. Future research should also investigate characteristics that predict adherence to the WMT program and change in near-transfer and far-transfer skills following training. Different types of WMT should be further investigated as well, because some types of WMT may be more effective than others for children with CIs. It is also important to note that the characteristics of effective WMT may be different for normal-hearing children and for prelingually deaf children with CIs. In addition to differences between CI users and NH peers in executive functioning, the relationship between executive functioning, concept formation, and spoken language skills may be different for CI users than for NH peers. Therefore, the core foundational influences on development of working memory, executive functioning, and spoken language likely differ for children with CIs compared to their normal-hearing peers. This dissociation in developmental influences on WM skills suggests that the characteristics of effective WMT for children with CIs may differ from those for NH peers.

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Auditory Training: Predictors of Success and Optimal Training Paradigms

19

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There has long been the question of how much people's speech perception can be improved. This question has led to efforts to develop training paradigms that can effectively improve speech perception. With the advent of cochlear implants (CI), these efforts have turned to improving the speech perception abilities of CI recipients after implantation. Here we argue that continued efforts to understand what characterizes successful vs. less-successful speech perception learners and to develop training paradigms that are matched to the ability levels of each learner are likely to result in most optimal speech perception outcomes. We first review extensive efforts to train second-language speech perception that have demonstrated that training paradigms using natural speech training tokens and that provide feedback are more likely to be successful. However, these training programs are marked by extensive variability in training outcomes. We then review recent efforts to identify the sources of variability and to develop individualized training paradigms than can result in optimal training outcomes. These efforts have indicated that some learners enter the training paradigm better able to perceive the acoustic differences among speech sounds. We conclude by describing work that indicates that for those listeners who have difficulty perceiving those same acoustic differences, simplifying the training paradigm to allow them to focus on the relevant differences among to-be-learned speech sounds can reduce post-training outcome variability.

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Effectiveness of Speech Perception Training

Speech Training in the Laboratory

Laboratory investigations into speech training studies often emphasize improvement of speech perception abilities in adults learning a second language. In particular, there is typically one phonetic contrast—though some studies focus on a few contrasts—that is especially difficult for the second-language learner and whose perception is meant to be improved following training (a classic example of this, and the focus of many training studies, is the difficulty native Japanese speakers have differentiating the English sounds /r/ as in "rock" and /l/ as in "lock"). These studies have found that paradigms using natural speech tokens produced by multiple talkers tend to be the most effective. The particulars of these paradigms can vary, but they all use real words that were produced by more than one talker, the to-be-learned contrast appears in multiple locations within the word, learners must make a forced-choice judgment (e.g., was the word presented "rock" or "lock"), and response feedback is given after each trial. Training is typically short, on the order of 5–10 days, but even such limited training is sufficient to produce speech perception gains (Iverson et al. 2011), which can be maintained for at least 3 months (Bradlow et al. 1999; Iverson and Evans 2009). Importantly, trainees learn to identify not only the talkers and speech stimuli used in training, but training generalizes to novel, untrained words and talkers (Logan et al. 1991).

This paradigm was originally developed to train native Japanese speakers to differentiate the English /r/ and /l/ (Bradlow et al. 1997, 1999; Logan et al. 1991; Iverson et al. 2005; Lively et al. 1993, 1994). It has since been applied to the

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training of linguistic tones from Mandarin Chinese to native English speakers (Wang et al. 1999), Hindi contrasts to English speakers (Pruitt et al. 2006), cross-language fricatives (Trapp et al. 2000), and cross-language vowels (Iverson et al. 2011; Iverson and Evans 2009; Nishi and Kewley-Port 2007, 2008). Given the success of the training paradigm in so many language applications, some of its hallmarks may be worth considering as we develop training paradigms for CI recipients.

Training CI Recipients

Compared to efforts to train speech perception in second-language learners, there are relatively few laboratory studies training speech perception in CI recipients. Additionally, these studies tend to lack a control group and train a very small number of listeners, limiting the generalizability of their findings. Nonetheless, they do provide evidence that speech perception training can be effective for CI recipients.

In the second-language learning scenario, the particular sounds that are difficult for the listener to differentiate tend to be consistent within a language group. This is a result of the fact that listeners are experts at perceiving the sounds of their first language and their speech perception categories reflect this expertise (Kuhl et al. 2006; Tsao et al. 2004; Werker and Tees 1984). The mismatch between the listeners' existing speech perception categories and the speech sound categories of the to-be-learned language results in speech perception difficulties (Best and Tyler 2007; Flege 2003) and this mismatch happens in predictable ways for pairs of languages. Conversely, for CI recipients, the particular sounds an individual listener may struggle with are less predictable. Each CI recipient has a unique experience with language, etiology of deafness, device, programming settings, etc. that combine to result in a set of sounds that are most difficult for each listener. Thus, one likely difference between second-language speech training and speech training for CI recipients will be the fact that whereas second-language studies train the same set of sounds for all listeners, CI studies may find more success by training those sounds difficult for an individual listener.

After determining which sounds were most difficult for each listener, however, the basic training model can be the same for all listeners. Fu et al. (2005) utilized some of the lessons from second-language training, developing a training paradigm that used primarily real words spoken by multiple talkers that placed the to-be-learned sounds in a variety of locations within the word. Listeners were asked to choose among confusable response options and received feedback after each response. Unlike the second-language training paradigms, response options became more confusable and the number of response options increased with increasing response accuracy; increasing response options also moved listeners away from real words to pseudo words. After 16 weeks of adaptive training, all listeners showed significant improvements over baseline performance. The same training paradigm was successfully used to train speech perception and lexical tone in native Mandarin Chinese-speaking CI recipients (Wu et al. 2007). A recent effort took a slightly different approach, working to improve speech perception in noise in CI recipient adults, instead of focusing on improving their ability to perceive individual speech sounds (Ingvalson et al. 2013a). Nonetheless, that study also used real words produced by multiple talkers, confusable response options, and feedback after each response. Also, like the other studies training speech perception in CI recipients, it used an adaptive paradigm where the number of items to judge increased and the level of background noise increased as accuracy increased. Significant improvements were found after only 4 days of training. Clearly, there is more work to be done to determine the effectiveness of speech training for CI recipients, but these studies provide preliminary evidence that such training can be effective and provide guidance for how such training might be best constructed.

Looking at the successful second-language and CI recipient speech training studies, we see some commonalities that routinely appear. These commonalities are likely to appear in future successful training paradigms to improve speech perception in CI recipients. In particular, we expect future successful studies to use real speech as training stimuli, present to-be-learned sounds in a variety of contexts, ask listeners to make responses from a set of confusable options, and give feedback after each trial.

Predicting Training Outcomes

The successful speech training paradigms improve speech perception on average; that is, the overall performance of a group will improve following training. However, there is extensive variability in the post-training performance. This level of variability is also typical of individuals who learn a second language or learn to hear with a CI outside of a training regimen, leading to the suggestion that these tasks are just hard and variability in performance is to be expected. While there is no question as to the difficulty of the speech learning task, the post-training variability also highlights the fact that some individuals are mastering the training rather easily whereas other individuals are struggling throughout training, leading to smaller post-training gains. If we are able to predict which listeners are those who are going to master the training rather easily, then perhaps we can identify what characteristics those listeners possess that makes speech learning easier for them than for other listeners. Transferring the characteristics of those who learn easily to those who struggle could result in better learning for those who struggle and could reduce the variability that has become a hallmark of speech training. The first step, of course, is to predict variability in outcomes using pre-training measures.

This pre-testing does not necessarily use speech materials, but may use pseudo-speech materials that capture the characteristics of the to-be-learned speech sounds that are particularly difficult for the listener. For example, in a study training the lexical tones of Mandarin Chinese to native English listeners, the pre-testing materials may use pitch patterns that have been taken out of a lexical context to determine listeners' sensitivity to moving pitch. Testing listeners before training with materials that highlight the difficult characteristics of the to-be-learned contrasts has shown, perhaps not surprisingly, that those listeners who are better at differentiating the difficult characteristics of the speech tokens pre-training show better training outcomes. This pattern of results has been found for native English listeners learning Mandarin speech sounds (Chandrasekaran et al. 2010; Ingvalson et al. 2013b; Perrachione et al. 2011; Wong and Perrachione 2007), native English listeners learning a Hindi contrast (Golestani and Zatorre 2009), and native Greek speakers learning English vowels (Lengeris and Hazan 2010). Not only does good pre-training performance predict good post-training performance, but good pre-training performance can predict more rapid learning and a shorter training duration (Golestani and Zatorre 2009). Though much of this work, as with much speech training work in general, has been done in the second-language learning domain, there is some indication that CI recipients with better speech skills early would show better gains following training. Using noise-vocoded speech to simulate a CI in normal-hearing listeners (Shannon et al. 1995), those listeners who were better able to understand the vocoded speech prior to training showed better training performance (Eisner et al. 2010).

Neural Correlates of Successful Learning

Together, studies looking to predict post-training outcomes from pre-training performance found that those individuals who were better able to perceive the relevant characteristics of the speech before training showed the greatest training benefit. This is consistent with the variability seen in speech training studies, which shows that some listeners master the speech learning task relatively easily whereas others continue to struggle. We have learned, then, that those listeners who are mastering the task easily show a pre-training aptitude for the sounds being presented, not a relatively greater ability to take advantage of the training paradigm. The question then is what provides this pre-training aptitude.

An obvious place to look for the source of these aptitude differences is in the neuroanatomy and neural activation of the listeners, particularly in areas associated with auditory perception. It has been established that there were neuroanatomical differences between congenitally deafened and normal-hearing adults (Emmorey et al. 2003; Penhune et al. 2003) and between musicians and nonmusicians (Gaser and Schlaug 2003; Schneider et al. 2005). The differences between musicians and nonmusicians were characterized by differences in gray matter volume in Heschl's gyrus, which has been shown to be important for pitch perception. If more successful learners' pre-training aptitude stems from a greater ability to utilize the acoustic cues differentiating the speech sounds, we might expect more successful learners to show greater gray matter volumes in Heschl's gyrus, especially when that acoustic cue is lexical pitch. We might also expect to see this larger volume in left Heschl's gyrus, as regions on the left side of the brain have been more associated with speech and language learning tasks. Tests of these expectations have been confirmatory. Larger left Heschl's gyrus volume was found in those native English listeners who were more successful learning Mandarin Chinese lexical tones (Wong et al. 2008). Beyond lexical tone learning, larger Heschl's gyrus volumes were found for native English listeners who were faster and more successful at learning a Hindi contrast (Golestani et al. 2002, 2006). Fast, successful learners of the Hindi contrast were also found to have a greater white matter density in left Heschl's gyrus which may result in more efficient transmission of neural signals relevant to speech perception.

The above studies used anatomical neuroimaging, which connects neural structural features to behavioral outcomes. Another method is functional neuroimaging, which connects neural activity to behavioral outcomes. Following the neuroanatomical results above, we would expect those listeners who have a pre-training aptitude for the to-be-learned speech sounds to show higher levels of activations in areas associated with speech and language. Looking again at native English listeners being trained to differentiate Mandarin Chinese lexical tones, before training successful learners showed greater activation in bilateral superior temporal gyrus (STG) affiliated with primary auditory cortex (Wong et al. 2007). After training, successful listeners showed more activation in posterior STG. Demonstrating that activation is related to the ability of listeners to perceive the relevant characteristics of speech sounds, Chandrasekaran and colleagues (2011) related the efficiency of nonlexical pitch pattern neural encoding in a midbrain structure called the inferior colliculus—known to be important for pitch encoding—to the fidelity of nonlexical pitch pattern representations. Not surprisingly, those listeners who encoded the nonlexical pitch patterns more efficiently and who had more accurate representations of the patterns performed better when trained on lexical tones. It appears, then, that those listeners who show more success in a speech training paradigm do so because they are better equipped to process the basic acoustic structure of the to-be-learned speech sounds before training. This may be because they have more gray matter volume in areas devoted to acoustics, more connections between auditory areas and the rest of the brain, more activation of areas relevant to a speech perception task, or more accurate encoding of the speech acoustics. These are abilities the listeners are

bringing to the speech training task, and those listeners who show less success after training are bringing a different set of skills that is less suited to speech learning. It is therefore on the trainer to identify those individuals who may not come to the speech training task with an existing aptitude and to develop a training that will utilize the skills that individual does posses to optimize learning for all listeners.

Optimizing Learning by Individualizing Training

Researchers have recently begun to investigate the potential benefits of matching listeners with a training paradigm that fits their ability level. We described above the most common second-language speech training paradigm, which trains all listeners using multiple speech tokens produced by multiple talkers. As mentioned, this paradigm has proven to be highly successful but outcomes are also highly variable. Knowing that pre-training nonlexical pitch pattern differentiation performance relates to neural correlates of lexical tone learning (Wong et al. 2007, 2008), Perrachione and colleagues divided listeners into those with a high pre-training aptitude for lexical tone learning and those with a low pre-training aptitude for lexical tone learning based on their pre-training performance (Perrachione et al. 2011). Listeners with a high pre-training aptitude for lexical tone learning performed best in the multiple speech token, multiple-talker paradigm described above. However, those listeners with a low pre-training aptitude performed better in conditions when they had the opportunity to focus more closely on the lexical tones outside of talker variability. This included hearing one talker for the duration of training, or having the multiple talkers separated into different blocks. It seems, then, that minimizing the variability in the training set that is irrelevant to the phonological characteristics that need to be mastered may help these listeners who may have more difficulty encoding the acoustics of unfamiliar speech sounds (though it is possible to take this idea too far (Ingvalson et al. 2012)).

If the reduced variability of single-talker paradigms is allowing listeners to pick up on the phonological characteristics of the to-be-learned speech which in turns allows them to make larger gains than seen in earlier studies, then perhaps providing these listeners with phonological training would result in even greater gains. A follow-up study supplemented the blocked-by-talker condition of Perrachione et al. (2011) with 3 days of phonological training (Ingvalson et al. 2013b). Specifically, listeners heard multiple speech tokens produced by multiple talkers, but talkers were blocked instead of intermixed. During the first 3 days of training, the listeners' task was to identify the direction of the pitch, focusing learners on the pitch dimension within the sound before introducing the lexical tone learning task. Learners identified as low-aptitude prior to training showed steeper training curves and better post-training performance following the phonological-plus-lexical tone training relative to similarly skilled listeners who received only lexical tone training. Thus, though some listeners may not have entered the training paradigm with the same ability to differentiate the pitch patterns that other listeners did, providing those listeners with a foundation for the speech learning task increased their learning success. As we begin to think of training paradigms for CI recipients, it may be wise to remember that not all of those listeners are created equal, either. Some of them are going to be more able to extract phonological information using their implants, whereas others will require more assistance to learn how to do this task. An understanding of the starting point of each individual listener will inform our awareness of what sort of foundation for speech learning already exists or may need to be developed through training. Providing listeners with the training that matches their ability level is more likely to result in optimal success than would using the same training paradigm for all listeners.

Conclusions and Future Directions

Though efforts to train speech perception have emphasized the use of multiple speech tokens produced by multiple talkers, research into understanding the variability that follows such training has revealed that it may not be optimal for all listeners. Looking into the neurological correlates of this variability suggests that some listeners are better able to perceive the acoustic characteristics that differentiate unfamiliar speech sounds. Further work is needed to determine what causes this enhanced ability to perceive the relevant characteristics of the speech token and whether it is possible to incorporate those causes into a training method for those individuals without such an ability. In the meantime, the evidence to date indicates that some listeners will struggle to differentiate to-be-learned speech more than others. We therefore need to develop good behavioral tests to identify those listeners for whom multiple speech tokens and multiple talkers may not be optimal. In the case of CI recipients, these tests will likely need to be all-encompassing, as the particular speech sounds that need to be trained will differ from listener to listener. Once this starting point has been established, the listener can be matched to a training paradigm that matches his or her ability level, likely resulting in more optimal learning outcomes. Ongoing work suggests that, for listeners who may struggle to perceive the phonetic characteristics of speech, reducing the degree of variability among tokens or introducing phonological training could be beneficial. Similarly, the preliminary efforts to train speech perception in CI recipients are inspiring in the fact that they are adaptive. Training that matches its difficulty level to the

ability level of the trainee is naturally individuated. As we move forward into developing more speech training paradigms to meet the needs of a growing CI recipient population, we advocate for a continued emphasis on the individual, both through pre-training assessments that match the learner with the appropriate training paradigm and through adaptive training that matches the learner's skill set throughout.

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Integrated Language Intervention for Children with Hearing Loss

20

Susan Nittrouer

Integrated Language Intervention for Children with Hearing Loss

Research on a variety of topics related to pediatric cochlear implantation has been discussed in this volume. Synthesizing results across these various topics allows us to make specific recommendations regarding how behavioral interventions should be implemented for children who receive CIs. Many of the ideas to come out of this effort are identical to those that would be recommended for any child born with any degree of hearing loss (mild to profound), or who might acquire such a loss early in life. Significant delays in language acquisition continue to be observed for children with only mild-to-moderate hearing loss, which is typically defined as auditory thresholds between 20 and 70 dB hearing level, in spite of advances in hearing aid technology (e.g., Briscoe et al. 2001; Davis et al. 1986; Delage and Tuller 2007; Wake et al. 2004). One finding of special interest coming out of that work is that mean performance levels obtained for children with mild-to-moderate hearing loss are commonly found to be one standard deviation below the means of typically performing children with normal hearing (e.g., Gilbertson and Kamhi 1995; Wake et al. 2004), which is strikingly similar to what is found for children with CIs: Consistently across studies of language acquisition for children with CIs, differences of that magnitude have been observed, as indicated by the studies listed in Table 11.1, as well as by others (e.g., Boons et al. 2012; Geers et al. 2003; Nittrouer et al. 2012). This level of mean performance marks significant improvement in language abilities for children with severe-to-profound hearing loss as a result of the availability of cochlear implants. But the finding that children with mild-to-moderate hearing loss are performing no better, on average, can be taken as evidence that improving implant technology alone should not be expected to close the gap in performance compared to children with normal hearing. These collective findings across studies emphasize that degree of hearing loss cannot predict language outcomes for children, a point explicitly discussed by several investigators (e.g., Davis et al. 1986; Gilbertson and Kamhi 1995; Norbury et al. 2001; Tuller and Jakubowicz 2004). In turn, that trend highlights the fact that there is more involved in learning language than simply being able to harvest linguistically relevant acoustic cues from the physical signal reaching the auditory system.

There are several ways in which hearing loss and subsequent cochlear implantation can negatively impact the development of language and literacy. The most obvious way is by diminishing the quantity and quality of the sensory input. As we move through our lives, we use sensations to inform us about events in our environment, as well as about the effects our actions have on that environment. Children recover information about the speech production patterns of others through their sensory systems, and refine their own production from the feedback they receive through those systems. Those interactions with the environment—both as perceiver and producer of spoken language—allow children to develop the linguistic elements that they will use in language and cognitive processes. Any degradation in sensory inputs can negatively impact the acquisition and refinement of these linguistic elements by diminishing the resolution of the representations. Where childhood hearing loss is concerned, degraded sensory input is responsible for the challenges children face in the acquisition of language. Accordingly, the dramatic improvements in language learning outcomes observed for severely to profoundly deaf children since cochlear implants became available are surely due to enhanced sensory inputs. Nonetheless, communication capabilities and language acquisition cannot be entirely explained

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by the quality of the input children have access to; if they were, degree of hearing loss would predict the lion's share of variance in the language capabilities of these children, and as was learned in the last paragraph, it does not.

Many more processes underlie communication and language learning than those associated with peripheral sensitivity to the sensory input. It is for this reason that factors related primarily to implants and implant surgery do not explain especially large amounts of variance in outcomes for children with severe-to-profound hearing loss. One nonsensory factor influencing communication abilities is that the perceiver must be able to attend to the information-bearing components of the sensory input, and ignore other inputs. Where speech perception is concerned, these strategies are known as *perceptual weighting strategies*. Investigations into this phenomenon reveal that listeners of different language backgrounds have different perceptual weighting strategies. What that means is that listeners selectively attend to different parts of the acoustic structure in the speech signal, depending on their native language background (Crowther and Mann 1992, 1994; Escudero et al. 2009; Flege and Port 1981; Iverson et al. 2003; MacKain et al. 1981). Because listeners in those experiments were all selected to have normal hearing, observed differences in attention could not be explained by differences in sensitivity to the relevant acoustic cues. In an especially stark demonstration of that discrepancy between sensitivity and weighting, Miyawaki and colleagues (1975) asked native Japanese speakers who learned English as a second language to discriminate a mid-frequency spectral glide that supports categorization of the phonemes [r] and [l] in English. They were found to be just as sensitive to this acoustic property as English speakers in a control group. However, when that short acoustic bit was merged with a more complete speech signal, the native Japanese listeners failed to use it to categorize [r] and [l]. That phonetic distinction is not present in Japanese, and apparently these speakers never learned to attend to the acoustic property on which it is based.

In addition to language background, the age of the listener plays a critical role in how the acoustic cues of speech get weighted. Much of the work demonstrating that point has been done in this laboratory, and shows that children initially attend strongly to the time-varying spectral structure of the signal arising from changes in shape and size of the vocal tract. That attentional strategy is different from what is generally found for adults, who attend more strongly to temporally restricted sections of acoustic structure. Illustrating these age-related phenomena are past experiments involving fricative-vowel stimuli. In a series of experiments, stimuli based on natural tokens of [ʃ]-vowel and [s]-vowel syllables were used (e.g., Nittrouer 1992; Nittrouer and Miller 1997; Nittrouer and Studdert-Kennedy 1987). Figure 20.1 displays these syllables with the vowel [ɑ], and shows that two kinds of acoustic structure, or cues, are clearly associated with the different places of constriction for these syllable-initial fricatives. First, the aperiodic fricative noise is lower in frequency for [ʃ] than for [s], a difference arising because the cavity in front of the constriction is larger for [ʃ]. In addition, the vocalic formants differ in onset frequency, direction, and extent of change, depending on place of constriction of the syllable-initial fricative. In particular, the second and third formants start at similar frequencies for [ʃ], but not for [s]. Consequently, the third formant rises after voicing onset for [ʃ], but falls following [s]; the second formant is higher at onset for [s] than for [ʃ]. Results of labeling experiments have consistently revealed that children weight the formant transitions more than adults when presented with these sorts of stimuli, and weight the static fricative noises less (Mayo et al. 2003; Nittrouer 1992; Nittrouer and Miller 1997; Nittrouer and Studdert-Kennedy 1987; Siren and Wilcox 1995).

Similar age-related differences in perceptual weighting strategies have been found for decisions regarding the voicing of syllable-final consonants (Greenlee 1980; Krause 1982; Nittrouer 2004; Wardrip-Fruin and Peach 1984). In this case, the two cues to voicing are the duration of the vocalic segment preceding the final consonant and the offset frequencies of the formants, especially the first formant. Children show the same preference for the time-varying formant patterns with these stimuli as they show with the fricative-vowel stimuli. Results across contrasts and experiments have led to the suggestion that children's perceptual attention changes with development and language experience, an idea termed the *developmental weighting shift* (Nittrouer et al. 1993). The explanation provided for this developmental change hinges on the notion that formant transitions span temporal stretches of the speech signal affiliated with more than one phonemic segment. One of the first tasks facing the child when it comes to language learning is discovering how to parse the signal into linguistic units such as words and syllables. Consistent patterns of formant change can mark these linguistic units, helping the young child learn how to divide the signal into meaningful units. As children get older, perceptual attention becomes increasingly focused on temporally discrete parts of the signal more closely affiliated with individual phonemic segments. That perceptual change accompanies the developmental enhancement of attention to word-internal phonemic structure observed for children across the first decade of life (e.g., Liberman et al. 1974; Walley et al. 1996).

Besides weighting acoustic cues according to language-specific strategies, it is essential that language users are able to integrate those cues in order to recover linguistic form accurately and efficiently. This process entails a phenomenon known as *perceptual organization*, defined as the strategies involved in blending sensory information into coherent

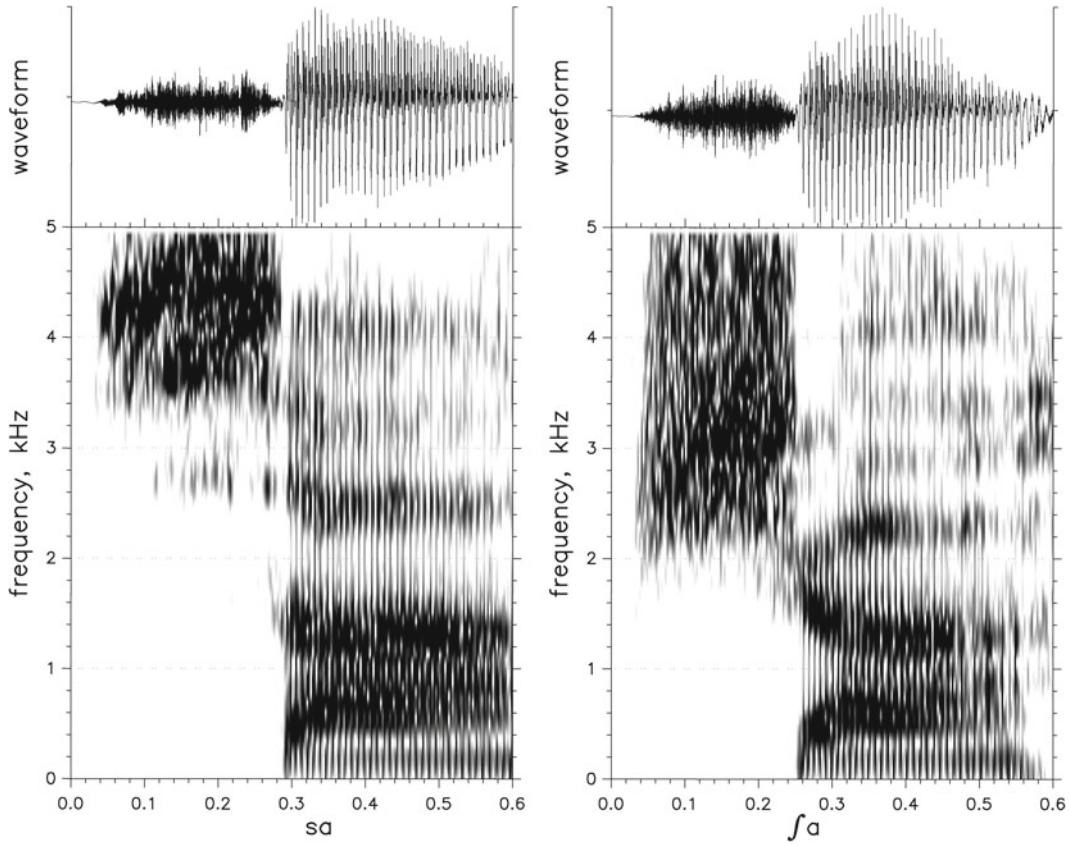

Fig. 20.1 Spectrogram of the syllables *sa* (*left*) and *sha* (*right*) spoken by a man, illustrating that both the spectral structure of the fricative noise and the formants differ depending on the initial fricative

units (Kimchi 2009). This phenomenon is the focus of a great deal of investigation in the visual domain where it can readily be illustrated. A simple example is provided by the well-known Rubin's vase. From the pattern of light and dark present in that simple drawing can be recovered either two faces in profile on either side of the image or a single vase in the middle. In either case the same sensory information is reaching the visual cortex; the form recovered is determined by how the perceiver organizes that information.

It is no coincidence that the visual images used to illustrate perceptual organization commonly involve degraded signals, typically patterns formed by various shades of gray. Degraded signals make is much easier to evoke alternative forms, and that point is relevant to appreciating what must be achieved by the child learning language through a cochlear implant because these prostheses provide only degraded auditory input. The child with normal hearing might not be totally immune to disorders of perceptual organization; in particular, a disorder of this nature has been suggested as underlying developmental dyslexia (Nittrouer and Lowenstein 2013). However, the probability of that kind of problem arising for children who have access to highly refined sensory information (i.e., those with normal hearing) is lower than it surely is for children with hearing loss, especially if they use cochlear implants. Thus, another challenge facing the child with a CI is learning appropriate perceptual organization strategies for speech. The emergence of such strategies cannot be assumed to be automatic for children with CIs because they have access only to a degraded representation of the speech signal.

The way in which sensory information comes to be organized is critically important. Among early theories of speech perception was the idea that listeners recover the articulatory gestures involved in producing the signal heard, with special reference to their own vocal tracts (Liberman et al. 1967). While that particular idea, known as the motor theory of speech perception, has not withstood the test of time, the general idea that speech perception is a sensorimotor process has received continued support (Kuhl 2010; Liberman and Mattingly 1985; Todd et al. 2006). In particular, there is clear evidence that the supplementary motor as well as the lateral premotor areas of the cortex are activated when listeners hear speech signals. Figure 20.2 illustrates that this process develops over the first year of life, with a concomitant diminishment in activation of the primary auditory cortex (Kuhl 2010). This figure shows images from magnetoenchephalography

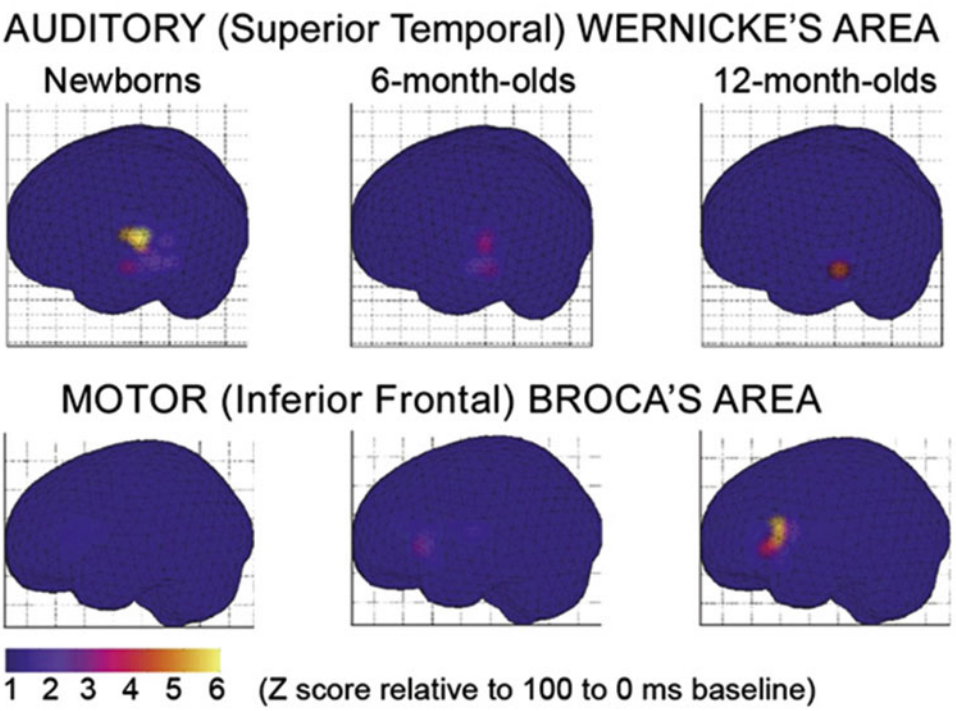

Fig. 20.2 Images from a magnetoenchephalography study of infants listening to speech across the first year of life, conducted by Imada et al. (2006). This image is reprinted from Kuhl (2010)

(MEG) recorded in the auditory (top) and motor (bottom) regions of the cortex in response to speech sounds. It shows how cortical responses to speech become isolated to the motor region. In that work, no similar shifts in the brain regions responding to nonspeech sounds were observed (Imada et al. 2006).

Behavioral evidence for the suggestion that perceptual organization of acoustic signals differs depending on whether those signals are processed as speech or nonspeech comes from several sources. For one, studies of sine-wave analogs of speech signals demonstrate the disparity in organizational strategies. These analogs eliminate most kinds of structure from the speech signal, except for the long-term trajectories of the first three formants. The center frequencies of these formants are tracked, and represented as separate sine waves in the generation of the analogs. Thus these signals are auditory analogs to ambiguous visual signals, such as Rubin's vase. In the very first experiment conducted with sine-wave speech, listeners were given no description of what they would be hearing prior to presentation. When queried after hearing them, many listeners reported hearing whistles or bird chirps or some other form of nonspeech sound. However, when listeners were instructed that they would be hearing degraded speech signals, they all were able to perceptually integrate these three disparate sine waves in such fashion as to recognize sentences (Remez et al. 1981). This dichotomy in how signals are organized as a function of expectations has been well replicated (e.g., Remez et al. 2001). Furthermore, patterns of phonetic labeling are rarely found to be explained by auditory sensitivity to the pertinent acoustic properties manipulated in the stimuli used in those experiments (e.g., Miyawaki et al. 1975; Nittrouer 1996; Nittrouer and Crowther 1998; Rosen and Manganari 2001).

In sum, three ways that hearing loss and subsequent cochlear implantation might hinder the development of age-appropriate language and literacy have been discussed. First, diminished access to sensory information can interfere with language learning. However, the finding of a lack of correspondence between degree of hearing loss and degree of language deficit reveals that other perceptual processes come into play. The second way that hearing loss and subsequent cochlear implantation might hinder the development of age-appropriate language and literacy concerns the fact that the child must discover what components, or cues, in the signal should be weighted strongly. That process requires a critical amount of experience hearing the ambient language, and such experience can be constrained by hearing loss. Finally, the child must be able to organize those cues appropriately in order to recover linguistic units, something that is a perceptually slippery undertaking when a degraded sensory input is involved. These considerations should help shape the way that we design intervention for children with CIs.

Principles of Integrated Language Intervention

The factors reviewed above concerning how speech signals are processed can and should be used to derive general principles for designing intervention programs for children with CIs. In this section, specific principles for designing integrated approaches to intervention are described that emerge from the basic science on how linguistic signals are processed, as well as from outcome studies of current language and literacy performance of children with CIs.

Use Sufficiently Long Signal Stretches

Some approaches to intervention for deaf children start by trying to elicit isolated segments or words from the children, and then seek to build language systems by training children to combine those smaller units. This approach fosters a concept known as *generalization*, meant to refer to a process in which the child first masters production of small units acquired in isolation and subsequently learns to incorporate those units into longer stretches of language. The speech therapist's job is seen as twofold: first, teaching children to produce isolated segments, and then training them to move each of those segments to broader language contexts. Unfortunately that approach is an example of the proverbial placement of the cart before the horse.

The first goal of intervention, especially with young children and infants, should be to generate attention to speech in order to facilitate the attainment of appropriate perceptual organization. Children with CIs must learn to recognize speech as such—a signal generated by human speakers—and learn to organize that structure according to speech-relevant strategies. These goals are best realized by using long signal stretches in clinical and educational efforts with children. Intervention to correct errors at the segmental level should be implemented only after a child demonstrates a desire to communicate with spoken language, and is producing—or attempting to produce—speech in order to express needs, feelings, and wishes. According to this approach, intervention to improve production of smaller units serves to polish what the child is already attempting to produce. Thus an appropriate conceptualization of the therapy process might be one of *progressive refinement*, indicating that children's attention should first be directed to global structure, with gradual redirection to increasingly detailed structure. This approach matches children's typical developmental refinement of attention from whole-syllable or word structure to word-internal phonological structure.

Use Speech Signals to Teach Language

Precisely because signal components are perceptually organized differently as a function of whether expectations are that they are part of speech or nonspeech signals, only speech should be used in language learning experiences with children with CIs. The use of nonspeech signals trains children only to be more attentive to some cues when listening to the signals as nonspeech structures.

Aids to Perceptual Organization

In 2009, Nittrouer and Chapman reported outcomes for a subset of children in the longitudinal study reported in Chap. 11 titled Early Development of Children with Hearing Loss, or EDCHL. In that study, it was observed that the children who wore a hearing aid on the ear contralateral to the ear that received a CI for a period of a year or more after receiving that CI demonstrated better language abilities across the board than children who discontinued use of a hearing aid upon receiving a CI. That was true, regardless of the degree of hearing loss in the ear with the hearing aid. Furthermore, these benefits were found to be long-term, with some positive effect of early bimodal experience seen in language abilities measured at kindergarten (Nittrouer et al. 2012).

In another experiment, one unaffiliated with the EDCHL study, we sought to verify the effect more generally (Nittrouer et al. 2014). To do that, stimuli were constructed to simulate the signal provided by a CI, both when presented alone and when presented in combination with an acoustic signal in just a very low-frequency range (i.e., below 250 Hz). Materials consisted of sentences and isolated words that were high-pass filtered with a low-frequency cutoff of 250 Hz and used to create four-channel, noise-vocoded signals that simulated CI inputs. Those signals were presented alone, as well as in combination with the original signal (i.e., not vocoded) below 250 Hz. In two separate diotic conditions, either just the CI-simulated signal was presented to both ears or the combination signal was presented to both ears. In two dichotic conditions, either the CI-simulated signal was presented to one ear only or it was presented to one ear with the low-frequency signal presented to the other ear. Both adults and children served as listeners. Results showed significant improvements in speech recognition for the combined signals, regardless of whether the two signals were presented diotically or dichotically, for adults and children alike.

Those findings might seem surprising because in all cases in the simulation experiment, and in many cases for the children wearing CIs, the limited spectral structure available through the hearing aid (or simulated hearing aid) was not

sufficient to provide any explicit linguistic information. Listeners could not understand any words with the low-pass signal alone. Nonetheless, immediate improvements in speech recognition were observed in the simulation study, along with long-term benefits to children using the bimodal prosthetics. The most likely explanation for those benefits is that the very-low-frequency, naturalistic speech signal spurred recognition of the entire complex (low-frequency + vocoded signal) as speech. Consequently, perceptual strategies promoting the organization of the various signal components into speechlike form were easily invoked. That kind of effect means that the low-frequency acoustic signal can be viewed as an aid to perceptual organization: It reduces ambiguity about how signal structure should be organized.

Another factor that should similarly facilitate speechlike perceptual organization is audiovisual presentation. Being able to see the speaker's face should evoke speechlike strategies for the degraded signals provided by cochlear implants. But at least one approach to early intervention has long advocated against allowing infants and young children with hearing loss to see the talker (Beebe 1978; Estabrooks 2001; Luterman 1976; Pollack 1970, 1984; Power and Hyde 1997). It is an approach based on perspectives of sensory development and processing dating back to the nineteenth century contending that transmission of sensory information through each modality is encapsulated from the periphery to the brain, and any use of a different modality would diminish the entrainment of sensory processing through the primary modality (e.g., Goldstein 1897). However, more recent views of the nervous system, based on imaging and electrophysiological evidence, indicate that there is much more integrated processing of sensory information across modalities than the earlier perspective recognized (Kayser and Logothetis 2007). For example, some experiments have explicitly shown that neuronal responses in the primary auditory cortex are modulated (usually meaning they are enhanced) by simultaneous input from the visual system (Lehmann et al. 2006; Pekkola et al. 2005). At the same time, the evidence from the work of Imada et al. (2006), discussed by Kuhl (2010) and described earlier, indicates that input from any one modality can project to different parts of the cortex, depending on the nature of the signal: nonspeech signals are projected only to the primary auditory cortex, and speechlike signals tend to be projected to the motor cortex, as well. When it comes to training children with CIs to organize the degraded signals they receive through their implants according to speechlike principles, adding sensory input from the visual modality can surely promote the appropriate kind of organization.

The appeal made here for audiovisual speech input for children with CIs does not rest on traditional views of speechreading. Those older views suggested that listeners with hearing loss benefit from seeing the talker because specific features of phonemic categories can be obtained through vision that cannot be obtained through impaired audition (e.g., Erber 1972, 1975; Miller and Nicely 1955; Numbers and Hudgins 1948; Woodward and Barber 1960). According to that perspective, speechreading serves the purpose of providing information about place of articulation, which is hard to get through impaired hearing because it tends to be high frequency; amplified hearing provides information regarding voicing and manner of articulation, which can be derived from lower frequency signal components. Thus, according to that older perspective, listeners benefit from a process of sensory summation that increases the amount of information available. The argument made here is that providing the visual display of speech helps the child learn to perceptually organize the signal according to speech-appropriate strategies: the child becomes more certain that the signal is speech, so can process it accordingly. This latter effect was demonstrated in a study by Remez et al. (1998) in which visual information was supplemented by one of several sine waves, replicating either fundamental frequency or one of the three lowest formants. In that study, the greatest benefit of the audiovisual over the audio-alone condition was observed when the second formant was presented. That formant provides information primarily about place of articulation, which meant that the information provided by the visual signal and the audio signal was mostly redundant. Thus, the benefits of audiovisual presentation cannot simply be sensory summation. In this case, multisensory input led to sensory enhancement. Regarding their finding, Remez et al. concluded that "agreement between seen and heard speech promotes fusion" (p. 71), thus allowing the listener to process the stimulus as speech. That conclusion matches the notion of sensory integration proposed by Kayser and Logothetis (2007). These latter authors proposed that having redundant information from more than one modality can help perception by reducing uncertainty of the internal representation. It is critical that this kind of multisensory input is available to children with CIs who receive only a highly degraded signal through their prostheses. Rather than diminishing the integrity of the acoustic speech signal, providing a concomitant visual signal serves to strengthen that auditory representation.

Children Learn to Understand Speech by Producing Speech

The evidence presented above demonstrating that speech perception is a sensorimotor process can be used to support the suggestion that intervention with deaf children should involve ample opportunity to produce speech. This principle can also be illustrated with outcomes of the EDCHL study. At each age for which data were collected in that study,

Pearson product-moment correlation coefficients were computed between measures of speech intelligibility and several measures of language ability. The metric of speech intelligibility used was the Children's Speech Intelligibility Measure (CSIM), an instrument originally developed by Wilcox and Morris (1999) to investigate motor control and organization for speech production by children at risk of articulation disorder. In this task, children imitate 50 words. The instrument itself consists of 200 such word lists that are constructed from a master list of 600 words (50 sets of 12 possible words). Most words are of single syllable, but a few have two syllables. In this study, each word to be imitated was presented as an audio-video sample of a woman talking. Including the visual display meant that errors in recognition were minimized. All children's productions were audio-video recorded at the time of testing. Later, each child's productions were downloaded to a hard drive, and the child's word productions were separated into individual audio files. The video signal was discarded so that only audio samples of children's productions remained. Listeners unfamiliar with the speech of deaf talkers came to the laboratory and listened to these samples. The task of the listener was to select the word that was produced from the set of 12 phonetically similar choices. Each listener heard productions from only three children (with a maximum of two children with hearing loss) so that no listener would have the opportunity to become familiar with the speech of children with hearing loss. Two naïve listeners scored the samples from each child. Here we used the mean score from the two listeners for each child, and report these scores as the percentage of words the listeners identified correctly. These scores may be viewed as an index of how well the children were able to produce and organize the articulatory gestures required for clear production.

Table 20.1 shows the Pearson product-moment correlation coefficients between these CSIM scores and scores obtained on five other measures from the children with CIs, collected at both 48 months and second grade. These measures were selected because they were ones obtained at both test times. The only difference in tasks was that the auditory comprehension scores obtained at 48 months were from the Preschool Language Scales—4 (PLS) (Zimmerman et al. 2002), and those from second grade were from the Comprehensive Assessment of Spoken Language (CASL) (Carrow-Woolfolk 1999). It is apparent from this table that language scores at 48 months were highly correlated with speech production abilities. That outcome highlights the sensorimotor nature of processing for speech signals. By second grade, the relationship has diminished, which might be expected as more children with CIs develop good speech intelligibility. For these children, the mean speech intelligibility score was 57 % correct (SD = 18 % correct) at 48 months of age and 89 % correct (SD = 8 % correct) at second grade. There is much less variability at the later test age, and that truncation in variability might explain the diminished correlation coefficients. Nonetheless, these analyses suggest that there is a relationship between early motor control abilities for speech production and language learning for these children with severe-to-profound hearing loss.

In approaches that handle especially well this recommended teaching style of emphasizing speech production, therapists and teachers require complete morphosyntactic forms from children in all communications. Not only does producing speech help children learn about the organization of articulatory gestures, but also generating morphosyntactic forms helps to solidify that structure for the child. Children need to create complete and accurate utterances as often as possible, even when it means that the child needs to repeat using an extended form an utterance originally produced as an abbreviated form. This aspect of a well-designed intervention program might be the component that feels most unnatural to novice speech-language pathologists and teachers, but it has great payoffs.

Closely tied to the principle of requiring children to produce complete morphosyntactic forms as often as possible is the idea of recasting. This term refers to the practice of recreating in more complete and syntactically accurate form an utterance that a child tried to say. Thus, a complete sequence of events combining this technique and the one above would consist of the child trying to produce an utterance

Table 20.1 Pearson product-moment correlation coefficients between speech intelligibility scores from the Children's Speech Intelligibility and other language measures for children with CIs in the Early Development of Children with Hearing Loss (EDCHL) study, described in Chap. 11

	Expressive vocabulary	NDW	Auditory comprehension	MLU	Pronouns
48 months ($N=58$)	0.698[a]	0.669[a]	0.638[a]	0.701[a]	0.552[a]
Second grade ($N=50$)	0.232	0.429[a]	0.254[b]	0.318[c]	0.353[c]

Expressive Vocabulary represents standard scores from the EOWPVT; NDW is the number of different words in the first 100 utterances of a narrative sample; Auditory Comprehension represents standard scores from the PLS at 48 months and the CASL at second grade; MLU is mean length of utterance from the 20-min narrative sample; and Pronouns are the number of pronouns in the first hundred utterance from that narrative sample

[a] $p<0.10$ (2-tailed test)
[b] $p<0.05$ (2-tailed test)
[c] $p<0.01$ (2-tailed test)

(*More milk*, or even just *more*), the adult recasting it (*Oh, you want more milk*), and finally the child reformulating the original utterance according to the recast version (*I want more milk*). This exchange can and should be as natural as possible. The process should not involve the adult producing the exact version of what the child should say by using a directive (*Say,* "*I want more milk*"). This latter approach is sometimes used by well-meaning practitioners, but it results only in imitation on the part of the child. In fact, the goal is for the child to generate the correct morphosyntactic form on his own, with some appropriate prompting in the form of a recast.

Direct Language Instruction

Although the general perspective taken in this chapter is that language emerges in the child as a result of maturation and experience, children with CIs require some explicit instruction. Largely due to the diminished opportunity to access high-quality sensory input, children with CIs have decreased opportunity for the kinds of language experiences most children have. Background noise, room reverberation, and simple distance can all hinder a child's ability to hear spoken language, and so to have opportunity to generate responses. That experiential deficit forms the basis of the suggestion that children with hearing loss need specially enhanced experiences. Nonetheless, it would be a mistake to presume that extra-enriched opportunities for language experience alone would be sufficient to help children with CIs attain the same levels of language performance as children with normal hearing. In addition to the relatively natural experiences described thus far, children with CIs require direct language instruction.

This term, *direct language instruction*, is often invoked to refer to methods used with students who are second-language learners of English. Although the population of children being discussed here differs, the principles are the same. Essentially the term indicates that phonological, lexical, and morphosyntactic structure needs to be introduced explicitly to the student. General educational policies have moved away from this approach, placing an emphasis instead on naturalistic learning of the language needed for both casual and academic communication. That naturalistic approach is appropriate and sufficient for typically developing children without sensory deficits, precisely because language acquisition is such a natural process for them. However, children with hearing loss need direct instruction in order for them to learn explicitly linguistic forms.

In the preschool years, this kind of instruction can appear informal, involving games meant to introduce new vocabulary or morphosyntactic structures. For example, snack time can serve as an opportunity to teach the difference between mass and count nouns by varying the kind of food that is available: *I want a lot of pudding* versus *I want three crackers.* In the school years, the instruction can be more overt, with activities meant to help these children focus on phonological or morphological forms, or enhance their vocabularies. For example, learning Latin roots for English words can help children with CIs expand their knowledge of morphological structure. At all ages, however, it is essential that the direct instruction supplements naturalistic experiences, and is begun only after a child has started producing spoken language of substantial quantity.

At one time a popular method of teaching sentence construction to deaf children was the Fitzgerald Key, first developed by Edith Fitzgerald (1929). Those of us who worked in schools for the deaf prior to the 1990s recall the symbols that formed the basis of the Key, which was on chalkboards in every classroom. Other readers might recall seeing the Key on the chalkboard in William Hurt's classroom in the movie *Children of a Lesser God*, which came out in 1986. The Key had six columns and each represented a component of sentence structure. The first column was the nominal clause (Who, Whose, What). The second column represented the verb clause, marked with a special symbol (⌐¬). The third column was the objective clause, and so on. Using the Key, deaf children were taught to construct sentences through protracted curricula extending over several years. Again we find the cart positioned before the horse with this approach. Historically, methods such as this one likely contributed to the highly stylized language patterns that were characteristic of the speech of deaf children. Clearly the Fitzgerald Key is a method of direct language instruction that is too formal and poorly timed with regard to language development. Children with hearing loss need to be given opportunities to generate language naturally, with appropriate recasting, while direct instruction well timed from a developmental perspective is provided in the curriculum.

Continued Intervention Throughout Childhood

In 1968, Carol Chomsky completed her dissertation, titled The Acquisition of Syntax in Children from 5 to 10 (Chomsky 1969). This work was a demonstration of the kinds of complex morphosyntactic structures acquired after the start of elementary school by children learning language in typical fashion. These constructions often have to do with meanings that cannot be derived from the surface form. For example, the two sentences in each pair below share the same surface form, but the meanings are different:

John is easy to please.
John is eager to please.

Donald promises Mickey to do a somersault.
Donald tells Mickey to do a somersault.

In her investigations, Chomsky found that typically developing 7- to 8-year-olds often failed to comprehend the differences in these kinds of sentence structures when semantic and external cues were removed. Thus these are samples of language skills that do not usually emerge until after children start school. In complement to those syntactic trends, Liberman et al. (1974) demonstrated that typically developing children do not have sufficient sensitivity to phonemic structure to enable them to count the numbers of phonemes in monosyllabic words until they are in second grade, even though they are capable of counting the numbers of syllables in multisyllabic words at kindergarten. When it comes to lexical development, the term *restructuring* is commonly used to refer to the process observed for children in that 5–10-year-old range. In early childhood, children enter words into the lexicon using holistic forms. Gradually, up to roughly the age of 10 years, the lexicon is reorganized until it is eventually structured according to word-internal phonemic units (e.g., Ferguson and Farwell 1975; Storkel 2002; Walley 1993). In sum, there is a lot of language development that is not expected to happen until after children enter school.

No one involved in the care of children with CIs discounts the importance of starting intervention as early as possible. When implemented properly, that intervention results in the acquisition of language skills in roughly the normal range (i.e., better than 1 SD below the mean of typical children) for half the children born with severe-to-profound hearing loss by the time they are ready to enter school. However, based on these outcomes, an equally common view has evolved that deaf children are ready to graduate from early intervention prepared to acquire on their own the language skills typically learned after the start of regular school. But there is no basis for that assumption. There is no reason to suspect that children with CIs who acquired early language skills only with strong support will stop needing that level of support once they enter school. Special teaching and language experiences beyond what is afforded children with normal hearing need to be provided during the toddler-preschool years for these children to develop the skills that they do during that time. Similar kinds of support are required once they start school, as well, in order for them to continue the learning process. In their chapter on the syntax of deaf children learning English according to the oral method, de Villiers et al. (1994) listed three factors that are needed for the acquisition of a first language: (1) innate language acquisition mechanisms; (2) the natural unfolding of biological and cognitive factors with maturation; and (3) experience with high-quality inputs. There is no reason to suspect that the first two of these factors would be deviant in children whose only problem is a sensorineural hearing loss. These children have typical language acquisition mechanisms, and maturation of biological and cognitive determinants of language should unfold at the usual rate. However, the third requisite factor is more difficult to provide. It is critically important that the nature and timing of language input and experience be kept as close to a natural timetable as possible, if these children are to develop as their peers with normal hearing. That requires the provision of adequate support for children with CIs after they start school.

Another way to conceptualize the need for ongoing support for deaf children with CIs is by viewing speech and language learning as a series of sensitive periods. Currently, the notion of a sensitive period for language learning is viewed as one single entity that starts at or before birth, and narrows dramatically sometime prior to the start of regular school age. A visual representation of this phenomenon was developed by Tomblin et al. (2007), and is shown in the top of Fig. 20.3. But different language skills emerge at different ages in typical children, coinciding with the ontogeny of various biological, cognitive, and even social factors. Consequently there is no reason to envision a single, monolithic sensitive period. The characterization of serial sensitive periods, shown on the bottom of Fig. 20.3, might be a more realistic perspective (e.g., Newport et al. 2001). According to this view, different language skills emerge at different stages of childhood. Ongoing, strong support for language learning is required by deaf children with CIs to help them through all of these emergent processes.

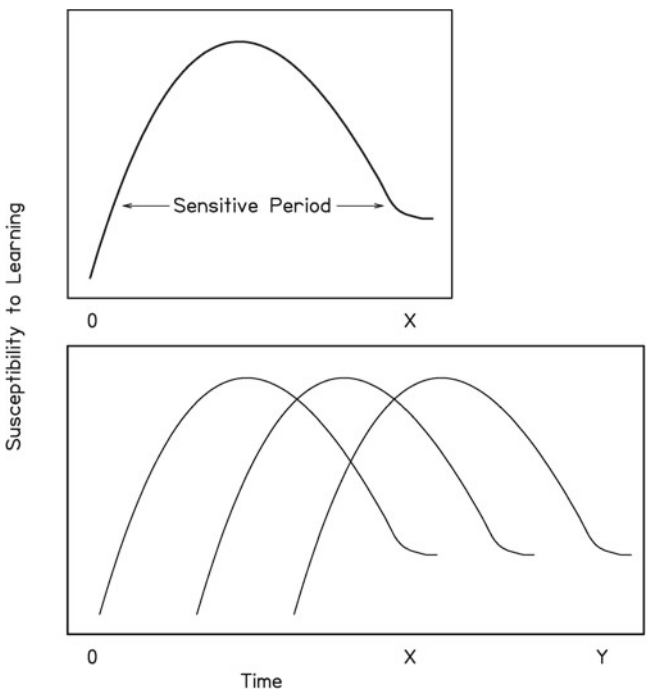

Fig. 20.3 Images illustrating the traditional conceptualization of a sensitive period for language learning, consisting of one such period extending from birth to roughly the start of school age (*top*), and a reformulation of this concept suggesting that a more appropriate perspective might view the phenomenon as serial sensitive periods extending into school age (*bottom*)

Other Treatment Considerations

Age of Implantation

One of the most highly debated treatment factors when it comes to children with CIs concerns how early a child should be implanted, specifically whether there is a need to push for very early implantation. There is widespread agreement that once unambiguous auditory thresholds have been obtained indicating hearing loss severe enough to warrant a CI, the child should receive it as soon as possible—assuming that there are no other medical considerations that might preclude implantation at that time. The issue in dispute specifically concerns how necessary it is to press to do the surgery very early—well before the first birthday—when there may be lingering diagnostic questions, medical concerns, or emotional issues on the part of family members.

Many studies of language acquisition in children with CIs report significant effects on outcomes as a function of the age of implantation for the first CI (Connor et al. 2006; Dettman et al. 2007; Kirk et al. 2000). For example, Geers and Nicholas (2013) showed that even after roughly eight and a half years of CI use, age of first implant still explained about 15 % of the variance in the latent language scores of a group of 60 children who received their first CIs between 12 and 38 months ($r = -0.396$). If there is a linear effect of age of first implant between 12 and 38 months, there is no reason to expect that relationship would be different below 12 months of age. Thus, "as early as possible" would seem the best policy when it comes to implants, and some investigators have explicitly reported benefits tied to implantation before the first birthday. For example, investigations by Houston and Miyamoto (2010) and Leigh et al. (2013) demonstrated better vocabulary scores for preschoolers who received first CIs before 12 months of age than for those who received first CIs between 12 and 24 months of age. However, these findings of significant effects of age of first implant are not consistently observed across studies. For example, Walker and McGregor (2013) failed to find any effect in a study of word learning by children with CIs.

In the EDCHL study, age of first implant was found to be a significant factor explaining language outcomes for children with CIs, but only for a subset of those children. Of the 50 children with CIs in that longitudinal study, 26 of them had continued to wear a hearing aid on the unimplanted ear for at least a year after they got their first implant. That group is referred to as the *some bimodal* group. The other 24 children ceased wearing a hearing aid around the time they received that first implant. Those children are referred to as the *no bimodal* group. Mean age of first implant was 22 months (SD = 14 months) for the children who had some bimodal experience at the time of their first implant, and 14 months (SD = 5 months) for the children who had no bimodal experience. The factor of whether children had some bimodal experience or not at the time of first implant turned out to be highly predictive of later language skills, and that effect could not be traced to other, potentially confounding factors such as socioeconomic status (Nittrouer and Chapman 2009). Table 20.2 shows Pearson product-moment correlation coefficients between each of the five language measures and age of first implant for each group. As can be seen, age of first implant explained significant amounts of variance only for the children with no experience wearing a hearing aid and a CI simultaneously. Based on these findings, the possibility presents itself that differences in whether or not age at first implant is found to explain significant proportions of variance in language outcomes across studies might be tied to whether the children included in the different samples tended to have some bimodal experience or not. That factor is rarely reported.

Bimodal Experience

The outcomes reported above regarding the effects of early bimodal experience are believed to reflect the important role that acoustic hearing—even if it is just a small amount—likely has on an individual's skill at perceptually organizing the degraded signal received through a CI. Even though the information provided is highly constrained, the amplified signal that infants with severe-to-profound hearing loss hear through high-powered hearing aids seems to be enough to help them learn to recognize speech signals as speech, and appropriately organize those signals. That experience with hearing aids may facilitate the shift in processing from the primary auditory cor-

Table 20.2 Pearson product-moment correlation coefficients between age of first implant and various language measures at second grade, for children who had some bimodal experience at the time of first implant and those who did not have any bimodal experience

	Expressive vocabulary	Auditory comprehension	MLU	Reading comprehension	Working memory
Some bimodal ($N = 26$)	−0.257	−0.339	−0.158	−0.094	−0.224
No bimodal ($N = 24$)	−0.404	−0.485*	−0.501*	−0.420*	−0.407*

Expressive Vocabulary represents standard scores from the EOWPVT; Auditory Comprehension represents standard scores from the CASL; MLU is mean length of utterance from the 20-min narrative sample; Reading Comprehension represents number of questions answered correctly about reading passages; Working Memory represents the number of words recalled in correct order. Correlation coefficients are all significant ($p < 0.05$) for the no-bimodal group; none are significant for the some-bimodal group

*$p < 0.05$ (2-tailed test)

tex to the motor cortex for speech signals, a shift observed by Imada et al. (2006) and described by Kuhl (2010) for children with normal hearing. It may be that children who either have only very limited experience with hearing aid use or discontinue wearing a hearing aid upon receiving a first implant must (re)learn how to organize the new signal they are hearing through their CIs, and (re)train the auditory system to project the input to the motor area of the cortex, starting from scratch. When no continued hearing aid use is provided, it makes sense that the earlier the first implant is received, the better. But children who continue to wear a hearing aid upon receiving a first implant may not have to go through the relearning process. It may be that consistent use of a high-powered hearing aid conditions the auditory system to handle acoustic inputs appropriately. As long as the new signal provided by the CI is accompanied by the hearing aid signal, it could be that the auditory system handles both inputs together, and according to the way the typically developing auditory system handles acoustic speech signals.

Of course, one potential challenge to the claim made above is that children who were given some amount of time with a bimodal configuration might have had better pre-implant auditory thresholds, thus biasing clinicians to provide that period of bimodal stimulation. And indeed that may have been the case for the children in the EDCHL study. Whereas the children who had no bimodal experience had mean pre-implant, three-frequency (0.5, 1.0, and 2.0 kHz) PTAs of 108 dB hearing level (SD = 11 dB), the children who had some bimodal experience had pre-implant PTAs of 97 dB hearing level (SD = 15 dB). Nonetheless, when Pearson product-moment correlation coefficients were computed between each of the language measures shown in Table 20.2 and pre-implant pure-tone average thresholds, none of these correlations were found to be significant. That lack of significance was observed when all children with CIs were included in the analysis. These correlation coefficients were also computed separately for the group of children who had some bimodal experience. Again, no significant relationships were obtained, indicating that even children with PTAs poorer than 100 dB hearing level stood to gain from a period of bimodal stimulation. Thus, in spite of the difference in pre-implant auditory thresholds, those thresholds are not able to explain any differences found for the two groups. It seems that even children with very little residual hearing in only the very low frequencies benefit from a period of wearing a hearing aid early in the language learning process.

Bilateral CIs

Another treatment option that is debated when it comes to children with severe-to-profound hearing loss involves bilateral CIs. There is a growing trend to give infants and toddlers with severe-to-profound hearing loss two CIs as soon as possible, a trend based at least partly on evidence from electrophysiological studies showing that having just one CI can lead to abnormal cortical organization in children (Gordon et al. 2007, 2008, 2013). However, where language is concerned, it is not clear what the effects of that abnormal organization might be. Certainly providing two CIs could be expected to promote auditory effects that derive from binaural listening, such as localization and spatial release from masking. Evidence of just those benefits has been observed, although these binaural effects are neither especially strong nor consistent across children with bilateral CIs (Grieco-Calub and Litovsky 2010; Misurelli and Litovsky 2012; Nittrouer et al. 2013). These diminished and inconsistent effects are likely attributable to problems with bilateral fitting (Kan et al. 2013), so research efforts are currently being undertaken to improve methods of bilateral fitting. Nonetheless, even if bilateral CIs are fit to maximize binaural effects, improvements in language acquisition are not assured. Each CI still provides spectrally degraded inputs. Children with unilateral CIs are already at risk for language delays, due precisely to the degraded quality of the input. It is not clear that having degraded signals at both ears should be expected to do anything to benefit language acquisition. Some studies have demonstrated better language scores for deaf children with bilateral CIs than for those with unilateral CIs (e.g., Boons et al. 2012), but that effect has not observed in this laboratory. Table 20.3 shows, in the top two rows, mean scores of language measures for children with one and two CIs at the time of testing. (Participant numbers in this table are fewer than in the last table because six children continued to use bimodal stimulation at the time of testing. Those children are not characterized as having either one or two CIs in Table 20.3.) In this table, a mean of the three phonological awareness tasks (initial consonant choice, final consonant choice, and phoneme deletion) was used as the metric of phoneme awareness, and is termed PA mean.

It appears from these data that children with two CIs performed better on most of the language measures. However, when scores from children with two CIs are separated into groups based on whether or not those children had some bimodal experience near the time of their first implant, shown in the bottom two rows of Table 20.3, it becomes clear that the advantage only extends to children with two CIs who had some bimodal experience. It is especially apparent from this table that scores for children with two CIs who had no bimodal experience, shown in the last row of the table, are similar to those for children with one CI, shown in the top row of the table. The children who performed the best on these language measures in this study were those who had some bimodal experience around the time of receiving a first implant and then went on to receive a second implant. These are the scores shown in the third row of Table 20.3.

Table 20.3 Means and *standard deviations* of language measures for children with one and two CIs are shown in the top two rows

	EOWPVT		CASL		MLU		PA		Comp.		W.R.		W.M.	
	M	SD	M	SD	M	SD	M	SD	M	SD	M	SD	M	SD
One CI ($N=13$)	89	14	91	13	5.01	1.14	44	21	15	5	99	13	38	11
Two CIs ($N=31$)	99	20	105	22	5.68	1.40	55	25	18	7	103	17	45	17
Some bimodal ($N=17$)	106	20	114	20	5.97	1.49	60	23	19	6	106	15	50	20
No bimodal ($N=14$)	91	18	93	21	5.33	1.26	48	26	16	7	100	19	39	10

Scores for children with two CIs are separated based on whether or not those children had some bimodal experience near the time of receiving a first implant, and means and *standard deviations* for those groupings are shown in the bottom two rows

Sign Support

Although this chapter has focused on children with CIs who have been growing up with the expectation that spoken English would be their first language, some parents elected to send their children to early intervention programs that supplemented spoken language input with sign language, either an English-based system or American sign language. A total of 17 children with CIs were in sign-supported programs during the preschool years. By second grade, however, no child remained in a sign-supported program or had a sign-language interpreter in school. Nonetheless, *t* tests were computed to see if there were differences in language performance based on early sign experience. The measures examined were expressive vocabulary (EOWPVT), auditory comprehension (CASL), MLU, phonological awareness (PA mean), reading comprehension (QRI), word reading (QRI), and working memory (number of words recalled in the correct order). The only measure to show a significant effect of sign exposure was MLU, $t(48) = 2.55$, $p = 0.014$: children who were in oral-only programs prior to starting kindergarten had a mean MLU of 5.76 (SD = 1.03) and children who had some sign exposure during those early years had a mean MLU of 4.79 (SD = 1.65). Those results were not differentiated based on whether the early sign system used was American sign language or an English-based system. (Some children with NH were also exposed to sign language early in life through the popular *Baby Signs* programs, but no differences in language abilities at this second-grade testing were observed for these children based on that sign exposure.)

Summary

Several broad ideas for intervention were discussed in this chapter. Based on empirical outcomes, it was recommended that children with CIs need intensive support for language learning throughout childhood. A model that carefully integrates enriched, naturalistic experience along with direct language instruction was recommended. The need for providing high-quality sensory input at all times was discussed. It was specifically recommended that the sensory input through the implant be supplemented by amplified acoustic hearing, at least for a while near the time of first implantation. An argument was made for providing visual input (i.e., speechreading) whenever possible. The importance of requiring children with CIs to generate and produce linguistic structures was highlighted. While cochlear implantation as early as reasonably possible is recommended, age of implantation for these children was not found to explain especially large proportions of variance in overall language outcomes. At least where language acquisition is concerned, no special benefits of two implants over one have been observed. At present, an intervention approach that provides a robust sensory input with a rich language environment offers the strongest means of helping children with hearing loss achieve their full language potential.

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Music as Communication and Training for Children with Cochlear Implants

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Kate Gfeller

The Role of Music in Childhood

Music, like speech, is a form of communication involving encoding, transmitting, and decoding of an intended message (Owens 2001; Gfeller 2008). The nature and functions of musical communication in the lives of children change along with their developmental capabilities and needs. Caregivers around the world use music to comfort their babies as part of parent–child bonding. Lullabies are made up of exaggerated but smooth pitch contours slightly larger than the pitch excursions in motherese and slow rhythmic sounds that promote joint attention, self-regulation, and emotional attachment (Gfeller 2008).

For toddlers, music is a common part of instructional and familial routines and informal play. The rich repertoire of children's songs includes lyrics that introduce vocabulary or concepts relevant to language development. From a pedagogical or habilitative standpoint, these playful and engaging childhood songs often provide ample repetition of vocabulary and concepts, and may be sung at a slower tempo than conversational speech. Action songs pairing lyrics with movements or gestures integrate auditory and motor systems. Psychosocial aspects of communication, such as turn taking and self-regulation, are supported through predictable but enjoyable routines (Gfeller 2008).

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As children mature, many families and schools encourage involvement in music instruction and ensembles. These experiences, which require personal discipline and adherence to social rules, promote cultural enrichment and self-expression (Gfeller 2008). From an auditory perspective, performing music involves ongoing exposure to rapidly changing fine-grained acoustic stimuli integrated with motor control. This multimodal experience places demands on a wide variety of higher order cognitive processes, and has been associated with experience-based plasticity in brain functions and structures (for review of music training and neural plasticity, see Herholz Sibylle and Zatorre 2012).

Adolescents are heavy consumers of music. It has been estimated that in adolescence, teens spend more time listening to popular music than they spend in the classroom from kindergarten through high school graduation (Davis 1985). Music plays an important role in psychosocial development through sense of identity, interpersonal relationships, and mood regulation (Behne 1997; Christenson et al. 1985; Christenson and Roberts 1998; North and Hargreaves 1999; North et al. 2000, 2004; Laiho 2004; Saarikallio 2007; Zillman and Gan 1997).

Because music is such a pervasive acoustic and sociocultural experience in most every culture, children who use cochlear implants are likely to be exposed to music on a daily basis. Acceptable music perception has practical implications for perceived benefit of the CI and full participation in society (Fujita and Ito 1999; Gfeller et al. 2000a; Gfeller and Knutson 2003). To what extent are different aspects of music experienced satisfactorily through the electrical stimulation of cochlear implants (CI)? The following section summarizes the technical characteristics of CIs in relation to key structural features of music, and the impact of electric hearing on perceptual accuracy and enjoyment of pediatric CI users.

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Music Perception and Enjoyment of Pediatric CI Recipients

Technical Characteristics of the CI in Relation to Music Perception

Music, like speech, is made up of complex sound waves with spectral and temporal envelopes that vary in time (Looi et al. 2012a; Limb and Roy 2014). However, music differs from speech with regard to perceptual requirements; this has important implications for CI users in relation to music listening. Extensive technical descriptions of CI technology can be found in Chap. 1 (Wilson, Dorman, Gifford, McAlpine), but briefly, several characteristics of CI design are poorly suited for transmitting important structural features of music.

One limitation is the range of frequencies transmitted by the device. Cochlear implants convey those frequencies considered most salient to speech perception, a range considerably narrower than frequencies produced by musical instruments (F0 and harmonics ranging from approximately 27 to 9300 Hz) (Limb and Roy 2014). Some recipients have described lower sounding instruments, such as the string bass or tuba, as "missing" or as sounding like short clicking sounds (Gfeller and Knutson 2003).

Another limitation is the manner in which the rich and complex elements of music are encoded by the CI (Looi et al. 2012a; Limb and Roy 2014). Present-day CI processing strategies usually remove the temporal fine-structure information in the stimulus wave forms and preserve the temporal envelopes extracted from 6 to 22 frequency bands; these are conveyed via the electrodes in the internal array. The electrode array has a small number of wide band-pass filters with fixed center frequencies, resulting in coarse spectral cues, and thus poor frequency resolution. This signal is effective in conveying speech perception in quiet as well as the rhythmic components of music. However, CIs are poorly suited for transmitting greater fine structure required for perception of pitch (which makes up melodies and harmonies) and timbre. In short, music perception of CI recipients is impacted significantly by the technical characteristics of the internal electrode array and signal processing strategies.

Interestingly, CI recipients who use similar technology differ considerably in perceptual acuity and enjoyment of music. Numerous factors, such as hearing history, residual hearing, hearing aid use, cognitive efficiency, and influential experiences (e.g., rehabilitation, education), contribute to this variability (Limb and Roy 2014; Gfeller et al. 2008, 2010; Hopyan et al. 2012).

Research Studies of Music Perception CI Users: Comparisons of Adults and Children

To date, most studies regarding music perception and CIs have been conducted with adult CI users (for reviews, see (Looi et al. 2012a; Limb and Roy 2014; McDermott 2004; Looi 2008)). A more modest body of research has focused on pediatric CI recipients, often comparing outcomes with those of adult CI users or children with typical hearing. Although pediatric CI recipients receive similar auditory input as adults, there are important differences to consider before generalizing adult research findings to the pediatric population.

Young pediatric CI users generally have greater neural plasticity than adults. However, their auditory pathways have developed primarily in response to electrical stimulation, which provides a degraded representation of spectrally complex features of music (e.g., pitch and timbre). Thus, pediatric CI users implanted early in life have few, if any, mental representations of how music sounds through a healthy hearing mechanism. These differences influence perceptual acuity as well as the types of contextual cues (top-down processing) that can be marshalled in listening to music. In addition, other maturational differences (e.g., cognitive and behavioral development) influence the scope and depth of music perception studies with children. The following section summarizes extant research with pediatric CI users.

Perception of Structural Features of Music by Pediatric CI Users

As noted previously, music is made up of complex and rapidly changing combinations of rhythm, pitch, timbre, and loudness. Of these features, rhythm is the component most effectively conveyed via the temporal envelope of the CI signal (e.g., (Looi et al. 2012a; Hopyan et al. 2012; Gfeller et al. 2012a; Hsiao and Gfeller 2011; Innes-Brown et al. 2013). *Rhythm*: Rhythmic components of music include tempo (i.e., slow, fast), the basic underlying beat (e.g., triple or duple meter), and rhythmic patterns (e.g., sequences of long or short notes) that provide durational organization to musical sounds. Pediatric CI users have similar perception as normal hearing (NH) children and adult CI users on tempo and rhythmic patterns (Innes-Brown et al. 2013; Stordahl 2002; Hsiao 2008; Olszewski et al. 2005; Mitani et al. 2007; Nakata et al. 2006). Thus, from a functional standpoint, pediatric CI users can perform similarly to NH peers on musical tasks such as clapping or dancing to a beat. Playing percussion

instruments can be a successful musical endeavor for children who have grown up using implants (Hsiao and Gfeller 2011, 2012).

Interestingly, perceptual accuracy does not guarantee optimal utilization of rhythmic cues within some musical tasks. For example, postlingually deafened adults are more effective than prelingually deaf CI recipients in using rhythmic cues to recognize familiar songs (Olszewski et al. 2006). Children may not realize the utility of rhythmic patterns in song recognition unless a direct prompt to do so is provided (Gfeller et al. 2011).

Pitch: Pitch, how high or low a note sounds, has been described as the most basic organizing structure in most musical cultures (Nakata et al. 2006; Patel 2008), and forms the basis for melodies and harmony. Perceiving the exact magnitude and direction of sequential or concurrent pitch relations contributes to recognition of melodic contours and familiar melodies, detection of errors in melodies, and perception of harmonies (Looi et al. 2012a; Gfeller et al. 2006). In short, the poor spectral resolution conveyed through the CI undermines accurate perception of pitch, and consequently melody and harmony (Looi et al. 2012a).

The largest proportion of extant pediatric music studies examines perception of pitch pairs, patterns, or melodies. These studies vary in stimuli presented, response tasks (e.g., recognition, discrimination), and extent of contextual cues [e.g., Hopyan et al. 2012; Olszewski et al. 2006; Vongpaisal et al. 2004]. Some studies have tested recognition of highly familiar songs, examining the listener's use of various structural features (pitch, rhythm, timbre). This includes recordings or synthesized versions of familiar children's songs such as "Twinkle, Twinkle" or "Happy Birthday" (e.g., Stordahl 2002; Olszewski et al. 2005; Hsiao 2008; Jung et al. 2012), or theme songs from children's TV programs, presented either as originally recorded or in manipulated versions (Mitani et al. 2007; Nakata et al. 2006; Scorpecci et al. 2012; Trehub et al. 2009; Vongpaisal et al. 2006).

CI recipients vary considerably on these pitch-based tasks; some perform at chance level or below, while a few "star" users have performed as well or nearly as well as some NH children in some tasks. In general, however, pediatric CI recipients as a group are significantly less accurate than NH children in recognition or discrimination of familiar melodies, melodic contours, or pitch ranking, especially in listening conditions reliant primarily upon pitch cues (e.g., no available rhythmic, timbre, or lyrics cues) (Mitani et al. 2007; Nakata et al. 2006; Olszewski et al. 2006; Scorpecci et al. 2012; Trehub et al. 2009; Vongpaisal et al. 2006). Thus, from a functional standpoint, musical tasks such as describing or responding to the directional changes of pitches (e.g., "Reach up high when the melody goes higher.") or recognizing melodies based upon pitch cues tend to be very difficult for many pediatric CI users (Hsiao and Gfeller 2011).

Pediatric CI users compared more favorably with NH peers in closed-set task responses, when perceptually accessible features are available (e.g., rhythm, song lyrics), or with cues learned through instruction or prior listening experiences (Hopyan et al. 2012; Mitani et al. 2007; Olszewski et al. 2006; Vongpaisal et al. 2004; Nakata et al. 2005). Thus, in real-life situations, instructors can support music listening through modifying the response task (e.g., closed-set tasks) or providing more accessible cues (e.g., visual input, rhythm, or lyrics).

Poor pitch perception also impacts pitch production. From a functional standpoint, tuning a musical instrument or singing in tune is typically quite difficult for children with CIs. Several studies (Nakata et al. 2006; Xu et al. 2009) indicate that young CI users (ages 4–9) are significantly less accurate than NH hearing peers in tasks such as pitch matching, accuracy in singing pitch contours, and singing exact interval changes. Therefore, musical activities that require in-tune singing (as opposed to singing for enjoyment or to encourage vocal use) may be among the most challenging or frustrating tasks for many pediatric CI users.

Timbre Recognition: Timbre, which comprises the unique onset transients, steady state, and decay of acoustic energy of the harmonics created by different musical sources, is important in identification of musical instruments (e.g., a flute vs. a piano) or singers (e.g., The Muppets vs. Justin Bieber). Pediatric implant recipients can often differentiate between two sounds with distinctly different temporal cues (e.g., piano vs. violin tone). In contrast, they may find it more difficult to differentiate between more similar sounds (e.g., saxophone vs. a clarinet) (Gfeller et al. 2011). Jung et al. (2012) found that timbre discrimination, while significantly poorer than adult CI users ($p = 0.039$), was significantly higher than chance level ($p = 0.027$). Innes-Brown et al. (2013) found that children using CIs were more accurate in identification of percussive as opposed to non-percussive instruments, and temporal cues can be useful in perception of timbre as well as rhythmic elements.

Timbre Appraisal (Sound Quality): Most people listen to music for enjoyment; therefore, from a functional standpoint, accurate timbre recognition is arguably less important than is pleasing sound quality. Interestingly, many pediatric CI users enjoy listening to music (Gfeller et al. 1999, 2012a), despite the atypical sound quality. This may be in part because prelingually deaf pediatric users have experienced music primarily through their CIs, and thus cannot make critical comparisons with music heard prior to hearing loss. Research indicates that some instruments sound more pleasant than others (Gfeller et al. 1999). Practically speaking, parents or instructors can encourage exploration to determine which instruments sound most pleasant to a given individual.

There should be a number of instruments with sounds sufficiently pleasant to CI users to support music enjoyment (Hsiao and Gfeller 2011; Gfeller et al. 2011).

Music Engagement and Participation

CI recipients vary considerably in music engagement (listening, participation, etc.) as well as perceptual accuracy. Some children dislike or lack interest, while others thoroughly enjoy and engage in music (Gfeller et al. 1999, 2012a; Gfeller 2000). Pediatric CI users enroll in music instruction, listen to music, watch music videos, attend concerts, and dance (Gfeller et al. 1999, 2011, 2012a). In a recent study of a cohort of pediatric CI recipients who have now reached adolescents or young adulthood, 66% described music as being important or very important in their lives (Gfeller et al. 2012a). Some CI users with greater residual hearing find that hearing aids used in conjunction with their CIs enhance sound quality (Gfeller et al. 2008; Looi et al. 2008; Gfeller 2009). Furthermore, greater familial involvement, peer group values, or exposure to musical training can all have an impact on music enjoyment (Gfeller et al. 2000b, 2012a; Stordahl 2002; Mitani et al. 2007; Hsiao 2008; Trehub et al. 2009; Vongpaisal et al. 2006; Nakata et al. 2005; Rocca et al. 2012; van Besouw et al. 2011).

Summary

Because music is so prevalent in educational, social, familial, and spiritual aspects of life, supporting pediatric CI recipients in their engagement with musical experiences can contribute to normalization and quality of life (Gfeller et al. 2012a; Hsiao and Gfeller 2012; Gfeller and Darrow 2008). Music does not sound the same as for NH children, but judicious choices of musical experiences and familial support can help pediatric CI users to have more positive experiences (Gfeller et al. 2012a). In addition, a modest body of research suggests that focused training can also enhance perceptual accuracy and enjoyment. Music training is the focus of the following section.

Training to Enhance Music Perception and Enjoyment of CI Recipients

An Overview of Research on Music Training for CI Users

To date, the majority of studies regarding music training of CI users have been conducted with postlingually deaf adults. Training protocols for adults have included computer-based programs or socially oriented support groups (for review, see Looi et al. 2012a). Through training, adult CI users have improved on melodic contour recognition (Galvin III et al. 2012; Galvin et al. 2007), complex (real-world) melody recognition (Gfeller et al. 2000b; Galvin et al. 2007), timbre (musical instrument) recognition (Gfeller et al. 2002; Driscoll 2012), improved appraisal ratings (sound quality) (Gfeller et al. 2002; Looi et al. 2012b), or general enjoyment and participation in music experiences (Looi et al. 2012b; Plant 2012). Thus, there is some evidence that CI users are capable of improving some aspects of music perception through focused training.

Comparisons Between Adult and Pediatric Training Studies

While studies with adults indicate that improved perception and enjoyment are possible, caution should be used in wholesale generalization of these studies to pediatric users. Many adult CI recipients use their memory of musical sounds prior to hearing loss to make sense of the signal (top-down processing). For pediatric CI users with severe congenital or prelingual losses, their auditory pathways have developed differently in response to electrical stimulation (Torppa et al. 2014). These children do not possess the spectrally rich mental representation of musical sounds that develop through many years of typical acoustic stimulation. Therefore, some perceptual tasks may be a poor "fit" for their mental representation of some sounds. In addition, training protocols that emphasize cues learned before hearing loss may be less suitable for pediatric CI users (Olszewski et al. 2006).

Developmental and motivational differences are also important to consider. Although children have the advantage of greater neural plasticity than adults, adults typically have greater maturation with regard to sustained attention and understanding of abstract concepts. Adult CI users may be intrinsically motivated to complete self-administered online training exercises that may or may not be "fun" (Gfeller 2001). In contrast, younger children may require protocols that are shorter in length, more playful, geared toward the individual child's attention and motivation, or occur within a socially motivating context (Hsiao and Gfeller 2011; Gfeller et al. 2011).

Some published accounts of music training for pre- and school-aged children have included playing of instruments, singing, listening, and moving to music, with an emphasis on exploration and enjoyment of sounds (Gfeller et al. 2012a; Hsiao and Gfeller 2011; Innes-Brown et al. 2013; Rocca et al. 2012; Abdi et al. 2001; Chen et al. 2010; Yucel et al. 2009). These protocols are the focus of the following section.

Published Studies on Pediatric Training to Enhance Music

To date, a small number of published studies have examined the effects of music training on pediatric CI users (Innes-Brown et al. 2013; Rocca et al. 2012; Abdi et al. 2001; Chen et al. 2010; Yucel et al. 2009). This is not surprising, given the considerable effort and resources required to implement and evaluate musical training. Parents of children with hearing losses have lives complicated by a host of educational and clinical concerns, which can undermine enrollment and persistence in longer term studies. Speech and hearing professionals may have limited knowledge of musical development and pedagogically sound music approaches associated with different ages (Hsiao and Gfeller 2011, 2012; Gfeller et al. 2011; Gfeller 2000). The development of protocols providing sufficient listening experience is time consuming and can be costly. The selection of assessment tools that are valid and reliable for children of various ages, and which reflect the varied aspects of music perception and production, is challenging.

As we examine extant research, some studies have enrolled children ranging in age from preschool to adolescence. Even for children with typical hearing, there are important developmental changes in music skills (as is true for speech and language) over a wide age span. As a point of comparison, consider the methodological and clinical challenges that audiologists and speech-language pathologists face when selecting suitable outcome measures of speech or speech/language training protocols for children ranging from age 2 to age 14. Similar concerns are associated with selecting appropriate music training and outcome assessments.

Although the developmental milestones of music perception and production are less fully and clearly defined than for speech and language, nevertheless, there are well-documented developmental changes in musical capabilities for NH children from preschool through high school (e.g., ages at which children can reliably differentiate timbre, pitch, rhythm patterns, tonal center, etc.). The sorts of musical concepts and response tasks (and thus valid and reliable outcome measures) that are realistic for 14-year-olds are quite different from those of preschool children (Gfeller 2008; Hsiao and Gfeller 2011). For example, we might expect a 3-year-old to detect the presence of sound or to discriminate between two different rhythm instruments with very different auditory characteristics. In contrast, a 14-year-old could be expected to use abstract notational systems in conjunction with complex patterns of sound. The 14-year-old would also possess much greater motor control and sustained attention required for more advanced musicianship. While a well-trained instructor or therapist can engage children of many different ages in musical play, the wide age range reported in some published studies brings up interesting questions with regard to implementation of training and interpretation of outcomes.

In a clinical narrative, Christine Rocca described that musical instruction for children aged 4–16 sustained over months or years, as part of ongoing deaf education (Rocca et al. 2012). In that curriculum, musical experiences were geared toward developing musical potential and sensitivity. Musical tasks were designed with different developmental stages and individual capabilities in mind. Thus, the training tasks and expected outcomes evolved as children matured and reached particular educational objectives. Rocca reported through narrative accounts that the children in their program have improved pitch matching, singing correct intervals, music listening, instrumental music performance, and general engagement in music. While current publications by Rocca have not focused on perceptual data, videos of Rocca's training presented at professional conferences have presented remarkable results by some children on tasks as difficult as vocal pitch matching (Rocca et al. 2010).

Innes-Brown and colleagues examined the impact of weekly music education classes (45 min per week) for groups of students (age 9–13 years) using CIs or hearing aids (HA) (Innes-Brown et al. 2013). Music instruction included vocal play and integration of aural, visual, and kinesthetic modes of learning—pedagogical approaches considered appropriate for children in upper elementary through middle school. The children were assessed before and after 24 weeks of training for discrimination of pitch and rhythm patterns and recognition of percussive and non-percussive instruments. Timbre recognition was greater for percussive instruments, suggesting greater usefulness of temporal cues. There were no significant perceptual improvements for pitch patterns, rhythmic patterns, or timbre that could be attributed to the training. However, the instructors reported that the children showed enhanced appreciation of music-based activities.

In another study, 23 children ranging in age from 2.5 to 12.5 years participated in weekly music classes (4 children per class) using Orff methodology and Se-tar lessons (Abdi et al. 2001). Details regarding the training protocol were not reported. The length of training varied from 3 to 12 months, depending upon the interest/motivation and availability of each child and their parents. Teachers rated each child on a scale from 1 to 10 on musical skill development and enthusiasm for music; no perceptual data were reported. Per these narratives, the children varied considerably in skills and interest, but all showed appreciable progress in instrument playing, albeit considerably slower than for NH children.

In another study, 27 CI recipients (ages 5–14 years; $M = 6.7$) participated in YAMAHA music instruction for between 2 and 36 months (mean of 13.2 months) (Chen et al. 2010). The authors described the training as "listening, singing, score reading and instrument playing" (p. 794). Following training, the children were tested individually on

a pitch ranking task of 49 tone pairs (256 Hz to 495 Hz) of prime (same note) to 11 semitones played on the piano by the test administrator. Individual accuracy on pitch ranking varied from 9.5 to 92.5 %. The duration of training was positively correlated with pitch perception accuracy ($r^2 = 0.389$, $p = 0.045$). There were no significant correlations between pitch accuracy and age at implantation, gender, or type of CI. However, children older than 6 years were more accurate than those younger than 6, which may reflect maturation as well as effects of training.

Another training protocol involved a home-based program for individual children, with parents implementing brief 10-min listening exercises (using electronic keyboard) distributed over a 2-year period (Yucel et al. 2009). Nine pediatric CI users (chronological age not reported) participated; their responses were compared with a control group of nine children not enrolled in music. Both training and control groups were enrolled concurrently in auditory-verbal training as part of their normal routine. The parents played pitch pairs, rhythm patterns, and short songs. The children also learned about sounds by exploring the keyboard. Diary entries completed by parents indicated that children engaged in a total of 116.87–175.42 (or approximately 2–3 h total) minutes of music training over the 2-year time frame.

The parents used a rating scale of one to five points to evaluate their child's responses to music at 12 and 24 months on the following: sound awareness, general reaction to music, voluntary participation in music outside of the training exercises, discrimination and identification of pitch and rhythm patterns, and emotional responses to music. No perceptual measures of music were taken. Speech perception (sound detection and identification, word and sentence identification) was measured prior to music training, as well as at 12 and 24 months. At the 12-month period, no differences were noted between the training and control groups on any measures. By 24 months, the music group had higher ratings for interest and awareness of music in daily life; no significant differences were found on any speech measures. The authors concluded that music training helped music appreciation and "may enhance their progress in other auditory domains" (p. 1043).

The training methods in these published studies would be difficult to replicate, given that the flexible and playful approaches used in the studies do not lend themselves to highly controlled training parameters (e.g., stimuli, format, exact length of training) more commonly used with adults. Furthermore, it can be difficult to interpret the outcomes, given the possible impact of physical, cognitive, and social maturation, or other influential factors co-occurring with music training (e.g., music in the home, private lessons, extent of parental input, SES, maternal education, differences in individual auditory factors). Consequently, many questions remain regarding what types or dosage of music training and outcome variables would be most effective or suitable, especially given the well-documented diversity among pediatric CI users on many parameters.

While more research may shed light on the most efficacious habilitative approaches, given the considerable variability among CI recipients in maturation and perceptual capabilities, one training approach will not be ideal for all children. Children and their families will also vary in their priorities with regard to music, with some families considering music a central part of their lives, while others may consider music of very limited import. Assessment of familial attitudes and resources as well as auditory capabilities is relevant when considering music-based training as part of habilitation plans.

Summary

To date, a modest body of studies has examined the impact of music-based training on perceptual accuracy and enjoyment. As a group, these studies indicate that children with CIs can engage in music, and some show enhanced enjoyment or perception. Particular musical features (e.g., temporal cues) appear more readily accessible. Interpretation of some studies is difficult, however, given the limited accounting of training protocols, the variability among participants, and limited perceptual data. More research is needed to confirm the type and extent of benefit from particular training methods with specific subgroups of the CI population.

In the past decade, interest in music training for CI users has expanded beyond improved music perception and enjoyment, toward possible transfer to speech and language development. Given perceptual limitations that CI users have for musical sounds, it may seem incongruous that music training could be an effective clinical tool. Therefore, the following section discusses research regarding convergences between perception of music and of speech, which has triggered interest in music-based training for CI recipients.

Music-Based Training to Enhance Speech and Language

A growing body of research has examined relations between musical and spoken communication. Three types of studies suggest possible generalization of music-based training to speech: (1) research with NH listeners implying overlap in brain networks that process acoustic features heard in both speech and music; (2) correlations between perception of music and speech, relative to electric hearing; and (3) common pairing of melodic and linguistic information.

Research with Normal Hearing Listeners Regarding Overlap in Neural Encoding of Music and Speech: Can Music Training Transfer to Non-musical Domains?

Like speech, music is made up of complex and dynamic modulations of acoustic parameters. Music activates a widespread bilateral network of brain regions (frontal, temporal, parietal, subcortical), which is associated with increased arousal and attention and draws upon working memory, semantic and syntactic processing, motor functions, and emotional response (Besson et al. 2007; Kraus and Skoe 2009; Moreno et al. 2009; Musacchia et al. 2007, 2008; Schön et al. 2008; Wong et al. 2007). Music listening and performance include tasks such as segregating a particular timbre, melodic or rhythmic pattern, from competing auditory input; this requires ongoing decoding of and comparisons with past structural features. The heightened fine-grained frequency discrimination in response to ongoing changes in acoustic parameters has been attributed with improved auditory working memory, attention, and more rapid spectro-temporal processes at various levels of the auditory system (Patel 2011). Furthermore, music involvement is often associated with arousal, reward, positive mood, and social factors, which increase motivation and may enhance perceptual efficiency. This can promote careful listening and contribute to sustained listening over time (Herholz Sibylle and Zatorre 2012; Patel 2011).

A growing body of research regarding experience-based neural plasticity, primarily conducted with NH listeners, suggests an overlap in brain networks that process acoustic features heard in both music and speech (reviews by (Herholz Sibylle and Zatorre 2012; Patel 2011; Shahin 2011). The higher perceptual demands along with motivational aspects of music listening may "fine-tune" the auditory system (Ingvalson and Wong 2013). The expanded prosody and rhythmic cues associated with melodic presentation of lyrics may benefit language learning. Several studies suggest that music experience/training may generalize to skills such as phonological processing, verbal memory, learning mechanisms for language, and lower perceptual thresholds for complex auditory input (Herholz Sibylle and Zatorre 2012; Besson et al. 2007; Kraus and Skoe 2009; Moreno et al. 2009; Musacchia et al. 2007, 2008; Schön et al. 2008; Wong et al. 2007, 2009; Patel 2011; Shahin 2011; Chermak 2010; DeThorne et al. 2009; Foregeard et al. 2008; Ho et al. 2003; Kraus et al. 2009; Parbery-Clark et al. 2009; Pior and Ortiz 2009; Strait et al. 2009, 2013; Thiessen and Saffran 2009; Chobert et al. 2014; Kraus and Chandrasekaran 2010; Tierney and Kraus 2013). It is important to emphasize that most of these studies included NH listeners engaged in extended training, often initiated in early childhood.

Researchers have suggested possible implications for music-based therapy approaches with persons who have communication deficits associated with dyslexia, specific language impairments, autism, aphasia, and hearing loss (Besson et al. 2007; Kraus and Skoe 2009; Moreno et al. 2009; Musacchia et al. 2007, 2008; Chermak 2010; DeThorne et al. 2009; Parbery-Clark et al. 2009; Thiessen and Saffran 2009; Overy 2000, 2003). At present time, very few formal studies have assessed the impact of music training on the speech and language development of CI recipients. One thread of inquiry suggesting possible benefits is correlations between perception of pitch-based structures in music and spectrally complex aspects of speech relative to electric hearing.

Perceptual Requirements of Music and Speech, Relative to Electric Hearing

As noted earlier in this chapter, the perceptual requirements for speech and music differ. Remarkably accurate word recognition is possible even with spectrally coarse information as long as the listening environment is relatively quiet (Wilson 2000). However, coarse spectral information (and thus poor frequency resolution) transmitted by the CI is problematic not only for pitch and timbre but also for perception of paralinguistic features of speech (e.g., lexical tones, linguistic or affective prosody) and speech perception in noisy listening conditions (Gfeller et al. 2007; Qin and Oxenham 2003).

Several studies with adult (Gfeller et al. 2007, 2009, 2012b; Wang et al. 2011) or pediatric CI users (Hsiao 2008; Torppa et al. 2014; See et al. 2013; Peng et al. 2008) have documented significant correlations between pitch or melody perception and speech in background noise, phoneme discrimination abilities, lexical tones, and prosodic features of speech; this suggests shared perceptual mechanisms in electric hearing. In relation to music training, Torppa et al. (Torppa et al. 2014) found significant correlations between pure-tone pitch discrimination, digit span scores, and sentence stress perception. They also found greater improvement longitudinally among pediatric CI users ($n=8$, ages 4–13) who had participated in ~16 months of music activities (e.g., singing, playing instruments, or dancing at least once a week) than 13 children in a control (no music) group. These findings suggest that enhancement of music perception may possibly generalize to some aspects of speech processing.

Functional Uses of Music That Commonly Pair Melodic and Linguistic Information

From a functional perspective, musical tunes are commonly paired with linguistic information (song lyrics) in social and educational situations (Gfeller 2008). Childhood songs are often packed with vocabulary and concepts important for children to learn, often repeated numerous times. For example, childhood favorites like "Old MacDonald Had a Farm" contain repetitions of each animal's name as well as speech sounds (e-i-e-i-o; baa, moo, etc.), and thus offer multiple opportunities to hear and produce target vocabulary. Songs can also be legitimately presented at a rate somewhat slower than conversational speech, providing additional time for auditory processing. The expanded rhythmic and prosodic cues of songs are believed to support language learning. In addition, the ritualistic nature of many children's songs helps youngsters to predict upcoming musical and speech patterns or behavioral transitions (Hsiao and Gfeller 2011, 2012; Gfeller et al. 2011; Gfeller 2000). The pairing of song lyrics with melodic patterns also has bi-directional benefits with regard to perceptual accuracy, in that the lyrics of songs may assist CI recipients in song recognition (Olszewski et al. 2005; Nakata et al. 2006; Gfeller et al. 2011; Hsiao 2008; Vongpaisal et al. 2006).

The ease or difficulty of recognizing or understanding lyrics in song recognition is influenced, however, by the familiarity of vocabulary in the lyrics, the tempo at which the lyrics are sung, modifications in phoneme production associated with singing (singer's formant, changes in production associated with particular vocal styles such as opera, rock music) (Gregg and Scherer 2006), or masking properties of instrumental accompaniments (Gfeller et al. 2007, 2008; Fetterman and Domico 2002). Each of these aspects can be modified by the clinician, educator, or parent to match the current functional level of each child, or to practice more challenging skills. Initially, it may be best to present new vocabulary without accompaniment. However, eventual addition of accompaniment challenges the child with a more complex listening task, which can help to fine-tune auditory processing skills.

Summary

Several bodies of literature suggest that music shares common perceptual processing requirements, and that music may have particular advantages as a habilitative tool. The following section will focus on those conditions under which music-based training is more likely to transfer to speech perception, and offers practical suggestions for clinical practice.

Applying Principles of Music-Based Training to Pediatric CI Users

Because habilitation tends to be a long and intensive process, clinicians and families are eager to identify efficient and effective clinical options that are sufficiently engaging to support persistence. A body of well-established habilitative tools/approaches developed by speech, language, and hearing professionals are available; consequently, it is important to consider whether music-based training, in general terms, could be a beneficial complement to accepted therapies.

From a theoretical perspective, as noted previously, there is growing evidence that the perceptual demands in music listening may have particular advantages in fine-tuning the auditory and cognitive processes that also apply to speech (Wong et al. 2007; Ingvalson and Wong 2013). However, as noted previously, the majority of studies regarding music training have focused on NH people with typical auditory development and long-term training, sometimes commencing in early childhood. Only a small proportion of CI users are likely to have had extensive music training prior to deafness (Gfeller 2001), and very early or intensive musical training post-implantation is not realistic for many families.

Generalization and interpretations of studies with NH listeners is complicated further by the many training protocols (e.g., auditory, multimodal, instrument playing, computerized listening tasks), variability among participants (e.g., age, auditory status), and outcome variables (e.g., MEG, fMRI, behavioral measures Herholz Sibylle and Zatorre 2012). Some training tasks and outcomes appear more relevant to clinical interventions and desired functional outcomes for CI users. Current evidence specific to CI recipients is preliminary in nature, especially for pediatric users (Looi et al. 2012a; Shahin 2011). More research is needed to establish if music-based training has particular advantages, and if so under what circumstances.

While future studies will hopefully provide clearer guidelines, in the short term, clinicians, educators, and families must, nevertheless, make decisions regarding habilitative options for their children. The applicability of music-based training for the CI population can be evaluated through clinical observations and ongoing dynamic assessment. Music-based training should be considered in relation to the unique strengths and needs of each pediatric user. For example, is music an important aspect of family life? If so, what aspects of music engagement are important for familial and social integration? Does the child respond positively to music? If so, might musical engagement provide motivation to persist in habilitation programs for speech as well as music? Because of the considerable variability among CI users on a host of factors (age, hearing history and profile, familial priorities, etc.), it is unlikely that all children will actually benefit, or benefit equally from music-based training.

One framework for evaluating application of music training can be drawn from the writings of cognitive neuroscientist, Aniruddh Patel, who outlines conditions that are essential for music-driven plasticity to generalize to linguistic outcomes (Patel 2011). According to Patel, music does not automatically provide a superior stimulus for auditory training relative to speech and language; rather, music-driven plasticity will be more likely to benefit speech processing networks if it occurs under particular conditions which he describes as his "OPERA hypothesis."

Conditions That Support Transfer to Speech

Patel's acronym, OPERA, refer to the following conditions: (1) *Overlap*: anatomical overlap in brain networks that process acoustic features used in both music and speech; (2) *precision:* music places higher demands on shared networks than speech with regard to precision of processing; (3) *emotion*: musical activities tend to elicit strong positive emotion; (4) *repetition*: musical activities engaging these neural networks are frequently repeated; and (5) *attention:* music activities are associated with focused attention. Let's consider these conditions in relation to music-based training for pediatric CI users.

Overlap: Preliminary correlational evidence relating pitch, melody perception, and speech measures (e.g., prosody, lexical phones, phoneme identification) suggest shared mechanisms through which speech and music are processed (e.g., (Torppa et al. 2014; Ingvalson and Wong 2013; See et al. 2013). These studies imply that Patel's first condition is fulfilled.

Precision: Because music requires greater precision in listening to fine timing cues (see Sect. "Research with Normal Hearing Listeners Regarding Overlap in Neural Encoding of Music and Speech: Can Music Training Transfer to Non-Musical Domains?"), Patel believes that music-based training can drive the system and benefit neural encoding of linguistic as well as musical sounds. Similar recommendations have been made by other hearing professionals and cognitive neuroscientists (e.g., Ingvalson and Wong 2013; Chermak 2010; Kraus et al. 2009).

Specific to CI users, this condition for benefit is complicated by the coarse representation of spectral information conveyed by conventional CI signal processing. This degraded input shapes the development of auditory pathways in pediatric CI users, and restricts access to fine spectro-temporal cues in music. Moreover, damage to the auditory system associated with hearing loss can further undermine utilization of advances in signal processing.

Available behavioral data from CI users reveal highly variable outcomes in extracting meaningful spectral information from signal processing (e.g., pitch or timbre perception) (Looi et al. 2012a; Gfeller et al. 2008, 2010). Some CI users appear to be able to extract sufficient fine structure to support perception of some spectrally complex sounds. For others, one could argue that the lack of fine structure and discriminable features in the CI signal is of sufficient consequence to undermine benefit from music training, as observed in NH populations. However, Ingvalson and Wong (2013) suggest that music training that provides practice with rhythm, timing, and sequencing may provide a good foundation for the development of auditory skills that are also integral to cognitive and language skills.

The use of spectrally rich (as opposed to pure tones) stimuli in training has been supported through several studies of adults with CIs (Galvin III et al. 2012) or NH persons training with CI simulations (Loebach and Pisoni 2008; Loebach et al. 2009). More complex training stimuli (e.g., melodic contours paired with masking, complex instrumental sounds) resulted in greater perceptual enhancement for some listeners, both for music and transfer to various linguistic tasks (Loebach and Pisoni 2008; Loebach et al. 2009).

Data from published music-based training with CI users indicate highly varied results (e.g., Galvin III et al. 2012; Chen et al. 2010), suggesting that some CI users may perceive enough discriminable features to achieve benefits from training. Other psychological factors such as attention, motivation, and use of contextual cues are also likely to influence training benefit. It is not yet clear whether CI users who access more fine structure in the first place, and are therefore more motivated to listen more frequently to music (which in turn promotes neural adaptation), or if greater exposure to musical experience can also help those with less robust auditory systems to improve. These questions will require ongoing research efforts; for the present, careful clinical observation of individual change over time should be monitored.

Next, let us consider three conditions of Patel's OPERA hypothesis that are inextricably related: emotions, repetition, and attention.

Emotion, Repetition, and Attention

Emotion: Music is not only a rich acoustic signal, but also a sociocultural phenomenon often associated with positive emotion; this in turn influences attention and persistence. In most cultures, music is a potent form of entertainment, and is associated with social and culturally significant events (e.g., graduation, weddings, sporting events, parties). For many, but not all children, musical activities, particularly when well matched to the child's capabilities and preferences, tend

to be associated with strong positive emotion and reward, such as social attention, praise, and the pleasure of hearing beautiful music (Gfeller 2008; Patel 2011).

However, as anyone who has ever taken music lessons can attest, practicing is not always enjoyable. Tedium or frustration can occur in the course of the numerous trials required to achieve satisfactory results. Frustration may be particularly problematic if the musical sounds themselves are not pleasurable, or if the desired outcomes are agonizingly slow to emerge. Given the technical limitations of the CI, one could easily assume that pediatric CI users would fail to garner positive emotions from music listening or participation.

However, as was noted in Section "Music Engagement and Participation," many pediatric CI users participate in music instruction or social uses of music, and consider music to be important in their lives (Gfeller and Knutson 2003; Gfeller et al. 2012a). Although the levels of musical engagement are not equivalent to NH peers, data indicate that at least some musical sounds or activities are enjoyable and mood enhancing. For those CI users who are less enthusiastic about music, other forms of habilitation may be more beneficial.

For clinical purposes, informal observation or brief questionnaires can provide evidence of which, if any, musical instruments or experiences promote positive emotions for individual children (Hsiao and Gfeller 2011; Gfeller et al. 2011), thus taking into account one of Patel's condition for training benefit. Musical sounds that are rewarding are also associated with increased behaviors (operant conditioning), or repeated listening or practice, which is discussed in the following section.

Repetition: Within the context of the OPERA hypothesis, Patel (2011) is referring to the inherently repetitive structures within music. Musical forms usually include repeated melodic, harmonic, and rhythmic patterns that organize and help listeners to predict musical events. Within the context of habilitation, repetition through practice and repeated trials is also essential for neural plasticity and learning. Thus, the inherently repetitive structure of music, especially if paired with emotional reward, can result in ample exposure to structurally complex sounds.

A review of extant research on music training for CI users has included protocols as brief as 10 min per day, with repetitions distributed over weeks or months (e.g., Driscoll 2012; Yucel et al. 2009) to 45-min class periods distributed over 24 weeks (e.g., Innes-Brown et al. 2013). While it is likely that more training will result in better outcomes, specific guidelines have not yet emerged from published studies regarding the amount of repetition that is sufficient to yield significant change, especially in light of the many other factors that influence training availability and persistence.

In addition, as noted previously, the benefits of music training are also associated with factors such as longer term music training. Only a small proportion of CI users are likely to have extensive music training prior to deafness. Interestingly, in our own implant center, CI users who have chosen to engage in years of instrumental music playing, pre- or post-implantation, have often developed surprisingly strong outcomes for both music and speech perception. It is not clear, as of yet, whether those CI users blessed with better perceptual outcomes are therefore more likely to tackle and persist at more demanding tasks such as playing musical instruments, or whether the engagement with music making has, over time, refined their auditory processes. This is yet another topic ripe for future investigation.

Attention: According to Patel's OPERA hypothesis, the positive emotions and social rewards associated with music listening and music making can enhance focused attention. Heightened attention can recruit more neurons to attend to subtle changes, and can increase synchrony of neural firing, and thus basic encoding of sound features (Herholz Sibylle and Zatorre 2012; Patel 2011). Although Patel approaches training from a cognitive neuroscience perspective, these suggestions reflect what good clinicians have implemented in therapy sessions from a behavioral standpoint for many years—therapy works best when the child is highly engaged, motivated, and attending to the appropriate stimuli.

The desired conditions of positive emotion, repetition, and focused attention, from a clinical standpoint, emphasize the importance of selecting developmentally appropriate and engaging musical stimuli that will motivate and foster focused attention. For children who do not respond positively or who lack heightened attention, music training may not be a good habilitative option. In short, the conditions described by Patel may not be fulfilled for all CI recipients. The unique auditory, personal, and environmental circumstances of individual CI users should be assessed with regard to specific aspects of music, and potential benefit of music training for music enjoyment or speech development. The following section provides some practical suggestions that can assist parents, clinicians, or teaching in appropriate use of music training for CI users.

Practical Suggestions for Music-Based Training

At present, from a research standpoint, more questions remain than have been answered regarding the efficacy and appropriate applications of music training for pediatric CI recipients. Given the considerable challenges involved in research protocols that are developmentally suitable for

young children, these questions will likely require a collective effort of many clinical research centers over the coming decades. For the present, dynamic assessment in clinical practice along with thoughtful analyses of available research can guide habilitative and educational choices. Musical activities and assessment tools should be chosen that reflect the desired outcomes (e.g., enhanced music perception, music enjoyment, or transfer to speech perception), and that are developmentally appropriate for the chronological and hearing age of the child. Taking into account current knowledge, the following section offers practical suggestions for enhancing music enjoyment or using music as part of habilitation for pediatric CI users.

Practical Implications Gleaned from Available Information

1. *Some aspects of music, such as rhythm, are more effectively transmitted by the CI.* The relative difficulty of different musical tasks should be taken into account when establishing educational or clinical objectives. More immediate enjoyment and understanding can be facilitated through selection of music with a strong rhythmic component, such as playing or listening to percussion instruments. In contrast, singing in tune is among the most difficult tasks for many CI users. Therefore, while singing can be enjoyable and promote vocal use, exact pitch matching is likely not a realistic expectation for most CI users (Nakata et al. 2006; Xu et al. 2009).

   For clinical interventions intended to increase perceptual abilities, more challenging musical elements (e.g., timbre, melody) can be gradually integrated into listening exercises. Initial discrimination tasks should begin with musical sounds that provide clearly different spectral shapes (e.g., the abrupt onset of a drum vs. the gradual onset of a maraca); more similar, and thus challenging, contrasts can be presented as the child's skills develop (Hsiao and Gfeller 2011; Innes-Brown et al. 2013; Gfeller et al. 2011; Gfeller 2000).
2. *The development of music skills, as is true for speech and language, will evolve as a result of physical, cognitive, and social maturation as well as life experiences.* Consequently, parents, teachers, and clinicians should consider "typical" developmental trends when choosing musical activities and expected outcomes. Consultation with music therapists or written resources on musical development in children (Gfeller et al. 2012a; Hsiao and Gfeller 2011, 2012) can be used to select musical activities appropriate for a child's chronological age and developmental and auditory profiles. Rates and extent of improvement will generally be less than those observed in children with typical hearing.

   Even though musical development may be atypical, music can nevertheless be enjoyed as part of everyday life. Playful and socially meaningful aspects of music can be naturally integrated into the daily routine in the home. Lullabies, play songs, exploration of rhythm instruments, or exploring sounds on the family piano are all natural opportunities for exploring sound informally within the normal milieu (Hsiao and Gfeller 2011, 2012; Gfeller et al. 2011; Gfeller 2000).
3. *CI recipients are highly variable in their perception and enjoyment of different aspects of music.* Therefore, aesthetic, educational, and habilitative goals and objectives should be individualized, based upon the unique auditory capabilities and personal and cultural interests of each child (Hsiao and Gfeller 2011, 2012; Gfeller et al. 2011; Gfeller 2000).
4. *Some aspects of music perception and enjoyment can be enhanced through focused music training, though the benefits from training vary considerably from one person to the next.* Furthermore, it is not yet clear what aspects of training are more efficacious for particular subgroups within the CI population. Consequently, clinical observation and documentation should be used to evaluate individual outcomes. For those aspects of music that appear less amenable to habilitation, accommodations or modified expectations should be considered (Hsiao and Gfeller 2011, 2012; Gfeller et al. 2011; Gfeller 2000).
5. *Because of the precision required to listen to the spectrally complex elements of music, music training has been advocated by some neuroscientists and clinicians as a beneficial stimulus for enhancing speech and language development.* However, there is currently limited empirical evidence specific to the CI population. Therefore, the following conditions should be considered before implementing music-based training for speech and language:

   Evaluate the emotional impact of music. Observe whether the child shows positive response and sustained attention in some musical sounds or activities. The musical sounds and activities should be sufficiently enjoyable to motivate sustained attention and involvement. Computerized training can include game-like applications that promote exploration and selection of preferred sounds. Playing real musical instruments can include experimentation and comparisons of different instruments. The social aspect of music making (e.g., singing or playing instruments in groups) can provide social motivation and encourage interactive spoken as well as musical communication (Gfeller 2008; Gfeller and Darrow 2008).

   Integrate sounds that are sufficiently complex and require careful listening. Gradually integrate musical structures that are spectrally rich, and thus require careful listening. For example, if enhanced perception of speech in noise is

the target of training, singing songs may start unaccompanied (*a capella)* and a background accompaniment can be added as the child's listening skills progress. Prosodic elements of speech can be reinforced by singing songs that imitate and exaggerate the natural inflections and stress of speech. Listening games that require discrimination, recognition, or production of pitch patterns or timbre contrasts should be included.

Provide ample repetition of musical patterns over time to facilitate learning. Persistence can be fostered through the choice of musical activities and sounds that are enjoyable and developmentally appropriate.

Pair songs and lyrics. Parents or clinicians should take advantage of the natural repetition of lyrics, rhythms, and melodies found in many songs, which provide ample repetition needed for learning. The rich repertoire of children's songs can be mined for those songs that present concepts and vocabulary that reinforce target goals and objectives for speech perception and language development. With a little creativity, existing song lyrics can also be modified to target vocabulary or speech sounds that are especially relevant for a given child (Hsiao and Gfeller 2011, 2012; Gfeller et al. 2011; Gfeller 2000).

Summary

In summary, music is a pervasive and natural part of children's lives. Even though CIs convey a degraded representation of pitch and timbre, music can be a personally and socially engaging and meaningful part of life. Furthermore, many aspects of musical sounds and functions offer potential as part of educational objectives and habilitative protocols that can be motivating and that encourage careful listening skills. Further research is needed to determine particular advantages of music-based training for musical, speech, and language development with this population, and under what circumstances training would be most beneficial.

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Part VI

Educational Management of Children with Cochlear Implants

Early Intervention Programs: Therapy Needs of Children Ages 0–3 Years Pre- and Post-cochlear Implantation

22

Maribeth Nelson Lartz and Tracy Meehan

Introduction

The decision has been made. After scores of medical, audiological, speech and language assessments, discussions with the surgeon, speech-language pathologist, educational specialist and audiologist, a hearing aid trial, and discussions of insurance coverage, costs, and schedules, the parents have decided that their young child will receive a cochlear implant. Now the work really begins. Although no one can predict with certainty how a child will grow and develop post-cochlear implantation, the therapy and the professionals who provide it will profoundly influence the benefits and outcomes of cochlear implantation for the child and his/her family.

For purposes of this chapter, the term *interventionist* will be used to describe early intervention professionals who work with the child with hearing loss and his/her family from the time of diagnosis to the time the child ages out of early intervention at age 3. Interventionists come from a wide variety of educational, developmental, and clinical professions and may have specialized expertise in deafness, child development, audiology, speech-language pathology, or other areas impacting development of the child with hearing loss. In this chapter, early interventionists planning for the development of listening, language, and literacy outcomes in young children with cochlear implants are highlighted.

This chapter provides a discussion of and rationale for quality birth-to-3 early intervention services for children with cochlear implants and their families. Included are pre- and post-implant therapy needs for the child and parent, and guidelines for the interventionist and recommendations for carryover activities that parents can engage in with their child during daily routines at home. A framework to be used for designing and delivering therapy content across a continuum of chronological and listening ages is shared, including strategies that enhance listening, language, and literacy, and activities professionals can utilize with parents and children who are pre- and post-cochlear implantation.

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Early Intervention Services

Professionals have been providing early intervention services to infants and toddlers with disabilities and their families as part of the *Individuals with Disabilities Education Act* (IDEA) Part C federal legislation since 1997 (IDEA 2004). The *IDEA* originally passed in 1990 to ensure that states provided special education and related services to children with disabilities beginning at age 3 and through their school years. In 1997, Congress added legislation (Part C) that authorized services for infants and toddlers with disabilities from birth through 2 years of age. Children who have developmental delays or have a diagnosed condition that has a high probability of resulting in a developmental delay are eligible for services through Part C of IDEA. Early intervention for families and children with hearing loss often includes services from the areas of audiology, assistive technology, education and family training, medical evaluation, service coordination, social work, and speech and language. Payment for these services comes from a variety of sources within states, including insurance, local agencies and schools, Medicaid, family cost share, and other sources.

Although infants and toddlers with hearing loss and their families have had access to services under Part C since its inception in 1997, prior to the establishment of the Early Hearing Detection & Intervention (EHDI) program and the resultant increase in newborn hearing screening at the birthing hospital, many deaf and hard-of-hearing children were not identified early enough to be enrolled in and benefit from the early intervention services that IDEA Part C provided

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(White et al. 2010). In 2000, Congress authorized the development of the EHDI program, and over the next several years EHDI programs across the USA began to screen and identify newborns and infant children with hearing loss and refer them to early intervention services. In 2010, President Obama signed the *Early Hearing Detection and Intervention Act of 2010* into law, which provided amendments pertaining specifically to the *training of qualified personnel* and the development of statewide systems for *appropriate educational, audiological, and medical interventions for children identified with hearing loss* (National Center for Hearing Assessment and Management 2011). With the advent of the EHDI screening process, the age of identification of hearing loss has moved from an average of 2–3 years to an average of 2–3 months of age (White et al. 2010) and, subsequently, an increasing number of children with hearing loss will enter the early intervention system at a much younger age.

Although the identification of so many children with hearing loss represents a major step forward, identification is only half of the equation; in all of the research documenting positive outcomes, it was the enrollment in *appropriate early intervention* that made the difference (Geers et al. 2009; Yoshinaga-Itano and Gravel 2001; Goldberg et al. 2010). The 1-3-6 model proposed by the Joint Commission on Infant Hearing (American Academy of Pediatrics 2007) in which infants are screened for hearing loss by 1 month of age, receive an audiological evaluation by 3 months of age, and, if hearing loss is confirmed, begin early intervention services by 6 months of age, attests to the importance of timely intervention.

IDEA Part C has at its core positive tenets for early intervention designed for all children birth–3 years of age, regardless of disability. These include:

- Development of the Individualized Family Service Plan (IFSP)
- Enhancement of infant and toddler development
- Development of collaborative partnerships between parent and professionals
- Provision of family-focused intervention and education

The original tenets of Part C have been strengthened through added regulations related to the provision of services in the child's *natural environments* and *measuring* and *improving child outcomes* (IDEA 2004). Additionally, numerous national organizations have developed professional standards to advance their implementation (Division for Early Childhood 2014; Head Start 2003), but recommendations designed for all children and professionals in early intervention do not provide the detailed knowledge and skills needed by professionals working with young children who have cochlear implants. As Susan Nittrouer offered, "In our efforts to provide intervention for all children with special needs, we have lost some of our skill at helping children with specific needs" (Nittrouer 2010, p. 35).

Some national organizations have developed knowledge and skill statements specifically for professionals working with young children with hearing loss (American Speech Language Hearing Association 2008; American Board of Audiology 2014; National Association of the Deaf 2010). The AG Bell Academy for Listening and Spoken Language has specifically established professional standards for knowledge and skills in the provision of listening and spoken language intervention for children with hearing loss whose families have chosen a spoken language outcome for their child (The AG Bell Academy for Listening and Spoken Language 2012).

Challenges to Appropriate Early Intervention for Children Who Are Deaf

The benefit of early intervention for children with hearing loss and their families will be maximized if the child is enrolled soon after diagnosis in an intervention program, if early intervention services are provided by qualified and experienced professionals, and if the services the family and child receive are designed to maximize listening, language, and literacy development that will carry over to the child's daily home and community environments.

In spite of the demonstrated efficacy of early intervention services for children who are deaf, several challenges exist to its provision. Based on data from the year 2009, the Centers for Disease Control reported that only 68 % of children with confirmed hearing loss were enrolled in early intervention services (National Association of the Deaf 2010). Unfortunately, a large percentage of families of children with confirmed hearing loss fail to enroll in the early intervention services for which they are eligible and are considered to be "lost to follow-up" (Centers for Disease Control and Prevention 2008). Although a thorough discussion of why many children are not enrolled and receiving early intervention services is beyond the scope of this chapter, the fact remains that listening, language, and literacy outcomes for these children are highly compromised without intervention before 3 years of age (Sharma et al. 2002). Another challenge to the provision of quality early intervention services for children with hearing loss is the shortage of providers. Across the nation, there is a serious lack of qualified personnel to work with deaf and hard-of-hearing children in the area of early intervention. Shortages cross multiple disciplines, including speech-language pathology, audiology, and education, and are growing at a critical level (Compton et al. 2009; Lenihan 2010; Moeller et al. 2006; Nelson et al. 2011).

Early intervention services for infants and toddlers with confirmed hearing loss should be provided by professionals

who have expertise in hearing loss (Muñoz et al. 2011) with specific training and/or experience to provide services to children with cochlear implants. Positive developmental outcomes for these children depend on the effective preparation and development of the professionals. Many speech-language pathologists, audiologists, and teachers of the deaf lack the professional training to prepare them to meet the current communication, audiological, and educational needs of children with hearing loss who now can be identified in the first months of life (Muñoz et al. 2011; Robbins and Caraway 2010). Children must have access to professionals who are qualified not only through knowledge of hearing loss and its impact on a child's development of language, communication, academic, and social skills, but also through clinical and professional experiences with young children with hearing loss, if they are to optimize listening and spoken language outcomes most effectively (Goldberg et al. 2010).

The third challenge to the benefit of early intervention services for deaf children is the quality of the content and the consistency of carryover to daily routines in the home and community. If parents desire listening and spoken language outcomes for their children, the content of the early intervention therapy services pre- and post-implant must include services that maximize listening, language, and literacy; provide early access to language and communication; promote consistent amplification and audiological management; and offer targeted aural (re)habilitation and continuous assessment of functional listening skills. To ensure that the child's listening and language skills continue to develop after the therapy session is complete, the early interventionist must educate and empower family members so they have the knowledge and confidence to embed listening and spoken language targets into daily interactions with their child. The interventionist who integrates listening and language targets into a family's routine activities at home or in other natural environments is designing intervention that will maximize carryover of newly acquired skills.

A Framework for Therapy Content and Carryover

For the infant or toddler who has or soon will have a cochlear implant, the importance of early intervention services cannot be denied. Children with early-identified profound hearing loss with early cochlear implantation and a *high-quality* auditory intervention program have spoken language expectations that are similar to children with early-identified mild-to-severe hearing loss and the use of conventional amplification (Yoshinaga-Itano 2006).

Best practice tenets appropriate for all children eligible for early intervention are an important foundation for professionals serving young children with hearing loss. Early interventionists must utilize the "general" best practices in addition to demonstrating competencies specifically related to supporting the youngest learners with hearing loss (Listening and Spoken Language Knowledge Center 2013). A framework that combines the basic principles of early intervention, best practices with specialized listening, language and literacy strategies, and activities needed for optimal development of listening, language, and literacy skills is presented.

The Framework Foundations

A therapy framework for delivery of services to young children with hearing loss and their families must include specific components to ensure successful developmental outcomes for the child. The framework suggested is designed for interventionists serving specific needs of children with cochlear implants. It includes the developmental domain, e.g., listening, language, and literacy, numerous developmental targets under each domain and rationale for each, target behavior expected from the child and interventionist during therapy session, instruction and feedback the interventionist will give the parent/caregiver, and suggestions for when and how the parent can carry over the skill during daily routines at home or in the community. The framework is designed to meet therapeutic needs of the child and family at both pre-implant and post-implant status. Although there are components of every therapy session that are the same, specific targets and activities will differ depending on the child's hearing level, developmental and listening age, and parent's knowledge and need.

The Framework Assumptions

Therapy Is Family Centered

The most effective intervention occurs in a family-centered model. When a diagnosis of hearing loss is delivered, parents enter a new world full of unknown terminology, new professionals and role models, and multiple appointments they had not anticipated when they decided to have a baby. A growing number of professionals are working with these families and their young children during a time when the critical relationship between child and caregiver is developing. Since optimal development occurs within the context of healthy social-emotional relationships with the family, attention to these relationships becomes an important component of quality early intervention services for infants and toddlers with hearing loss (Lartz and Meehan 2009). Early intervention services provided by knowledgeable and skilled professionals can support the family when dealing with stress, grief, and the emotional ramifications of a confirmed hearing

loss in a newborn infant. As stated by Christine Yoshinaga-Itano in the 2011 article by Felzien (Felzien 2011, p. 25):

> It is the job of the early interventionist to help the family recover the joy and celebration that they would normally experience about the birth of their infant, despite the audiology appointments, the decisions about communication choices, amplification (if necessary) and early intervention, as well as the many unexpected issues which the family may face.

Family-centered early intervention reflects the themes of (1) family involvement in the child's therapy, (2) family opportunity to participate in early intervention and learn knowledge and skills that promote development of new abilities, and (3) family and professional collaboration. It is a given that the family is an essential member of the team and that the team includes practitioners from multiple disciplines as needed (Division for Early Childhood 2014).

As assistive listening technology continues to provide early access to sound for the youngest children, early interventionists will be increasingly called upon to provide listening and spoken language skill development within the context of the family (Lartz and Meehan 2009). Family-centered intervention requires the professional to develop a positive collaborative relationship with the family, a relationship that is fostered when the professional shifts from sharing information to modeling and teaching. Rotfleisch shares: "Parent education becomes a focal point in intervention in the model whereby the caregiver is a critical partner in their child's development" (Rotfleisch 2009, p. 445). Professionals working with families of children birth-to-3 are not primarily "teaching" the child—they are really teaching the people interacting with that child. They bring their clinical and educational expertise to the arena, but they must be able to implement their practices in a variety of home, clinic, and community environments. In addition to utilizing evidence-based intervention to develop listening, language, and literacy, the intervention must be presented in family-friendly ways and be implemented within the child's everyday routines. Early family education must inform the parents about appropriate developmental expectations for their child and the steps to get there (Winter and Phillips 2009), should build confidence in the parents as their child's primary teacher (Lartz et al. 2014), and should inform parents of the importance of their child's early listening and linguistic environment and their positive role in it.

Early Intervention Positively Impacts Neurological Development

Evidence regarding neural development supports early intervention for the optimal development of language and hearing in infants. Development of visual attention, auditory attention, and all other precursors to language and communication between an infant and his/her family members must occur early in the infant's life for maximum development to transpire. Once hearing loss is detected in infants and enrollment in early intervention occurs, interventionists can begin to facilitate development and learning by providing rich auditory and linguistic experiences that enhance the billions of major neural connections being formed in the infant brain. The auditory system of infants is working to lay down the neural structure for processing of speech sounds very early in life, and those abilities will diminish quickly when auditory stimulation is absent (Cole and Flexer 2011). With the advent of cochlear implants, greater neural stimulation is available to young children who are candidates, but development of audition does not occur automatically following implantation and the access to sound it provides. Age of implantation, length of auditory deprivation, and the intensity and quality of auditory stimulation are strong predictors of outcome performance of children with cochlear implants (Geers and Nicholas 2013). If appropriate and sufficient acoustic stimulation is provided with a cochlear implant when the child is 3 or younger, the auditory cortex will develop similarly to that of a child who had typical hearing (Sharma et al. 2009), but intense auditory stimulation/habilitation must be provided in the same order of development as typically hearing children. For children to achieve maximum auditory comprehension, they must first master the basic auditory developmental steps:

1. Awareness—demonstrating the awareness of a particular sound or sounds, regardless of whether it is a speech sound or an environmental sound.
2. Discrimination—deciding whether two sounds are similar or different.
3. Identification—selecting a picture or object that represents the sound or word that is heard or imitating what has been heard.
4. Comprehension—demonstrating understanding of the input by answering a question or making a response that is different than the original auditory stimuli (McClatchie and Therres 2003).

Early intervention professionals with specialized training and experience in listening and spoken language development of deaf children are uniquely qualified to enhance the neurological and auditory development of deaf children with cochlear implants.

Early Intervention Occurs Within a Variety of Communication and Language Environments

Discussions continue between professionals about the best approach to promote linguistic competency in children who are deaf or hard of hearing. Parents receive and must evaluate information they are given from medical, social, educational, and community advocates. Without support, this information can be overwhelming and may thwart the family's ability to

move forward. An early intervention professional, in conjunction with the child's audiologist, provides information regarding cochlear implants as a potential treatment for severe-to-profound hearing loss. They can also share current cochlear implant candidacy requirements and encourage parents who desire listening and spoken language outcomes for their children to pursue a candidacy evaluation.

While technological advances in assistive hearing devices, including digital hearing aids and cochlear implants, provide improved access to auditory information, linguistic development of deaf children raised in hearing households still remains a laborious process (Bennet and Hay 2007). The language and communication environment may include various forms of visual and/or signed language and a variety of languages and modalities. Whatever language or communication method the family chooses for their child will impact the development of spoken language. Professionals must be sensitive and knowledgeable when sharing information regarding the impact of each child's linguistic environment on the child's potential for spoken language outcomes. For the early intervention professional serving families who desire a listening and spoken language outcome for their children, it is crucial to share the important role that the cochlear implant will serve in their child's journey toward that outcome.

Planning for Pre- and Post-cochlear Implantation

Much planning must occur both pre- and post-implantation so that therapy goals are developed to parallel typical developmental stages in auditory and language development. The therapy session should provide focused auditory and linguistic stimulation at the individual child's developmental level and the interventionist should present clear models of auditory and linguistic behaviors. Intervention strategies designed to increase joint attention and eye gaze and coordinate gestures and vocal play might be emphasized while a child is awaiting cochlear implantation. Imitating vocal play, expanding verbal exchanges, establishing reciprocity of communication, and promoting activities to move the child through the listening hierarchy might be emphasized after a child receives a cochlear implant. Regardless of implantation status, interventionists should purposely select listening, language, and literacy interventions that are linked to improved outcomes for deaf children (Listening and Spoken Language Knowledge Center 2013). Implementation of strategies is continuous and change occurs based on careful observation and assessment of listening and communication within the child's natural and functional listening environment. Engaging the parent in discussion about how these goals or targets will be carried over to family routines should be a major part of preplanning for each session for all interventionists. From assessment and ongoing observation, interventionists plan activities to promote child development in all developmental domains. At the same time, interventionists are pre-planning how their interactions with parents will promote successful carryover of the strategies into the family's daily routines.

Pre-implant Therapy Focus

Now that much more is known about brain development, executive functioning, and other neurological foundations of listening and spoken language, early intervention services must be designed to provide the developing brain of infants and toddlers with auditory and linguistic experiences that capitalize on unique developmental periods and characteristics of the child.

Family involvement and support are crucial for the development of children's cognitive, communicative, and social skills (Bennet and Hay 2007; Boggett 2013). For parents of children with hearing loss, the major concerns are in the domains of communication and language development, learning, social development, and the inclusion of their child in the family and community (Brown and Remine 2008). Because early interventionists are primarily trained and interested in the communication, cognitive, and social aspects of development, the parent-professional relationship can be a perfect match. Since many parents do not have the detailed level of knowledge related to the acquisition of an ordered sequence of skills in developmental domains, the early interventionist assumes the role of teacher. Additionally, in family-centered practice, the interventionist's teaching, modeling, and coaching are designed for the adult and child learning together.

Intervention for infants and toddlers with hearing loss occurs while a family is making decisions about ways their child can meet the outcomes they desire. Most families will acquire hearing aids for their baby and the new learning begins. For a family interested in cochlear implants and whether their child might be a candidate, there is additional learning. Interventionists need to reinforce information that the cochlear implant team has shared with the family; most notably, the potential outcomes for their child's auditory and linguistic development that are possible with consistent use of the device and adherence to the recommended schedule for follow up activities and appointments. During pre-implant therapy, continuous dialog will occur with the family regarding any cognitive delays, visual impairments, global developmental delays, or medical conditions their child may have. The interventionist should encourage a discussion of goals the family has in relation to the expected language and mode of communication the child will use, as

well as goals the family has for the child's use of the cochlear implant. Do the parents expect the child to develop spoken language and only use it for all communication or do they plan to use some form of visual support or language, e.g., Cued Speech, American Sign Language, et cetera, with the child? Intervention sessions with the family pre-cochlear implantation are an ideal time to provide information and resources on the variety of communication choices parents have for their children with hearing loss and to discuss the impact that intellectual and global developmental delays or various medical conditions may have on their child's rate of progress and level of listening, language, and literacy development.

During the early months of intervention, while the child and family are becoming adjusted to hearing aid use, intervention sessions look different from subsequent sessions when new skills are coming at a more rapid pace. Rhoades and Duncan remarked in their 2010 book (Rhoades and Duncan 2010, p. 140):

> A child is part of two systems: (1) the family, and (2) the larger social network of individuals, interacting families, and social institutions. However, it is a "family" that is the primary and most powerful system to which the child belongs.

Identifying routines that occur in this family will allow strategies brought to therapy for demonstration to be repeated regularly with child and family members. Recognizing how a family functions on a day-to-day basis will strengthen the likelihood of carryover of strategies developed to match a family's outcome of listening and spoken language for their child.

To prepare for family-centered therapy sessions, interventionists pre-plan in multiple areas. Using their knowledge of typical child development in auditory, language and literacy domains, previous coursework or training focused on developmentally appropriate curricula and activities for young children, and personal observation of the child and family, the professional selects a particular skill and activity for this current point in the child's development. The skill and activity are selected based on *developmen*tal age and not chronological age. The professional must be able to explain the skill and demonstrate it for the child and for the parent using a developmentally-appropriate activity. Through observation and coaching, the interventionist must be certain that the parent understands and is confident in implementing the strategy or skill. Finally, and of equal importance, the interventionist must dialog with the family about routines that occur often within the day, where this strategy or skill might occur naturally. One challenge is separating the activity used to model the strategy or skill from the strategy or skill itself. For example, if the skill being modeled is *vocal turn-taking*, this could occur in any number of daily activities. Vocal turn-taking will occur not only with the selected toy or book chosen for demonstration, it must occur across a wide variety of activities, in a variety of environments and with a variety of people to be fully mastered.

Table 22.1 contains a completed framework which could be used for intervention with a family and child pre-cochlear implantation. The table includes a separate example for each of the core developmental domains of listening, language, and literacy, and includes developmental targets and rationale for each, target behavior expected from the child and interventionist during a therapy session, instruction and feedback the interventionist will give the parent/caregiver about each strategy or skill used, and suggestions for when and how the parent can carry over the strategy or skill during daily routines at home or in the community. Italicized sentences under the *parent modeling and feedback* column represent the interventionist's actual words to the parent/caregiver.

Post-implant Therapy Focus

Now that implantation has occurred, the child and family need to shift expectations and the interventionist has to make a mental shift from pre- to post-implantation therapy. The interventionist will, in most cases, move from using visual and tactile strategies as cues for communication to more auditory-focused strategies. More emphasis is placed on modeling therapeutic techniques to the parents and observation becomes more focused on how the child's implant is working, how the child is bonding with it, and what functional listening skills the child is demonstrating. Many interventionists who have limited experience with cochlear implants find it difficult to modify their therapy to reflect the increase in the child's access to sound. Developmental targets have to shift and the interventionist has to have a secure knowledge of developmental expectations so that appropriate targets are selected for listening and language and any *red flags* in development can be identified quickly. In post-implant therapy there is a significant shift to listening—and learning language through listening. Whereas pre-implant therapy focused on access to language through a variety of modes and multi-sensory experiences, now the thrust of therapy is having the child use listening to learn language and "listening to learn." Parents need to be reminded that although the implant provides auditory access, the child may still need some form of visual communication to supplement the information received by listening alone. The interventionist continuously monitors communication and language growth, in addition to auditory development.

Table 22.2 contains a completed framework for intervention with a family and child post-cochlear implantation. The reader will note the shift to more auditory-focused activities and expectations. The table includes a separate example for

Table 22.1 Pre-implant therapy framework

Developmental domain (target and rationale)	Expectation for the child (C) and interventionist (I)	Parent modeling and feedback	Carryover of new skill to daily routines
Listening			
Consistent amplification Sets the stage for auditory imprinting Allows observation in different environments Gives good data to audiologist for better programming	I: Expect use of amplification during therapy and longer each day in between therapy I: Communicate with other therapists on the early intervention team to make sure success of hearing aid use is seen in those sessions, if appropriate C: Depending on the age, can help put on and show early sound awareness by making vocalizations when the device is turned on	*Where do you think Amy can put her hearing aid when she takes it off? Would a small plastic bowl work here on the table? Practicing this together will give Amy some independence and will help you not be so worried about losing the hearing aid(s).* *Let's think about when you come home from work and the different rooms and listening scenes Amy hears.*	Morning routine—all waking hours Afternoon routine—all waking hours Evening routine—all waking hours Where are problem times when it might be difficult to keep the hearing aid(s) on the child?
Observing listening behaviors in different acoustic environments Data necessary for hearing aid trial during pre CI candidacy	I: Observing and exposing child to sounds and speech in varying environments Outside—noting background noises Inside—different levels and positions and distance of listening Different therapy sessions Observe in the different places where the child spends time	*When we are more than a yardstick away from Amy, our speech drops off. Let's see how taking a walk outside will impact this.* *Amy can't tell us with words what she is hearing or not hearing. This is why we must be good observers because babies have a lot to say if we can pay attention. Have you seen her "telling" you that she hears in different places? What does she do that makes you realize she is listening?*	Watch for alerting when 3 feet and then 6 feet away from speaker Watch her body and eyes when sounds occur *Is she recognizing the difference in people's voices? How do you confirm that she is identifying? Does she search for them; stop and pause her play; get up and go look?*
Conditioned response To obtain accurate audiological information during each visit Child needs to know what is expected in familiar environment before she can perform in the booth with an audiologist	C: Plays by doing a "drop it in task" I: Pair the "drop it in" task with a verbal cue. Cue to child, "I hear that," then put it in. Encourage the task in play	*Let's practice so Amy understands that hearing means she does that action. Remember, big smiles when she drops it in. We want her to learn that this is important to us, and praising her and being excited will show her we like what she is doing. Learning what it is we want her to do is important. I wonder how you will feel when Amy cooperates better during the sound booth testing and you get a good report from the audiologist next time you take off work for that appointment?*	Many times we are "dropping it in". Practicing listening first, then performing the action helps familiarize the child with the expected task. Cue her with "I hear that," then "drop it in." Think about adding a verbal prompt when putting clothes in a laundry basket; loading the diaper bag; putting blocks into a shape sorter; putting pieces of small items into small bowls
Language			
Eye gaze and visual attention Allows you to enter into the child's space and see the physical objects they choose and to be able to share the experience Promotes opportunities to overhear when visual attention is obtained first	I: Setting out preferred toys or familiar objects Follow child's gaze then hold and comment C: Allow to make choices and pass back and forth	*See Amy look to the Sesame Street rattle? Hold that up close to your mouth and let her look at you. Take turns shaking the rattle. See how Amy and you are looking at the same thing?* *When could Daddy or Grandma do this?*	Use good observation skills during daily routines to see things of interest to the child. Finger feeding—where do Amy's eyes land? Diaper changing-what can you share? Hide and seek with a blanket and hiding a toy

(continued)

Table 22.1 (continued)

Developmental domain (target and rationale)	Expectation for the child (C) and interventionist (I)	Parent modeling and feedback	Carryover of new skill to daily routines
Reciprocal communication The way parents interact and the frequency of these interactions will strongly influence outcomes Pacing and waiting might not seem natural at first	C: Makes a sound and looks up when it is made back to her I: Expect a response from the child when participating in exchanges I: Modeling activities where wait time is pre-planned	*Parent: She is always so independent that I hardly ever need to get things for her* *I: Yes, but remember, we need to increase the times we interact with Amy each day so we might need to participate with her when she is helping herself to things. Let's think about her getting the ice cream. To participate with her we can say …. Brrrr that ice cream is cold. Brrrrr. You like this ice cream? WAIT* *Brrrr. You think it is cold too? I'm excited to hear your voice go up and down. This excited Amy and made her want to talk back to you*	Say something, show something and say it again. Then WAIT Key is expecting a response and showing excitement when that response comes Think about simple routines; getting something out of your purse; cleaning up toys and saying "all done" when each one is put away; opening up a bag of fruit snacks
Sensory experiences Infants must experience tummy time to be able to have a sense of where they are in the world Directly enhances cognitive function so that better attention, and memory for longer parts of communication is stored and is able to be used later	I: Repeating experiences in same sequence helps build predictability Physical contact as tolerated by individual child Observe physical environment-too stimulating? Not stimulating enough?	*Amy doesn't like to be held, so let's think of ways to soothe her with touch or things she likes. Have you tried light rubbing like a massage? Let's look at how you enter Amy's room to pick her up. Remember, Amy might not hear you walking down the hallway or hear you calling her until she sees you*	Using pillow or rolled up towel under chest during tummy time to keep head up off the ground. Remembering that Amy takes in information primarily through her eyes and can feel alone and frightened during tummy time Keep a box or bowl with different washcloths, wipes, blankets to try with touch and for play
Literacy			
Using vocal inflection and expression Parents are imprinting the baby's brain with the sound code of language	I: Using a melodious voice and adding inflection and expression will increase a child's interest and attention	*Listen to your voice when you read Brown Bear …. What do I see? What do you notice here with your voice?* *That is what we want to capture during other times of the day*	The inflection of asking a question goes beyond simply reading the story and shared literacy experience. Now, take this rhythm and inflection and use it at other times during the day
Increased attention Sharing the reading allows for more interaction and not just listening	I: Share the book-pointing to pictures. Depending on the age of the child, you may be commenting together on each page and not necessarily reading the actual print	*Can you share page turning and pointing to pictures with this book?* *We are sharing this experience so each of you will have a chance to talk. Let's see what Amy has to say about the textures on this page*	Building attention to what you are saying takes repetition. Joint attention is achieved when you follow the child's lead and talk about what is happening that seems of interest to her

each of the core developmental domains of listening, language and literacy, and includes developmental targets and rationale for each, target behavior expected from the child and interventionist during therapy session, instruction and feedback the interventionist will give the parent/caregiver about each skill or strategy used, and suggestions for when and how the parent can carry over the skill or strategy during daily routines at home or in the community. Italicized sentences under the *parent modeling and feedback* column represent the interventionist's actual words to the parent/caregiver.

The interventionist will modify the contents of the framework frequently as the needs of the child and family change, but the core components of developmental domain target, child and interventionist expectations, parent modeling, and feedback and plans for carryover of skill to the family's daily routines should always be present. Challenges are sure to arise in providing early intervention services to children

Table 22.2 Post-implant therapy framework

Developmental domain (target and rationale)	Expectation for the child (C) and interventionist (I)	Parent modeling and feedback	Carryover of new skill to daily routines
Listening			
Associating sound with objects Scaffolding skills means sounds must be associated to objects first before the actual word or name for object is used	I: Identify toys that "talk and make sound" that child has been playing with using hearing aid. Observe her behaviors when toy is activated now using her cochlear implant. Remember to select developmentally appropriate activities while matching her much younger listening age I: Build sets of props where sounds of different lengths, a variety of vowels, loud versus soft, and some initial consonants can be used	*We assume Amy has heard this horse noise that her favorite toy makes, but don't know for sure that she knows that this favorite toy makes sound at all! We do know that it will sound different now that she has an implant. Let's watch Amy's reaction when she hears this sound now with her CI. Then, we can attach a speech sound to this horse too Matching sounds to objects are first steps for learning the words and using the words later*	Sound—object association is enhanced by using the same sound to match with an object. These activities are helping babies grasp the patterns of language spoken around them every day We have long sounds, "neeeeeighhhhhhhh" like our horse, and then the shorter sounds like Amy's puppy makes when he says "woof woof woof." Let's hide these objects in your diaper bag, or your laundry bag when you go to the Laundromat. This gives her extra practice hearing these sounds while you are doing routines you do naturally. At home, hide them in a toy bin so the family can use these short and long sounds in different places
Child responds to voice used to get attention without using visual cues Want to get the child's attention first by using voice	I: Note how far away you are from child when calling her name. C: Looks toward speaker's voice.	*We must be careful to continue giving Amy reinforcement as she turns when you say her name at different distances without seeing you. Unless there is a reason for her to turn and she is reinforced when she does turn when she hears your voice, she may lose interest. We might need to start by standing close to Amy and showing her we are here when we call for her attention. But, soon, we will be around the corner calling for her attention -- this is going to be exciting!*	What are things we might use for reinforcing a child when we call her name? Smiles, hugs, high five hand taps, being picked up, being tossed in the air? Practice and see what she enjoys. Have a toy in your hand, then call to the child. She will be surprised to see you with a favorite item, and eager to continue looking when she hears your voice the next time
Language			
Embellished environment Often the typical number of interactions or the acoustic environment needs to be enhanced	I: Recognize and discuss with the parent that many of the therapy ideas do not "look natural". To obtain steady progress, there will be some catching up to do for a child using listening for learning. Increasing the opportunities for the child to wear her cochlear implant and interact with people is your goal	*You've done great in helping Amy bond with her CI. She's wearing it all day and asking for it after nap. She is getting so independent and helping herself to many things since she is walking around now. Remember that Amy needs more conversations between her and the adults that are close to her. She is not overhearing yet, so we need to increase these opportunities*	When a child is in daycare, planning and collaboration to create an embellished environment must occur. Activities to consider when interactions between adult and child occur in close proximity to other people: teacher sits next to child during circle time and highlights events as they occur; review of the schedule that occurs from drop-off to pick-up, ensuring that one of the childcare teachers can interact with the child individually and provide increased interaction opportunities

(continued)

Table 22.2 (continued)

Developmental domain (target and rationale)	Expectation for the child (C) and interventionist (I)	Parent modeling and feedback	Carryover of new skill to daily routines
Recognizing child's communication and expanding upon it Children who are talking and putting a few words together can benefit from these strategies. Children will want to continue the discourse if what they say is recognized. The expansion incorporates part of their talking and provides a more syntactically correct, and/or semantically richer sentence	I: Set up opportunities for child to share a thought through play or with a single toy or book I: Be prepared and ready to recognize the child's start of a conversation. You are planning your acknowledgment or recognition that they said something purposeful. What will you model for the parent? "Yes," "Right," "Un Huh," "Really?"	*Amy is starting to label things now and giving us some connected words during her play. Let's stay with an activity or conversation she starts and give her a few more words in response. You will see how we can do this easily and increase Amy's interest in continuing the conversation*	Identify and listen for the child to start a conversation. "Boots" she says. "Right, you see Mommy's boots" You recognize what she said, then gave her an expansion. Extending this later would include "Right, you see mommy's boots. They are dirty"
Literacy			
Positioning Auditory imprinting continues during shared reading and child needs clear audible input	C: Chooses a book to share I: Sits on the side with the child's cochlear implant. Watch for child's interest on each page. Verbal presentation should be first for listening only	*Now that Amy has more listening ability while wearing her CI, we want to build her confidence about getting information through listening only. By sitting next to her or holding Amy on your lap, you are sharing the book- reading experience plus allowing her to hear your voice over and over again*	Books can be made with pictures, can include experiences the family has had and can also include books obtained from a library. Shared reading allows the adult to sit close and be able to capture a child's interest on each page. Language used by the adult will vary by age, but even young infants benefit from early literacy exposure Using a container to hold the books may help your child predict events that are happening. If sitting at a restaurant and waiting for food is a concern, consider using the same bag for these outings and packing it with a book before leaving the house
Reading aloud This activity allows the child to use her imagination, to increase enjoyment of books, and helps create background knowledge. Reading aloud provides opportunities to introduce descriptive and grammatically correct models in the native language	I: Model fluency using a slightly slower rate of speech than typical conversation Use intonation and expression	*Amy has some favorite books that you have been reading. Now we will continue with these same books and watch for Amy to begin predicting or reading along. When you are reading, you can pause on a page and show Amy you are interested in her thoughts. Begin with the familiar phrase in the book, then pause and let's see if Amy will begin to fill in the missing next piece*	Finding times during the day to read together may be the challenge. But, by increasing a child's pleasure through shared interactions a parent will begin to see different skills emerge and will feel empowered to continue. Help families find props that match the story so that they are available to use when they sit and read with their child
Acoustic highlighting Drawing attention to sounds or words used while speaking interests the listener and increases attention to the speaker and to a variety of reading materials	I: Different loudness levels of your voice can help a child identify who is talking in the story. Emphasizing key words or phrases will draw the child's attention to the page and make shared reading fun!	*Every time we read this book, we will change our voice a little to match to the character. We can use a soft little voice for the baby and then a loud slow voice when the daddy comes on the page*	This strategy of highlighting can be done with or without the book. Match loudness of a voice with a person in the family. When dressing, emphasize a sound as many times as possible. For instance, sssssssocks go on your feet. Here are your sssssocks. Can you help put on your sssssocks?

while meeting the diverse needs of families with varying cultural, extended family or caregiving dynamics, social, and educational backgrounds (Desjardin 2009). Children with dual diagnoses of hearing loss and other disabilities such as autism spectrum disorder or visual impairments experience multiple threats to typical development of listening, language and literacy and require a cadre of professionals and an integrated system of intervention to meet their needs (Wiley and Moeller 2007; Malloy et al. 2009; Young et al. 2012). With rising numbers of premature births and earlier identification of syndromes associated with hearing loss, more children with disabilities in addition to their deafness

will likely enter early intervention programs. In spite of additional disabilities and challenges for these children, as stated in the Joint Commission on Infant Hearing position statement of 2013 (Listening and Spoken Language Knowledge Center 2013, p. 10) "Regardless of the primary disability, however, it is critical to recognize the primacy of communication for learning and the impact of communication delays on other developmental domains."

As more research is published indicating the importance of early brain development, executive functioning processes and the relationship these processes have on the development of listening and spoken language skills, the more specialized the therapy must be for young children with hearing loss. Due to the necessity of specialized and carefully coordinated services for many children, home-based, center-based, or teletherapy (Edwards et al. 2012) may be appropriate to meet a family's outcome for their child.

Conclusion

Interventionists must continue to keep abreast of new research findings that have the potential to improve their service delivery and positively impact the children and families with whom they work. The most effective interventionists are those who consciously and continually modify their pre- and post-implant therapy in order to implement evidence-based intervention strategies that positively influence the family and lead the child with cochlear implant(s) to maximum developmental outcomes in the critical domains of listening, language, and literacy.

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Educational Considerations: Supporting Children with Cochlear Implants in Mainstream Schools

23

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Introduction

Cochlear implantation (CI) is now the most common response to a child's diagnosis of severe-to-profound hearing loss in most developed nations (Punch and Hyde 2011). Increasing implantation rates have been accompanied by a shift in educational placement, as more children are moving from schools for the deaf and self-contained placements into mainstream classrooms where they are educated alongside typically developing peers for part or all of the day (Francis et al. 1999; Punch and Hyde 2010). A corresponding shift in classroom communication modality has occurred, from placements including sign to speech-only settings; cochlear implantation has been identified as a differentiating factor in this migration (Allen and Anderson 2010).

The River School in Washington, D.C. was founded in 1999 to capitalize on the promise of CI technology. The River School seeks to normalize and optimize the education of young children with CIs by providing an inclusive educational model tailored to their needs. Children with CIs make up 15 % of the student body; they are educated alongside their hearing peers from birth through third grade. The school uses small class sizes; a co-teaching model that pairs a master's level educator and a speech-language pathologist in each classroom; and a transdisciplinary support team to ensure that children with CIs achieve academically and socially at a level commensurate with their hearing peers.

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The authors of this chapter are River School professionals with expertise in education, speech-language pathology, developmental psychology, occupational therapy, and audiology. Over the past 15 years, we have worked with hundreds of children with CIs who have gone on to success in mainstream public and private schools. On average, our students with CIs achieve age-appropriate skills in language, social, and academic development within 3 years of River School intervention.

Shifts in the Education of Children with Hearing Loss

Historically, most children with hearing loss, especially those with severe-to-profound losses, were educated in separate classrooms or schools. In 1975, a federal law was enacted entitled the Education of All Handicapped Children Act (PL 94-142), later renamed the Individuals with Disabilities Act (IDEA). The legislation introduced the terminology "Least Restrictive Environment (LRE)," which mandated that children with disabilities be educated in environments allowing the greatest possible access to typically developing peers (Turnbull et al. 2007). Five years later, the first child in the world to receive a CI received a single-channel House device (Eisenberg and House 1982). These two developments, one a legislative action and the other a technological innovation, changed the landscape for children born with severe-to-profound hearing loss. In 1975, when PL 94-142 was enacted, 48 % of students with hearing loss were enrolled in residential or day schools for deaf students; by 2000 that number had been cut in half (Karchmer and Mitchell 2003).

More than 95 % of children with deafness are born into hearing families (Mitchell and Karchmer 2004), who may consider the CI a tool, rather than a cultural choice. A CI can provide significant advantages for a child including easier access to language and academic skills (Marschark et al. 2007),

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a full range of educational and vocational options, and participation in the family's culture and community.

Mainstreaming rates for children with cochlear implants are fairly consistent internationally. In Australia, 83 % of children with CIs are in the mainstream (Punch and Hyde 2010). In Finland, 90 % of children with profound hearing loss receive cochlear implants; most are mainstreamed (Huttunen and Valimaa 2012). Data from the US Department of Education (2009) indicates that 87 % of children with hearing loss, including children with CIs, spend part or all of the day in general education classrooms (Berndsen and Luckner 2012).

Mainstreaming

For children with CIs, factors that promote mainstreaming include parent education level, communication mode, early implantation, and duration of CI use (Yehudai et al. 2011). Decreased need for support services correlates with both length of implant use and the rate of full-time mainstream placement (Francis et al. 1999; Venail et al. 2010). Moog et al. (2011) reported that 95 % of their longitudinal cohort, who had used CIs for more than 10 years, was mainstreamed for more than half the day by high school.

In mainstream settings, children with CIs perform best socially and academically when their spoken language skills are close to those of their hearing classmates. Early experience in an auditory-oral environment has been correlated with later academic success. Geers and Hayes (2011) concluded that children with cochlear implants in early elementary grades who relied on spoken language exhibited better verbal rehearsal skills and better speech perception, intelligibility, language, and literacy in high school. In France, Venail et al. (2010) noted that earlier implant age and early oral education improved academic outcomes in 100 mainstreamed students who received CIs before age 6. This cohort eventually achieved educational and employment levels similar to their hearing peers. Mainstream classrooms offer children with CIs consistent access to hearing peers who can scaffold their development of social, communication, and academic skills and lay the foundation for their participation in the wider community (Yehudai et al. 2011).

Beginning in early childhood, a child's social and language development will benefit from interactions with peer models. Learning to establish positive relationships and interact with peers at a young age can improve overall academic achievement (DeLuzio and Girolametto 2011). More exposure to typically developing hearing peers can lead to more peer acceptance overall (Leigh et al. 2008). In addition, pragmatic communication develops best with steady exposure to advanced linguistic experiences such as discourse with hearing peers (Most et al. 2010).

While children with CIs have unprecedented access to sound, they still face potential challenges in the classroom. Despite age-appropriate speech and language skills, some children continue to struggle socially and academically compared to their hearing peers (Marschark et al. 2007; Geers and Hayes 2011; Punch and Hyde 2010, 2011). Children from a nationwide sample of adolescents who had used CIs since preschool reported increased participation in mainstream academic settings; growing use of spoken communication; and movement toward age-appropriate language, academic, and social skills (Geers et al. 2011). However, by high school only about half of these students achieved scores in the normal range for reading, and only 38 % reached age-appropriate levels of written expression (Geers and Hayes 2011). Similarly, Punch and Hyde (2011) used teacher ratings to track 151 students with CIs in Australian classrooms; 70 % of the students fell below the median in academic achievement, including literacy and numeracy.

Students with cochlear implants have experienced varying periods of auditory deprivation and consequent language delay (Punch and Hyde 2011). Educational programs that include children with CIs must consider providing thoughtful supports across the domains of development as early as possible, beginning with supports for the child's family.

Parent-Child Interaction

The importance of early intervention for children with CIs, especially for fostering spoken language, has been well documented (Moeller 2000; Moog and Geers 2010). While professionals play a critical role in early intervention, there is increasing evidence that facilitating parent engagement results in improved child outcomes (DesJardin and Eisenberg 2007; Zaidman-Zait and Most 2005; Moeller 2000; Houston and Bradham 2011; Cruz et al. 2013). Niparko et al. (2010) reported that higher parent-child interaction scores were significantly related to improved spoken language comprehension in a longitudinal study of spoken language outcomes of young children with CIs. Parental use of higher-level facilitative language has been found to contribute significantly to children's development of literacy skills (DesJardin et al. 2009).

Fleming et al. (2011) advocate for a model that includes parent participation in audiologic habilitation, but studies show that many early intervention settings use a traditional child-centered service model (Peterson et al. 2007; Campbell and Sawyer 2007). Parent interaction training that incorporates didactic and/or hands-on experiential coaching should be considered an essential component of hospital-based CI programs and schools. Training can take many forms. Educational environments may offer parent-infant programs that model effective interaction and/or teach structured

therapeutic techniques. At The River School, parent workshops, presentations, panels, and support and discussion groups provide families with direct instruction on nurturing language, social, and behavioral development during everyday activities and routines.

Quittner et al. (2013) evaluated the role that parent-child interactions play in child outcomes. They found that maternal sensitivity predicted significant increases in language growth for children with CIs, and that linguistic stimulation was related to the child's language growth *only* in the context of high maternal sensitivity. Notably, the effects of both maternal sensitivity and cognitive stimulation were similar to the effects found for age at implantation, long considered one of the most powerful predicators of CI outcomes. Other studies have also supported the importance of emotional availability and maternal responsiveness on language learning and developmental play for children with hearing loss (Pressman et al. 1999; Spencer and Meadow-Orlans 1996).

Cochlear implant programs can likely improve child outcomes if maternal sensitivity training is incorporated into the intervention (Quittner et al. 2013). In addition, parents may benefit from learning specific strategies to manage their child's behavior, given that behavior issues are often associated with language delays (Beitchman et al. 1996). With improvements in behavior, children are likely more receptive to intervention in language, literacy, and social development.

Parent-Child Interaction Therapy (PCIT) is an empirically supported treatment that works with parent-child dyads to promote child development and foster parent-child relationships (Eyberg 1988; Zisser and Eyberg 2010). The program has been used successfully at The River School with hearing parents and their young children with CIs. Parents are taught skills to promote development, establish a nurturing and secure relationship with their child, increase their child's prosocial behavior, and decrease negative behavior (Bell and Eyberg 2002; Allen and Marshall 2011). The treatment is structured in two parts: Child Directed Interaction (CDI), which is similar to play therapy and engages parents and children in play situations; and Parent Directed Interaction (PDI), which teaches parents behavior management techniques nurtured during play (Schuhmann et al. 1998).

PCIT was initially developed to support children with behavioral difficulties, but has since been used with a range of populations (Storch and Floyd 2005). It has been shown to improve language and prosocial behavior and to increase positive parent/child interactions as oppositional behavior decreases (Allen and Marshall 2011). PCIT has demonstrated statistically and clinically significant improvements in the oppositional behavior of participating children (Rich et al. 2002) and important changes in the interaction style of parents and caregivers, including increases in key interaction patterns such as reflective listening, physical proximity, and prosocial verbalization; and decreases in criticism (Rayfield et al. 1999). Parent benefits include significant changes on self-report measures of personal distress and parenting locus of control. PCIT can be important for parents of children with hearing loss, since these children may benefit from increased time in play environments that promote development (Spencer and Meadow-Orlans 1996). Positive parental interaction and play, and specifically PCIT participation, may improve language growth, particularly in pragmatic language (Allen and Marshall 2011).

In typically developing populations family participation in education has been found to be twice as predictive of students' academic success as family socioeconomic status (Henderson and Berla 1994; Kellaghan et al. 1993). The more intense the parent involvement, the more benefit. For a child with a CI, parent participation and support may critically influence outcome (Moeller 2000; Niparko et al. 2010). Parents, however, may need individual support and training to successfully manage the stress associated with raising a child with hearing loss.

Emotional Supports for Parents

Parents may benefit from supports for their psychological well-being as they manage the period of grief and adjustment that may accompany a child's diagnosis of hearing loss. Support services for parents have been cited as particularly important for healthy interactions between parents and their children with CIs. Interactions in these dyads are rated as more sensitive after interventions that include parent psychological counseling, when compared to families who do not receive such services (Greenberg 1983). When hearing mothers are happy with their social support, improvements in maternal sensitivity are noted (Meadow-Orlans and Steinberg 1993).

Studies of patterns of parental response to diagnosis are varied (Koester and Meadow-Orlans 1990; Kurtzer-White and Luterman 2003; Meadow 1968). McCracken and Sutherland (1991) note that it is difficult to predict how any particular family will react, because the response depends on a variety of factors: social support, access to information and services, and parental temperament. Parents of newly diagnosed children sometimes experience initial strong emotional reactions including confusion, sadness, hopelessness, and despair (Kurtzer-White and Luterman 2003). Parental responses may be affected by issues such as disruption in communication and the need for medical and educational decisions (Spencer 2000, 2002).

A diagnosis of hearing loss may be particularly difficult for hearing parents who lack knowledge about deafness (Luterman 1999; Mitchell and Karchmer 2004; Moores et al. 2001).

Hearing parents naturally base their expectations for their child on the assumption that their child can hear; diagnosis can overturn these assumptions (Kampfe 1989). Just as some deaf parents hope to have deaf children who can share their experiences, language, and culture (Lane and Bahan 1998), hearing parents have similar expectations (Mellon 2009).

Family and peer support, stress, and psychological health have all been identified as variables influencing the adjustment of hearing parents who have children with hearing loss (Dix 1991; Goldberg et al. 1990; Krech and Johnston 1992; Peterson et al. 1994). Support is critical to promoting healthy child-parent interactions and well-adjusted children. Parents who are psychologically healthy and well supported by family and friends typically report lower levels of stress and are better able to meet parenting demands (Dunst et al. 1986; Sheeran et al. 1997). Educational settings can serve as a vehicle for parents to access emotional support by providing parents with group and individual supports and mentoring. Connecting with other parents of children with CIs may be particularly helpful in building and maintaining networks.

Classroom Supports

In order to be fully included in the general education classroom, children with CIs must be considered full members of the class, not visitors. The design of the program and the teacher's perspective on the child will determine the student's status in the classroom. Educators need to create a classroom atmosphere of belonging, anticipating the struggles children may face and implementing specific support strategies (Antia et al. 2002).

Despite impressive performance in one-to-one communication, children with CIs may struggle to hear peer discussions or teacher directions, depending on classroom acoustics, teaching practices, group size, and classroom dynamics. They may need support to navigate socially in certain learning environments, such as cooperative learning groups that require students to discuss, negotiate, and create as a team (Punch and Hyde 2010). These challenges can negatively impact a child's academic and social success and may be overlooked by professionals.

Two areas—theory of mind and discourse—may underlie the social delays that have been noted in children with hearing loss (Wauters and Knoors 2008). Lack of exposure to hearing peers, as well as difficulty with theory of mind and discourse, may impact the pragmatic development of children with hearing loss (Most et al. 2010). Particular difficulty has been noted for contingency—extending a conversational exchange by contributing an on-topic response. Some early implanted children educated in auditory-oral environments appear to show progress in these skills. Recent findings include:

- Most adolescents in the longitudinal cohort self-reported high self-esteem and well-developed social skills (Moog et al. 2011).
- Peer ratings and peer nomination of children with deafness in inclusive settings were comparable to hearing peers on measures of peer acceptance and friendship, but lower on social competence (Wauters and Knoors 2008).
- Five- and 6-year old children with CIs showed strong performance on a peer entry task. Better performance was associated with longer duration of implant use and higher self-esteem (Martin et al. 2010).

Understanding the Listening Environment

Although early implantation leads to better language, a key component of success in the classroom, students may continue to struggle in noisy environments (Vermeulen et al. 2012). Student progress should be carefully monitored. Each child should be observed and assessed before recommending classroom accommodations. FM systems that provide a direct link to the speaker make listening easier; however, they may override informal discussion or conversation taking place among peers. While some children might benefit from an individual FM system, others might find it difficult to manage or socially isolating. Sound Field systems can increase the teacher's vocal volume without blocking out peer conversations.

When possible, carpet and drapes should be used in the classroom to decrease ambient noise. Children with CIs should be given preferential seating, with clear visual access to the teacher. Older students can use recording devices to revisit lectures without depending on peer note takers. The purpose of any assistive technology should be to increase independence without hindering social interaction. In order to learn most effectively, children should be encouraged to become as independent as possible with their devices, and to advocate for themselves.

Ecological Inputs

Language and cognitive outcomes can be improved by ecological factors such as peer language level, the quality of the teacher talk in the classroom, and teachers' behavioral management strategies, all of which can either inhibit or intensify the impacts of intervention. Mashburn et al. (2009) examined associations between the expressive language abilities of peer models and the receptive and expressive language development of children with relatively poorer language. In all, they studied 1812 four-year-olds in 11 states with public prekindergarten programs. Higher peer expressive language abilities were positively associated with the

children's development of receptive and expressive language. Peer-to-peer conversations were found to be important to language development, as were frequent, rich and informal conversations between teachers and children. The positive association between peers' expressive language abilities and the children's receptive language development was stronger for children within classrooms characterized by better classroom management. A well-managed classroom in which children listen respectfully to the teacher and their peers, and discussions are conducted in such a way that only one student speaks at a time, are more conducive to learning and participation (Berndsen and Luckner 2012).

Classroom Practice

Every classroom is comprised of students with a variety of individual needs who can all benefit from tailored supports. For children with CIs in mainstream settings, it is even more critical to assess individually their need for extra classroom supports, using both formal and informal measures. Kane et al. (2009) documented strategies that have been found to successfully support children with CIs in classrooms at The River School. These include differentiated instruction, longer teacher wait times, visual cues, a play-based approach, therapeutic bombardment, acoustic highlighting, social scripts, social stories, repair strategies, thematic curriculum, scaffolding, pre-teaching, and opportunities for cooperative learning.

Differentiated instruction is the umbrella term that describes how teachers adapt all areas of their practice to meet individual needs. Teaching strategies such as *wait time* and *visual cues* may help one child with hearing loss, while another might benefit more from support for maintaining attention or modeling of specific behaviors. Children with CIs should be exposed to a variety of environments in order to practice listening and responding in both quiet and noise. Prompting and modeling for appropriate eye contact and vocal volume, and practice responding to peers should occur when situations arise naturally in the classroom.

A *play-based approach* for young children is often desirable since children learn most effectively through play (Vygotsky 1978; Westby 1988). Play allows the teaching team to embed vocabulary and concepts throughout the classroom and across the day. The natural conversations between peers and with teachers that occur during play allow for frequent repetition and modeling of vocabulary and grammatical structures, as well as practice in pragmatics (Most et al. 2010). Teacher talk that embeds vocabulary multiple times in all of the activities and conversations across the day is referred to as *therapeutic bombardment*. Because it occurs in the natural environment of the classroom, it is easily generalized into children's functional repertoire (Kane et al. 2009). Teacher talk should incorporate *acoustic highlighting* techniques, wherein key vocabulary and concepts are emphasized in speech by inserting brief pauses before and after salient items. Acoustic highlighting techniques can also enhance comprehension by using repetition and rephrasing (Kane et al. 2009).

Vocabulary size is continually enhanced and assessed in the classroom. Lederberg and Spencer (2009) examined the word-learning abilities of a group of 98 children with hearing loss and found that lexicon size was more strongly correlated with word-learning abilities than age. Because children with hearing loss may need to hear new vocabulary many times in different contexts to incorporate words in their lexicons (Yoshinaga-Itano 1999), special emphasis should be placed on the acquisition of vocabulary through multiple exposures, direct teaching, and rich verbal environments (Lederberg and Spencer 2009). Once new vocabulary is introduced it should be used repeatedly across many settings to promote retention. When children fully understand a new word, they can explain its meaning, demonstrate how it relates to other words and concepts, and use it in their own speech.

At The River School teachers facilitate play-based scenarios by modeling *social scripts* that extend play, and by helping children formulate and use *repair strategies* during communication breakdowns with peers. *Social stories* also help create visual supports, with logical sequences and outcomes. Children can learn perspective taking from the repeated use of social stories that highlight desired behaviors and extinguish undesired ones. Vocabulary, key concepts, and social scripts are embedded in each story to help children internalize them.

Thematic curriculum is an approach that incorporates theme-related vocabulary and materials throughout classroom activities for several weeks at a time. The theme provides students with an overall high-interest learning topic; meaningful, realistic materials to work and play with; and the opportunity to delve into a subject in greater depth. Students guide theme selection, but teachers have multiple opportunities to embed specific skills, ensuring that academic curricular areas are threaded into the common topic. The theme offers a useful vehicle for bombarding children with targeted vocabulary.

As students encounter more rigorous academic goals with more complex language demands, they may benefit from *scaffolding* that connects new academic information with their existing bank of knowledge. Teachers may provide scaffolding by breaking down directions into a few clear steps, sitting in direct proximity to the child with hearing loss, or restating and reframing the instructions. This level of support is needed for children whose language delay places them behind the academic language level of their classmates.

Pre-teaching of content can be a critical support that prepares students for the challenge of new learning in the classroom. It allows students a preview of the information that will be taught, promoting greater understanding of concepts and increased self-confidence in the classroom. Some students with CIs will require less support than others, and teaching strategies and interventions can be modified accordingly.

Children with CIs often benefit from direct support for pragmatic language and conversation (Most et al. 2010). These skills can be coached in both small group settings and in whole group discussions. Figurative language and social turn taking can be practiced in small groups and individually with the teacher. If needed, teaching teams should work one-on-one with students with CIs to practice reciprocal conversation and prepare them to engage successfully with peers, increase confidence, and directly affect overall motivation.

In a school setting, children must be able to work collaboratively to problem-solve and share ideas; take others' perspective; summon background knowledge; and ask pertinent and salient questions. *Cooperative learning* requires students to follow group conversations in noisy environments, negotiating and listening, as well as partitioning tasks to group members. With teacher facilitation, real-time experience promotes the development of cooperative learning skills that children can apply throughout their school careers.

Building a Foundation for Literacy Development

Children with CIs often have delays in phonological awareness when compared to hearing peers (Ambrose et al. 2012; Marschark et al. 2012), and these delays can negatively impact reading outcomes (Harris and Terlektsi 2011; Dillon et al. 2012). One study of achievement results for a national normative group of students reported that only about half of 18-year-olds with hearing loss were reading above a fourth grade level (Traxler 2000). Similarly, only half of the cohort of 181 experienced CI users in Geers' (2003) report read at or above age level. Geers and Hayes (2011) reported an average reading delay of 4–5 years for students with CIs entering high school.

Explicit teaching of phonological awareness skills can play a prominent role in preparing children with hearing loss for reading (Archbold et al. 2008; Moog and Geers 1985; Geers 2003). Even very young children can benefit from explicit teaching of the skills needed for reading (Justice and Pullen 2003). Kane et al. (2009) described a multisensory program that is used at The River School to help children acquire a well-rounded, in-depth foundation in phonological awareness and literacy skills. The River School's "Mouth Time" and "Sound Spot" programs enable children to build skills across several domains, including vocabulary, oral motor skills, speech production, auditory processing, and comprehension (see Fig. 23.1 for parameters of the program).

"Mouth Time" is an early language arts program for children ages 18 months to 5 years of age. The five parameters of Mouth Time include oral motor, auditory processing, speech production, vocabulary and concepts, and phonological awareness skills. Developmentally, very young children are not ready to connect the abstract and arbitrary symbols that comprise English graphemes (letters) with speech sounds. Mouth Time turns many of these abstract graphemes into concrete mouth shapes (e.g., a flat line representing that your lips are closed when producing the "mm" sound). The mouth shapes (e.g., flat line or square lips) are paired with simple directions, such as "Put your lips together." When children see the concrete symbols and pair them with tactile and proprioceptive feedback, they begin to learn that symbols have meaning and represent certain sounds.

Unlike graphemes, Mouth Time symbols represent placement of articulators; therefore, children are able to easily decode symbols and symbol sequences by matching the symbol with the position of their lips, tongue, and teeth. Using easy-to-understand mouth shapes allows young children to establish sound-symbol correspondence, a critical foundational skill for reading. If children are not able to form the correct placement, daily oral motor practice can help them improve their articulatory strength, coordination, and range of movement, thus improving speech. The use of tongue twisters and other silly sayings can help the group practice sounds and improve co-articulation.

Mouth Time includes sound discrimination, an effective tool to help children learn to produce sounds accurately. Viseme pairs such as /p/ and /b/ look similar on the lips; helping children hear and feel the difference promotes clearer speech. Auditory discrimination tasks build into higher-level skills in auditory processing and comprehension. Using theme-based activities during Mouth Time helps highlight key vocabulary and concepts and supports understanding and carryover. By age 4 or 5, once phonological awareness skills are firmly in place, children are ready to make the leap to English letters, and Mouth Time symbols are used as a bridge. During transition, the concrete symbol (e.g., a flat line) is paired with the grapheme (/m/).

Children who have participated in the Mouth Time program have a strong foundation of sound-symbol association. A systematic approach, "Sound Spot," is then used to teach children to make letter-sound correspondences, blend sounds, practice reading words, and identify word families. In order to become fluent readers, children must employ strategies effortlessly. Explicit instruction is used to draw children's attention to the phonemic rules of the English language, consistent with the Mouth Time/Sound Spot approach.

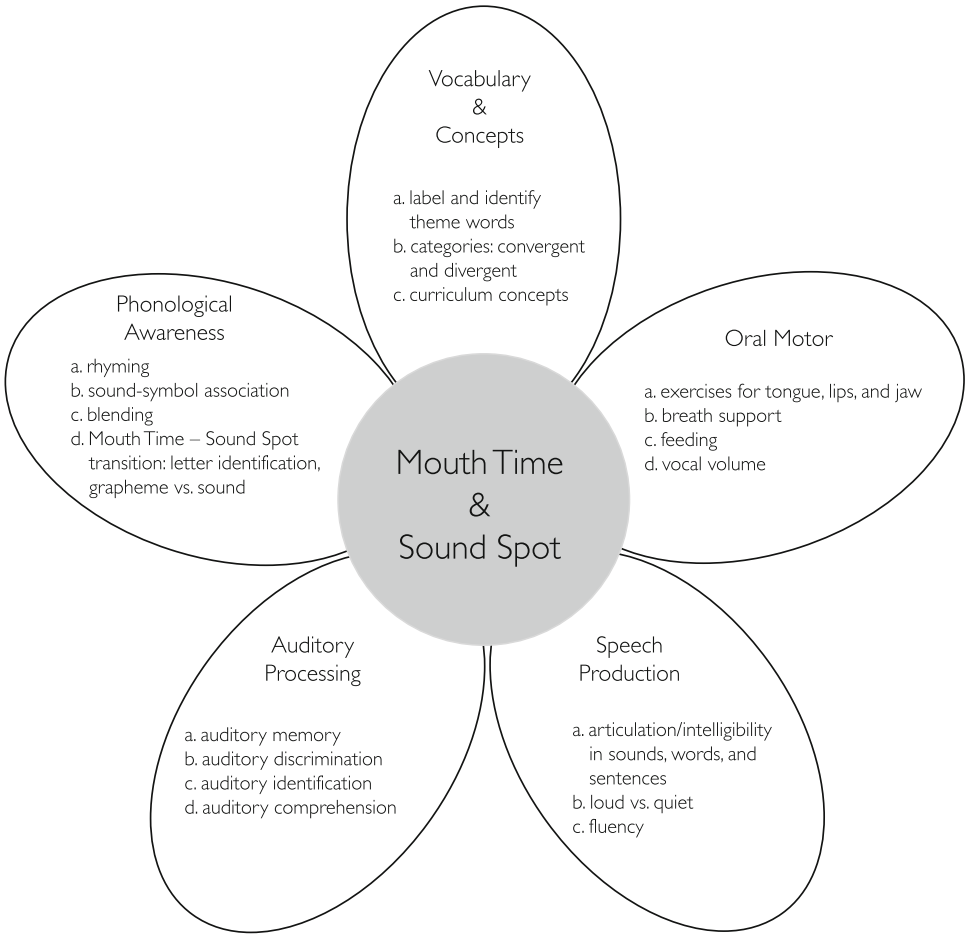

Fig. 23.1 Mouth time and sound spot parameters

During Sound Spot, students learn to mentally manipulate sounds: holding sounds in working memory, sequencing them, and then blending them to form words. Some students require extensive practice, with visual and tactile supports, in order to complete these skills.

Reading Comprehension

Luckner and Handley (2008) reviewed the literature on reading comprehension outcomes for children with hearing loss published between 1963 and 2005, and identified common areas of deficit. These include vocabulary, word recognition, fluency, comprehension, topic knowledge, syntax, and understanding of figurative language and text organization. Vocabulary size has been found to predict literacy outcomes in this population of children (Dillon et al. 2012; Schopmeyer 2009). Children with CIs have been found to acquire vocabulary at a slower rate than their hearing peers and to exhibit problems with retention of new words (Walker and McGregor 2013). Houston et al. (2012) noted similar word-learning delays in their cohort of children with CIs compared to children with normal hearing with the important exception of children who received their implants prior to their first birthday. Word learning appears to be constrained by a sensitive period that is even earlier than for speech perception and central processing auditory skills (Houston and Miyamoto 2010). Houston et al. (2005) noted that children with CIs in mainstream settings would likely continue to require additional supports, such as practice and repetition with any new material, because they have more difficulty learning and retaining novel associations between words and objects.

While many children with CIs in mainstream classrooms have shown improvements in attaining literacy skills (Geers and Hayes 2011), others continue to need support and can benefit from specific teaching strategies that target these capacities (Harris and Terlektsi 2011). Specific strategies that improve comprehension include pre-teaching, in which the teacher previews literature and important academic questions, allowing the child to practice ahead of time. When the material is then covered in the classroom, it is familiar and thus better internalized.

At The River School emphasis is placed on comprehension throughout the day and across many settings. Teachers

read aloud on a regular basis to support early literacy skills such as print referencing, rhyming and letter identification, and to model fluency, sentence construction, word pronunciation, and appropriate use of expressions and phrases. After a few repeated listening experiences, children practice the language of a book by pretending to read the book on their own or with a peer. Teachers read aloud in order to introduce difficult content or new genres, including informational or nonfiction books. When the teacher serves as the reader and interpreter, children are able to understand above their current independent reading level. With teacher modeling, children learn to make connections to the text, and to predict and summarize. Comprehension may be practiced in larger-sized groups in order to provide children with CIs with "think alouds": sharing the thinking process by talking about it explicitly. Comprehension is also addressed in small groups or one-on-one, perhaps during a Reader's Workshop conference. In order to use comprehension strategies, children need multiple exposures to teacher think alouds, both guided and independent practice, and opportunities to reflect on the ways in which "thinking about their own thinking" aids in comprehension.

Written Expression

Writing can be an area of relative weakness for children with CIs (Geers et al. 2011). Reading and writing are complementary tasks; listening to stories helps make children better writers. In the early years, the focus is on talk. The language used to talk about the book builds language skills in children and helps them bridge what is happening in the story to what is happening in their own lives. Through early and constant exposure to print, children learn that it is the print, not the picture that carries most of the meaning. They learn that letters correspond to speech sounds. They also learn directionality concepts that matter in print: left to right and top to bottom. As with the reading process, listening to stories exposes children to different text forms as they experiment with different types of writing, such as informational writing or poetry.

In order to write effectively, children must have something to say, or a purpose for writing. They must have the vocabulary to express their thoughts, the ability to spell, an audience or point of reference, and knowledge of written structure. As early writers, children write to communicate with themselves and others about content that is important to them. Teachers of emergent writers often serve as scribes, as children tell stories in pictures and/or scribbles and letter-like symbols. The writer's thinking is captured in repeated readings of the story to self and/or others. Many discrete processes are involved in writing: thinking of an idea, organizing one's thoughts, connecting speech to written words, and recalling letters and sounds.

Emergent or early writing is one of the best predictors of children's later reading success. Children need time to explore the function of writing and practice their skills through guided conversations about their writing. At The River School daily writing activities include journaling, shared writing, pretend writing, and picture captions. In shared writing, children dictate stories about a personal experience. The personal connection makes it easier for them to predict unknown words when they are rereading the story. The teacher writes the story, demonstrating the relationship between oral and written language, and allowing the young writer to observe the structure and conventions of written language: spacing between words, left/right and top/bottom orientation, and the use of punctuation. Children with hearing loss often present writing composed of shorter sentences and word omissions or substitutions. Shared writing removes mechanical barriers, allowing children to focus on content.

The classroom environment should support young writers by providing a print-rich environment, a word wall, and writing centers. An individualized approach is used to scaffold each child's level of development after the child's current writing level is established through observation. Teachers can model that they value writing attempts by displaying work prominently in the classroom, by modeling their own writing, and by producing class books. Children learn that while spoken language allows the speaker to influence the listener in the present moment, writing reaches beyond the immediate to many moments and readers. A hierarchy web can be used to explain this idea visually. Once a child demonstrates mastery, explicit instruction on intended audience is sometimes necessary.

Co-teaching in the Classroom

Many classroom teachers have little experience or knowledge of children with hearing loss. Ideally, co-teaching is used, and both teachers receive professional development relating to the technology and recommended supports for students. Jiminez-Sanchez and Antia (1999) documented the benefits of a *team teaching* approach in integrated classrooms of students with and without hearing loss. They note that having two qualified educators in the classroom provides twice the expertise, attention and opportunities for intervention, and an environment where teachers can model the negotiating skills they need to solve problems in the classroom.

Co-taught classrooms with deaf educators or special educators have been shown to provide significant benefits (Wauters and Knoors 2008). Antia et al. (2002) reported that a co-taught classroom, especially where one of the teachers was well versed in the potential challenges faced by students with hearing loss, led to higher expectations for chil-

dren that in turn resulted in better outcomes. Huber and Kipman (2012) compared the performance of 40 children with CIs ages 7–11 who were implanted before age 5, with 40 of their normal hearing peers on a variety of cognitive measures. The presence of a co-teacher in the classroom was found to correlate with higher scores on digit span, comprehension, vocabulary, and visual memory span. Co-teaching provides all students with more exposure to teacher talk and more individualized attention and support in the classroom.

Children with delays in spoken language have been shown to benefit from high quality parent and teacher talk and from the presence of higher peer language models in the classroom (Hart and Risley 1995; Mashburn et al. 2009). These conditions promote the use of open-ended questions that foster critical thinking and are essential in gauging a child's understanding of the classroom material. Additionally, systematic use of acoustic highlighting, rephrasing, and repeating may be needed throughout the day. The need for these supports will vary, and teachers need to make real-time ongoing assessments of each student's needs. When a full-time co-teacher is not an option, students may benefit from the supportive services of a resource teacher, teacher of the Deaf, speech-language pathologist, occupational therapist, physical therapist, or psychologist, depending on the individual child.

Integrating Social Emotional Learning Curricula

Children with hearing loss are at risk for delays in social development as a consequence of delayed language acquisition (Wauters and Knoors 2008). Aside from the benefits to their self-esteem and experiences with peers, intervention in the social domain can improve academic performance. Social competence in childhood has been cited as a powerful predictor of academic achievement (Wentzel 1992; Berghout-Austin and Draper 1984; Green et al. 1980). When social emotional learning (SEL) programs and approaches are implemented effectively, children's academic achievement increases, problem behaviors decrease, and the child's relationships with others improve (Elias 2004). Children who are accepted by their peers and demonstrate prosocial behaviors tend to be high achievers, whereas socially rejected children are at high risk for academic failure (Wauters and Knoors 2008; Wentzel 1991).

SEL intervention enhances academic achievement; benefits persist over time and positively affect students in multiple areas (Durlak et al. 2011). After intervention, students demonstrate positive attitudes, more competent social behaviors, fewer conduct problems, and lower levels of emotional distress. SEL interventions require time during the school day, but they do not detract from students' academic performance. Rather, students in SEL programs show academic improvement of up to 11 percentile points on standardized achievement tests, a significant gain relative to peers not receiving the intervention (Durlak et al. 2011).

Children with CIs often present with delays in social development, including the quality of reciprocal social interactions, the ability to comprehend the feelings and emotions of others, and the development of Theory of Mind (ToM) (Peterson 2004; Peterson and Seigal 2000). The child's ability to understand that others have different thoughts, perspectives, and feelings apart from their own is compromised; and further compromises the development of specific aspects of social emotional functioning.

Delays in language acquisition and subsequent delays in exposure to mental state language may cause an atypical developmental sequence of ToM in children with CIs when compared to hearing peers (Remmel and Peters 2008). A child's participation and engagement in pretend play also influences the acquisition of ToM; young children with hearing loss may be more vulnerable to delays due to later acquisition of a common language and less opportunity to engage in social pretend play experiences (Brown et al. 1997). Peterson and Wellman (2009) documented two critical differences between children with and without hearing loss: understanding pretence (imaginary, representational situations) occurs at a later age for children with hearing loss, but earlier within the sequence of emergent understanding of the thoughts of others. These findings suggest that cognitive development relating to complexity and executive function may also play a role in ToM development (Halford et al. 2007). The differences are attributed to the variability of social interactive experiences between children with hearing loss and those with normal hearing.

At The River School children with CIs benefit from interventions that target social communication, perspective taking, mental state language, and reciprocal social interactions. SEL supports include facilitation during social interactions at school; modeling prosocial behaviors during peer-based social experiences; and direct teaching of social skills (see Box 23.1 for more on classroom strategies to support social emotional learning). Thoughtful integration of SEL curriculum can maximize early learning for children with CIs in the context of play and social interactions with hearing peers, and lay the foundation for the acquisition of additional skills that enhance a child's emotional intelligence and overall social functioning.

Box 23.1 Social Emotional Learning Strategies in the Classroom

- Use social stories (individualized with familiar photos, school scenes, classroom props).
- Practice attending to multiple sources of auditory input: several people talking at one time and paying attention to the flow of conversation; active listening.
- Support group discussions: giving enough background, considering audience, and taking on others' perspectives, understanding common vs. individual goals.
- Practice working together a group: team work, cooperation, and importance of being flexible.
- Direct teaching of empathy-use of sarcasm, humor, idioms, and figurative language.
- Use of role-playing: expand vocabulary for emotions, personal reflection, and mental state language regarding how others feel.
- How to ask questions—"just me questions" (does your questions pertain only to you).
- Self-expression and clear communication, use of "I statements".
- Social scripts: highlighting faces, body language intended messages.
- Support of specials: drama to act out expressions, body language, art to explore emotions and feelings.
- Explicit teaching of presentation skills (face your audience, use a "speaking voice," monitor volume and prosody).
- Importance of eye contact for understanding social meaning and joint referencing.
- Use of reflections at the end of writing time to allow children to share their work and engage in authentic talk time about their writing.
- Teaching using peers as models.
- Think aloud to teach strategies for implicit questions and developing deeper understanding of self and others.
- Direct and indirect experiences through reading of books and writing of own experiences.

Supports for Working Memory and Other Cognitive Skills

When assessing the cognitive development and educational progress of students with CIs, the impact of early auditory deprivation on the developing brain should remain a constant consideration. Sound is experienced as a temporal and sequential signal and hence may scaffold general cognitive abilities that rely on sequential and temporal patterns (Conway et al. 2009). Sequencing skills are critical to functional short-term and working memory, as well as the subsequent encoding and retrieving of information. Beer et al. (2011) assert that the establishment of early neurocognitive function is inextricably impacted by prelingual deafness, as well as compromised auditory experience. These early neurocognitive functions are essential to early learning and can be delineated into specific areas: working memory, short-term memory, retrieval systems, phonological awareness, sequential processing, metacognition, and the capacity for fluid reasoning.

Children who experience an early period of auditory deprivation are especially vulnerable to disturbances and delays in verbal short-term and working memory processes including rehearsal and retrieval of verbal information (Pisoni et al. 2010), and deficits in phonological storage capacities (Lyxell et al. 2008). Verbal rehearsal speed largely explained the performance variability across a wide range of outcomes used to assess speech and language benefit after cochlear implantation in a large CI cohort (Geers et al. 2011). The ability to encode, process, and learn serial patterns is also impacted (Marschark 2006; Rileigh and Odom 1972; Todman and Seedhouse 1994), as is auditory memory as measured by digit span (Harris et al. 2011). Digit span forward scores have been associated with speech and language outcomes in adolescence, and backward digit span correlates significantly with measures of higher-order language functions (Pisoni et al. 2011).

Children with CIs often benefit from supports for working memory, which enables temporary storage and manipulation of information needed for such complex cognitive tasks as language, comprehension, learning, and reasoning (Baddeley et al. 1998). Fagan et al. (2007) demonstrated the far-reaching implications of working memory deficits by associating poor performance on a nonverbal visuospatial neuropsychological design-copying task with poorer performance on measures of receptive language and phonological awareness. Working memory can predict outcomes independent of IQ and is an essential component of executive function (Alloway and Alloway 2010). It is critical for following spoken directions and multiple step commands, performing the mental arithmetic in word problems, and processing sequential steps in daily living skills, as well as for more complex learning. Children need to be able to retain in memory what was just read or heard, and recall what was previously read or heard, in order to make sense of information being read, spoken, or taught (Harden 2011).

Working memory is a support for complex cognitive processes such as metacognition and fluid reasoning. Emerging in the first 2–3 years of life, fluid reasoning encompasses the capacity for logical thought and problem solving in novel

situations. Fluid reasoning requires cognitive processing "in situ"; it allows for the flexible application of existing thought to new problems or situations. Fluid reasoning requires short-term memory for aspects of information storage and for the attentional control aspects of working memory (De Abreu et al. 2010), and provides a framework that enables children to acquire new and more complex cognitive abilities (Blair 2006; Cattell 1987); specific suggestions for supporting the development of cognitive skills are detailed in Box 23.2.

Box 23.2 Cognitive Processing and Memory Strategies for the Classroom

- Provide children with additional time to think flexibly or creatively. Provide wait time to process information through their working memory system; longer pauses in group discussions, longer periods to process auditory information, and extra time to organize their thoughts and verbal responses.
- Provide visual supports for memory. A "word wall," manipulative materials, facts charts, and calculators, for mathematical concepts, such as number lines, calendars, visual schedules of activities and classroom routines can facilitate organization without taxing memory.
- Reduce the overall amount of material to be stored (e.g., shorten written sentences or number of items to be remembered), particularly when cognitive processing is a required component of the task.
- Increase the meaningfulness and familiarity of the material to be recalled\association with real life experiences, visual representations, and presentation of the same concepts in multiple ways.
- Simplify the linguistic structures of verbal language and written language material, such as in homework assignments (Gathercole and Packiam-Alloway 2007).
- Support memory through use of memory rehabilitation strategies, including use of memory cues, mnemonic devices, strategies such as repetition, visualization, grouping or chunking of data, and incorporating sensory or motor modalities, such as movement or singing into learning activities.

Facilitating Fluid Reasoning Abilities for Metacognition

- Use of research-based strategies for organizing and teaching such as those produced by University of Kansas Content Enhancement.
- Teaching children routines for planning and organization of content and materials.
- Experiential or problem-based learning and explicit and systematic teaching of metacognitive skills involved in flexible thinking through modeling, perspective taking, and guided practice with specific feedback.
- Use of peer tutors, group learning, and reciprocal teaching strategies to model thinking and collaborative approaches to problem solving.
- Use preview, review, and repetition to learning of broader concepts.
- Use graphic organizers to demonstrate relationships, for brainstorming and organizing ideas, as well as to guide students through the writing process.

Facilitating Sequential Processing in the Classroom

- Teach attending to details; utilize memory strategies and memory cues for immediate details to facilitate encoding and retrieval.
- Teach categorical separation first, then apply sequential structure to the categories; support through functional daily living skills, as well as in more advanced educational activities.
- Provide multiple opportunities for serial ordering moving from simple three place ordering to more advanced structured ordering, such as alphabetizing, ranking, and complex categorization.
- Mental planning: overall organization of thoughts can be structured via verbal planning, written steps, and graphic organizers.
- Organization of activities via structured, verbal and written planning applied to daily activities, weekly and monthly activities and applied to daily agendas, weekly and monthly calendars.
- Provide ample opportunities to learn how to sequence: utilization of sequential formats, model sequencing by logical order, and provide written or pictorial steps in a process.
- Utilization of an organized, systematic, step-by-step manner when teaching.
- Incorporate multisensory activities to facilitate learning and encoding.
- Teach long-term projects through specific steps and endpoints (Dawson and Guare 2010).
- Provide frequent sequencing activities that include manipulation of pictures, letters, words, sentences, and paragraphs to build literacy skills (Shank and Tazewell 2012).
- Utilize functional methods of sequencing: color-coding, underlining, numbering, spatial organization, outlining.

(continued)

Box 23.2 (continued)

- Apply written, step-by-step numbered formulas for learning how to problem-solve complex mathematical computations.
- Use timelines to illustrate historical and current events, paired with visual imagery to support encoding into memory.
- Provide deliberate sequential presentation of directions and information.
- Ensure daily practice with multistep directions in multiple modalities: visual, oral, and tactile, then visual and oral, then just oral.

Academic performance in school can be accurately predicted by fluid reasoning ability in childhood (Gottfredson 1997). Abilities associated with fluid intelligence continue to develop well through adolescence and are used during any processing that requires information retrieval (Ferrer et al. 2009). Notable gains have been reported on specific tests of fluid reasoning abilities by young children just one year after cochlear implantation (Edwards et al. 2006), but the close association of fluid reasoning and memory functions remains an area for educational vigilance and support. Children learn new and innovative ways to solve problems by coupling flexibility in new thinking with previous experience, thus relying on the ability to "think about thinking." Given these well-documented vulnerabilities, schools educating children with CIs alongside their hearing peers must be prepared to provide supports for these specific cognitive skills that support learning.

Supporting Motor Development and Sensory Integrative Needs in the Classroom

Language and motor development have been hypothesized to have a synchronous relationship (Magrun et al. 1981) with language developing in combination with other domains, such as motor, cognitive, and social (White 1975). As a consequence, children with CIs can benefit from assessment and support for sensory and motor development. Developmental differences have been documented in the areas of balance, complex motor sequencing, and sensory and vestibular processing (Cushing et al. 2008; Schulmberger et al. 2004; Suarez et al. 2007). Clear disturbances in motor sequencing tasks have been noted, which have been significantly correlated with language outcomes (Pisoni et al. 2011). Children with CIs may be at risk for sensory processing disorders, with deficits in vestibular, regulatory, and gross/fine motor skills that impact classroom success (Bharadwaj et al. 2009).

Vestibular deficits, specifically diminished postural control, balance, and eye tracking, have been identified in children deprived of auditory stimulation (Brey et al. 1995; Enbom et al. 1991; Selz et al. 1996). Children with CIs experience higher rates of difficulty in tasks related to vestibular system function and typically perform poorly on tests of dynamic standing balance when compared to hearing peers (Cushing et al. 2008). As a result, a child's equilibrium and ability to maintain postural control can be compromised, resulting in poor head and body righting reactions. Vestibular processing deficits, because they result in diminished ocular and postural control, balance, interpretation of body position in space, bilateral coordination, and motor sequencing tasks, can impede a child's ability to develop language, interact with peers, and feel physically and emotionally secure. Other observed patterns of vestibular dysfunction include lower muscle tone (hypotonicity) with poor co-contraction of muscle groups; poor stabilization of the head, neck, shoulders, eyes, and trunk; and postural-ocular deficits (difficulty with visual tracking and poor conjugate eye movements). Children with hearing loss can often benefit from assessment and support for their sensory and motor development and for challenges posed by vestibular deficits (Mellon et al. 2010).

Assessment and Intervention Considerations

At The River School, an occupational therapist (OT) provides individual classroom support and modification through collaboration with educators and if needed, direct intervention to help address students' needs and maximize learning; specific supports for motor and sensory development are detailed in Box 23.3. Educators typically initiate a request for an OT evaluation when a significant discrepancy exists between the student's expected and actual performance in the classroom. Assessments of functional gross and fine motor skills, balance, and sensory processing can provide valuable information about a child's ability to learn and to navigate the environment. In addition to standardized testing, clinical observations of a child's performance on a variety of tasks, and parent and teacher questionnaires with a focus on student performance patterns help identify treatment needs. Services may be delivered in the classroom, on the playground, or in a therapeutic gym, as well as community locations. Individual OT services take place in a sensorimotor gym equipped with suspension equipment to engage all sensory systems.

Box 23.3. Classroom OT Strategies That Enhance Learning

The American Occupational Therapy Association 2009 recommends the following sensory strategies for use in the classroom:

- Designing sensory-enriched classrooms with a variety of seating options and opportunities for movement and proprioceptive experiences throughout the day to help enhance body awareness.
- Structure the sensory environment to meet the student's needs (e.g., reducing distractions and improving attention to salient auditory and visual information).
- Maintain ability to organize behavior by providing scheduled sensory breaks.
- Maintain peer relationships by supporting and compensating for motor planning needs in age-appropriate games and sports.
- In consultation with educators, develop strategies for modifying the sensory, motor, or praxis demands of assignments to increase student activity.
- Prevent inattention, poor posture, and restlessness when sitting for prolonged periods by modifying seating options, allowing sensory and movement breaks, and allowing the student to work in various positions.
- Prevent social isolation by providing motor planning and social strategies to participate with peers.

Movement Activities for the Classroom

- Participate in various "animal walks," jumps, and wheelbarrow walking down the hallway during transition times to emphasize weight-bearing and joint compression.
- Classroom job assignments that embed proprioceptive work, such as helping the teacher erase chalk/dry-erase boards, classroom door opener, and helping to take chairs off of desks, are beneficial.
- Weighted vests, weighted lap pads, and weighted blankets are useful during circle time, seated work, and during naptime to help maintain a continual stream of input.
- Bumpy seats, therapy balls, or t-stools can help improve attention and give children an opportunity for movement while engaging in active listening.
- Participate in wall push-ups to "make the classroom bigger," and "dance parties" to encourage movement.
- Tying sensory bands to chair legs for opportunities throughout the day to push or pull with feet and hands or using a squeeze/fidget toy that can provide input.
- Provide children with adequate opportunities during the day for playground breaks, emphasizing jumping, running, and climbing on playground equipment.
- Provide "chewy" breaks, emphasizing the use of chewy foods, such as gum, fruit leather, and "chewy" necklaces, if needed, that provide resistance.
- Use handwriting curriculums that rely on a multisensory and developmental approach, to provide students with an opportunity to move, touch, feel, and manipulate objects, which provides kinesthetic feedback and helps children who have body awareness challenges.
- Provide deep pressure squeezes to limbs, back, and torso throughout the day.

Improving Body Awareness, Postural Motor Control, and Bilateral Coordination

- Have children sit on carpet squares or bumpy seats to help inform bodies of where they are positioned in relation to peers and teachers.
- Provide opportunities to engage trunk muscles during the day to enhance postural control (encourage sitting up tall while on the floor instead of leaning on furniture; backless chairs or therapy balls can target this as well).
- Encourage participation in activities that require both hands working together, such as rolling out play dough, cutting shapes, digging through sensory bins.
- Encourage ascending and descending the stairs and riding tricycles and bicycles to target reciprocal leg movements.
- Engage in simple games such as "Simon says" or "hokey pokey" to encourage imitation of posture and to enhance limb/body awareness.

Treatment Using a Sensory Integrative Approach

Sensory Integration (SI) theory is a comprehensive framework for assessing and treating children with underlying processing deficits. SI theory describes information processing as a neurobiological function requiring the detection, assimilation, organization, interpretation, and use of sensory input, allowing individuals to interact effectively with the environment in daily activities at home, school, and other settings (Ayres 1972b).

The ability to process sensory information has been linked to academic achievement in children with learning disabilities (Ayres 1972a, b, 1975, 1979, 1989) and contributes to a child's ability to learn and acquire motor skills (Nackley 2001). Achievement in arithmetic and reading in school-age children has also been correlated with SI function (Parham 1998); and praxis, the ability to conceive, plan, and execute motor skill, has been consistently identified as the most powerful predictor of academic achievement (Bundy et al. 2002). Deficits in integrating sensory information are often complex and include various patterns of perceptual, motor, and praxis difficulties affecting the speed and accuracy of learning and variations in sensory responsiveness that in turn affect emotional well-being and social competencies, including play (American Occupational Therapy Association 2009).

Sensory and Motor Deficits in the Classroom

Children with deficits in vestibular processing, including balance, may exhibit behaviors such as falling out of chairs or leaning on peers when sitting unsupported, as well as tiring easily with physical activity. They may appear clumsy due to difficulties with spatial orientation, accurate perception of movement, and body awareness. Developmental motor milestones, such as sitting and walking independently, may be delayed, limiting engagement in gross motor play and classroom activities. Vestibular deficits can impact reading, writing, cutting, and copying. Safety, particularly on stairs and uneven surfaces, may be a concern. Poor laterality (awareness that there are two sides of the body) and diminished bilateral integration and sequencing are also associated with poor vestibular processing. Children with CIs may have difficulty using both sides of the body in a coordinated fashion and sequencing motor actions. Often they do not cross midline and have difficulty using two hands together cooperatively. They may also display poor rhythmic and coordinated symmetrical and asymmetrical movements and movement sequences of arms, hands, and feet. Games and playground activities require adequate coordination of both sides of the body; thus, children with bilateral coordination and sequencing disorders may perform poorly in games and sports and may sit on the sidelines or avoid challenging age-appropriate activities. This can hinder peer interaction and socialization, negatively affecting self-esteem and self-concept.

Individual Treatment Considerations

Individual OT treatment is warranted when motor delays and difficulty processing sensory information impact a child's performance of daily tasks. Treatment objectives often include improving balance and postural control, ocular motor skills, bilateral coordination, and body awareness; and helping compensate for lower muscle tone through generalized strengthening. Treatment targets affected classroom skills, such as writing, cutting, drawing, and tying, as well as moving safely through the environment. A child-directed approach is recommended, including experience with a variety of props, materials, and equipment.

Balance is targeted through activities rich in vestibular and proprioceptive input, including suspended and specialized equipment such as swings, trapeze bars, zip lines, and crash pads. Swinging in different positions, when combined with a target activity such as throwing beanbags into a bin, fosters postural responses while requiring the child to maintain a stable visual field. Strength, postural control, and tone can be improved with weight-bearing, engaging the child in therapeutic positioning and handling such as work on a therapy ball.

For children with CIs who present with diminished body awareness and bilateral motor coordination, intervention goals include improving their sense of their bodies' location in space; rhythm and movement sequencing; and coordinated use of both sides of the body. Proprioceptive activities, such as pressing limbs, trunk, and back with a large bolster, and multistep obstacle courses can improve overall body awareness. Activities that require symmetrical patterns such as pumping swings with legs, catching balls, and rowing can be used to improve bilateral motor coordination. Activities that require alternating motor patterns such as tug of war, wheelbarrow walking, and swimming can improve bilateral body coordination.

Conclusion

The needs of children with CIs in mainstream settings are varied, individual and sometimes complex. Ideally, two teachers will share responsibility for classroom learning. If one of them is a speech-language pathologist, as in The River School model, then speech, language, literacy, auditory, and social goals can be easily integrated into the general education classroom (Mellon et al. 2010). With early, intensive, and individualized intervention children with CIs increasingly can succeed socially and academically in mainstream schools.

Current school culture is shifting from a strict focus on mastering content across academic domains toward developing learners who can communicate and work effectively in teams (Costa and Kallick 2008). The definition of academic success has expanded to include the coupling of academic achievement with personal skills, such as self-confidence, the ability to work collaboratively, and the technological proficiency to acquire knowledge with cognitive flexibility

and creativity. In order to acquire these capacities, children with CIs will need strong spoken language and literacy skills and well-developed social behaviors.

Schools that include children with CIs in the general education setting must hold high expectations for children's classroom performance. With improvements in early identification and intervention, and with earlier access to improved iterations of CI technology, more children with CIs will achieve academic and social outcomes commensurate with their hearing peers. At the present time, those successes are often the result of years of careful support and timely intervention—supports that should not be automatically discontinued once children who appear to have overcome early lags in speech and language enter mainstream classrooms. Only the right balance of challenge and support will allow the promise of CI technology to be fully realized, giving children with severe-to-profound hearing loss the opportunity to reach their full potential.

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