CI 2013 Symposium/October 24–26, 2013 Proceedings of the First Annual Symposium of the American Cochlear Implant Alliance

Introduction

Emerging Issues in Cochlear Implantation

Craig A. Buchman, Donna L. Sorkin

The American Cochlear Implant Alliance was established as a non-profit organization in October 2011 by a group of clinicians, scientists and educators to address the low utilization of cochlear implants and the barriers to access. It was widely felt that a new organization was needed to provide greater attention to cochlear implantation.

A board of directors was elected, bylaws were adopted, an executive director was hired and the ACI Alliance moved forward with a set of targeted activities designed to help make the cochlear implant intervention more widely accessible to children and adults who may benefit from it. The organization operates in an inclusive and collaborative manner with existing organizations in the field of hearing loss while at the same time providing a dedicated focus on cochlear implant access.

The CI 2013 Symposium "Emerging Issues in Cochlear Implantation" was the first annual conference of the American Cochlear Implant Alliance. Held in Washington, DC on October 24–26, 2013, the symposium brought together individuals who address cochlear implants in a range of settings—universities, hospitals, private clinics, non-profit organizations, schools, and governmental agencies.

The overarching goal of the Symposium was to encourage improved performance of cochlear implants through advances in the technology and expanded candidacy. Combined electrical and acoustic stimulation, through hearing preservation surgery, has now been studied extensively and found to be an approach that has merit for certain patients. Implantation in the very young as well as in older adults appears to provide performance advantages not previously recognized. The burgeoning topic of cochlear implantation among individuals with single-sided deafness is an exciting means to restore binaural auditory function. Technology promises to further shape the field by allowing remote testing and long distance therapy as a solution to limited access because of geographic distance or patient mobility.

These six topics were explored in a clinical research context by presenters who were drawn from a range of disciplines and also across the continuum of care for cochlear implantation. Speakers included basic scientists, ENT surgeons, audiologists, speech language pathologists, educators, pediatricians, gerontologists, and preventive medicine physicians. We are grateful to the individuals who shared their knowledge and experience as presenters, panelists, and audience participants.

Craig A. Buchman
Chair, Board of Directors
American Cochlear Implant Alliance and
Chair, CI2013 Organizing Committee
Vice Chairman for Clinical Affairs
University of North Carolina Otolaryngology/Head and Neck Surgery

Donna L. Sorkin
Executive Director
American Cochlear Implant Alliance

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Cochlear Implantation of the Very Young

Co-Chairs: J. Thomas Roland Jr.[1], Teresa A. Zwolan[2]
Presenters: J. Thomas Roland Jr.[1], Holly Teagle[3], Teresa Caraway[4], Ivette Cejas[5], Derek Houston[6]
[1]New York University, [2]University of Michigan, [3]University of North Carolina, [4]Learning Innovation Associates, [5]University of Miami, [6]Indiana University

Research on the effect of age on outcomes has helped drive a lowering of the age at which children are receiving cochlear implants. Evolving clinical practices have allowed surgeons to minimize surgical challenges and risks for children who receive an implant when they are less than 12 months of age while improvements in objective and behavioral audiological procedures have resulted in an improved ability to evaluate candidacy and map the speech processors of very young patients. Electrophysiological research has provided additional support regarding the benefits of early implantation. We also know that children who receive an implant early require appropriate parent-centered therapy to facilitate language development. This section will review research on the effects of early implantation on children's language, behavioral and social development. Research will be shared that compares word-learning skills in children implanted between 6 and 12 months of age with those of children implanted between the ages of 12 and 18 months. The key role of early intervention services in providing information and support to allow early implantation will be discussed.

Surgical Management of Cochlear Implants in the Very Young

J. Thomas Roland, Jr.

Cochlear implantation of very young children, especially children under age one, is regularly performed at many centers in the United States. The initial inspiration was parent driven.

The data presented in Figure 1 was published in Pediatrics in 2006 and shows 6 and 12 month IT-MAIS results for a cohort of children who received a cochlear implant prior to their first birthday compared to the trajectory of scores for children with normal hearing on this measure (Waltzman & Roland, 2005). The implication is that children implanted under age one did not start with a deficit and were on par with children with normal hearing. It was this early data, coupled with parental requests, that led our center to initially consider implantation of children less than one year of age.

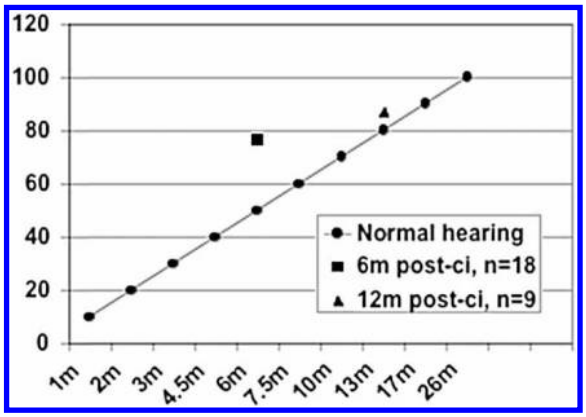

Figure 1 ITMAIS results

Considerations for cochlear implantation in the very young differ from considerations in older children in a number of ways, and often include issues related to the certainty of test results. Genetic testing may also weigh heavily in the determination for cochlear implantation vs. continued observation with traditional amplification. Although some clinicians may have concerns regarding programming, objective procedures are available that can facilitate accurate programming with this group. Anesthetic and surgical risks exist and very young children might be at higher risk of complication. The content of this presentation focused on surgical and anesthetic considerations encountered with implantation of the very young child.

Anesthesia issues: One concern is the pulmonary maturity and response to gases and the cardiac reflexes to intravascular volume changes. In the very young child the cardiac reflexes to significant blood loss do not present until very significant and perhaps life threatening loss has occurred. However the issues normalize around 6 months of age. It is for this reason that we do not implant below the age of 6 months. Additionally, data has shown that the increased risk of anesthesia is minimized when a pediatric anesthesiologist is handling the surgery.

Surgical Risks: A very young child has a small blood volume so extreme care must be taken when dealing with blood loss at the skin level, with emissary veins and with bone marrow in the immature mastoid. Strict attention to hemostasis is a necessity and even small bleeding sites, that in an older child or adult might seem insignificant, must be addressed. Diamond burs are used in the mastoid drilling to prevent blood oozing; blood loss of 35 cc might represent 10% of a child's total blood volume.

Infection concerns are an issue in the very young child receiving a cochlear implant. Young children in general are at a higher risk of meningitis. Additionally, some suggest that traumatizing the cochlea also increases this risk. Lastly, we are implanting early in the otitis media prone years, which may also increase meningitis and other infection risks. If we are considering bilateral cochlear implantation are we doubling the risk? Therefore, immunizations with Prevnar 13, HIB vaccines and, at two years of age, immunization with pneumovax 23, are extremely important to minimize the infectious risks in this age group. Peri-operative antibiotics and diligent sterile surgical technique is the rule. CI surgeons and teams must communicate to pediatricians that early and aggressive treatment of otitis media is essential in children implanted at a young age. There is very good evidence that the judicious implementation of tympanostomy tubes is safe and efficacious when indicated. Good surgical technique should minimize cochlear trauma and manufacturers are producing ever more atraumatic electrode designs.

Scalps are very thin in young children so it is this author's opinion that larger bony wells that accommodate the receiver-stimulator depth are important. The device can be depressed and recessed against the dura and fixed in place with a non-absorbable suture. The lower profile results in less tension on the delicate scalp tissues and will likely minimize untoward wound issues. Fixation minimizes device migration, especially since the skull is a dynamic organ that rapidly changes size in this age group.

Consideration of device location and orientation is important so that the magnet does not reside on the top or back of the head. We often orient the devices more vertically in young children. The facial nerve can be only one centimeter away from the cortex in this age group and can be very close to the surface at the mastoid tip. Knowledge of anatomy and good surgical training and experience operating on very young ears are prerequisites when tackling CI in the very young.

The aforementioned concerns and challenges could result in higher complication rates in this population. We reviewed our published data (Wang et al. 2010) as well as a number of other reports (James & Papsin, 2004; Waltzman & Roland, 2005; Colletti et al., 2005; Miyamoto et al. 2005; Dettman et al. 2007; Valencia et al. 2008), and found a very favorable complication rate after implantation in the very young child.

The chart in Figure 2 details complication rates in our first 50 subjects for children implanted prior to their first birthday compared to two other reports in older subjects (Ovesen & Johansen 2009; Bhatia et al. 2004).

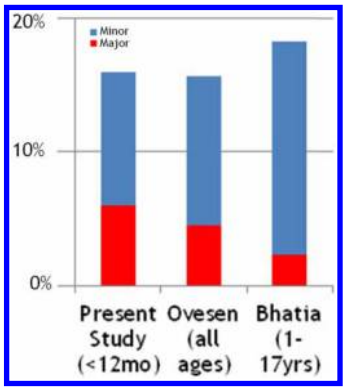

Figure 2 Complication rates in children implanted prior to their first birthday compared to two other reports in older subjects

In summary, with appropriate attention to detail and experience, cochlear implantation can be safely performed in the child under age 12 months with good results and low complication rates. Also presented was data from a number of other studies with a cumulative patient cohort of over 200 subjects. The safety and efficacy, as well as the trend toward improved outcomes when compared to later implanted children, was additionally supported in these studies.

Audiological Management of Cochlear Implants in the Very Young

Holly Teagle

Three areas of patient management that fall within the domain of the audiologist on a pediatric cochlear implant team are 1) evaluation of candidacy, 2) device programming and 3) ongoing audiological care. The decision to move forward with cochlear implantation of the very young child hinges on early identification and a collaborative team effort to identify the child's needs and resources. Candidacy criterion has evolved since multi-channel cochlear implants became available for children, and we have learned over the years that earlier implantation is desirable (Fryauf-Bertschy et al. 1997; Dettman et al. 2007; Kirk et al. 2002; Niparko et al. 2010). However, there are often obstacles to achieving the ideal time line of early identification, intervention, assessment and preparation. These obstacles may include auditory factors, medical issues or parental factors. A review of cochlear implant patient records at UNC revealed that 96% of children with "no response" ABR results eventually obtained a CI. While there are intrinsic benefits to behavioral testing and optimized hearing aid fittings, in the presence of a "no response" ABR, questions related to residual hearing should not delay cochlear implantation beyond the time it takes to resolve other considerations (Hang et al. in press).

Related to device programming, expert management and optimization of cochlear implant software and hardware is essential. Audiologists must continually update and use all tools available. Objective measures alone are not sufficient for best outcomes; behavioral measures are still the gold standard for fitting. Audiologists should resist one-size-fits-all thinking, recognize that not all solutions are answered by technology, and continuously counsel and educate our patients and families. We need to be critical observers of patient performance and use standard measures to identify red flags, to support individual progress, and to plan for future needs. Continued team efforts are important to plan and vision for the individual child as well as the next generation of candidates.

Role of the Speech-Language Pathologist (SLP) in Management of Very Young Children Who Receive Cochlear Implants

Teresa Caraway

The value of a multidisciplinary team in cochlear implantation for children has long been recognized. Neurotologists, Audiologists, Speech-Language Pathologists, and Educators of the Deaf each have unique knowledge and skills to maximize auditory outcomes for a child. It is critical for each team member to provide current information to families on the outcomes possible for today's young child receiving cochlear implant(s). Today, children born profoundly deaf who receive cochlear implants early and appropriate auditory based early intervention can develop age appropriate spoken language and literacy skills (Ching et al. 2009; Colletti 2009; Dettman et al. 2007; Niparko et al. 2010; Miyamoto et al. 2008; Wie 2010) An infant who is diagnosed with hearing loss should be immediately referred to the Speech-Language Pathologist so that intervention can begin as early as possible to take advantage of the neuroplasticity of the brain for learning. The role of the Speech-Language Pathologist is to partner with the family providing support and education to best position the infant and the family to reach their desired communication outcome. Following diagnosis, the family must learn a great deal of information and make decisions. In implantation of the very young child, family education and decisions occur within a shortened timeline thereby placing greater demand on the essential knowledge and skills of the SLP. Children with hearing loss cannot capitalize on the access to sound and spoken language without professionals who are knowledgeable and skilled in assessing and developing listening and speaking skills (McConkey-Robbins 2009). Guidelines, technical reports, position statements and resources delineate broad areas of expertise and clinical competencies that SLPs should have in working with children with hearing loss (A G Bell Academy 2007; American Speech-Language-Hearing Association 2007; Muse et al. 2013). The AG Bell Academy for Listening and Spoken Language® (AG Bell Academy) has defined nine domains of essential knowledge for professionals serving children who are deaf or hard of hearing as: Hearing and Hearing Technology; Auditory Functioning; Spoken Language Communication; Child Development; Parent Guidance, Education and Support; Strategies for Listening and Spoken Language Development; History, Philosophy and Professional Issues; Education and Emergent Literacy. The AG Bell Academy offers a certification as a Listening and Spoken Language Specialist (LSLS)™ with two designations: LSLS Certified Auditory-Verbal Therapist and LSLS Certified Auditory-Verbal Educator recognized world-wide as the gold standard for a professional serving children and their families. In implantation of the very young child, the SLP is a critical member of the CI team evaluating and monitoring child functioning in all areas of development through standardized & criterion referenced assessments, observation, and parent reporting including: auditory skills, receptive and expressive language, speech, cognitive skills, adaptive skills, play, emergent literacy, fine and gross motor skills and the child's disposition. However, standardized evaluation measures are lacking for this age of child thereby magnifying the importance of the SLP's expertise and experience. An SLP with particular expertise in auditory skill development can facilitate and monitor an infant's functional auditory skill development relative to audiological findings. Through weekly early intervention sessions, the SLP's role is to build capacity in the family's ability to be their child's first teacher and provide continuity of care. The SLP equips families with the strategies and techniques to create auditory living for their child during daily, play and social routines. Additionally, the SLP provides parent support and education and connects parents to other parents of children who are deaf or hard of hearing. Since early intervention sessions are diagnostic in nature, the SLP closely monitors the child's development and can help identify any issues or concerns that may have emerged and may need to be addressed by the CI team. Additionally as a team member, the SLP can support the audiologist during evaluations, activations and subsequent programming sessions as he or she knows the child's alerting responses, interests, and disposition while simultaneously gain valuable diagnostic information about the child's auditory skill development. Teamwork is critical in cochlear implantation of the very young child. There is a need to develop and validate robust evaluation and intervention measures for this

population to maximize auditory outcomes for the infant and family.

Effects of Cochlear Implants on Young Deaf Children's Language, Behavioral, and Social Development

Ivette Cejas

Extensive literature exists on the auditory and linguistic benefits of early cochlear implantation (Niparko et al., 2010); however, little is known about the effects of a cochlear implant on social and behavioral development. Children with hearing loss are at increased risk for behavior problems, attention difficulties, and deficits in social skills. The purpose of the study was to evaluate the effects of cochlear implantation on social and behavior development. Children were part of the Childhood Development after Cochlear Implantation study, a multi-site, longitudinal study of 188 severe-to-profoundly deaf children who received cochlear implants (CI; mean age 2.2 years) and 97 hearing controls (NH; mean age 2.3 years; Fink et al. 2007). Assessments were performed prior to implantation and at 6, 12, 24, and 36 months post-implantation (Quittner et al. 2004). Data indicated that prior to cochlear implantation, deaf children had higher rates of externalizing behavior problems, more attention difficulties, and deficits in social competence. At baseline (prior to implantation) language was significantly related to behavior problems and visual attention (Barker et al. 2009; Quittner et al. 2007). Further, language at baseline was predictive of improvements in social competence at 12 months post-implantation. Overall, results showed that by 24 months post-implantation, CI children were catching up to their hearing peers; behavior problems decreased and social competence improved. Thus, our results suggest that early cochlear implantation has beneficial effects on behavior and social development. CI clinics should consider screening children annually for behavior problems and social skills deficits to help identify children who need additional interventions. Further, research should focus on developing evidenced-based interventions to improve behavior and social skills in children with hearing loss.

The Role of Early Auditory Experience on the Development of Word-Learning Skills after Cochlear Implantation

Derek Houston

Ever since the FDA lowered the approved age for cochlear implantation to 12 months of age in 2000, scientists and clinicians have wondered whether implantation before 12 months should be considered. A growing number of studies have assessed speech and language benefits of implantation before 12 months with mixed results. At least 13 studies have shown better language (receptive and/or expressive) and/or speech intelligibility outcomes in children implanted under 12 months compared to children implanted between 12 and 24 months (Colletti et al. 2005; 2009; 2011; Dettman et al. 2007; Holman et al. 2013; Holt & Svirsky 2008; Houston et al. 2003; 2012; Houston & Miyamoto 2010; Leigh et al. 2013; Miyamoto et al. 2005; Nicholas & Geers 2013; Schauwers et al. 2004). However, no study (out of at least 7 so far) has found better speech perception – word recognition or speech discrimination – in children implanted under 12 months (Holt & Svirsky, 2008; Horn et al., 2007; Houston & Miyamoto, 2010; Leigh et al., 2013; Lesinski-Schiedat et al., 2004; Miyamoto et al., 2005; Schauwers et al., 2004). One possible reason for this pattern of findings is that implantation before 12 months does not improve hearing or speech perception compared to implantation during the second year; rather, it may facilitate children's ability to integrate sound with what they experience through their other sensory systems. We investigated this possibility by assessing novel-word-learning skills in ten children implanted between 6 and 12 months of age and 10 children implanted between 12 and 16 months of age. We used an intermodal preferential looking paradigm (Houston et al., 2012). Both groups were tested 12 to 18 months after implantation. The groups did not differ significantly with respect to pre-CI hearing, communication mode, number of bilateral recipients, or maternal education. We found that the earlier implanted group performed significantly better on the novel-word-learning task than the later implanted group. Moreover, performance on the task predicted vocabulary and language outcome measures two to four years later. The findings suggest that early access to sound may facilitate important multimodal integration skills that are fundamental to learning novel words and developing age-appropriate language.

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Cochlear Implants in Multiply Involved Children

Co-Chairs: John Oghalai MD[1], Nancy Young MD[2]
Presenters: Nancy Young[2], Teresa Zwolan[3], Susan Wiley[4], Jill Ellis[5], Heather Bortfield[6], John Oghalai[1]
[1]Stanford University, [2]Northwestern University, [3]University of Michigan, [4]Cincinnati Children's Hospital, [5]Center on Early Intervention on Deafness, [6]University of Connecticut

Children who are cochlear implant candidates often have multiple disabilities, including those that raise questions about cognitive potential and prognosis for spoken language. The role of a cochlear implant in the care of these children is an important issue, especially in light of the increasing survival of very premature infants at risk for multiple disabilities. Historically most implant centers did not consider children with developmental delays to be acceptable candidates. Implant candidacy selection was influenced in part by disagreement over whether cochlear implantation is an effective and appropriate intervention for deaf children. The result was typically an implant candidacy process strongly biased toward children expected to develop spoken language equivalent to their hearing peers. Herein, we summarize the variety of perspectives presentations at the recent CI 2013 conference regarding this diverse population of deaf children. We hope to provide professionals with a new framework in which to consider these children as cochlear implant candidates.

Introduction: The decision of whether or not to perform cochlear implantation with a child with multiple disabilities is a difficult one (Corrales and Oghalai 2013). The challenges facing implant programs serving the deaf plus population are many. They include more time consuming and difficult hearing aid fitting and determination of aided benefit as well as more challenging programming. Current measures to determine benefit and progress post implantation are also not designed for this population. Measurements demonstrating improved quality of life that includes assessments of the quality and variety of social engagement are lacking (Pierson et al. 2007). Even the terminology is difficult. Here, we use multiple disabilities, developmental delays, "deaf plus" as interchangeable although we recognize that perhaps there are certain stigma associated with some terms but not others. In any case, implantation and outcomes evaluation of children with developmental delays have lagged behind that of their typically developing peers.

The purpose of this manuscript is to summarize the variety of perspectives presentations at the recent CI 2013 conference regarding this diverse population of deaf children. We hope to provide professionals with a new framework in which to consider these children as cochlear implant candidates. An important goal of the session is to encourage further research in regards to implantation of children with developmental delays.

Unlocking Hidden Communication and Cognitive Potential

Nancy Young

Cochlear implant programs may be hesitant to implant children with multiple disabilities for a variety of reasons including concern about cognitive impairment limiting or precluding benefit. Implant teams focused on spoken language as the primary outcome measure may view children with poorer cognitive potential, especially those with significant motor impairment and oral motor dysfunction, as unfavorable candidates. The Lurie Children's Hospital of Chicago Cochlear Implant Program has been implanting complex infants and children with conditions such as CHARGE, severe cerebral palsy and history of extreme prematurity with developmental delays for more than a decade. Our experience tells us that improved hearing through cochlear implantation is invaluable in helping many of these children to develop their full communication and cognitive potential. Evaluations currently available cannot accurately predict post implantation outcomes nor does our standard testing capture improvements in quality of life experienced by the child and family. We have observed that multiply involved children typically benefit from improved detection of sound as it enables more meaningful engagement with family and caretakers. Measurable word recognition ability may emerge, often after prolonged period of device use. Similarly, expressive language that may include spoken words, sign and/or use of augmentative communication devices, may develop depending on the child's unique constellation of complicating conditions.

Comprehensive evaluation of intelligence is often a valuable pre-operative tool. It provides a snap shot of current skills in multiple domains such as language, attention, memory, social responsivity, and motor skills. However, using this type of evaluation as a determinant of implant candidacy is not recommended as these assessments are not predictive beyond a year nor do they predict outcomes after cochlear implantation or other interventions. These evaluations may also be compromised by the impoverished language of many deaf children and often are further limited when applied to children with motoric disorders who are unable to manipulate objects used in non-verbal tests of reasoning. These factors may contribute to underestimation of cognitive potential.

On the other hand the potential of cochlear implantation to improve these children's language and cognition is significant. Language is critical to development of cognition because children need an internal language process to translate their experience. Better language builds reasoning and cognition which in turn improves language. Earlier intervention to improve language through hearing is advantageous for both typically developing and multiply involved deaf children because language and cognition boot strap one another. Cochlear implantation significantly and positively impacts development of sign and spoken language (Robbins et al. 1997). Thus early implantation has the potential to impact both language and cognition during critical years of brain development. Not to consider implantation based primarily upon limited cognitive potential should be done with great care as an opportunity to improve quality of life, if not language and cognition may be lost.

Determination of Candidacy and Post-Operative Management

Teresa Zwolan

The safety, efficacy, and benefits of cochlear implants in children have been well documented, opening the

door for greater numbers of children with additional disabilities to receive cochlear implants. Advances in technology, such as NRT, NRI, and ART, make it possible for clinicians to objectively set speech processor levels in this difficult to test population while increased experience working with such children has clearly demonstrated that many children with additional disabilities receive great benefit from a CI.

Pre-operatively, audiological management of children with additional disabilities (AD) is challenging as children with AD are frequently unable to participate in standardized test protocols. When this happens, clinicians must develop creative procedures for evaluating benefit from amplification or from the CI. Often, clinicians rely on the results of parental questionnaires that inquire about the effect the hearing aid or CI has on behavior, communication, production, eye contact, play, family and social interactions, quality of life, and parental satisfaction, to name a few. Preoperatively, it is important to counsel families regarding the possible identification of learning difficulties and mild neurological deficits when children receive an implant at a young age. When a disability is known, it is important for the clinician to warn parents about the effect the disability may have on performance and for the clinician to refer the patient to other professionals to facilitate development of realistic expectations.

There are several things the clinician can do to facilitate receipt of reliable responses from the child. These include having adequate seating that supports the child's head and neck so the clinician can see the child's face and view their response to sound; utilization of a test assistant to manage behavior and help evaluate the child's responses; scheduling of additional appointments or reduced appointment time to ensure accuracy of the information received and reduce patient fatigue, and minor adjustments in the test setting such as relocation of Visual Reinforcement Audiometry (VRA) equipment and dimming of lights to make the visual reinforcements more visible or appealing to the child. Additionally, stimuli routinely delivered via tape can be administered live voice to reduce the formality of the test situation while pre-teaching can be used to ensure the child understands the task. Importantly, clinicians should never assume that a child with additional disabilities cannot perform a task. Each appointment with the child should be viewed as an opportunity to evaluate performance and to work with the child to facilitate future responses. Close monitoring of and interaction with all specialties involved in the child's care is needed to wholly manage the patient and his/her needs. Additionally, regular communication with the child's school can result in an improved understanding of the benefits of CI and provides an opportunity for others to provide suggestions that may aid in programming the device.

Although audiological management of a CI in children with additional disabilities can be challenging, provision of sound via a cochlear implant can provide such children with improvements in communication, cognitive function, social and relational skills, quality of life, and may increase the likelihood of future independence.

Cochlear Implant Candidacy for children who are multiply involved: A developmental-pediatric perspective

Susan Wiley

Due to the high rate of additional disabilities among children who are deaf/hard of hearing (Boyle et al. 2011; Gallaudet Research Institute Report 2011), recognizing developmental trajectories and patterns is important in order to provide appropriate supports for all a child's needs. Etiology of hearing loss is not always a good indicator of risk for additional developmental concerns.

Furthermore, as young children are receiving implants at very young ages, developmental concerns may not be evident at the time of implantation or candidacy evaluation. Thus, ensuring on-going surveillance for children who have received an implant is important in order to adapt interventions and supports appropriately to meet all of a child's needs, not just those related to hearing.

We should also be cognizant of the current limitations in the literature about this group of children as rarely have comparison groups included children with disabilities, but rather has compared children to typically developing children who are deaf/hard of hearing. Merely using this comparison group will likely always indicate that the child would not make the same rate of progress. However this approach does not allow clinicians or families to understand what are expected benchmarks and rates of progress for an individual child.

As many children with implants and additional disability have a variety of medical and developmental needs, partnering with families to identify areas of priority as well as providing appropriate support and adaptations will ensure children will make meaningful progress as they grow and develop.

Meeting Early Education Needs of Deaf Children with Additional Medical Needs and Developmental Delays

Jill Ellis

Teachers and therapists working in early intervention programs must consider not only the etiology and severity of a child's hearing loss, but are also charged with creating a holistic approach to neuro-development when determining best practices for implementing a family based individual education plan. For children who are "deaf plus" these considerations must be expanded to address the impact of both environmental and genetic influences. Examples of the deaf plus population include children whose conditions include profound deafness, autistic spectrum disorder, cerebral palsy and sensorimotor integration challenges. Best practice techniques include a sensory integration diet; tips and prompts to promote effective learning outcomes for children who have autism, effective positioning for children who have motor challenges; the influence of inborn temperament traits for observing behavior and determining appropriate curricula and positive reinforcements, and strategies for utilizing effective preference assessments techniques. Routinely including patient-specific information from the child's teachers in the cochlear implant team decision-making process also is beneficial to patient outcomes.

How is Parent-Child Communication Affected in Deaf Plus Children?

Heather Bortfield

In recent years, researchers have begun to document a relationship between joint attention and language development in children. This work has played a critical role to the development of new therapeutic interventions for children with developmental disorders, such as Autism Spectrum Disorders. Given the asymmetry in primary communication modalities in deaf children-hearing parent dyads, therapeutic approaches that emphasize the establishment and maintenance of joint attention in these dyads may likewise facilitate language development. However, we first need to document how joint attention is established in deaf children-hearing parent dyads and how this compares to the process in hearing children-hearing parent dyads. The current study compared joint attention behavior during free play sessions between primary caregivers and children who were candidates for cochlear implantation. Typically developing hearing children were recruited as age-matches to the deaf children for comparison purposes. Data suggest that the communication between hearing parents and their deaf children is aided by the establishment of joint attention, particularly when parents use multiple communication modalities (e.g., touch, vision) to supplement their speech. We advocate moving beyond standardized clinical measures in an effort to obtain rich data on these more subtle aspects of parent-child communication, which may provide evidence of an important scaffold for subsequent language learning (e.g., via a cochlear implant).

Deaf and developmentally delayed: What is the physician's role?

John Oghalai

Children with special needs require complex, individualized therapy to maximize their long-term quality of life. One subset of children with special needs includes those with both developmental delay and deafness. Currently, there is little compelling evidence supporting the idea that cochlear implantation provides benefit to children that don't have the cognitive potential to develop normal speech and language. As well, pediatric cochlear implantation does entail a certain level of risk. Therefore, many centers will not implant deaf children if they have severe cognitive delays. Alternatively, some centers implant nearly every child referred to them regardless of the child's developmental status. This often leads to parental disappointment and disenfranchisement when the cochlear implant does not resolve the myriad of disabilities facing the child. It is critical for the parents to have appropriate expectations regarding outcomes, but in this population data is lacking.

A common way of treating hearing loss to use powerful hearing aids to achieve vibrotactile sensations or acoustic sensitivity to low frequency sounds. The typical clinical outcome is sound awareness, i.e. the ability to detect environmental sounds and voices. A cochlear implant can improve auditory thresholds and word recognition ability, but because of the lower intelligence of patients with developmental delay, this value of this additional information is questionable.

Our preliminary data demonstrate that linguistic and cognitive development increase in children with developmental delays after cochlear implantation (Oghalai et al. 2012). Based on these retrospective data, we hypothesize that development and quality of life will improve more in deaf children with developmental delay after cochlear implantation compared to when they were initially treated with hearing aids prior to cochlear implantation.

We are performing a multi-site, prospective, longitudinal study to answer the question of which intervention provides more benefit to this population of children using validated, norm-referenced tests. All children eligible for cochlear implantation will undergo a

cognitive evaluation (Caudle et al. 2012; Katzenstein et al. 2009). Patients can be given either hearing aids or a cochlear implant per the clinical recommendations of the study site's cochlear implant team and followed longitudinally for two years. If our hypothesis is correct and cochlear implants significantly improve development and quality of life in deaf children with developmental delay, our study will provide essential evidence to support clinical decision-making in this population.

Thus, our long-term goal is to develop guidelines that may help when selecting a treatment for hearing loss in a child with developmental delay. This proposal is significant because children with special needs are deserving of evidence upon which to base treatment decision-making, but remain underrepresented in the medical literature and are often not studied. This research is designed to meet the criteria for the NIH roadmap because it will generate this type of objective evidence that can directly improve patient care.

Conclusion: Increasingly, pediatric cochlear implant programs are viewing implantation as a means of helping deaf children to achieve their full communication potential. This view does not exclude those children whose other disabilities may limit their cognitive potential or ability to develop spoken language. Early access to hearing may positively impact language, both spoken and sign that in turn supports further cognitive development. Denying this intervention to young children based on concerns about cognition has risks. First, it may guarantee that the ultimate cognitive level that the child might reach is stunted. As well, measuring intelligence in deaf children is difficult because many of the commonly-used measurement tools test how children respond to verbal commands. Underestimating intelligence is even more likely in deaf children with motor and visual impairments who are also unable to complete nonverbal tests, such as those based on motor skills. However, beyond the usual risks associated with cochlear implantation in any child, there are additional risks to implanting a child with developmental delays. It may lead to increased stress and reduce the time available for other therapeutic efforts, such as physical therapy. If a child does not respond to non-auditory stimulation nor demonstrate social responses to caregivers despite appropriate therapies, the likelihood of benefit is minimal. Therefore, it is important for the pediatric cochlear implant team to carefully assess, recognize, and weigh the potential risks and benefits of cochlear implantation in children with multiple disabilities. Major challenges remain in developing and using validated test measures that accurately assess the degree of a child's disabilities and their impact on outcomes after cochlear implantation.

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Hearing Preservation Cochlear implantation

Co-Chairs: Bruce Grantz[1], Oliver Adunka[2]
Presenters: Renee Gifford[3], Larry Lustig[4], Christopher Turner[1], Kate Gfeller[1], Camille Dunn[2], Margaret T. Dillion[5], Bruce Grantz[1], Oliver Adunka[2], Adrien Eshragi[5]
[1]*University of Iowa,* [2]*University of North Carolina,* [3]*Vanderbilt University,* [4]*University of California, San Francisco,* [5]*University of Miami*

Over the past decade, hearing preservation and subsequent combined stimulation has emerged as a new stimulation paradigm to improve cochlear implant performance. Specifically, the combination of electric and acoustic hearing typically allows patients to better utilize their cochlear implant for everyday listening situations such as music perception or speech perception in noise. As clinical applications for this new strategy broaden and patients with greater degrees of residual hearing are considered for cochlear implantation, it appears paramount to clearly define the clinical applications, patient populations, expected outcomes, and future research required. Since this strategy potentially includes a new patient population, cost effectiveness should be reviewed and reimbursement strategies should be developed.

With the expansion of cochlear implant candidacy criteria, more hearing impaired individuals have access to this technology. However, instead of simply replacing acoustic hearing with electric stimulation through the

implant, electric-acoustic stimulation provides an added benefit especially in challenging listening environments while maintaining residual acoustic hearing. The preservation of hearing has thus emerged as a key topic in cochlear implantation and recent research has been able to confirm the various benefits of non-traumatic cochlear implantation even in candidates without residual hearing.

Introduction
Bruce Gantz

Hearing preservation in cochlear implantation and subsequent electric and acoustic stimulation have evolved over the past decade. As such, hearing preservation rates have been carefully documented utilizing various electrode carriers and surgical techniques in both Europe and North America. A careful analysis of the data shows that shorter electrode carriers typically carry a higher rate of hearing preservation. Electric stimulation, alone, on the other hand, may be limited using very shallow insertion depths. However, once hearing preservation efforts have been successful, the ipsilateral (and contralateral) combination of both electric and acoustic stimuli typically provides individuals with improved speech discrimination abilities especially in noise. Progressive loss of residual hearing has been observed and long-term data continue to evolve (Gantz et al. 2009; Turner et al. 2008; Gantz et al. 2006; Gantz et al 2005; Gantz & Turner 2004).

Hearing Preservation Cochlear Implantation – An Overview
Renee Gifford

An overview of the clinical outcomes data available to date was provided. Specifically, Dr. Gifford showed data from her own lab testing Hybrid and EAS patients from multiple institutions. The data demonstrated the performance differences between bimodal patients and Hybrid patients and emphasized the importance of proper audiological management of these patients, which is typically quite complex. Many topics related to combined stimulation remain unstudied to date (Carlson et al. 2011; Gifford et al. 2010; Dorman & Gifford 2010; Gifford et al. 2008).

An Overview of Current Clinical Systems
Lawrence Lustig

Various clinical systems are currently available. However, at the time of the meeting, all systems have remained under clinical investigation with the FDA. Cochlear Corporation's Hybrid system utilizes relatively shallow electrode insertion lengths of 10 mm. The electrode is typically inserted through a round window related cochleostomy. Hearing preservation rates with this electrode carrier are very favorable with hearing preservation rates of well above 90 percent. The MED-EL EAS system utilizes a modification of their standard cochlear implant electrode array. This electrode, however, features a compressed electrode distance and is thus designed for shorter electrode insertions of about one full cochlear turn. Such insertions have been demonstrated to result in hearing preservation rates of slightly above 90 percent. Also, speech perception results via electric stimulation only utilizing this electrode show very good results comparable to conventional cochlear implantation.

Both manufacturers have bimodal speech processors combining hearing aid and cochlear implant technology into one device. These devices are key in providing the added benefit of preserved acoustic hearing. Cochlear Corporation's combination processor will likely be the standard speech processor for all cochlear implant recipients underlying the future importance of this technology (von Ilberg eta al. 1999; Kiefer et al. 1998; Gantz & Turner 2003; Woodson et al. 2010).

Benefits of Combined Stimulation for Speech Perception in Quiet and Noise
Christopher Turner

There appear to be various reasons as to why cochlear implant performance scores continue to evolve over the past decade: improved peripheral resolution in the newest patient populations and decreased duration of deafness with Hybrid or EAS patients essentially having no time of complete auditory deprivation.

Various sets of data support these theories. Spectral resolution, for example, predicts speech perception outcomes. Hybrid patients, for example, typically have very good peripheral resolution despite their much shorter electrode insertion depths and thus perform exceptionally well. In fact, many patients with short electrodes can perform equally well when compared to long electrodes despite the much closer electrode spacing in short electrodes.

Music Perception with Combined Stimulation
Kate Gfeller

Low-frequency information available through preserved acoustic hearing can assist CI recipients in segregating the target speech signal from background noise. However, low-frequency information can also assist in the perception of salient features of music such as rhythm, timbre, and pitch as well as the lyrics in vocal music.

Music and speech have very different sound features thus providing multiple technical difficulties.

Specifically, music has a much greater frequency spectrum than speech, the spectrum of music is typically highly variable and changes rapidly and the intensity of a speech signal is much less variable than that of music. Hence, music requires a much better resolution of pitch and spectral shape than speech.

Various features of music have to be extracted before it can be interpreted and/or enjoyed. Pitch ranking remains fundamental and Hybrid patients remain superior to conventional cochlear implant recipients with regards to this task. Accordingly, simple and complex melody recognition tasks demonstrate that hybrid patients approach performance scores of normal hearing listeners. Similar findings were documented for various other tests such as interval normalization, lyrics recognition, and timbre recognition. Accordingly, hybrid subjects demonstrated improved speech perception with background noise.

The ability to perceive pitch correlated with speech perception with background noise, recognition of prosody marking of linguistic contrasts, tone discrimination in Mandarin Chinese, and in talker discrimination. Thus, preserved residual hearing benefits in pitch and timbre perception and in an improved extraction of a target speaker from background noise or music.

Sound localization with Combined Stimulation
Camille Dunn

Localization of sound is the individual's judgment of direction and distance of a sound source. Proper sound localization skills are important for various tasks and have been linked to quality of life measures. Cues include timing and level information on the sound source. Previous studies are inconclusive on whether or not contralateral use of cochlear implant and hearing aid (bimodal hearing) improves sound localization abilities. In fact, it appears that patients with bilateral cochlear implants have better sound localization abilities than most bimodal users. The difference appears to be due to the fact than various devices might not accurately convey differences in level and timing.

The potential localization benefit of electric acoustic or hybrid stimulation is that similar signal processing across ears likely results in improved coding for level and timing cues. Also, preserved low-frequency hearing may provide an additional advantage for sound localization. In fact, sound localization was studied in 11 adult Hybrid short electrode cochlear implant recipients, who wore a contralateral hearing aid. Each subject was assessed using various set-ups including testing in combined mode, ipsilateral Hybrid mode, and in a bimodal listening condition without the hearing aid on the cochlear implant ear.

The results demonstrated no significant difference between the combined mode and the listening condition using two hearing aids, only. However, the bimodal listening condition showed significantly poorer localization abilities when compared with the combined mode utilizing bilateral hearing aids and a cochlear implant.

Thus, bimodal users (cochlear implant and hearing aid on contralateral ears) do not benefit in terms of localization abilities. However, with the addition and utilization of ipsilateral low-frequency hearing after cochlear implantation, localization abilities improved statistically.

Quality of Life After Hearing Preservation with Cochlear Implantation
Margaret T. Dillon

Various quality of life measures have been obtained after cochlear implantation. There are reported improvements in communication abilities, reduction in feelings of isolation or being a burden to others, and improved relationships with loved ones. These subjective improvements are commonly experienced equally by older and younger adult recipients.

As part of a large EAS clinical trial, two subjective benefit materials were recorded, the hearing device satisfaction scale and the abbreviated profile of hearing aid benefit (APHAB). Each material was collected pre-operatively, at initial EAS stimulation and at 3, 6, and 12 months post EAS fitting. Assessment of satisfaction was performed focusing on different features of the device such as cosmetics, sound quality, manipulation, and effectiveness in noise. Both materials demonstrate a statistically significant improvement between the pre-operative and the 3 months data assessment. Data continued to improve after this interval, although the improvements were not statistically significant.

For the APHAB material, the greatest advantages were seen in ease of communication, background noise, and reverberation. There were no recorded benefits in aversiveness of device use. Other previously published reports also document the improvement at the 3-month interval. Also, there is a previously reported improvement in quality of life in hearing preservation cases.

Thus, there appears to be a generalizable improvement in device satisfaction whereas the largest gains were documented for challenging listening environments such as in movie theaters according to the HDSS material. There were social, emotional, and physical improvements but no benefits in cosmetics, visibility to others and in manipulations such as insertion/removal of the device. The APHAB showed benefits for ease of communication, listening in background noise, and reverberation categories but did not document a difference on the aversiveness subtest.

Consideration for Progressive Hearing Loss Over Time

Bruce Gantz

Hybrid or electric acoustic stimulation of the auditory system heavily relies on successful hearing preservation. As previously detailed, hearing preservation rates are very favorable, at least when viewed in the short term. The underlying pathology responsible for the hearing loss, however, may cause a progressive loss of hearing over time.

Various studies have been undertaken to evaluate this effect. There appears to be some loss of low frequency acoustic hearing in the first few months following in some subjects; however if the acoustic hearing is preserved it appears to be maintained for up to twelve years. The cause of this loss is unknown, but might be related to some reaction within the cochlea associated with the interaction of electrical and acoustic hearing. Loss of hearing following implantation can occur, but most of the loss becomes apparent after the implant has been activated. Results of clinical trials with different length electrodes also suggests that longer electrodes (20 mm) induce more hearing loss than shorter lengths (16 mm and 10 mm) If acoustic hearing is lost the cochlear implant typically provides proper electric stimulation to maintain the individual's communication abilities (Gantz et al. 2009).

Physiologic Consequences of Intracochlear Electrode Placement

Oliver Adunka

Development of a real-time monitoring system might help to improve various aspects of electrode placement. If successful, such a system may improve hearing preservation rates while optimizing electric stimulation. Multiple approaches have been proposed many of which do not allow for real-time feedback during the electrode insertion process.

The proposed method features a recording system built into the cochlear implant device. In contrast to neural response telemetry systems utilized in current cochlear implant systems, responses to acoustic stimulation may more accurately reflect the hair cell and neural substrate for combined stimulation.

An animal model was established to simulate cochlear implant electrode insertion under changing conditions including various levels of sensorineural hearing loss. These results demonstrated the feasibility of this approach and subsequent human experiments conducted during cochlear implantation confirmed these findings. Specifically, acoustically evoked potentials remained robust even in the vast majority of both pediatric and adult cochlear implant recipients. Furthermore, even greater responses were obtained when the electrode was placed within scala tympani as opposed to the round window. Interestingly, these recordings correlated well with postoperative performance measures. Future efforts will focus on the translational aspect to incorporate this technology into future cochlear implants (Demason et al. 2012; Choudhury et al. 2012; Suberman et al. 2011; Choudhury et al. 2011; Campbell et al. 2010; Adunka et al. 2010).

Biologic aspects of hearing preservation

Adrien Eshraghi

Cochlear implant trauma (EIT) causes not only direct tissue trauma and cell losses, but also generates molecular events that may initiate programmed cell death (PCD) via various mechanisms such as oxidative stresses, release of pro-inflammatory cytokines; activation of the caspase pathway which can result in apoptosis; generation of pro-apoptotic signal cascades via, for example, mitogen-activated protein kinases/c-Jun-N-terminal kinases (MAPK/JNK) within the damaged tissues of the cochlea which can lead to a loss of residual hearing. Animal studies were performed to dissect the mechanisms involved in loss of hair cells and support cells. We found that the molecular damage encountered in cochlear implantation trauma involves different type of activation of program cell death in hair cells and support cells. These mechanisms are different in support cells as opposed to hair cells. Caspase-3 activation was not observed in HCs of any turn at 12 and 24 hrs, but p-c-Jun labeling was observed at 12 hrs in both HCs and SCs of middle and basal turn and in HCs of all turns at 24 hrs. There is also a window of opportunity to treat the cochlea with appropriate otoprotective drugs before onset of cell death in HCs.

Recent research of relevance and key findings

Recent research findings on hearing preservation and combined electric acoustic stimulation can be summarized as follows:

Outcomes research has demonstrated various benefits of combined stimulation:

- Improved speech perception in noise
- Improved music appreciation
- Improved quality of life (various measures)
- Improved sound localization (over bimodal users)

Outcomes research has documented the clinical results from hearing preservation:

- Hearing preservation can be successful in about ninety percent of subjects.
- Shorter electrodes are typically more successful in preserving residual hearing when compared to long electrode insertions.
- Currently, free-fitting lateral wall electrodes are utilized. Histological evidence points to a greater insertion trauma when pre-formed arrays are used.
- Long-term hearing preservation data are mixed.

Intraoperative monitoring

- Intraoperative monitoring seems feasible (acoustic stimulation and intracochlear recording). Clinical algorithms are still lacking and human experiments are ongoing
- Might be able to measure the appropriate electrode insertion depth instead of using various insertion length without a true physiologic marker
- Inhibition of apoptotic pathway and other adjunct measures seem very promising.

Major issues or challenges remaining

- Clinical protocols are similar but not standardized
- No standardized electrode insertion lengths
- Audiological follow-up remain extremely time consuming and complex
- Need to better understand fitting parameters
- Intraoperative monitoring not yet clinically available

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Single Sided Deafness and Cochlear Implantation

Co-Chairs: John K. Niparo[1], Jill Firszt[2]
Presenters: David Ryugo[3], Ann Marie Tharpe[4], John K. Niparo[1], Jill Firszt[2], Bruce Gantz[5], Mario Svirsky[6]
[1]University of Southern California, [2]Washington University, [3]Garvan Institute of Medical Research, [4]Vanderbilt University, [5]University of Iowa, [6]New York University

Individuals who listen with just one ear have significantly reduced communication abilities. Recent studies have suggested that cochlear implantation can benefit those with unilateral hearing loss (UHL) when one ear hears normally and one ear has profound hearing loss, also referred to as single sided deafness (SSD). Typically these recipients have been adults with sudden sensorineural hearing loss who have affected ear duration of deafness of ten years or less. This session brings together biological, historical and clinical perspectives of SSD along with current findings in individuals who have received cochlear implants. The invited speakers and panel members address questions of candidacy, benefit, limitations, central auditory system effects, cost effectiveness, and future research needs in this clinical population.

Sensory Experience and Brain Development
David Ryugo

Auditory nerve input to the brain is normally dense and highly organized. Hearing loss (HL) reduces signal transmission, which causes quantifiable changes to excitatory and inhibitory circuits. Auditory input is not only depleted with HL but less organized resulting in more difficult speech comprehension. In normal hearing animals, the large synaptic endings of auditory nerve fibers (endbulbs) ensure that information is transmitted without fail and with high fidelity to the next neuron in the auditory nerve circuitry chain. Analysis of cochlear nucleus neurons shows abnormal primary afferent and reduced inhibitory input in mice with both congenital and acquired HL. Reductions of sensory input fundamentally alter brain organization at the earliest processing center in the central auditory pathway. These changes in sensory circuitry represent key issues that challenge hearing aid and cochlear implant (CI) effectiveness. In a recent study, differences were observed in the inferior colliculus of unilaterally deafened rat pups compared to normal hearing (NH) controls, including dendrite alignment changes which could translate to poorer frequency resolution. Hearing loss at any age results in loss of sound details, timing jitter, collateral sprouting of normal fibers into denervated regions, and possible dendrite rearrangement at higher levels. Effective strategies for creating or improving assisted hearing devices should consider how the brain changes due to HL.

Historical Perspective on Unilateral Hearing Loss in Children
Anne Marie Tharpe

Until the mid 80s, audiologists and otolaryngologists generally had little concern for UHL in children. An early study demonstrated that medical and educational histories for 60 children with UHL (6–18 years) revealed that for more than 70%, the HL was not identified until age 5 years or later (Bess and Tharpe, 1986). Almost half of these children had failed at least one grade in school and/or required educational resource help for at least one year. Subsequent studies throughout the U.S. and the world also demonstrated that a greater number of children with UHL had educational difficulties than their NH peers (Oyler et al., 1988, Jensen et al., 1989, Bovo et al., 1988). A study of 25 children with UHL and age-matched NH children found children with UHL had poorer sound localization and speech recognition abilities and were perceived by teachers to have increased behavioral problems (e.g., withdrawal, inattention) than their normal hearing peers. But, the groups had fewer differences on standardized speech/language tests (Klee and Davis-Dansky, 1986). More recent studies showed that children with UHL have poorer language scores than NH siblings (Lieu et al., 2010) and increased educational difficulties compared to NH peers (Kesser et al., 2013). However; little is known about the reasons some UHL children have significant academic difficulties whereas others do not. Potential contributing factors include variability in listening conditions and required listening effort, lack of early or aggressive intervention, concomitant otitis media, or etiology. Additional study is required to understand the variability in academic performance observed among children with UHL.

Single-Sided Deafness - Challenges and Outcomes with Current Technology
John K. Niparko

The binaural hearing mechanisms of head shadow, squelch and diotic summation that are necessary for sound localization/spatial hearing and unmasking of speech in noise are not available for SSD individuals. Various contralateral sound routing approaches have been used including contralateral routing of signal (CROS) amplification, deep canal hearing aids, Audiant implantable hearing aids, and bone anchored hearing aids (BAHA). Ten SSD adults (mean age 45 years, mean SSD duration 5 years) underwent a CROS hearing aid trial followed by a BAHA (Niparko et al., 2003). Perceived benefit was greater with the BAHA than the CROS, but neither provided directional hearing. Sentence understanding in noise findings were mixed and depended on noise source direction. A review of nine published articles with SSD individuals who received a CI (144 patients total) suggests 80–100% of patients achieve improved soundfield expansion, speech understanding and sound localization with three articles reporting approximately 2 dB benefit from squelch. Another role of cochlear implantation in SSD patients is tinnitus suppression. Data pooled across nine articles (n = 89) found tinnitus resolved for 19%, improved for 74% and showed no change for 7% of patients (Arts et al., 2012). In summary, published articles that have evaluated the impact of intervention and SSD with BAHA (15 articles) and CIs (9 articles) indicate that head shadow is effectively managed by both technologies, summation is somewhat more pronounced with a CI, and localization does not occur with BAHA but is present for high frequency inputs with a CI. Squelch effects are minimal with a CI and absent with a BAHA. Tinnitus suppression and overall subjective impressions are positive with both interventions, although auditory benefits with SSD appear to be more robust with cochlear implantation based on the literature to date.

Effects of Unilateral Hearing Loss in Adults and Children: Implications for CI Candidacy

Jill Firszt

Many adults and children have asymmetry in hearing between ears, the extreme of which is UHL where one ear hears in the normal range and the other ear has severe to profound hearing loss. This population has been somewhat underserved, due to limited understanding of UHL consequences and lack of treatment guidelines. A recent study in adults compared those with UHL to those with NH who were listening either with one or both ears. Among measures of localization and speech recognition in quiet and noise, a notable finding was the great variability seen for unilateral listeners. In children, while UHL is known to affect academic performance and language outcomes, the specific effects on localization and speech understanding in noise remain understudied. Recent experimental work in children demonstrated, as expected, poorer abilities of children with UHL compared to those with NH bilaterally. In addition, the variability in performance among the children with UHL was pronounced. Reasons for this variability in both adults and children are currently unknown and being investigated. Although a small number of individuals have received a CI in the poorer ear to treat UHL, there is minimal information about best treatment options or CI candidacy guidelines for people with UHL.

Results in Patients with Single-Sided Deafness and Cochlear Implants

Bruce Gantz

Twenty-nine patients at University of Iowa have undergone CI for SSD, with relatively short length of deafness. Twelve participants had simultaneous labrinthectomy and CI for treatment of Meniere's disease. Data from 19 participants with post-operative scores suggested varied individual results for CI-alone speech recognition in quiet. A more consistent trend was observed for sound localization where most participants demonstrated greater accuracy with the CI and NH combined than in the NH-alone condition. A subset of participants was tested with sentences presented from the front and noise from the front, the better hearing ear side, and the CI side. Results suggested summation and head shadow effects for understanding sentences in noise at improved signal-to-noise ratios. Squelch effects benefits were observed for some participants. All participants reported CI satisfaction and, except for one individual, used their device during all waking hours. Patients with Meniere's disease were relieved of vertigo while gaining useful hearing.

Validation of Acoustic Models of Auditory Neural Prostheses

Mario Svirsky

Acoustic models have been used in numerous studies to simulate percepts elicited by auditory neural prostheses. SSD patients allow direct comparison of these models to actual CI stimulation. Although signal processing is very similar in CI and acoustic models (e.g. lack of fine structure, limited number of channels), NH speech perception using an acoustic model (simulated CI) is better than average CI speech perception (Dorman et al., 1998, Dorman and Loizou, 1998, Friesen et al., 2001). CI pitch perception results can also be very different than using acoustic models (Laneau et al., 2006). Initial work with six SSD CI recipients compared varied acoustic models with participants' CI speech processors. Sentences were presented via acoustic models (noise band and acoustic tone) to the NH ear and via a speech processor to the CI ear. Acoustic models had the same channel number and frequency filters as the participants' CI. Participants selected the model that sounded most similar to the CI. For the majority of participants, the self selected model was a better match (based on questionnaire and word identification results) to the CI than the standard acoustic model used in many published studies. In general, the standard models were not accurate: they sounded better and allowed better speech perception than the CI. SSD patients who receive a CI provide the ability to compare various acoustic models to electrical stimulation and to offer insight about what CIs really sound like, as well as about the way human listeners adapt to distorted auditory input.

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Cochlear Implantation in Older Adults

Co-Chairs: Frank Lin[1], William Shapiro[2]
Presenters: Frank Lin[1], Larry Humes[3], Jace Wolfe[4], Linda Daniel[5]
[1]Johns Hopkins University, [2]New York University, [3]Indiana University, [4]Hearts for Hearing, [5]Dallas Ear Institute

Current population estimates in the United States indicate that nearly 1% of all adults older than 70 years would likely meet audiologic criteria for cochlear implantation but less than 1 in 20 of these individuals have a cochlear implant. While the reasons for this low rate of uptake are multifactorial, the number of older adults with severe-to-profound hearing loss will continue to rapidly increase over the next decade with the aging of the current generation of baby boomers. Increasingly, we are also understanding the potential public health implications of age-related hearing loss and its effects on cognitive, social, and physical functioning. What are the challenges that face our field of cochlear implantation in older adults with respect to audiologic, rehabilitative, and surgical issues? How can we understand and study the impact that cochlear implantation has on the functioning of older adults, particularly in the context of establishing the health economic basis for the interventions and services that we provide? What steps do we need to take to refine our approach from simply fitting a cochlear implant to ensuring that the older patient can effectively communicate in all settings? This session brings together a group of speakers and panelists to discuss these critical issues and develop recommendations for clinical, research, policy, and industry initiatives going forth for cochlear implantation in older adults.

Hearing Loss in Older Adults – Implications for Healthy Aging

Frank Lin

Age-related hearing loss (ARHL) in older adults is often perceived as being a relatively inconsequential part of aging, but this perception is not rooted in evidence. There has been little research exploring whether ARHL may be independently associated with outcomes critical to successful aging such as maintaining cognitive and physical functioning. A conceptual model through which ARHL could be mechanistically associated with these downstream outcomes hypothesizes that non-mutually exclusive pathways of increased cognitive load, alterations in brain structure, and loss of social engagement could mediate effects of ARHL on cognitive and physical functioning (Lin 2012). Recent epidemiologic studies supporting independent associations of hearing impairment with cognitive functioning (Gallacher et al. 2012; Lin et al. 2011; Lin et al. 2013), physical functioning (Viljanen et al. 2009a; Viljanen et al. 2009b), health economic outcomes (Saps et al. 2013), and mortality (Karpa et al. 2010) are consistent with such a model. Hearing loss is highly prevalent in older adults with nearly two-thirds of older adults 70 years and older having clinically-significant hearing loss but with less than 20% receiving any form of rehabilitative treatment. Broader public health initiatives are needed to inform the public and medical providers of the importance of ARHL and how to optimally address hearing loss through a combination of concerted counseling, awareness, and proper fitting of hearing aids and other rehabilitative devices. Further research investigating the mechanistic basis of these associations and whether hearing rehabilitative therapies could affect these outcomes are critically needed.

Speech Communication and Higher Level Processing in Older Adults

L. Humes

It is broadly acknowledged that many older adults have difficulty understanding speech in a background of noise. This difficulty is most often worse when the background signal is other competing speech. There is evidence that many older adults have peripheral hearing loss, primarily impacting the cochlea or spiral ganglion. In addition, some older adults may experience age-related cognitive declines, especially for processing-sensitive measures, such as working memory, speed of processing, inhibition of distracting information, and executive function. Although the evidence in humans is not compelling to date, there is evidence in laboratory animals of higher-level auditory deficits as well, due either to aging alone or to the presence of the high-frequency sensorineural hearing loss common in many older adults. Humes & Dubno (2010) recently reviewed the evidence supporting the peripheral, central-auditory, and cognitive hypotheses noted above. Generally, the bulk of the evidence primarily supports peripheral factors underlying individual differences in speech perception for unaided listening at conversational levels and in relatively simple backgrounds (quiet, steady-state noise). When amplified speech is considered, however, the

peripheral hearing loss plays a relatively minor role and higher-level auditory and cognitive processing factors take on increasing importance. This is especially evident in backgrounds employing speech or speech-like competition. This was affirmed most recently in a study of 98 older adults from our laboratory (Humes et al. 2013).

The Use of Hearing Assistance Technology to Optimize Performance and Benefit of Elderly Cochlear Implant Recipients

Jace Wolfe

Although older adult cochlear implant recipients frequently understand speech very well in quiet environments many continue to experience great difficulty understanding speech in challenging listening environments e.g., background noise, television, telephone, etc. (Gifford et al. 2008). The use of hearing assistance technology is likely the most effective means to improve speech recognition in difficult listening situations. Wireless remote microphone hearing assistance technology systems are comprised of a transmitter that is coupled to a microphone and a receiver that is coupled to the recipient's cochlear implant sound processor. Research has shown that use of remote microphone technology typically improves speech understanding by as much as 50 percentage points at challenging signal-to-noise ratios (e.g., 5 dB.) (Schafer & Thibodeau 2004). Wireless remote microphone technology has evolved significantly over the past ten years. Research has also examined performance obtained with remote microphone systems using both directly-connected radio receivers (via direct auditory input) and radio receivers that are coupled to induction neckloops which convey the signal of interest to the sound processor telecoil via magnetic induction. This research has indicated that directly-connected systems provide better speech understanding when compared to performance with induction neckloop systems (Wolfe et al. 2013). Over the past few years, manufacturers have developed hearing assistance technologies that utilize digital adaptive radio transmission, Bluetooth, short-range digital magnetic induction, and proprietary digital streaming. More research is needed to understand the benefits and limitations of each of these systems as well as to identify clinical strategies to appropriately select hearing assistance technology that best meets the needs of an individual adult recipient.

Cochlear Implantation of Older Adults: Role of the Aural Rehabilitation Professional in Patient Selection and Follow-up Care

Linda Daniel

The Dallas Ear Institute, in conjunction with HEAR In Dallas, uses a team approach on their cochlear implant program. The neurotologist, audiologist, neuropsychologist, radiologist and aural rehabilitationist (rehabilitative audiologist or a speech-language pathologist) evaluate every cochlear implant candidate to bring potential issues to the physician's attention. The aural rehabilitation professional evaluates the patient's reasons for and accuracy of knowledge about cochlear implants, communication needs, support system and ability to do independent home practice. Post-activation rehabilitation sessions are scheduled on programming days. Family members are included to assist the recipient in managing the technology and practicing listening via the individualized home program. Sessions primarily focus on education and counseling: post-linguistically deafened patients typically need little formal listening training. If direct listening therapy is needed, a top-down approach is used, moving from auditory-plus-visual (hearing, speech reading, print) tasks to auditory-only tasks. This gives the individual a feeling of success when adjusting to the signal. Advanced listening is practiced in noise, at a distance and over the telephone. The aural rehabilitation professional provides feedback regarding patient performance to team members as needed. (S)he notifies the programming audiologist of patient response to MAP modifications and relays patient concerns. The neuropsychologist or physician is contacted if related issues arise. The aural rehabilitation professional plays an important function in the evaluation and treatment of older CI recipients. (S)he provides direct therapy and education to the patient and significant others and coordinators patient needs with other team members. A well-coordinated team contributes to patient satisfaction.

Conclusion: The aging of the U.S. population and a better understanding of the impact of hearing impairment on the cognitive and physical functioning of older adults necessitates comprehensive approaches to addressing age-related hearing impairment at the public health level as well as at the individual level. Multidisciplinary approaches for care of individuals with severe hearing impairment requires an understanding of how different factors (cochlear implant programming, rehabilitation, use of assistive device, intrinsic cognitive substrates for language use) contribute to the ability of an individual to communicate successfully.

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Telehealth and Cochlear Implantation

Co-Chairs: K. Todd Houston[1]
Presenters: Gary Kahn[2], Michelle L. Hughes[3], Hannah K. Eskridge[4], K. Todd Houston[1]
[1]University of Akron, [2]University of Colorado/Denver, [3]Boys Town National Research Hospital, [4]University of North Carolina

As telecommunication and distance technology continue to evolve, new opportunities to provide diagnostic, treatment, and (re)habilitation services to meet the medical and communication needs of children and adults with cochlear implants are increasing. Physicians, speech-language pathologists, audiologists, and other service providers should be aware of these forms of service delivery – from telehealth and telemedicine to telepractice and teleintervention. Likewise, mobile health (mHealth) applications, especially through smartphones and tablet computers, are impacting the delivery of healthcare, including how practitioners interact with the patients they serve. Consumers continue to embrace telehealth, telepractice, and mHealth, and they are demanding that these service delivery models be available to them. This session addresses current applications of telehealth, telepractice, and mHealth to meet the diagnostic, treatment, and habilitation/rehabilitation of patients who use cochlear implants.

Telemedicine in Otology and Cochlear Implantation
Gary Kahn

Telemedicine, using information and communication technology to deliver healthcare services, is not new. Teleradiology, for example, has transformed traditional radiology practice by using digital information and communication technology to transmit and remotely interpret images–vastly improving both quality and efficiency. As the use and evolution of CI technology illustrates, Otololaryngology has long been an early adopter of technologies to improve patient outcomes, including telemedicine (Garritano & Goldenberg 2011). Because they practice in areas with large, underserved rural communities, Otolaryngologists from Alaska and Australia have been the pioneers in telemedicine. In a 2008 study, Kokesh and colleagues, established that telemedicine was reliable for ensuring tympanostomy tube placement and patency (Kokesh et al. 2008). An Australian study, of a otolaryngology telemedicine consult service demonstrated 99% agreement between telemedicine and face-to-face diagnoses, and 93% agreement in terms of the management plans Smith et al. 2008). Falling costs and improving technology are compelling practitioners and policymakers alike, to go beyond the focus on improving rural access, and realize the promise of telemedicine services to deliver higher quality, more efficient, and more cost-effective patient-centered care (American Telemedicine Association State Policy Toolkit). To accomplish this, the focus is shifting from overcoming technology barriers to overcoming regulatory (licensure, liability) and reimbursement barriers (H.R.3306: telehealth enhancement act of 2013). TACIT (Telehealth And Cochlear Implant Therapy) is a newly-funded, multi-center clinical trial being conducted by researchers at the University of Colorado comparing the traditional face-to-face approach with a telehealth approach for listening and spoken language therapy in children following CI. This research utilizes behavioral and neurophysiological outcomes to evaluate the differing service delivery models.

Validation of Audiological Measures via Telepractice for Cochlear Implants
Michelle L. Hughes

Cochlear implants (CIs) represent a specialty service within audiology, which means that CI clinics tend to be somewhat sparsely located. Further, numerous visits are required within the first year of implantation for programming, speech/language/listening sessions, and medical follow-up, with ongoing visits thereafter. Clearly these combined factors can create significant burdens to families in terms of travel expenses,

missed school or work, and lack of immediate access to clinical care. CI service delivery via remote technology (telepractice) is an attractive alternative to these obstacles. Initial studies have focused on comparing map levels for adult recipients for in-person versus remote programming (Hughes et al. 2012, McElveen et al. 2010, Ramos et al. 2009, Wesarg et al. 2010). Results show that map levels are not significantly different when obtained remotely versus traditionally. Similar results have been reported for other clinical measures such as electrode impedance and neurophysiological measures (Hughes et al. 2012). Speech perception, on the other hand, has been shown to be poorer in the remote condition for more realistic local test environments (i.e., no sound booth) (Hughes et al. 2012, Goehring et al. 2012). Alternative solutions such as acoustic room treatments, direct-connect testing, or perhaps development of correction factors for specific noise and reverberation levels need to be further explored and validated for remote speech-perception testing. For full implementation of telepractice in CIs to be successful, all aspects of patient care should be validated for remote service delivery. Other areas that remain to be validated include equipment troubleshooting, pediatric mapping, sound-field threshold testing, and aural (re)habilitation. In sum, telepractice is a viable option for CI service delivery; however, additional validation studies still are needed.

Praises and Pitfalls: Teletherapy for Children with Hearing Loss

Hannah K. Eskridge

Private therapists, schools and hospital clinics throughout the United States have begun to use teletherapy to provide services to children with hearing loss. The data to support or oppose this method of service delivery for children learning to listen and talk is unavailable nor have best practices for practitioners been established. The University of North Carolina Ear and Hearing Center, which includes the pediatric cochlear implant team and intervention program, CASTLE, has begun a pilot program to measure outcomes for children ages 0 to 3 receiving services through teleintervention. Research has documented pros and cons of teleintervention for other medical practices (Saab et al. 2004, Peddle 2007, McDuffie et al. 2013). Cons include access to technology and computer literacy, staff frustration with troubleshooting and technical difficulties. The UNC CASTLE staff also report they have concerns regarding parent and therapist learning style, incorporating goals in a naturalistic way, and managing behavior. Documented pros to this approach include increased access to more families, providers can reach more patients to reduce strain due to provider shortage and families can receive services in the naturalistic environment of their home (Saab et al. 2004, Peddle 2007, McDuffie et al. 2013) . The UNC CASTLE staff also report these pros along with the challenge professionally of learning something new and that this method is the truest form of 'coaching the parent to be the primary teacher' a principal of auditory-verbal therapy. Data presented includes six children enrolled in REACH (our teleintervention program) and 4 children receiving direct services as controls. With a year of data, all children are testing within normal limits on language using the Preschool Language Scales-5. Parent behavior was the primary outcome we wanted to track as we believe parents are their child's primary teacher. Parents in both the REACH group and the control group increased their confidence in understanding and addressing issues relating to their child's hearing loss. They report on average to be somewhere between confident and very confident. We also conducted a video analysis of parent skills over time. Targeted behaviors went from an average of rarely observed to emerging. In conclusion, early data is demonstrating the effectiveness of teaching children who are deaf or hard of hearing to learn to listen and talk through the use of teleintervention. Continued research needs to be conducted to include strategies and skills of the therapists and children with multiple disabilities.

Connecting to Communicate: Telehealth, mHealth, and Telepractice: Applications in Cochlear Implantation

K. Todd Houston

Physicians are providing diagnostic, treatment and patient counseling through models of mHealth, telehealth and telemedicine, and pediatric audiologists and speech-language pathologists are embracing telepractice to provide services to a range of patients who have hearing and communication-related delays and/or disorders (Houston 2014). Models of telepractice are being used to provide remote cochlear implant mapping/programming, often with excellent results for the patient. Likewise, teleintervention, a specific program of early intervention provided through distance technology, provides family-centered services to infants, toddlers, and young children with cochlear implants and allows the speech-language pathologist to model and coach parents in language facilitation techniques. For adults with cochlear implants, finding well-trained rehabilitation practitioners is often a challenge, and telepractice is viewed as a viable, efficient, and effective means to connect service providers to patients. As telemedicine, telepractice, and teleintervention become more common and integrated into standards of care, cochlear implant professionals – physicians,

mapping audiologists, and speech-language pathologists – will be tasked to incorporate these service delivery models into their programs.

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