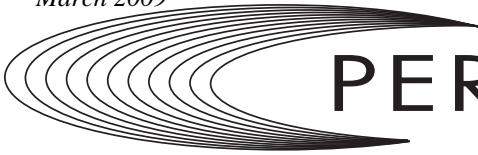


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AMERICAN SPEECH-LANGUAGE-HEARING ASSOCIATION **DIVISION 9**

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Coordinator's Column

Gayla Hutsell

Has the first quarter of 2009 really already passed? I guess time must really fly when we're having fun, right? In a similar vein, has it really been almost 20 years since the FDA approved multichannel cochlear implants for children? As a young audiologist, I was thrilled to hear stories on popular media about a "new and remarkable technology" that would allow people who are deaf to hear speech and environmental sounds. At the time, nobody was quite certain how much the cochlear implant would improve auditory, speech, language, and academic outcomes for children with profound hearing loss. Now, of course, we know from numerous research studies that many children who have severe to profound hearing loss and use cochlear implants achieve speech, language, and literacy outcomes that are similar to their peers with typical hearing.

Cochlear implant outcomes have been so successful and cochlear implant technology has improved so much that the past few years have seen an expansion in the number of children who are candidates for cochlear implantation. Children with greater amounts of residual hearing are receiving cochlear implants as are children with auditory neuropathy spectrum disorder (ANSO) hearing loss. Also, there is a trend toward children receiving cochlear implants at younger ages. These rapid changes in clinical practices and vast improvements in technology have left many professionals and families of children with hearing loss with questions about the cochlear implant and how it can be used most effectively to maximize outcomes for children with hearing loss. What components should be included in the evaluation process to determine whether a child should receive a cochlear implant? What is the earliest age at which we should consider providing a cochlear implant for a child? Will children with ANSO or multiple disabilities benefit from a cochlear implant? How should we program a cochlear implant and then provide therapy for the child? What about hair cell regeneration? How does it affect the decision to recommend an implant for a young child?

The next two issues of *Perspectives* seek to provide answers to these questions and others that challenge professionals who serve children who may benefit from a cochlear implant or those who already have one. This issue highlights contemporary evaluation and management protocols for pediatric cochlear implantation, and the September issue will feature several articles discussing pediatric cochlear implantation in special populations, contemporary rehabilitative approaches, and the potential impact of inner ear therapies on cochlear implant candidacy and outcomes. An absolute "who's who" of researchers and clinicians has shared expertise in manuscripts which provide cutting edge information for professionals who provide services for children with cochlear implants.

In this issue, Terry Zwolan and Ellen Thomas kick things off with an excellent article that thoroughly describes the comprehensive, interdisciplinary cochlear implant evaluation protocol used at the University of Michigan's Cochlear Implant Program. This article is a must-read for pediatric audiologists and speech-language pathologists alike.

Sticking with matters pertaining to the cochlear implant evaluation process, Suzanne Purdy and Kirsty Gardner-Berry discuss how auditory evoked potentials, and in particular the cortical auditory evoked potential, can be used to determine whether a

child should receive a cochlear implant. Their article is guaranteed to make you think about recommending cortical auditory evoked potential assessment for the children you see in your practice.

Oliver Adunka and Craig Buchman, both otologists at the University of North Carolina-Chapel Hill, describe the cutting edge protocol they use to ensure the absolute best medical care for children with cochlear implants or those who are being considered for cochlear implantation. Their article describes the fascinating work being done at UNC, which serves over 2000 children with hearing loss. All professionals involved in pediatric hearing health practice should be familiar with the various components that ensure the optimal well-being of the children whom they serve. This article aptly describes gold standard medical management of the child with hearing loss and includes information that you will likely want to share with the families and professionals with whom you work.

Division 9 Steering Committee (SC) member Tamala Bradham and her colleagues from Vanderbilt University have written a superb article describing in detail their entire management protocol from evaluation to intervention for children with cochlear implants. They put a lot of work into ensuring that their protocol is evidenced-based and reflective of the most current information, practices, and technology available today; they are very gracious in their willingness to share it with us. Whether you work in a large cochlear implant program or you are still waiting to serve your first child with a cochlear implant, you will definitely want to read this article.

Finally, as part of her dissertation project, Yell Inverso created the Nonlinguistic Sound Test (NLST) to evaluate cochlear implant recipients' ability to discriminate non-speech sounds such as animal, nature, and machinery sounds. Dr. Inverso points out that such a test is needed because although cochlear implant technology and evaluation tools have historically focused on the recognition of speech, many recipients desire to discriminate non-speech sounds as well. In her article, she discusses the development of the original NLST, which was intended for adults, and also a newly developed pediatric version of the NLST.

We hope you enjoy the excellent contributions of these fine authors. Please look forward to the September issue, and also please be certain to share your thoughts and suggestions with the Division 9 SC. Along with our Editor, Jace Wolfe, we are committed to providing the best possible information to our membership. We want to hear from you! With the way time flies, it's never too early to begin thinking about topics for our 2010 *Perspectives*. Cheers!

Contemporary Protocols for Evaluating Cochlear Implant Candidacy of Children

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Abstract

Candidacy criteria for determination of cochlear implantation for children have changed significantly since cochlear implants were first introduced. The contemporary evaluation process includes initial consultation, audiological assessment, speech and language assessment, preoperative counseling, selection of the ear to implant, medical evaluation, and additional assessments as needed. Ideally, an interdisciplinary team of professionals is involved in this evaluation process. The following article discusses procedures used in our clinic to determine a child's candidacy for a cochlear implant.

Introduction

In the United States, the Food and Drug Administration (FDA) oversees the selling, distribution, labeling, and marketing of drugs, medical devices, and other products and determines if the specific wording used in device labeling, such as information regarding indications for use, is appropriate. Cochlear implant systems provide information regarding FDA-approved indications for their use on labels that are provided in packaging of the internal array. Clinicians use the FDA-approved indications as a guide when determining if they should recommend a cochlear implant for a child. The Current Candidacy Criteria for Children describes indications for use of the Nucleus Freedom cochlear implant system (Cochlear Americas, 2009):

- Severe to profound bilateral sensorineural hearing loss (unaided thresholds should be greater than or equal to 90 dB HL beyond 1000 Hz). In younger children, the hearing loss should be confirmed using electrophysiologic measures such as the Auditory Brainstem Response (ABR) test.
- 12-18 months of age or older.
- Little or no benefit from hearing aids. In young children, this is demonstrated by lack of progress in the development of simple auditory skills in conjunction with appropriate amplification and participation in an intensive auditory habilitation program. This is also demonstrated by parental response to client-administered questionnaires such as the MAIS (Robbins, 1998) or the IT-MAIS (Zimmerman-Phillips, Osberger, & Robbins, 1998). In older children, minimal benefit from amplification is demonstrated by minimal scores on open-set speech recognition measures such as the Lexical Neighborhood Test (LNT) (Kirk, Pisoni, Osberger, 1995) or the Multisyllabic Neighborhood Test (MLNT) (Kirk, Pisoni, Osberger, 1995). A 3-6 month hearing aid trial is required for children with no previous hearing aid experience.

- No medical or radiological contraindications to surgery.
- Placement in an educational setting that is able and willing to provide a concentrated auditory skill development program.
- Motivated family and consent of the child if appropriate.

The FDA-approved criteria for cochlear implants in children have changed greatly since they were first introduced. A comparison of FDA-approved wording from 1990 (when multichannel cochlear implants were first FDA-approved for use in children) and today reveals a drop in the minimum age for implantation (12 versus 24 months), expansion to include children with severe-to-profound sensorineural hearing loss, and expansion of speech perception skills to include children who demonstrate “limited” benefit from appropriate binaural hearing aids (versus the previously indicated “little or no benefit”). Additionally, the types of tests used to evaluate candidacy have evolved to include measures of open-set speech recognition, indicating an expansion to include children with greater preoperative speech recognition.

In many ways, determination of implant candidacy is simpler today than in the past. This is because a great deal of data exists regarding average performance levels of children with cochlear implants. Although a child’s performance levels with an implant cannot be predicted preoperatively, such average CI performance data provides us with benchmarks that can guide us as we decide whether or not to recommend an implant. Available benchmarks include speech/language skills, speech perception skills, auditory detection, and academic performance, to name a few.

When recommending an implant for a child, it is important to keep in mind that such surgery often results in complete elimination of the child’s residual hearing in the implanted ear. Thus, if a recommendation to implant is made, one should be certain that the child’s hearing potential is greater with an implant than it would be if he or she continued to use a hearing aid. If the child demonstrates greater auditory potential with one ear over the other, then this should play in the decision of which ear to implant if a unilateral implant is recommended.

In the case of young children, we use audiometric, speech and language, and speech perception data to estimate the child’s potential for developing or continuing to develop, spoken language skills with sustained hearing aid use. If this potential looks poorer than one would expect from a cochlear implant, a recommendation to provide the child with a cochlear implant should be made. If the young child demonstrates a potential for success with hearing aids that is equal to or better than the results anticipated with a cochlear implant, a recommendation to not implant but to continually monitor and evaluate spoken language skills should be made. Monitoring is important because a recommendation to implant may be made at a later date, if appropriate progress has not been made by the child.

Thus, one of the most important steps an implant program can take to improve their ability to identify candidates for an implant is to continually monitor the post-operative progress of children who receive cochlear implants at their facility. Doing so will not only facilitate awareness of the progress that patients are making, but such data will also provide a benchmark against which the preoperative results of children who use hearing aids can be compared. Such monitoring should include both speech perception and speech/language results, as both factors contribute to spoken language. In this article, we will discuss the procedures used in our clinic to determine a child’s candidacy for a cochlear implant, keeping in mind that such determination is made on a case-by-case basis. This review will focus on the roles that the audiologist and speech-language pathologist (SLP) play, but will include a brief description of the role that other professionals may also play in the decision process.

Referrals to the Implant Program

Many professionals contact our clinic to inquire about the best time to refer a child for a cochlear implant evaluation. If the child has a severe-to-profound hearing loss that was identified soon after birth, we recommend he/she be scheduled for an implant evaluation soon after obtaining hearing aids. Ideally, such appointments take place by the time the child is three months of age. Such an early referral enables the professionals in the CI program to evaluate the child's progress with hearing aids over time and facilitates early implantation if it is determined that the child is a candidate.

With older children, a referral to the implant program should be made if the child has a severe-to-profound hearing loss or if the child experiences a drop in hearing that causes a plateau or decrease in speech perception or speech/language skills. In all instances, professionals and parents should be encouraged to contact the CI program to discuss the child's case if they are questioning the appropriateness of such a referral.

Initial Consultation, Case History, and Questionnaires

The implant evaluation process begins with an initial consultation. During the scheduling process, parents should be instructed to bring copies of all pertinent medical records to the appointment. Some clinics find it helpful to send the parents a copy of the case history questions ahead of time to help them prepare for the appointment.

In addition to case history questions, clinician-administered questionnaires can be used to determine how the child uses hearing in his/her daily life. Audiologists often administer the Infant-Toddler Meaningful Auditory Integration Scale (IT-MAIS) (Zimmerman-Phillips et al., 1998) to parents of children who are less than 2 years of age and administer the Meaningful Auditory Integration Scale (MAIS) (Robbins, Renshaw, & Berry, 1991) to parents of children who are older than 2 years of age. Both scales rate parental responses to questions regarding the child's meaningful use of sound in everyday situations.

Similarly, SLPs often administer questionnaires to evaluate the milestones of auditory function and spoken language that occur prior to the child's first production of words. The following inventories, which rely on parent report and clinical observation, are used with children approximately 3 years of age and less: The Rossetti Infant and Toddler Language Scale (Rossetti, 2005) and The Cottage Acquisition Scales of Listening, Language, and Speech (Wilkes, 1999). Additionally, parents may be asked to complete the MacArthur-Bates Communicative Development Inventory: Words and Gestures (MacArthur-Bates, 2007). This inventory provides a baseline for vocabulary development. In our clinic, we ask parents to first complete the inventory for words understood using spoken language alone. If the family uses sign language with the child, we ask them to also indicate the words the child understands when signed.

Preoperative Counseling

Counseling, an essential part of the preoperative process, may include topics such as device options, cochlear implant technology, candidacy requirements, expectations of performance, appointments involved in the evaluation process, and financial obligations. If the family is unaware or uninformed about the communication options available for their child, we provide them with resources such as the publication, *Opening Doors: Technology and Communication Options for Children with Hearing Loss*, developed by The U.S. Department of Education (2006), the book *Choices in Deafness* by Sue Schwartz (2007), and a referral to the Beginnings Web site (<http://www.ncbegin.org/>). Lastly, we often make arrangements for parents to meet with parents of a pediatric implant recipient to discuss the implant evaluation process.

Audiological Testing

Audiological testing is performed to evaluate the type and severity of the child's hearing loss. Objective Measures used in the implant evaluation process can include Immittance testing, Auditory Brainstem Response (ABR), Otoacoustic Emissions (OAEs), and Electric Auditory Brainstem Response (EABR). Auditory Brainstem Response (ABR) testing can be used to supplement and verify threshold information obtained with behavioral testing. Additionally, the results of ABR testing, when used in combination with otoacoustic emissions, can be used to diagnose auditory neuropathy/dysynchrony (AN/D). In cases of AN/D, the child will demonstrate a negligible or abnormal ABR combined with a normal or partially normal OAE reading. A diagnosis of AN/D will have a great influence on the decision regarding candidacy, particularly if the child demonstrates poor speech perception skills and a speech/language delay. Some clinics utilize the Electric Auditory Brainstem Response (EABR) test when there are concerns regarding the absence/presence of an VIIIth nerve or regarding electric stimulability of the ear to be implanted (Kileny et al., 1994) and will only proceed with cochlear implantation if a positive EABR is obtained.

Behavioral Testing

As part of the cochlear implant evaluation process, a complete audiometric test battery should be performed on each ear and should include acoustic immittance testing, unaided bone conduction thresholds, unaided air conduction thresholds, speech discrimination, speech reception threshold (if possible), and speech detection threshold. It often takes more than one appointment to obtain all of the above-mentioned information for a young child.

Aided testing should also be performed for each ear individually and in a binaural condition, and should include aided soundfield thresholds 250-4000 Hz, aided SRT (if possible), aided SDT, and aided speech perception testing. Test results should be evaluated to determine if the child receives appropriate and optimal gain from the hearing aids. If he/she does not, appropriate hearing aids should be fit to the child and the child's hearing should be re-evaluated. Thresholds obtained with the hearing aid should be compared to thresholds traditionally received with a cochlear implant (10-40 dB HL across the frequency range of 250-4000 Hz) in order to determine if the implant will facilitate improved detection +for the child.

Speech Perception Testing

The ability of the child to perceive speech when using hearing alone is an important factor that needs to be considered when determining candidacy for a cochlear implant. The specific tests used to evaluate speech perception vary among clinics, and also vary as a function of the patient's age and linguistic level. When administering speech perception tests, patients should be seated in a sound treated room that contains minimal visual and auditory distractions. The presentation level should be calculated using a calibration microphone placed at the center of the listener's head. Test materials should be presented a single time only and feedback should not be provided. The recommended presentation level for test stimuli is 60 dB SPL (Firszt et al., 2004). Although most clinics administer speech perception tests in quiet, many clinics also administer test materials in the presence of background noise or at various presentation levels (i.e., 50 dB SPL) as doing so can provide information regarding how the child perceives speech in less than optimal listening conditions. A greatly reduced score in difficult listening conditions may expedite the decision to implant.

Speech perception materials used to evaluate candidacy can be broken down into two categories based on the type and number of choices provided to the listener. First, closed-set materials provide the listener with a set of choices from which to select

a response. The difficulty of the closed-set task and its chance level are determined by the number of choices available to the listener. For example, a test item containing two possible responses will have a 50% chance of being correct and is easier than tests containing four possible choices and a chance score of 25%. Secondly, open-set materials provide the listener with a completely “open” set as no choices are provided. Therefore, chance scores for open-set tests are 0%. Both open-and closed-set tests provide valuable information that can help professionals determine candidacy for a cochlear implant.

Children Younger Than 2 Years

Very few formalized speech perception tests are available for use with very young children due to a child’s reduced language skills. In many cases, candidacy for an implant is based primarily on the results of objective and behavioral audiometric tests combined with the results of the questionnaires and surveys administered during the case history. Some children in this age group can participate in the Ling Six Sound Test (Ling, 1976; 1989). This is an informal test that determines if the child can detect sounds that lie within the speech spectrum of hearing. The test is usually performed live voice with the presenter asking the child to detect or identify the following six sounds: /m/, /oo/, /ah/, /ee/, /sh/, and /s/. The level of difficulty can be varied depending on the skills of the child and can include detection, discrimination, identification, or comprehension. Children who are candidates for an implant will demonstrate a variety of responses on this test, ranging from good to poor detection of speech sounds.

Additionally, the low verbal version of the Early Speech Perception Test (ESP) (Moog & Geers, 1990), can sometimes be used with very young children as it was designed for assessing young children with limited verbal ability. It is easier than the standard version of the ESP as it provides the child with stimuli in smaller sized closed sets (4 choices versus 12 in the standard version) and utilizes objects instead of pictures. The subtests evaluate the child’s ability to identify words that vary in number of syllables or stress patterns (level 1), to discriminate between various spondaic words (level 2), and to discriminate monosyllabic words (level 3).

Children Ages 2 Years and Older

Most formal speech perception tests are designed for use with children ages 5 years and above. Children who are 2 to 5 years of age can sometimes participate in formal speech perception testing. If they are not able to participate in formalized testing, the procedures described above for younger children are used and candidacy is primarily based on the results of objective and behavioral audiometric testing and parental questionnaires.

Numerous open-set tests are available to evaluate the speech perception skills of older children. The open-set tests used most often include recorded versions of the Multisyllabic Lexical Neighborhood Test (MLNT) (Kirk, Pisoni, & Osberger, 1995) and the Lexical Neighborhood Test (LNT) (Kirk, Pisoni, & Osberger, 1995). These tests are cited in the FDA-approved indications for use of contemporary cochlear implant systems (see Table 1). Other open-set tests utilized by clinics include the PBK-50 word list (Haskins, 1949), BKB Sentences (Bench, Kowal, & Bamford, 1979), and GASP Words and Sentences (Erber, 1982). In general, a cochlear implant will be recommended if the child demonstrates minimal open-set speech skills, and if the clinician feels that speech perception skills will improve with a cochlear implant.

It should be noted that caution must be exercised when open-set tests are administered to a child with poor speech production skills. Closed-set tests are useful with such children as they can provide information regarding the child’s ability to perceive speech without being influenced by the child’s poor speech intelligibility. Closed-set tests require the child to point to a picture rather than to repeat a word.

Closed -set tests used to evaluate candidacy include the Early Speech Perception Test (ESP; Moog & Geers, 1990), the Northwestern University Children's Perception of Speech (NU-CHIPS) test (Elliott & Katz, 1980), and the Word Intelligibility by Picture Identification test (WIPI, Ross & Lerman, 1979). The results of some closed-set tests, such as the Minimal Pairs test (Robbins et al., 1988), can be examined to determine specific features of speech that are or are not being perceived by the child.

Clinicians may want to consider administering both open-and closed-set tests to evaluate lipreading skills and lipreading enhancement. Evaluation of a lipreading-alone score will provide the clinician with information regarding the child's lipreading skills, and subtraction of a score obtained when using lipreading-alone from lipreading plus amplification will provide an indication of lipreading enhancement that can be attributed to use of hearing aids.

Speech and Language Evaluation

The speech/language evaluation is an essential part of the evaluation process for determining candidacy for a cochlear implant for children because development of spoken language is greatly dependent on hearing. This evaluation helps determine if speech/language development is delayed due to poor audibility or clarity of speech when using hearing aids. If such skills are delayed, and if it is felt that the ability of the child to develop spoken language will improve with the increased audibility of speech that the implant will provide, an implant will be recommended for the child. If, however, the child is making adequate progress with spoken language with hearing aids, a cochlear implant will not be recommended.

The speech/language evaluation should always be preceded by an appointment with the audiologist to determine if the child is using appropriately fit hearing aids and to determine if the aids are in proper working order. In our clinic, the SLP begins each evaluation and each therapy session by performing a listening check of the hearing aid or implant followed by the Ling 6 sound test (Ling, 1976; 1989) to evaluate the audibility of sounds across the speech spectrum.

The specific procedures used to evaluate speech and language skills will be determined by the chronologic and linguistic age of the child. Many milestones occur in auditory function and spoken language development ahead of a child's first words. Between the ages of birth and 3 months, infants typically learn to discriminate speech from other types of sounds, and most important of all, to recognize their mother's voice. Infants and toddlers with profound sensorineural hearing loss, however, are often not able to achieve these early listening milestones. This is often documented by the results of inventories that rely on clinical observation and parent report when assessing speech and language in a child under the age of 3 years. As stated previously, the Rossetti Infant and Toddler Language Scale (Rosetti, 2005), the Cottage Acquisition Scales of Listening, Language, and Speech (Wilkes, 1999) and the MacArthur-Bates Communicative Inventory (MacArthur-Bates, 2007) are tools that we frequently use with children who are 3 years of age or younger.

Children Age 3 Years and Older

For children in the 3-6 age range, our clinic also uses the Clinical Evaluation for Language Fundamentals Preschool 2-Revised (Wiig, Secord, & Semel, 2004). Areas difficult for individuals with less than optimal hearing to acquire, such as sentence structure, word structure, and following directions, are assessed by this measure. It is not uncommon for our preoperative patients to be unable to complete any of the items on this test. This test is particularly useful with children with Auditory Neuropathy/ Dysynchrony as they often have relatively good detection of sound, demonstrate good speech at the single word level, but often show poor performance on measures that look at sentence-level auditory comprehension and word structure.

With children ages 6 years and above, auditory comprehension is assessed using various subtests of the Woodcock-Johnson Tests of Achievement (Woodcock, McGrew, & Mather, 2001). Subtest #4, "Understanding Directions", assesses the child's ability to follow increasingly complex oral directions. Subtest #15, "Oral Expression", evaluates auditory processing by asking the child to complete a fill-in-the blank statement. Subtest #9, "Passage Comprehension", is administered to obtain information regarding the child's literacy level. Preoperatively, most patients are unable to achieve more than 1 or 2 items correct on each subtest. A child with a progressive hearing loss may show better performance on the passage comprehension subtest than on the listening comprehension subtests, indicating difficulty processing spoken language.

Vocal control is often difficult for individuals who are unable to monitor their speech via an auditory-feedback loop. Thus, vocal control should be part of the speech/language evaluation. The structure of the oral mechanism is evaluated and noted, and parents are asked about the child's voice quality, including questions about ability to monitor pitch and volume. When a child has sufficient speech to supply or imitate words, the Arizona Test of Articulation 3 (Fudala, 2000) is administered and results are evaluated using Long, Fey, and Channell's computer profiling program "CPROPH" (Long et al., 2004). This program can be used with any articulation measure, and provides a percentage of vowels and consonants produced correctly.

Finally, standardized tests, such as the Expressive Vocabulary Test (EVT) -2 (Williams, 2007) and the Peabody Picture Vocabulary Test (PPVT-4; Dunn & Dunn, 2007) are administered to evaluate receptive and expressive vocabulary. Both tests are administered via spoken language and a standard score is obtained. If the child uses sign language and is not able to name pictures using spoken language, signed responses to stimulus pictures will be noted. These responses do not yield a standard score but do provide insight into the child's language skills.

Other Professionals

Other professionals play a role in determining candidacy for a cochlear implant. Importantly, the implant surgeon obtains a complete medical history and determines if the child is healthy enough to participate in surgery. The surgeon will attempt to determine the cause of the hearing loss if it is not already known and will determine if treatment options other than a cochlear implant are more suitable for the child. The physician will request preoperative imaging, such as computed tomography (CT) or magnetic resonance imaging (MRI) of the temporal bone to visualize development of the mastoid and inner ear structures in order to determine if there are any additional findings that may complicate the surgery or subsequent patient management (Fishman & Holliday, 2006) and may influence the decision regarding which ear to implant.

An evaluation of the child's cognitive status should be performed by a psychologist if the audiologist or SLP is concerned that factors other than hearing impairment are hindering the child's auditory development. Such an evaluation should include non-verbal assessment of cognitive, social, emotional, and adaptive abilities. Depending on the patient's age and the presenting concerns, the ability of the child to attend to and remember information may also be assessed and recommendations may be made regarding educational services. The presence of a cognitive impairment may impact the child's ability to develop spoken language and should be taken into consideration when making a decision regarding implant candidacy. The input of the psychologist is essential when determining if referrals to other professionals are necessary prior to and after the child receives an implant. Preoperatively, identification of a cognitive impairment will influence the counseling provided to parents regarding the expected outcomes for their child. Additionally, parents of very young children should be informed that some psychological deficits (e.g., autism) are not typically

identified until the child is two years of age or older, and that performance with the device may be hindered if a cognitive impairment is identified as the child ages.

When handling older children, many clinics communicate with the child's school as part of their evaluation. School personnel, such as teachers, teacher consultants of the hearing impaired, speech pathologists, and education audiologists can provide important information regarding the child's use of hearing aids and assistive devices in the classroom along with information regarding his/her compliance with device use. Such professionals can also provide feedback regarding the impact the hearing loss has on the child's social and academic skills.

Determination of Which Ear to Implant

The determination of which ear to implant can be influenced by several factors. Many clinics routinely implant the ear with the least amount of residual hearing, whereas other clinics prefer to implant the patient's "best" hearing ear. In our clinic, the "poorer ear" is selected for implantation if we feel that continued use of a hearing aid in the child's better ear will result in better performance than use of an implant alone, or use of an implant in combination with the use of a hearing aid in the poorer ear. Some clinics leave the decision of which ear to implant up to the patient and/or the parents. Research indicates there are no known factors that help professionals predict which ear will respond best to the implant.

Recently, many clinics have begun to offer bilateral implants, eliminating the need to make a decision about which ear to implant. A cochlear implant can be placed in each ear during a single surgery (simultaneous bilateral implantation) or during separate surgeries (sequential bilateral implantation) that take place weeks, months, or even years apart. Recent studies indicate numerous benefits of bilateral implantation, including benefits for speech perception resulting from overcoming the head shadow effect, improved speech understanding in noise, and improved sound localization (Patrick, Busby, & Gibson, 2006). One of the benefits of bilateral simultaneous implantation is that such a strategy ensures that the better ear is always implanted.

Summary of Preoperative Testing

In summary, the preoperative evaluation helps us address several questions that help us determine if a recommendation for a cochlear implant should be made. Comparison of audiological test results obtained with a hearing aid to those obtained by existing patients with cochlear implants helps determine if provision of an implant will improve the child's auditory detection skills including improving his/her ability to detect speech. A more difficult question that needs to be addressed is, "Do we think that provision of a cochlear implant will improve the patient's ability to develop spoken language when compared to skills obtained with a hearing aid?" Speech perception tests provide information regarding features of speech that are and are not being perceived by the child, and the results of speech and language tests determine the impact that the child's hearing loss has had on development of spoken language thus far. Input received from other professionals, such as that of the implant surgeon, psychologist, and school personnel, all contribute to this important decision. Because the primary goal of the implant is to improve the child's communication skills, it is important to monitor the performance of children who have received this technology; such monitoring helps us learn from our decisions and helps lay the groundwork for future decisions as the candidacy for cochlear implants continues to evolve and change.

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Auditory Evoked Potentials and Cochlear Implants: Research Findings and Clinical Applications in Children

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Abstract

Auditory evoked potentials can be used to objectively assess hearing sensitivity, central auditory processing, and neural encoding of speech sounds up to the level of the auditory cortex. Evoked potentials have been of interest to clinicians and researchers in the cochlear implant field for a long time because of their potential for objectively predicting cochlear implant outcomes, as well as improving candidacy determination, and implant programming. Neural response telemetry and intra-operative electrical auditory brainstem recording have been routinely performed by implant programs for many years. Recently, there has been great interest in potential clinical applications of cortical auditory evoked potentials in the implant field. Research and clinical applications are reviewed and case studies are presented that illustrate clinical applications of cortical evoked potentials in children before and after implantation.

Background

The advent of universal newborn hearing screening and the need for early effective intervention for young children with hearing loss has led clinicians and researchers to become increasingly interested in the use of objective methods to evaluate hearing aids, determine cochlear implant (CI) candidacy, and optimize CI settings. Auditory evoked potentials (AEPs) are a useful objective measure because they can be recorded using instrumentation that is available in many audiology clinics and they can provide an objective indicator of both auditory detection and discrimination. In the following sections the literature on the use of AEPs to assess CI candidacy and outcomes is reviewed. The focus is on the auditory brainstem response (ABR) and cortical auditory evoked potentials (CAEPs).

Auditory Evoked Potentials

AEPs are usually categorized based on their time course or latency, but can also be separated into obligatory AEPs, which depend primarily on the characteristics of the stimulus, versus discriminative, which result from a change in stimulus characteristics. Obligatory AEPs include the auditory brainstem response (ABR), the electrocochleogram (ECoG), the middle latency response (MLR), and cortical auditory evoked potentials (CAEP). Obligatory CAEPs are evoked by delivering a series of auditory stimuli (clicks, tonebursts, or speech sounds) while the person listens passively. Discriminative CAEPs are recorded in response to a different (deviant/oddball) stimulus in the midst of a train

of standard acoustic stimuli or in response to a change within an acoustic stimulus. Discriminative potentials include the mismatch negativity (MMN) and P3 (Martin, Tremblay, & Korczak, 2008), recorded during passive and active listening, respectively. The acoustic change complex (ACA) is another discriminative cortical potential that occurs in response to a change in an ongoing sound (Ostroff, Martin, Boothroyd, 1998; Tremblay, Kalstein, Billings, & Souza, 2006).

Because sleep state affects cortical activity, CAEPs are not reliably present during sleep (Cody, Klass, & Bickford, 1967). Whereas ABR and auditory steady state response (ASSR) evoked potential testing is usually performed during sleep, CAEPs are recorded while the listener is awake. Adults and older children would typically watch a silent subtitled video during CAEP recording whereas young infants are distracted using age-appropriate toys and books (Purdy, Katsch, Dillon, Storey, Sharma, & Agung, 2005). CAEPs can be recorded at near-threshold levels (Davis, 1965; Tsui, Wong, & Wong, 2002); however, the evoked potential of choice for estimating hearing sensitivity in infants would usually be the ABR or ASSR. Currently, CAEPs are primarily used for objective assessment of central auditory function/neural encoding of speech sounds (Martin et al., 2008); for these applications stimuli are typically presented at suprathreshold intensity levels.

Auditory Brainstem Response

In infants who are too young for reliable behavioral audiometry, the toneburst ABR (or ASSR) audiogram is the main indicator of hearing sensitivity. Accurate prediction of audiometric thresholds in order to prescribe appropriate amplification is a very important first step in determining CI candidacy. Stapells (2000, 2002) reviewed the literature on toneburst ABR and provided recommended frequency-specific correction factors to apply to the toneburst ABR in order to predict pure tone audiometric thresholds. This information can be accessed by following the Clinical ABR/ASSR Tips link on Stapells's Human Auditory Physiology Lab Web site (<http://www.audiospeech.ubc.ca/haplab/>).

The click-evoked ABR is another important measure when determining CI candidacy because of its important role in the diagnosis of auditory neuropathy spectrum disorder (ANSD). The term ANSD has been adopted recently and replaces auditory neuropathy/dys-synchrony and the variations on this term used previously (Roush, 2008). Children with ANSD have an abnormal (delayed latency, abnormal morphology) or absent ABR, absent acoustic reflexes, and present cochlear microphonic (King, Purdy, Dillon, Sharma, & Pearce, 2005). Otoacoustic emissions are present in about a half of ANSD cases (Rance et al., 1999). Children with ANSD are of particular interest when considering CI candidacy because speech perception outcomes with hearing aids are generally poorer (but not always), and are more difficult to predict than for children with sensorineural hearing loss (SNHL) (Rance & Barker, 2008).

Pre-implant prediction of CI outcomes and objective measurement of progress with a CI are clearly of interest to both clinicians and researchers. The electrically-evoked ABR (EABR) has been used for this purpose in several studies that have shown correlations between EABR characteristics and speech scores in groups of adult CI users (Abbas & Brown, 1991; Gallégo et al., 1998; Maurer et al., 2002). Gordon and her colleagues (2006, 2007) found that children's EABR latencies improved with CI experience. Thus, monitoring EABR latencies could be a clinically useful objective method for determining auditory plasticity in children with CIs who are too young or who have other disabilities that make it difficult to measure speech perception outcomes. There have been attempts to use EABR thresholds for objective setting of CI current levels, however, intra-and inter-subject variability has limited the success of this approach (Brown, Hughes, Lopez, & Abbas, 1999; Hughes, Brown, Abbas, Wolaver, & Gervais, 2000).

Cortical Auditory Evoked Potentials

In adults, the CAEP waveform consists of a series of peak or troughs (labeled P1, N1, P2, N2) that occur at about 50-250 ms. In infants and young children the CAEP waveform has a different morphology and is dominated by a large positivity (P1) at about 100-250 ms followed by a late negativity at about 250-400 ms (Gilley, Sharma, Dorman, & Martin, 2005). Although researchers have found correlations between discriminative CAEPs (MMN and P3) and speech perception in CI users (Groenen, Beynon, Snik, & van den Broek, 2001; Makhdoum, Hinderink, Snik, Groenen, & Van den Broek, 2002; Micco et al., 1995), these techniques have not been adopted clinically, in part because it is difficult to reliably record responses in individual participants (Dalebout & Foxe, 2001). There are several recent studies that have explored the use of the acoustic change complex in adult CI users (Friesen & Tremblay, 2006; Kim, Brown & Abbas 2008; Martin, 2007) that indicate that this may be a useful future tool for measuring speech sound discrimination in children with a CI.

Most recent studies exploring objective measures of CI function have focused on obligatory cortical responses. Several studies have shown correlations between CAEP latencies or amplitudes and speech scores in adult CI users (Firszt et al., 2002; Kelly et al., 2005; Maurer et al., 2002). Gordon and colleagues (2005) found a relationship between CAEP morphology and speech perception outcomes in children with CIs. CAEP latencies reduce with CI experience in children, particularly in early-implanted children (Sharma et al., 2002a, b). There is also some evidence that CAEPs are predictive of speech perception and functional outcomes for children with ANSD (Pearce, Golding, & Dillon, 2007; Rance et al., 2002). Thus, CAEPs show promise as a clinical tool for either predicting CI outcomes or optimizing CI settings in children with severe-profound SNHL or ANSD.

Using CAEPs to Evaluate Hearing Instruments

Some years ago, a number of researchers explored the use of ABR to evaluate effectiveness of hearing aid fitting in children (e.g., Gorga, Beauchaine, Reiland, & 1987). This work was largely unsuccessful because the ABR is best evoked by a brief stimulus such as a click or toneburst. This type of stimulus can generate stimulus artifacts when transduced through a hearing aid and may not trigger a hearing aid's compression circuitry. Longer duration stimuli including speech sounds can be used to evoke CAEPs, and hence, there has been recent interest in using CAEPs to evaluate hearing instrument effectiveness in children (Dillon, 2005). The goal of this approach is to determine whether speech sounds at conversational levels are effectively transduced by the child's hearing instruments and detected at the level of the auditory cortex. This is akin to a "cortical-evoked Ling sound test (Agung, Purdy, & Kitamura, 2005)." According to Ling (1976, 2002), if a child can detect conversational-level speech sounds that span the speech frequency spectrum with their hearing instruments, then they have the capacity to develop auditory/oral communication skills, provided there are no other factors that would impact on this development.

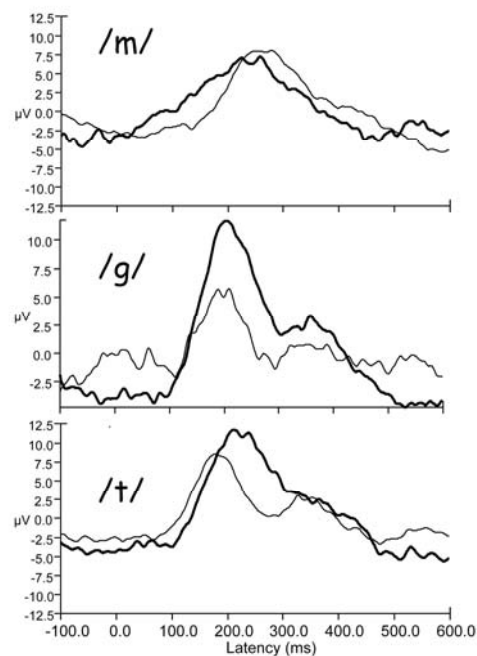
Speech-evoked CAEPs have been used to objectively determine whether a child with severe-profound hearing loss is detecting speech sounds and processing them at the level of the auditory cortex. Sharma and her colleagues (2002a, b) have compared pre-implant aided P1 latencies to post-implant latencies and find that P1 latencies reduce significantly after implantation. The improvement in P1 latencies with a CI depends on the amount of prior auditory deprivation and CI experience (Dorman, Sharma, Gilley, Martin, & Roland, 2007). Sharma and her colleagues have predominantly used one speech sound (/ba/) for CAEP recordings. Researchers at National Acoustic Laboratories (Purdy et al., 2005; Golding et al., 2007; Pearce et al., 2007) have used a range of speech sounds spanning the speech spectrum to evoke

CAEPs in children with typical hearing and with moderate to profound hearing loss. These studies and others have shown that obligatory CAEP recordings provide a reliable objective measure of neural encoding of speech sounds in children with hearing loss, and that CAEP presence/absence is correlated with speech perception or functional outcomes with hearing instruments (Rance et al., 2002; Golding et al., 2007; Pearce et al., 2007). The following case examples illustrate several aspects of CAEP testing in children before and after cochlear implantation.

Case Example 1: Absent Speech-Evoked CAEPs While Wearing High-Powered Hearing Aids Facilitates an Early Decision about CI Candidacy

Figure 1 shows CAEP recordings from an infant with profound SNHL bilaterally who was being evaluated for CI candidacy. The top and bottom traces show CAEP recordings (three replications per condition) from three scalp locations (C3, Cz, and C4, referenced to the right mastoid) over the left hemisphere, vertex, and right hemisphere, respectively. The bottom traces are similar across electrode locations; usually clinicians and researchers would only record from Cz. The top overlaid traces indicate the average waveforms recorded in response to a loud speech sound (/g/ at 75 dB SPL; 78 ms duration; 1125 ms interstimulus interval, ISI) while the child was wearing a high powered hearing aid at maximal gain in the left ear; no repeatable CAEPs are evident at C3 and Cz, although there is a possible peak at C4. Based on the CAEP findings, combined with other functional indicators of poor hearing aid outcomes, this child was implanted with a Nucleus device. The bottom trace shows robust cortical responses recorded to /g/ at a conversational speech level (65 dB SPL) obtained while the child was wearing the CI a short time after implantation. This child has made good progress with his CI.

Figure 1. Cortical auditory evoked potentials (CAEPs) recorded at three electrode locations (C3, Cz, C4, referenced to the right mastoid) in response to the speech sound /g/ recorded in an infant wearing (A.) a high-powered hearing aid in the left ear pre implantation (/g/ at 75 dB SPL), and (B.) a Nucleus CI in the left ear (/g/ at 65 dB SPL). CAEPs are absent with the hearing aid (although there is a possible peak at C4) and robustly present with the CI. Speech stimuli (78 ms duration, 1125 ms ISI) were delivered via a loudspeaker located in front of the child. P1 and N250 peaks are labeled. The sharp peak prior to P1 in the lower traces indicates stimulus artifact that often is present in CAEP recordings from CI users.

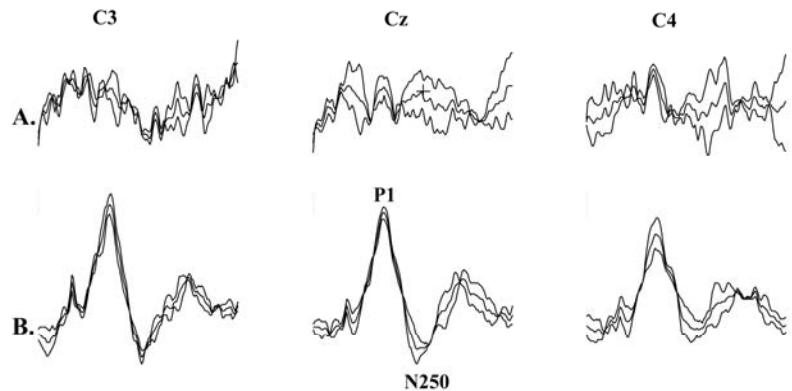


Case Example 2: Unaided and Aided Speech-Evoked CAEPs in a Child with Auditory Neuropathy Spectrum Disorder (ANSD) Facilitates the Decision to Continue with Low-Powered Hearing Aids

Figure 2 shows CAEPs recorded in response to three speech sounds (/m/, /g/, /t/) that span the speech spectrum in a child with ANSD. In this child CAEPs are evident for both aided and unaided conditions. Pure tone audiometry obtained when the child was old enough for visual reinforcement audiometry indicated a moderate bilateral hearing loss, but initial ABR thresholds were consistent with severe-profound hearing loss bilaterally (thresholds greater than 95 dB nHL). This child was born at 24 weeks and had prolonged oxygen therapy whilst in intensive care. In this case the presence of

robust CAEPs to moderate level speech sounds (65 dB SPL) led to the decision to continue with bilateral hearing aids rather than a referral for CI candidacy evaluation.

Figure 2. CAEPs recorded at one electrode location (Cz, referenced to the right mastoid) in response to 65 dB SPL speech sounds (/m/, /g/, and /t/) recorded in an infant with ANSD who was either unaided (solid line) and/or wearing a moderate-powered hearing aid (thin line). CAEPs are present both aided and unaided. Toneburst ABR thresholds (> 95 dB nHL) were consistent with a severe-profound hearing loss bilaterally.



Behavioral audiometry results

when the child was old enough for visual reinforcement audiometry (VRA) testing were consistent with a moderate hearing loss bilaterally.

Case Example 3: Stimulus Artifact Can Make it Difficult to Identify CAEP Peaks in a Child With Cochlear Implants

Figure 3 shows CAEPs recorded in response to 500, 1000, 2000, and 4000 Hz tonebursts (60 ms duration) delivered via a frontal loudspeaker to a 10-year old child using a Nucleus CI in her right ear. Stimuli were delivered at a “loud but OK” level. CAEPs were recorded from seven scalp locations referenced to the earlobe opposite the CI (left). C3, F3, and T5 are left hemisphere scalp locations and C4, F4, and T6 are right hemisphere scalp locations. The electrical artifact caused by the CI is most evident at T6, adjacent to the right ear implant. The observed peaks at C3 (on the side opposite the CI and hence less likely to be contaminated by stimulus artifact) may reflect the CAEP alone, or the CAEP plus some stimulus artifact. Figure 4 shows CAEPs recorded from a child of similar age with typical hearing, showing the change in waveform as stimulus repetition rate slows down (from 910 to 3640 ms ISI). Slowing down the stimulus rate should impact on the neural response rather than the stimulus artifact and hence this might be one way to ensure a cortical response, rather than an artifact-contaminated response, has been recorded. Gilley and colleagues (2005) reported changes in the CAEP waveform with slowing of the ISI similar to those seen in Figure 4. Recently, Gilley and colleagues (2006) described two approaches that may prove useful for separating stimulus artifact from CAEPs in children with CIs, but further work is needed to establish the clinical utility of these approaches.

Figure 3. CAEPs recorded from seven electrode locations (all referenced to the left ear) in response to 500, 1000, 2000, and 4000 Hz tonebursts (60 ms duration, 1125 ms ISI) delivered via a frontal loudspeaker to a 10 year old with a right Nucleus CI. The electrical artifact is greatest at the T6, the electrode location closest to the implant, and smallest at C3 on the side opposite the implant. The CAEP recorded from a child of similar age with typical hearing is shown in Figure 4 for comparison.

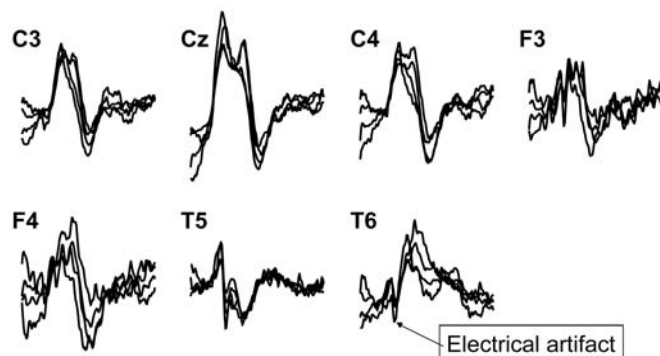
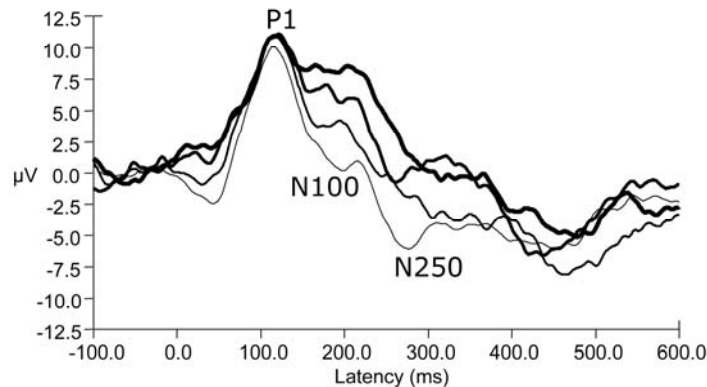


Figure 4. CAEPs recorded at Cz (referenced to the right ear) from an 8 year old child with typical hearing showing the enhancement of P2 as the stimulus presentation rate slows. As the line gets thicker, interstimulus interval increases progressively from 910 to 1820ms to 2730 to 3640 ms ISI. The CAEP waveform resembles that recorded at C3 in the (early-implanted) 10 year child with a CI illustrated in Figure 3.



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Medical and Surgical Evaluation Prior to Pediatric Cochlear Implantation

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Abstract

The evaluation of children for cochlear implantation requires a multidisciplinary effort among several groups of professionals. Given the recent trend towards very early identification of hearing loss, clinicians have begun to evaluate these children and make intervention recommendations in the first few months of life. In addition to an accurate audiological assessment, a search for the etiology of the hearing loss as well as associated medical conditions is critical and frequently affects the management paradigm. The evaluation usually requires a number of studies, including imaging of the temporal bones and brain, electrocardiograms, genetic testing, and careful review of medical records. The timely identification, and management of, confounding otological problems such as otitis externa and otitis media is frequently needed. This presentation will focus on an otologist's perspective in the multidisciplinary evaluation prior to the implantation process.

Background and Protocol

Hearing impairment is common among newborn infants with an incidence of 3-4/1,000 in live births. Approximately one quarter of this group of children have a severe to profound hearing impairment, which often times results in cochlear implantation. Untreated, severe or profound levels of hearing loss markedly reduce sound and environmental awareness. This may result in delayed or even absent speech and language acquisition and impaired communication abilities. These individuals frequently attain lower levels of educational achievement than the typically hearing population, thereby adversely affecting employment opportunities and quality of life.

Since the inception of universal newborn infant hearing screening, there has been a dramatic increase in the number of children presenting for comprehensive hearing evaluation and management. The age at identification has decreased from 24-30 months to 2-3 months with the use of reliable, automated technologies (Harrison, Roush, & Wallace, 2003). With these advances in newborn infant screening and electrophysiological techniques for assessing hearing (Gorga et al., 2006), there has become a strong desire to proceed with cochlear implantation at an earlier age. Thus, prior to discharge from the nursery, after a family learns that their baby has "failed" the newborn hearing exam, diagnostic testing in the form of conventional auditory brainstem response (ABR) and/or auditory steady-state response (ASSR) audiometry may be carried out in the first month or 2 of life. When the results of these test(s) are consistent with a severe to profound or profound hearing loss, families are interested in pursuing cochlear implantation rapidly in an effort to prevent the detrimental effects of auditory deprivation. In fact, recent data have again demonstrated that age at implantation is one of the most important factors for achieving speech perception and

production skills and spoken language outcomes similar to typically hearing peers (Hayes, Geers, Treiman, & Moog, 2009).

Caution must be exercised when cochlear implantation is recommended for a child based on the results of ABR and/or ASSR alone. There remain a number of potential shortcomings in the ability of these technologies to accurately predict auditory thresholds (Tlumak, Rubinstein, & Durrant, 2007). This can make decision-making, based on electrophysiological profiles alone, imprecise. Behavioral testing using visual reinforcement audiometry (VRA) is usually not reliably attainable before 6-8 months of age. With these constraints in mind, cochlear implantation before this time should probably be a rare occurrence because this procedure usually results in a loss of native, residual hearing. Especially important is the avoidance of bilateral implantation in these very young children because the opportunity for further utilizing residual hearing might forever be lost. Thus, there remains constant concern during the early assessment period for children with suspected severe to profound loss and the timing of intervention. While cochlear implant surgery at 6 months of age might be anatomically feasible, this timeline might not allow for an accurate audiological assessment or a sufficient opportunity to undertake a reasonable hearing aid trial. Given these issues, implantation around 12 months of age is probably a more practical and realizable goal.

Currently, our center is serving more than 2,000 children with hearing loss (roughly 650 of them using cochlear implants). Over the years, we have developed a standardized clinical protocol for the evaluation and treatment of these children. This endeavor focuses on the multidisciplinary input from the child's family and a diverse group of professionals with expertise in a variety of hearing-related fields. The professionals hail from the following areas: audiology, medical/surgical, speech-language pathology/auditory-based intervention, genetics, and education. It is common that certain disciplines might predominate during one phase of the process while others frequently become more active at another point. In an effort to create a timely diagnosis and early intervention, we have created a timeline for the events of the first year of life:

- Birth-Day 1-7 = Newborn infant hearing screening
- 2-4 Months = Diagnostic ABR, ASSR, OAE/History and Physical/Medical evaluation (EKG, imaging, genetic testing)/Initiation of hearing aid trial/Early intervention services with auditory-based therapy
- 7-9 Months = Behavioral audiometric testing to confirm thresholds/Auditory-based Therapy/Evaluation of hearing aid trial/Consider CI evaluation
- 11-14 Months = Cochlear Implantation/Auditory-based therapy.

While not set in stone, this serves as a rough guide for the events of the first year of life especially when cochlear implantation is considered.

The otologist's role in caring for the child with hearing impairment is to

1. Diagnose hearing loss by identifying
 - a. Etiology and severity,
 - b. Specific anatomical relationships to functional findings,
 - c. Identification of associated problems, and
 - d. Referrals to related professionals.
2. Treat hearing loss by providing medical and/or surgical interventions.
3. Refer for amplification and/or speech therapy.
4. Prevention and Education of parents, children, and other health care providers on issues surrounding hearing loss
5. Communicate with professionals on the hearing loss team.

The otologist's role usually commences once a hearing disorder has been identified. One exception might be the child that requires diagnostic audiology, but is either unable to be tested under natural sleep or sedation because of associated medical conditions or when it is apparent that middle ear effusions need to be addressed in order to garner accurate test results. These children are taken to the operating room for general anesthesia. In this setting, the ears are examined using the operating microscope and a determination regarding middle ear status is made. When fluid is present, we prefer to place tympanostomy tubes, and the audiological testing protocol is subsequently carried out. Ear canal bleeding must be avoided because this might negatively affect testing results. Placement of otic drops is deferred until after the auditory testing has been concluded. If a hearing impairment has been identified, ear canal impressions for future hearing aid molds are usually taken during the same setting for convenience. Findings are subsequently entered into the newborn infant hearing screening database.

Medical Diagnostic Evaluation

In general, the medical evaluation of hearing loss focuses on trying to identify an etiology for the hearing loss and associated problems that may negatively affect communication or other health issues. Implicit is the fact that a detailed understanding of the causes of hearing loss in children is needed to identify the salient issues in a particular patient. An excellent review of the potential etiologies of hearing loss has been previously published (Morton & Nance, 2006). In addition to searching for the etiology of hearing loss, one must conduct a careful evaluation to identify disorders in vision, craniofacial malformations, and primary speech and auditory processing disorders to allow a comprehensive approach to the communication needs of a child and his/her family. Referrals among a variety of medical professionals are often needed.

There are more than 300 distinct hearing loss syndromes that have been identified by their association with other clinical features. Syndromic forms of hearing loss are less common than the non-syndromic counterparts, accounting for only 10-20% of new cases. However, identification of syndromic features might affect decision-making, including those related to cochlear implantation. Thus, a thorough understanding of these syndromes allows for relatively simple and rapid identification of the genetics and associated co-morbidities that might affect a particular child. For all types of hearing impairment in children, evaluation by a geneticist with expertise in pediatric hearing loss can provide additional information for families.

History and Physical Examination

A careful history, physical examination, and selective use of imaging studies and laboratory testing can identify the etiology of a child's hearing loss in many cases. In addition to the details of the newborn infant screening and diagnostic auditory testing, the medical history should be thorough in the areas of pregnancy and complications, past medical/surgical history, and family history. Details of the child's history that should be extracted include

- Did the pregnancy progress to full term? Were there associated complications such as eclampsia, fetal distress, oligo- or polyhydramnios, bleeding, Rh incompatibility, premature rupture of membranes, preterm labor?
- Was there perinatal infection, such as toxoplasmosis, herpes simplex, rubella, syphilis, cytomegalovirus infection, Group B streptococcus, or other infection (TORCHeS)?
- Did the child spend time in the neonatal intensive care unit (NICU)? If so,

- What was his/her birth weight? (Below 1500 grams)?
- Was the child on a ventilator? What duration?
- Were high oxygen concentrations needed?
- Were there blood transfusions?
- Was there intracranial hemorrhaging?
- Was there necrotizing enterocolitis? This might be associated with the use of aminoglycoside antibiotics.
- Was there retinopathy of prematurity?
- Were there heart defects? If so, what type?
- Was the child jaundiced? If so, how high was the bilirubin concentration, for what duration, and how was it treated?
- Did the child have meningitis? Bacterial? If so, what organisms were present?
- Did the infant require any surgeries?
- What other medical disorders does the child have?
- Are there difficulties with vision, feeding, or with other bodily functions?
- In addition to the usual medical history regarding medicines, allergies, and past surgeries, it is important to assess other family members with hearing disorders and/or disorders related to hearing loss.

The physical examination is focused on trying to identify syndromic features, associated ear-specific disorders, and anatomic situations that would adversely affect communication. Features of the physical examination that might point to specific conditions include

- General appearance traits, such as wide set eyes, pigmentary changes such as heterochromic irides and a white forelock of hair, suggesting Waardenberg syndrome
- Cervical fistulas and pits with ear deformities, suggesting brachio-oto-renal (BOR) syndrome
- Cleft lip/palate, down slanting eyes, coloboma, low set small external ears, and mandible and maxillary hypoplasia in association with a conductive type of hearing loss, possibly suggesting Treacher-Collins syndrome
- Palatal and lip clefts in association with choanal atresia, external ear deformity, and facial paralysis might raise the suspicion for CHARGE association or similar syndromes.
- Microcephaly might be seen in association with perinatal CMV or rubella infection or other events such as birth asphyxia or brain underdevelopment.
- The general neurological status of the child should be assessed. Although this may not provide direct evidence for the etiology of the hearing loss, global neurological and cognitive impairment might clearly influence the effectiveness of a variety of interventions.
- Otitis externa and otitis media should be assessed in all cases as these can adversely affect precise hearing loss assessment and the institution of amplification.

Radiographic Imaging

Radiological imaging is critical in the assessment of all children with hearing loss. In our program, imaging is recommended immediately after the diagnosis of hearing loss has been established by electrophysiological measures. Early anatomical assessment of the temporal bones, auditory, vestibular, and facial nerves, as well as brain, may further characterize the etiology of hearing loss. Also, imaging can identify morphological features that have been associated with progression or poor prognosis from the various interventions, such as hearing aids or cochlear implants.

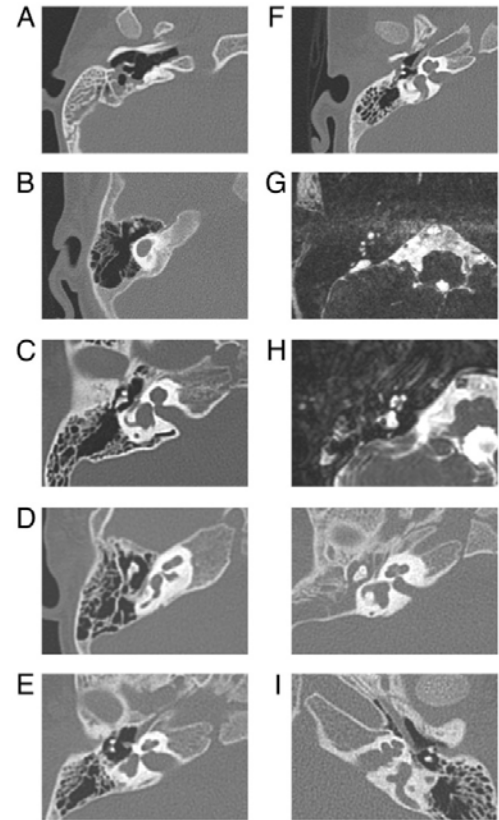
Classical studies of temporal bone and ear morphology have been carried out using histological/pathological techniques in a variety of conditions (Merchant et al., 1993). Thus, the structural characteristics of many of the hearing loss syndromes have been described and can broadly be classified into those with or without radiographically detectable abnormalities. Patients with isolated inner ear cellular or membranous labyrinthine disorders are currently not identifiable based on current imaging resolution. Conversely, labyrinthine malformations of the external, middle, and inner ears and internal auditory canal (IAC) are clearly detectable using currently available imaging. Structural anomalies of the nerves of the IAC and brain are also resolvable in some cases. In general, high resolution computed tomography (HRCT) is well suited for assessing the osseous structures (external auditory canal and middle ear), while magnetic resonance imaging (MRI) provides excellent soft tissue detail for looking at the cranial nerves and brain. The inner ear is well visualized using either MRI or HRCT. HRCT shows the osseous labyrinthine shell well, while MRI shows the fluids within the inner ear that conform to the otic capsule outline. The protocols that we use for these studies have been described previously (Adunka et al., 2007; Adunka et al., 2006; Buchman et al., 2006).

There currently remains some debate regarding which of the various imaging modalities is most appropriate for assessing children with hearing loss (Adunka et al., 2007; Adunka et al., 2006; Buchman et al., 2006; Parry, Booth, & Roland, 2005; Trimble, Blaser, James, & Papsin, 2007). This controversy stems mostly from otologists and radiologists familiarity in interpreting HRCT for inner ear morphological changes. For cases of aural atresia and other conductive hearing losses, HRCT remains superior to MRI for assessing bony detail. Conversely, we prefer MRI rather than HRCT in all children with newly identified sensorineural hearing loss because it allows direct imaging of the cochlear nerves and fluids of the inner ear as well as the brain. The consequences of missing either isolated cochlear nerve deficiency or unsuspected retrocochlear/brain pathology could be profound and might ultimately result in inappropriate treatment of the child. For example, cochlear implantation in an ear without a cochlear nerve or in an ear affected by a tumor could be devastating for the child and family. In cases of sensorineural hearing loss, we use supplementary HRCT only in cases where (a) semicircular canal defects are identified so that the anatomy of the facial nerve is determined, (b) inner ear obstruction is evident on MRI to further determine if the lesion is osseous or fibrous (post-meningitis), (c) the IAC is narrow to determine patency of the bony cochlear nerve canal, and (d) temporal bone pathology has been identified such as in cases of tumors (Adunka et al., 2007; Adunka et al., 2006; Buchman et al., 2006).

Inner ear malformations that are detectable on imaging studies are common in children with SNHL. Some studies have estimated that 20-30% of children with SNHL have some morphological abnormality of their inner ear (Coticchia, Gokhale, Waltonen, & Sumer, 2006). These cases can be conveniently divided into abnormalities of the cochlea and vestibular apparatus or abnormalities of the neural structures. Cochlear abnormalities can take the form of aplasia (absence or Michel aplasia; Figure 3A), hypoplasia (small cochlea; Figure 1E), or dysplasia. Cochlear dysplasias are usually

characterized as an incomplete partitioning as in the classical Mondini malformation (Figure 1G, H) or modiolar deficiency as in X-linked stapes gusher syndrome (Figure 3I). Vestibular morphological variants can also have aplasia, hypoplasia or dysplasias and can affect the semicircular canals, otolithic organs, and the vestibular aqueduct.

Figure 1. Different labyrinthine malformations: (A) HRCT of a right temporal bone depicting a typical case of Michel aplasia, (B) cystic common cavity malformation, (C) cystic cochleovestibular anomaly (CCVA), which shows a separation of cystic cochlear and vestibular structures, (D) absent lateral semicircular canal as part of the hypoplastic inner ear malformation spectrum, (E) hypoplastic cochlea, often combined with other hypoplastic inner ear malformations including absent cochlear nerves, (F) enlarged vestibular aqueduct (EVA, LVA), (G) MRI of enlarged endolymphatic sac, (H) Incomplete cochlear partitioning; MRI & HRCT findings, (I) typical radiologic finding for X-linked stapes gusher syndrome.



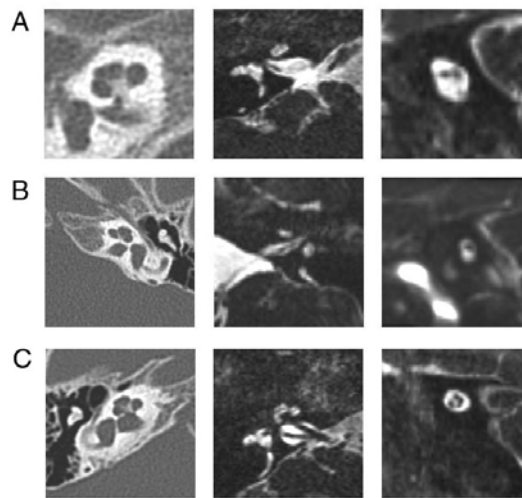
While many children with inner ear or neural malformations have no identifiable clinical syndrome, some will. For instance, absent semicircular canals (Figure 1D) with or without cochlear hypoplasia and cochlear nerve deficiency are very common in children with CHARGE association, VATER syndrome, as well as BOR syndrome. An enlarged vestibular aqueduct (and endolymphatic duct) when seen in isolation or in association with an incompletely partitioned cochlea (Mondini's deformity; Figures 1G, H) might indicate that Pendred's syndrome is present. Children with Waardenburg's syndrome might also have inner ear malformations along the Mondini spectrum of findings as well. A bulbous or dilated IAC that widely communicates with a deficient cochlear modiolus is suggestive of the X-linked stapes gusher syndrome (Figure 1I; Morton & Nance, 2006).

Children with hearing loss can also have a variety of congenital and acquired changes to the central nervous system that are evident on imaging. While a detailed discussion of the central nervous system pathologies associated with hearing loss is beyond the scope of this work, some examples include Dandy-Walker syndrome, congenital CMV, meningitis, neurofibromatosis type II, and changes related to prematurity and kernicterus.

In addition to changes in the brain, anatomical deficiency of the cochlear nerve can be identified on MRI (Adunka, Jewells, & Buchman, 2007; Adunka et al., 2006; Buchman et al., 2006). While this disorder was originally described in children with inner ear malformations and very narrow IACs on HRCT, it has more recently been identified in children with normal inner ears and IACs. Moreover, it has been associated with a variety of syndromes such as CHARGE and VATER and can present in ears with electrophysiological evidence of auditory neuropathy on ABR testing. Figure 4 shows an example of cochlear nerve deficiency (Adunka et al., 2007; Adunka et al., 2006; Buchman et al., 2006).

Figure 2. Cochlear nerve deficiency, axial HRCT images (left), axial CISS sequence MRIs through the level of the internal auditory canal (IAC), and parasagittal reconstructions perpendicular to the axis of the IAC: (A) absent cochlear nerve and normal size internal auditory canal (IAC), (B) single nerve within a small IAC; especially evident on the parasagittal reconstruction. Also, the bony cochlear nerve canal (BCNC) usually seen on axial HRCT images is closed. (C) Normal anatomical situation of the internal auditory

canal. All 4 nerves (facial nerve: anterior superior; cochlear nerve: anterior inferior; superior and inferior vestibular nerves: posterior) can be seen in the parasagittal reconstruction.



Laboratory Assessment

In addition to imaging, several laboratory tests are available for children with confirmed SNHL (see list below). The set of laboratory exams recommended is, in part, dictated by the child's presenting situation. In general, an EKG, CMV, and PCR testing of the Guthrie card and connexin testing are offered to all families, while an eye examination to detect Usher's syndrome should be considered. While the Jervell and Lange-Nielsen's syndrome is exceedingly rare, a properly performed EKG can identify some cases. Because there are treatments for this disorder that can be life saving, this simple and inexpensive test appears justified for all children with SNHL (Morton & Nance 2006). For Usher's syndrome, the hearing loss usually presents prior to the onset of visual changes, making detection in an infant difficult without an electroretinography (ERG). VDRL is offered to families to detect congenital syphilis only in children who have been adopted, where the background of the parents might be unknown. Tests listed as #5 and #6 below are considered in older children or adults with progressive hearing loss when autoimmune disorders might occur. Finally, renal ultrasound is used in children with the clinical stigma of BOR where pits, ear tags, and microtia/atresia exist. Routine screening for Alport's is not currently carried out since the proteinuria/hematuria is usually later in onset (Morton & Nance). Recommended laboratory tests to assess SNHL include

1. Electrocardiogram (EKG) [Jervell and Lange-Nielsen's]
2. Guthrie card PCR for CMV infection
3. Connexin 26 and 30 mutation testing
4. VDRL (syphilis)
5. Erythrocyte sedimentation rate (ESR), complete blood count (CBC)
6. Rheumatoid factor (RF), antinuclear cytoplasmic antibody (ANCA), antinuclear antibody (ANA), and anticardiolipin antibody
7. Renal ultrasound (BOR) and urinalysis (Alport's)
8. Eye examination/electro-retinography (Usher's)

Cochlear Implant Indications and Timeline

Cochlear implants are reserved for those children with a severe-to-profound sensorineural hearing loss (>90 dB HL) in the presence of an anatomically intact cochlea and cochlear nerve. These children should also be enrolled in an educational program committed to an auditory-oral approach and demonstrate limited progress with speech and language development while using appropriately fit amplification. Since the hearing loss is severe to profound, high gain amplification is required, and these devices must be fit using real ear measurements and according to prescribed Desired Sensation Level (DSL) targets.

In the best scenario, children are identified following birth by a newborn infant hearing screening program. Following verification in the first month of life, the degree of hearing loss is estimated by way of electrophysiological testing, and the trial of amplification is instituted. At the same time, the child is followed by a trained speech-language pathologist with experience in auditory-oral approaches. By 6-9 months of age, behavioral audiometry confirms the degree of hearing loss and the speech-language

pathologist can provide feedback regarding auditory awareness and the development of the earliest vocalizations, such as canonical babbling. When progress is evident, continued observation and amplification occurs. Conversely, when the child is making no or limited progress, cochlear implantation is considered with the goal of getting the device implanted around the end of the first year of life.

Factors that might delay this timeline include delayed or inaccurate diagnosis, inappropriate or delayed amplification, inappropriate speech-language pathology treatment, medical co-morbidities that delay diagnosis or preclude surgical intervention, conditions such as severe motor or cognitive developmental delays that hinder accurate auditory assessment, diagnosis of auditory neuropathy spectrum disorder (ANSD), and lack of commitment by the family. Later ages at implantation are also expected in children with progressive hearing loss because these children achieve the severe-to-profound benchmark later in life and usually have better speech and language development due to their residual hearing.

The surgical procedure itself takes about 90 minutes to 2 hours and is almost always performed through a post-auricular incision (~4 cm) and transmastoid, facial recess approach. For most cases, device choice is left up to the patient, with assistance provided by the cochlear implant team. In instances where inner ear malformations or cochlear luminal obstruction have been identified on preoperative imaging, special electrode configurations are available and are considered on a case-by-case basis. Intraoperative electrical telemetry is typically used to interrogate the device for integrity and to roughly estimate a starting point for programming in the postoperative period. While complications are possible, they remain very unusual (Francis et al., 2008). Most children are implanted on an outpatient basis with anesthesia provided by a pediatric anesthesiologist. The children wear a head bandage for 3 to 5 days and return for a check approximately 1 week postoperatively. The device is usually activated 2 to 4 weeks postoperatively.

Bilateral Cochlear Implantation

Are bilateral cochlear implants better than unilateral implants in children? Recent evidence suggests that binaural implantation in adults who are postlinguistically deafened provides significant improvements for hearing in noise and sound localization abilities (Buss et al 2008; Grantham, Ashmead, Ricketts, Labadie, & Haynes, 2007). In children, data are only recently starting to emerge but similar conclusions seem evident (Litovsky, Johnstone, & Godar, 2006; Peters, Litovsky, Parkinson, & Lake, 2007). Only further research will determine whether there is a critical time window for developing these binaural skills in prelinguistic infants. Whether binaural implants will allow children to develop speech and language faster and to a higher level remains the critical unanswered question. What is clear regarding binaural implantation is that having a second device provides a backup should there be trouble with the equipment in one ear. Should the device problems require surgery, having the backup or second side device will prevent unwanted time “off the air.”

Cochlear Implants & Meningitis

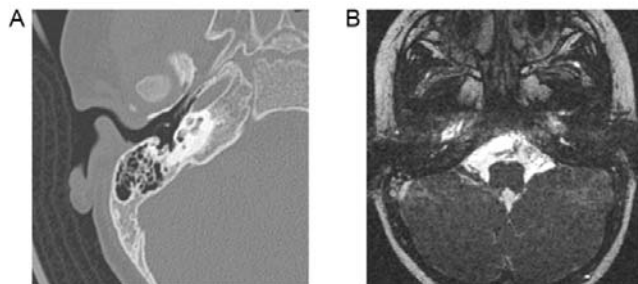
Recent evidence suggests that children with sensorineural hearing loss with or without cochlear implants are at a higher risk for developing bacterial meningitis than the general population (Biernath et al., 2006; Parner et al., 2007; Reefhuis et al., 2003). It also appears from these data that cochlear implants impart an additional risk for contracting meningitis beyond that of having hearing loss alone. While the factors responsible for this increased risk have not been completely elucidated, one particular model of cochlear implant device that used a separate electrode positioner was implicated as predisposing to meningitis at a much greater rate than other models. This positioner has been removed from the market. The findings of these studies also

prompted the United States Food and Drug Administration (FDA) and the Centers for Disease Control (CDC) to recommend for the routine vaccination against the common bacteria implicated in cases of meningitis for all cochlear implantees. Thus, streptococcus pneumoniae (i.e., pneumococcus) and hemophilus influenzae type B vaccination are indicated in all children with cochlear implants. The pneumococcal vaccines include the heptavalent conjugate vaccine (Prevnar[®], Wyeth, Madison, NJ) for children younger than age 2 years and the 23-valent polysaccharide vaccine (Pneumovax[®], Merck & Company, Whitehouse Station, NJ) after age 2 and again 5 years later (before age 10). The precise recommendations of the CDC and FDA vaccine programs are available on the Web (<http://www.fda.gov/cdrh/safety/101007-cochlear.html>). Currently, this is required prior to cochlear implantation in all children and adults in our program.

Conclusions

With the institution of universal newborn infant hearing screening programs across the United States, the evaluation of pediatric hearing loss has changed substantially. The young age of the affected children provides quite a few challenges, especially for those with severe or profound hearing loss where cochlear implantation is being considered. In those cases, a time efficient diagnostic process has to be initiated to ensure implantation around the age of one. To meet the goals for the first year (identification, diagnosis, intervention, education), this process typically involves a group of several professionals including pediatric audiologists, speech-language pathologists, social workers, and otologists, among others. Adequate communication between these groups is fundamental in the process and the otologist usually plays a pivotal part especially prior to cochlear implant surgery.

Figure 3. HRCT & MRI in early intracochlear ossification due to labyrinthitis ossificans after bacterial meningitis: (A) axial HRCT shows increased density of basal intracochlear tissues consistent with early intracochlear ossification; (B) MRI demonstrating fluid sparing of the basal cochlear turn adjacent to the round window region. This indicates intracochlear fibrosis or ossification.



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Current Practices in Pediatric Cochlear Implantation

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Abstract

Technological advances, specifically cochlear implants, have significantly impacted the treatment of children with severe to profound hearing loss. There are, however, very few professional guidelines or resources providing direction for hearing healthcare providers who are serving children with cochlear implants. The following article discusses a comprehensive management protocol for interdisciplinary teams providing cochlear implant services for children.

Introduction

Technological advances, specifically cochlear implants, have significantly impacted the treatment of children with severe to profound hearing loss. Many organizations, including the American Speech-Language Hearing Association (ASHA), the National Association of the Deaf (NAD), and the National Institutes of Health (NIH), and the William House Cochlear Implant Study Group, all have position papers or guiding documents pertaining to cochlear implants; however, there are no published “best practices” or “standard of care” guidelines for cochlear implants. NIH published a consensus document on cochlear implants in 1995 when Nucleus was the only FDA approved cochlear implant system. A technical report on cochlear implants was written in 1986 and then revised in 2003 by ASHA. The Joint Audiology Committee on Clinical Practice, of which ASHA was a member, also published The Joint Audiology Committee Clinical Practice Statements and Algorithms, which included statements on cochlear implant assessments for adults and children (ASHA, 1999).

With the lack of evidence based-standard of care, patients are seen as needed by the audiologists and the schools or early intervention systems are responsible for developing and implementing the aural (re)habilitation program. Cochlear implant programs typically use the Food and Drug Administration (FDA) guidelines and the cochlear implant manufacturer’s recommendations as much as possible in developing their cochlear implant program (see Table 1). In this paper, we will present a best practice model for teams to consider when implementing a pediatric cochlear implant program. We will describe the candidacy, follow-up management, practice management, and outreach/marketing issues that should be considered.

Table 1. FDA Guidelines for the Current Cochlear Implant Systems

Company	Age	Hearing Loss	Sentence Scores	Word Scores	Auditory Development	Hearing Aid Use
Advanced Bionics						
Harmony	12-23 months	Profound SNHL AU (90 db HL)			Lack of auditory development as indicated on IT-MAIS or MAIS	3 months

	2 years - 3 years, 11 months	Profound SNHL AU (90 db HL)		20% or less on MLNT (live voice)	Lack of auditory development as indicated on IT-MAIS or MAIS	6 months
	4- 17 years	Profound SNHL AU (90 db HL)	30% or less on HINT-C	12% or less on PBK		6 months
Cochlear Americas						
Freedom	12-23 months	Profound SNHL AU (90 db HL)			Lack of auditory development as indicated on IT-MAIS or MAIS	
	2 years - 4 years, 11 months	Severe to Profound SNHL AU (70 dB HL)		30% or less on MLNT (live voice)	Lack of auditory development as indicated on IT-MAIS or MAIS	
	5 - 17 years	Severe to Profound SNHL AU (70 dB HL)		30 % or less on LNT	Lack of auditory development as indicated on IT-MAIS or MAIS	
Med El						
Maestro	12 months - 4 years, 11 months	Profound SNHL AU (90 db HL at 1k Hz and above)		20% or less on MLNT	Lack of auditory development as indicated on IT-MAIS or MAIS	
	5 years to 17 years	Profound SNHL AU (90 db HL at 1k Hz and above)		20% or less on LNT	Lack of auditory development as indicated on IT-MAIS or MAIS	

Candidacy

Marketplace for Children

Despite the substantial benefit the procedure can provide recipients, cochlear implantation is an underutilized service. To address this fact, one of the Public Health Application and Outreach goals from Healthy People 2010, Objective 28:13b [8], is to increase the number people who are deaf or very hard-of-hearing who use cochlear implants. The NIDCD's Healthy Hearing Progress Report (2004) notes that only 2 of every 1,000 adults who are deaf or very hard-of hearing received a cochlear implant. For children, Bradham and Jones (2008) reported that only 55% of the children who are candidates for cochlear implantation between ages birth through 6 were recipients of this technology.

Evaluations needed

The cochlear implant candidacy assessment varies from center to center. Based on the current FDA guidelines (see Table 1), children must present with a significant hearing loss, demonstrate a lack of auditory development with appropriately fit hearing aids, and have no medical contraindications for surgery. Due to this, a cochlear implant program needs at least an audiologists and a surgeon. The minimal tests needed to determine candidacy are audiological testing at two points in time to demonstrate degree of hearing loss and (lack of) auditory development. The patient must also undergo a medical evaluation.

Evidence based practices tells us though that this is not enough to achieve the potential outcomes that cochlear implant technologies can provide. Each child must also undergo a speech-language-auditory evaluation by a highly qualified speech-language pathologist in the area of pediatric deafness; educational assessments which

may include a visit with the early interventionists or school personnel; psycho-educational testing to help with establishing appropriate expectations; occupational therapy assessment because many of these children also have other sensory-motor issues; and a family assessment by a social worker. If the following assessments were not completed at the time of identification of hearing loss, then they may also be recommended, as needed, during the candidacy testing: ophthalmology assessment, vestibular assessment, cardiology evaluation, genetic counseling, developmental pediatrician evaluation, and neurological assessment.

Due to the FDA guidelines, audiological testing for determining if a child would benefit from a cochlear implant is relatively straight-forward. Specifically, each child should have audiometric information and speech perception measures for each ear (see Table 2). Speech-language-auditory evaluation should also be completed prior to implantation to obtain a baseline measurement of the child’s current functioning level in these skill areas. Testing protocols should be established and implemented at cochlear implant centers and include vocabulary, language, articulation, and auditory comprehension testing (see Table 3). In addition, academic testing may be included if applicable so that additional recommendations can be formulated for educational agencies. Following implantation, both formal and informal testing should be completed at 6-month post-activation and at yearly intervals thereafter. Given pre- and post-testing information, the implant team is able to measure benchmarks for success in all areas.

Table 2. Audiological Procedures Used During the Cochlear Implant Process

Visit	Description	Procedures	CPT Codes	Time
Inquiry	CI Packet mailed Case history Insurance and release forms Cochlear implant information			15 min
Candidacy Assessment	Audiological Assessment	Otoscopic examination Tympanograms Ipsilateral and contralateral acoustic reflexes OAE, limited Ear Specific information (Pure tones and speech) Comprehensive audiometry OR Conditioned play audiometry OR Visual reinforcement audiometry ABR (if one has not been performed, or poor reliability)	92567 92568 92587 92557 92582 92579 92585	1 hour Separate appointment
	Evaluation of Hearing aids and Counseling	Electroacoustic Analysis of Hearing Aids Binaural Monaural Hearing Aid Check Binaural Monaural Real Ear Measurements (to verify appropriate amplification and settings) Evaluation of Aural Rehabilitation Status	92595 92594	1.5 hour

		<p>Aided thresholds binaural, right ear only, left ear only</p> <p>Best Aided Condition - Aided Speech Recognition (60 dB SPL; Words - Closed set, Words - open set, and sentences)</p> <p>Questionnaires</p> <p>MAIS or IT-MAIS</p> <p>Expectations Questionnaire</p> <p>Meadow-Kendall Questionnaire</p> <p>Case History</p> <p>Review device(s), expectations, candidacy, and evaluation process</p>		
	Educational Consultation	<p>Questionnaires with EI or school provider(s)</p> <p>Site Visit, if possible</p>		1-3 hours
Day of Surgery	Audiological Testing and Consultation	<p>Auditory Evoked Potential Testing in OR</p> <p>Equipment orientation</p> <p>Warranty information</p>	92585, 95920	1 hour
Programming	Initial Activation (2 day process) At post medical surgeon's visit – 2 weeks post operative	<p>Day 1</p> <p>Telemetry Test</p> <p>Program Sound Processor(s) (using VRA, CPA, or Standard methods)</p> <p>Neural Evoked Resources and/or Electric Evoked Acoustic Reflexes</p>	<p>Under 7 yrs, 92601; over 7 yrs, 92602</p> <p>92584</p> <p>92868</p>	1.5 hrs
		<p>Day 2</p> <p>Telemetry Test</p> <p>Program Sound Processor(s)</p> <p>Neural Evoked Responses and/or Electric Evoked Acoustic Reflexes</p>	<p>Under 7 yrs, 92603; over 7 yrs, 92604</p> <p>92584</p> <p>92868</p>	1.5 hrs
	Two Week Follow-up	<p>Program Sound Processor(s)</p> <p>Neural Evoked Responses and/or Electric Evoked Acoustic Reflexes, if needed</p>	<p>Under 7 yrs, 92603; over 7 yrs, 92604</p> <p>92584</p> <p>92868</p>	1 hour
	One Month Follow-up	<p>Evaluation of Aural Rehabilitation Status</p> <p>Aided air conduction thresholds (use C to mark responses on audiogram)</p> <p>SAT/SRT</p> <p>MAIS or IT-MAIS and/or other questionnaires</p>	92626	1 hour

		Telemetry Test	under 7 yrs, 92603; over 7 yrs, 92604	
		Program Sound Processor(s), if needed		
Two Months Follow-Up, if needed		Aided air conduction thresholds Telemetry Test	Under 7 yrs, 92603; over 7 yrs, 92604	1 hour
		Program Sound Processor(s)		
Three Months Follow-Up		Case History and/or questionnaires (assessing for red flags) Aided air conduction thresholds Telemetry Test	Under 7 yrs, 92603; over 7 yrs, 92604	1.5 hours
		Program Sound Processor(s) If needed, Neural Evoked Responses and/or Electric Evoked Acoustic Reflexes	92584 92868	
Six Months Follow-Up		Evaluation of Aural Rehabilitation Status Aided air conduction thresholds (use C to mark responses) SAT/SRT MAIS or IT-MAIS and/or other questionnaires Aided Speech Recognition (60 dB SPL; Words - Closed set, Words - Open set, and sentences) Telemetry Test	92626	45-60 minutes
		Program Sound Processor(s) If needed, Neural Evoked Responses and/or Electric Evoked Acoustic Reflexes	under 7 yrs, 92603; over 7 yrs, 92604 92584 92868	45 minutes

Table 3. Annual Comprehensive Speech-Language and Auditory Skill Assessment Protocol

Age range	Areas of Assessments						
	Voice	Fluency	Oral Mechanism	Auditory	Articulation /Phonology	Language	Cognition/Academics
Birth -3 years	Informal Assessment	Informal Assessment	VBWC Speech Mechanism Screenings Oral Speech	Auditory Development Checklist (VBWC) ²	Phonemic Inventory ² Arizona Articulation Proficiency	Rossetti Infant Toddler Scales ² MacArthur Index of Communicative	Sensorimotor Tasks (Miller et al., Uzgiris and Hunt) Cottage Acquisition Scales for Listening,

			Mechanism Screening Examination – Revised	Early Speech Perception (ESP) Test ²	Scale – Third Revision (as applicable)	Development ² Communication/ Language Sample Bloom-Lahey Semantic Analysis – one and two word utterance stage Language Assessment and Remediation Screening Procedure (LARSP) – above two-word utterance stage Record of Communicative Functions	Language & Speech ¹ Pre-Verbal (0-12 months) Pre-Sentence (12-24 months) Simple Sentence (24-48 months)
3-5 Years	Informal Assessment	Informal Assessment	Oral Speech Mechanism Screening Examination – Revised	ESP Test ² Auditory Perception Test for the Hearing Impaired (APT/Hi) Red Flags ¹	Arizona Articulation Proficiency Scale – Third Revision Identifying Early Phonological Needs in Children with Hearing Loss ¹	Battery 1 MacArthur Index of Communicative Development (as applicable) Reynell Developmental Language Scales Battery 2 Expressive One-Word Picture Vocabulary (EOWPVT-R) Receptive One-Word Picture Vocabulary (ROWPVT-R) Communication/ Language Sample ² Bloom-Lahey Semantic Analysis – one and two word utterance stage LARSP – above two-word utterance stage Assigning Structural Stages – complex sentence structures Record of Communicative Functions VBWC Semantic Idiosyncratic Language Pattern	Bracken Basic Concepts Cottage Acquisition Scales for Listening, Language & Speech ¹ Simple Sentence (24-48 months) Complex Sentence (48+ months) Sounds & Speech Teacher Assessment of Grammatical Structures (TAGS) ¹ Test of Early Reading Ability (TERA-3) ¹

						Checklist	
						Test of Early Language Development (TELD-3) ¹	
5-18 Years	Informal Assessment	Informal Assessment	Oral Speech Mechanism Screening Examination – Revised	ESP Test ² APT/HI Test ²	Goldman Frostoe Test of Articulation KLPA-2: Khan Lewis Phonological Analysis-2 nd Edition (as applicable)	Battery 1 Clinical Evaluation of Language Fundamentals (CELF-4) Oral Written Language Scales Written Portion Only (OWLS) Battery 2 Comprehensive Assessment of Spoken Language (CASL) OWLS Communication/ Language Sample ² Assigning Structural Stages – complex sentence structures Record of Communicative Functions VBWC Semantic Idiosyncratic Language Pattern Checklist At the six month (mid-year) assessment time CASL or CELF-4 (opposite of what you did at the annual evaluation session) ¹ If you cannot give the CASL, then give EOWPVT-R and ROWPVT-R ¹	Bracken Basic Concepts Diagnostic Achievement Battery (DABS-4) as applicable The Phonological Awareness Test (for children who are not reading yet) Gray Oral Reading Tests (GORT-4) Cottage Acquisition Scales for Listening, Language & Speech ¹ Simple Sentence (24-48 months) Complex Sentence (48+ months) Sounds & Speech TAGS ¹

Note: ¹Only at 6 months, ²Both six months and annually

Determining Candidacy

The candidacy criteria have changed dramatically since the first multichannel system was approved. Once FDA approves the device and it is commercially available,

the surgeon can determine how to best use the product. For example, the cochlear implant is approved for all people with significant permanent hearing loss including children down to 12 months of age. Many children younger than 12 months of age, however, are being implanted as well as people who have more residual hearing. The “clinically acceptable” candidacy criteria are evolving with improved technologies and outcome based studies. Due to this, when determining candidacy criteria, the professionals should ask the following questions:

1. “Is physical implantation of the device possible and/or advisable given the medical status of the patient?”
2. “Is it likely that an individual will receive more communication benefit from the cochlear implant than from a hearing aid or alternatively from no hearing prosthesis at all?”
3. “Do the necessary supports exist in the individual’s psychological, family, educational, and rehabilitative situation to keep a cochlear implant working and integrate it into the patient’s life? If not, can they be developed?” (ASHA, 2004).

Another question that implant programs are starting to ask is “Is it cost effective to implant this patient?” There is enough evidence-based research that can assist the teams in answering this question. For example, we know that the age of implantation has a significant effect on the child’s outcome in successful usage of the cochlear implant system.

After asking all the above questions, the cochlear implant team members should be able to determine if the patient is a cochlear implant candidate. The team may also want a more formal tool to use in determining candidacy such as the Children’s Implant Profile (Hellman, Chute, Kretschmer, Nevins, Parisier, & Thuston, 1991), Cochlear Implant Candidacy – Children (CICC; Bradham, Lambert, Turick, & Swink, 2003), Graded Profile Analysis (Daya, Figueirido, Gordon, Twitchell, Gysin, & Papsin, 1999), Modified CHIP (Barnes, Lundy, Schuh, Foley, & Maddern, 2000). The information from this form can be helpful in 1) identifying areas for further counseling to the patient and 2) providing an overall “objective” number, which can be used in the team report for insurance authorization.

Follow-up

Surgery

Once a child is considered to be a cochlear implant candidate, it is recommended that they visit the hospital child life program, if available. These highly specialized providers will help prepare the child and his/her siblings for the surgery by explaining what will happen on the day of the surgery using developmental age-appropriate materials and games to prepare them. Today, many surgeons no longer shave the hair around the surgical site. Furthermore, the surgery for a single-stage procedure now takes less than 2 hours though it will be longer until the family sees their child in recovery. Most centers continue to use a “pressure” type of head dressing on the surgical site. It is important for the child to wear the dressing around the surgical site for 24 hours to minimize potential swelling or bruising. During the surgery, the audiologist can perform auditory evoked potential testing using the cochlear implant system to determine the presence of neural responses. Additionally, implant programs are starting to use this time while the child is in surgery to counsel the family on the equipment use and care, warranty information, and to review what to expect on the initial stimulation visit to help prepare them for that special day.

Audiological Services

At the post operative medical appointment prior to seeing the surgeon, we recommend conducting the initial stimulation of the device. This first visit should include auditory evoked potential testing and behavior responses using Visual Reinforcement Auditory or Conditioned Play Audiometry if possible. If the child is a recipient of simultaneous cochlear implantation, then we recommend only stimulating one device the first day, load the same program on all positions, and review product information. When they return the next day, the second cochlear implant system should be programmed, the first implant should be reprogrammed with multiple programs, and any additional counseling needed. The changes in the programs require frequent visits to the cochlear implant system during the first month. Depending on the family needs, a two month post initial stimulation visit with the audiologist may be needed. An example of one audiological protocol is presented in Table 2.

Speech-Language-Auditory Therapy

Therapy services established prior to the child receiving a cochlear implant is highly recommended. The therapy routine can be established, children and their families understand the expectations and counseling and guidance are provided before, during and after the procedure. Individual therapy pre-and post-implant should emphasize developing listening skills for learning. Parents/caregivers and all family members are encouraged to be equal partners in the therapy process. This equal partnership promotes carry-over of activities to other environments and provides families with the tools they need to develop listening and spoken language skills. The therapist should be continually assessing progress during each session in order to achieve maximum benefit from the device.

Red Flags

Because of ongoing assessment and diagnostic therapy, the team is able to constantly monitor progress through established benchmarks. Areas of concern or “red flags” may arise during this monitoring. These warning signs may include a notably slower rate of progress than anticipated, the child refusing to wear the device during all waking hours, and regression of skills. If “red flags” are uncovered, the implant team begins the investigation or search for the possible root causes. In the newly revised edition of The Auditory Performance Test – Hearing Impaired Revised test, there is a section on identifying red flags that may be helpful to the cochlear implant team. If red flags are identified, the team may recommend reprogramming the device or an integrity check of the internal equipment, utilizing behavior charts to increase the amount of time the child is wearing the device, incorporating different therapy strategies, increasing the amount or type of therapy being provided, adding sensory modalities, consulting with colleagues or referring to other professionals.

Program Management

FTE for Cochlear Implant Program

According to Backous and Littman (2003), they reported from a national survey that 44% of the centers surveyed had 1 FTE, 27% had 2 FTE, and 12% had 3 FTE to run their cochlear implant programs. Forty-four percent scheduled 2.5 to 3 hours for an initial stimulation and 47% saw their patient back the next day for additional programming. Carolyn Brown, Former Director of the Children Cochlear Implant Program at UNC in Chapel Hill, reported that to have adequate staff, a pediatric program should have approximately 80 cochlear implant patients per audiologist (personal communication, March 9, 2004). Further inquiry revealed that a program should have one audiologist to 100 total adult cochlear implant patients (Advanced Bionics, personal communication). In 2002, Garber and colleagues reported an average of approximately 2.5 FTE audiologists dedicated to cochlear implants programs. Furthermore, the study reported approximately 5.5 hours being used for audiological

candidacy testing per patient, approximately 2.5 hours per programming session, and the mean number of follow-up programming visits per patient was 9.3. Due to changes in technology, 2 hours would be more appropriate for programming appointments: 30-45 minutes for history, aided audiogram, and speech perception tests; 30-45 minutes for programming sound processors; and 15-30 minutes for report writing, follow-up phone calls, and repairs. For initial activations, the audiologist would spend approximately 45 minutes programming, 30 minutes counseling on how to use the system, and then 15 minutes for report writing and filling out warranty information.

Marketing

There are many ways to market a program: forums and community talks, newsletters, articles in the newspapers, television interviews, the internet, and word-of-mouth from patients and their families. These are all ways to help educate people about hearing loss and cochlear implants. It is important to repeat these efforts on a continuous cycle because technology changes. Additionally, each cochlear implant manufacturer has a “find a clinic near you” on their Web site. These are very helpful to both professionals and consumers who are looking for a center for cochlear implant services.

Future of Cochlear Implants

As technology is refined, there will be less audiological follow-up needed to provide appropriate care for cochlear implant patients. There is discussion of the development of remote and/or self programming techniques, some of which are currently being used in pilot programs. As the age of implantation decreases, less speech-language services will be required due to being able to take advantage of natural language development. The educating of children with hearing loss will also change. There is much discussion on literacy development in children with hearing loss, in which more research is needed. There will probably be a shift in the future that the SLPs and teachers of students who are deaf will need to focus more on the “other disabilities” rather than the hearing loss. There will be an increase case-load of multiple disabilities where there will need to be more collaboration with occupational and physical therapists. Furthermore, with reimbursement for services continuing to decline, more innovative practice patterns will have to be employed for cochlear implants to remain open for services.

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Pediatric Assessment of Nonlinguistic Sound Perception: A Look Back and a Look Ahead to a Pediatric Nonlinguistic Sounds Test

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Abstract

The overwhelming majority of test measures to assess adult and pediatric cochlear implant candidacy, efficacy, and progress are based on speech perception. Nonlinguistic sounds have received comparatively little attention, despite their central importance for incidental learning, daily living and environmental sound awareness. The purpose of this review is to: 1. Highlight the importance of nonlinguistic sound perception, 2. Discuss currently available pediatric nonlinguistic sound perception profiles and behavioral measures, and 3. Describe both the Nonlinguistic Sounds Test (NLST) for adults and adolescents as well as the pediatric Picture-identification Nonlinguistic sounds Inventory for Children (PicNIC).

Introduction

Nonlinguistic sounds (NLS) allow individuals, both adults and children, to feel safe in, as well as connected to, the environment that surrounds them (Ramsdell, 1978). Comprehension of sound, and information about sound sources, has practical significance for the listener. Listeners can alter their behavior depending on the sound-producing objects in their immediate environment. For example, knowing that a bus or train is approaching or hearing the honk of a car horn or the growl of a nearby dog can help the listener avoid danger or respond to an immediate threat. Even in non-emergency situations, being able to identify the source of a sound allows an individual to respond appropriately. Identifying the sources of nonspeech sounds is a perceptual task that is routinely performed by individuals with hearing that is within normal limits. Survey research has shown that both prelingually and postlingually deafened cochlear implant (CI) candidates describe the reception of environmental sounds as one of the desired benefits of a cochlear implant (Wendt-Harris, Pollack, & Lassere, 2001; Zhao, Stephens, Sim, & Meredith, 1997).

The early goals of implantation were modest, focusing on rudimentary environmental sound perception and an aid for speech-reading; however, the overwhelming success of multichannel CIs led appropriately to a focus on speech perception. The emphasis on speech perception has been so dominant that the initial goals of implantation—environmental and other nonlinguistic sound perception—have largely been ignored. Speech perception measures constitute the bulk of test methods used to assess subject candidacy, post-implant performance, and rehabilitation progress. Implant technology has been optimized and specially designed for speech perception such that modern multichannel CIs are essentially speech processing implants, with nonlinguistic stimuli processing achieved through algorithms geared

towards speech. As a result of this emphasis on speech, high-level speech performance is now routinely achieved in individuals who are postlingually deafened. Many implantees in fact consider language perception significantly easier than environmental sound perception (Parkinson et al., 1998).

For children, nonlinguistic sounds (NLS) help to shape their auditory environment through incidental learning. Just like speech, nonlinguistic sounds can bring enjoyment as well as meaning when identified and paired with something in a child's environment. In the case of infants and young children with severe to profound hearing loss, these sounds can be lost, and therefore, many important auditory connections are not made. For adults who are postlingually deafened, these connections are formed when hearing is within normal or near-normal limits. When hearing is lost and later "re-gained" through amplification or cochlear implantation, it has been shown to be difficult for them to identify nonlinguistic sounds out of context (Inverso, Bickley, & Limb, 2007). Many would argue that for a young child, implanted at or around 12 months of age, nonlinguistic sounds such as environmental sounds would be learned via the cochlear implant; however, acoustical research has shown that CI signal processing is designed to transmit the important spectral and temporal cues of speech specifically which does not necessarily translate to perception of all NLS (Bickley & Inverso, 2008). In fact, until recent years the signal processor of a cochlear implant system was called a speech processor, and often still is.

In part, the relative neglect of nonlinguistic sound perception may have to do with the lack of standardized test materials, as well as a lack of recognition that nonlinguistic sounds have both practical importance (Gygi, 2001), as well as acoustic features that differ significantly from those of language. However, as a result of the broad success of implantation, two trends have emerged: an ever increasing incidence of cochlear implantation in the population of individuals who are prelingually deaf as well as greater expectations in those individuals of what CIs are able to provide. Both of these trends encourage clinicians and scientists to re-examine the issue of nonlinguistic sound perception more focally.

Regardless of the importance of NLS, or the desire of CI users to regain their ability to hear and identify the sources of sounds in their environment, there is a lack of testing materials that evaluate NLS perception, especially for a pediatric population. Additionally, there are an increasing number of very young children and children with multiple disabilities being assessed for and implanted with cochlear implants. There are however, too few measures appropriate for assessing young children who are profoundly deaf or multiply involved children with limited or no linguistic skills. This type of evaluation is especially crucial in the early months following implantation when progress must be monitored closely. The goal of this review is to examine the ways in which nonlinguistic sound perception is evaluated as well as to describe and discuss a new evaluation tool called the Nonlinguistic Sounds Test (NLST).

The assessments currently available and that have been used in the past, can be divided into two categories. First, there are profiles, questionnaires that are filled out by parents, audiologists, teachers, and aural rehabilitationists. These profiles typically offer a list of milestones or behaviors that are used to assess abilities or progress before and after a given treatment. This type of informal assessment goes beyond what can be learned in the clinic and weighs heavily on the perception of the individual being assessed. The second type of assessment is a more traditional measure of sound perception conducted by an audiologist to determine what a child is hearing and/or comprehending.

Currently Available NLS Profiles and Questionnaires

One evaluative tool used to examine perception of sound in children is called the Listening Progress Profile (LIP). The LIP, developed by Archbold (1994), is a profile designed to monitor changes in early auditory perception of young children, and is most commonly used with children using cochlear implants. Two types of nonlinguistic sound perception abilities are monitored using this profile: environmental sound awareness and environmental sound discrimination. LIP also monitors speech perception abilities; however, for the purpose of this review only the sections on environmental sounds will be highlighted.

The LIP is commonly completed by the parent, audiologist, teacher, or aural rehabilitation therapist of the implanted child. The LIP identifies three different skills: response, discrimination, and identification. The response skill is used to describe the detection of a sound. Discrimination is used to describe the ability for the child to choose correctly between two different sounds. Identification is used to describe the ability to correctly choose the target sound from an open set of sounds. The child then is scored on each of these skills in the following way: N (Never/not known), S (Sometimes), and A (Always). The profile is structured as a list of behaviors or skills such as "Response to Environmental Sounds". For this example, if the child sometimes shows awareness of environmental sounds shown by spontaneous response, then the implant teacher recording the profile would mark "S" for sometimes. There are six skills pertaining to nonlinguistic sounds, the example above, "Response to a drum", "Response to a musical instrument", "Discrimination between 2 different instruments", "Discrimination between a loud and quiet drum", "Discrimination between a single and repeated drum", and the "Identification of environmental sounds". Overall, the LIP is a useful tool for determining how well a child is responding to environmental sounds in their environment according to a professional working closely with them; however, it is much more qualitative than quantitative in nature. Additionally, it offers little information on the types of sounds that the child can identify or comprehend.

The Meaningful Auditory Integration Scale (MAIS, Robbins, Renshaw, & Berry, 1991) is a parental interview with ten questions. The interview evaluates the meaningful use of sound in everyday situations such as, attachment to the hearing instrument of the CI, the ability to alert to sounds, and the ability to attach meaning to sounds. The MAIS is designed for children ages 3 years and older. Parents are the interviewee, however, they are not permitted to fill out the form themselves nor are they to give yes or no answers. The final evaluation is based both on parent report as well as clinician observation. The MAIS details ten areas that are probed and then are given specific questions to be asked within those ten areas. The audiologist is to write the responses and examples given by the parent word for word and then choose a specific response for the area. The options are: 0= Never, 1= Rarely, 2= Occasionally, 3= Frequently, and 4= Always. There are four specific questions that address responses to nonlinguistic sounds: "Does the child spontaneously alert to environmental sounds (doorbell, telephone) in the home without being told or prompted to do so?", "Does the child alert to auditory signals spontaneously in new environments?", "Does the child spontaneously recognize auditory signals that are part of his or her school or home routine?", and "Does the child spontaneously know the difference between speech and nonspeech stimuli with listening alone?" The results of this questionnaire are subjective and are meant to compare the child only to himself or herself as an indicator of progress.

The Infant-Toddler Meaningful Auditory Integration Scale (IT-MAIS, Zimmerman-Phillips & Osberger, 1997) is a questionnaire that is used with parents to assess how they feel their very young child is hearing with the cochlear implant. It surveys spontaneous auditory behaviors that children present in daily living, using examples in three different hearing ability developmental areas. These three areas include vocalization changes associated with using the device, alertness to

environmental sounds, and attribution of meaning to sounds. Using information provided by parents, an examiner scores each question, according to the occurrence and frequency of the behavior, from 0 (“never showed this behavior”) to 4 (“always showed this behavior”). The maximum IT-MAIS score is 40. The areas that are related to NLS perception are alertness and attribution of meaning to environmental sounds. In this way, the IT-MAIS is very similar to the MAIS; it is only the age of the children the questionnaire was designed for that is different. The questions about environmental sound perception for the IT-MAIS are: “Does the child spontaneously alert to environmental sounds (dog, toys) in the home without being told or prompted to?”, “Does the child spontaneously alert to environmental sounds in a new environment?”, “Does the child spontaneously recognize auditory signals that are part of his/her everyday routine?”, and “Does the child spontaneously know the difference between speech and nonspeech stimuli with listening alone?” Similarly to the MAIS and the LIP, the IT-MAIS is a qualitative questionnaire that only compares the child’s performance to themselves at a different time. It also does not offer much information regarding which types of sounds are being heard; therefore, it offers little specific information, for example, to help a cochlear implant audiologist re-map accordingly.

Currently Available Behavioral Measures of NLS Perception

The Minimal Auditory Capabilities (MAC; Owens, Kessler, Telleen, & Schubert, 1981) battery is a collection of tests used to evaluate patients who are profoundly deaf and cannot perform on tests of speech perception. The MAC was designed for use with adults who are postlingually deafened; however, a subset of the test has been used with children. There are 13 auditory tests in the revised MAC battery; the following relate to NLS perception: The everyday sounds test is an open-set task. The patient must identify 15 familiar sounds such as a doorbell, people talking, dog barking, etc. This test has not been normalized on young children; however, the portion of the test which evaluates identification of everyday sounds has been used in the past as a language-independent evaluation.

The Test of Auditory Comprehension (TAC; Trammell, 1976) is a closed-set evaluation of both environmental sound perception as well as speech perception. The TAC has ten subtests ranging in difficulty from the ability to discriminate between speech and nonspeech sounds, to the ability to comprehend speech in the presence of a competing signal. The test is appropriate for children as it uses picture identification as the method of response, meaning the child points to a picture to identify their response. The TAC was standardized on a national sample of children with moderate through profound hearing loss, ages four to seventeen years old. The results of the test produce a profile of the child's performance on a continuum of auditory tasks, and provide a basis for instruction with curriculum, as developed by Los Angeles County. The TAC also allows comparison of results by age, degree of hearing loss, and type of educational placement. The TAC is also widely used, for children and adults, as a pre-and post-assessment of the effectiveness of cochlear implants or other auditory devices.

The area of nonlinguistic sound perception is attracting more attention in recent months from the research community. There are several tests currently in development that have been used for researching the perception of nonspeech sounds by cochlear implant users (Arnephy, 2008; Kaga et al., 2008; Shafiro, 2008; Shafiro, Gygi, Cheng, Mulvey, & Holmes, 2008). This list is specific to evaluations of nonmusical perception, as CI-mediated perception of music is an additional topic receiving more attention and evaluation tools than in recent years.

The Nonlinguistic Sounds Test (NLST)

The Nonlinguistic Sounds Test (Inverso, 2008), was originally developed to evaluate NLS perception by adults who are postlingually deafened and cochlear implant

users. The goal of the NLST was to add a simple, clinically useful test to the audiologist's arsenal that was not based on speech perception.

In developing the NLST, we reasoned that it should only include tokens that were correctly evaluated by a pilot group of typical hearing listeners (that is, typical subjects should score essentially perfect on the test), and that it should systematically represent the wide range of sounds present in the auditory world. To increase clinical applicability, we aimed to create a test that is straightforward to administer, not overly long to take (thereby minimizing effects of subject fatigue, particularly in the context of other auditory rehabilitation methods), and that could be used by CI users with a wide variance in performance.

Five categories of nonlinguistic stimuli important for everyday life were identified: (a) Nature (Inanimate) – sounds from nature (e.g., thunder); (b) Animal/Insect (e.g., crickets chirping, dogs barking), (c) Mechanical/Alerting (e.g., telephone); (d) Human Nonlinguistic (e.g. coughing) and (e) Musical Instrument (e.g. piano). These categories were felt to accurately represent the wide range of nonlinguistic sounds commonly encountered, and were comparable to stimuli used in other studies of nonlinguistic sounds perception (Ballas, 1993; Gygi, 2001; Marcell et al., 2000; Shafiro, 2004; Shafiro, 2008). Synthetic sounds, such as science-fiction or special-effects type sounds were excluded, as they were not considered representative of environmental sounds encountered during everyday life. NLS tokens were comprised of monophonic samples compiled from a range of sources (e.g. royalty-free sound effects libraries, audio editing software) with a sampling rate of 44.1 kHz and quantized at 16 bits. For musical instrument stimuli, a timbre task of instrument identification was selected as comparable to the sound object identification task used for the other categories. Professional-grade instrument samples were recorded using the Logic Pro sequencing environment (Apple, Cupertino, CA) as MIDI tracks and rendered into audio at 44.1 kHz at 16 bits. All musical stimuli consisted of one of ten musical instrument samples (flute, saxophone, clarinet, trumpet, guitar, xylophone, harp, violin, piano, organ) playing an ascending and then descending C major chord arpeggio at 100 beats per minute in eighth notes (5 seconds total/stimulus). These instruments were selected as representative examples of all of the major musical families.

Using these categories and specific tokens, the original version of the NLST was designed to have both closed set (category identification) and open set (token identification) properties, thereby allowing testing of a diverse patient population with variable clinical performance. This allowed us to determine whether subjects perceived the general nature of the sound (e.g. animal) and to see if they were able to differentiate the specific source (e.g. cat meow vs. dog bark). It was felt that sounds within categories were more likely to be confused with one another, and that correct category identification provided a meaningful index of nonlinguistic sound perception. Ten sounds from each of the five categories comprise a 50 item list. A total of three lists were generated for use in the original adult version. Each list is in a simple CD format with score sheets and answer keys that match each CD for the audiologist to use to score the patient's performance.

An investigation using the NLST revealed that overall, adult CI users who are postlingually deafened performed poorly compared to typically-hearing listeners, and a great deal of variability was noted among study participants. Postlingually deafened CI users who participated in the study performed at an average level of less than 50% correct, and typically-hearing participants all scored 100% correct for identification of NLS during the pilot study. This finding suggests that NLS perception is difficult for most CI users. The low level of performance recorded in this study is concerning in light of reports emphasizing the importance of environmental sound perception for CI users (Dorman, 1993; Tyler, 1993; Zhao et al., 1997; Zwolan, Kileny, & Telian, 1996).

The Picture-identification Nonlinguistic sound Inventory for Children (PicNIC)

The use of a language-independent test of CI-mediated perception would be an invaluable tool for the pediatric cochlear implant audiologist. The NLST was developed for adult use; however, the pediatric version called the Picture-identification Nonlinguistic sound Inventory for Children (PicNIC) is in development (Inverso, in progress). The PicNIC will use sounds from the same five categories as the NLST: animal, mechanical/alerting, human nonlinguistic, nature, and musical instruments. The sounds were chosen based on familiarity to children and ability to be recognized by picture as well as by audition. The child will hear a sound and have a closed-set of pictures representing sounds from within and outside of that sound's category. Therefore, the scoring will be very similar to that of the NLST. The PicNIC will be a simple and fast test to administer and can serve to evaluate auditory performance for children ranging from typically hearing to both pre-and post-cochlear implantation.

Often, children with significant disabilities such as autism, severe motor and mental retardation, and severe learning disabilities are not identified or considered cochlear implant candidates because testing with behavioral measures can present a challenge. One issue surrounding cochlear implants in children with multiple handicaps is the qualitative benefits he or she receives in communication skills. Additionally, the benefits of implantation can commonly not be determined because of the lack of standardized achievement tests that allow for the great heterogeneity of this population. The PicNIC is language-independent; therefore, it can be used with children who do not respond to traditional auditory testing using speech sounds. The PicNIC will be especially suited for children: who have yet to develop speech or language, with multiple disabilities, who are not native English speakers, etc. In children with profound hearing loss, additional handicaps will be present 41% of the time (McCracken & Bamford, 2000). These numbers are significant because a large number of children and young adults with these audiologic results may be considered for a cochlear implant. Children with profound hearing loss and multiple handicaps present a unique range of challenges including difficulty obtaining complete and reliable audiologic results using standard methods. The use of the PicNIC with this population could greatly increase our ability to determine need for and progress with a cochlear implant.

Conclusion

The success of newborn hearing screening programs across the United States has led to an increase in early identification of children with hearing loss. This has led to a greater number of children being managed and treated with hearing aids, cochlear implants, and audiologic rehabilitation. Additionally, the once very fuzzy line between severe hearing loss and other disabilities in children is becoming clearer. Many children with multiple handicaps are never able to communicate with spoken language even with the use of a cochlear implant. However, the cochlear implant may allow them to be more alert of nonlinguistic sounds. The ability to hear these sounds may alert the child to dangerous environmental situations, strengthen emotional relationships, and lead to an improved attentiveness in both school and home activities (Hamzavi, Baumgartner, & Egelierler, 2000). Children in general, with or without multiple disabilities, may receive benefits from a cochlear implant which may not be observed using standard speech-based measures and parent/teacher profiles and questionnaires. In conclusion, the NLST for older children/adults and the language-independent PicNIC for the pediatric population, are greatly needed to evaluate cochlear implant-mediated perception of nonlinguistic sounds.

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