Pediatric cochlear implantation – II: postoperative follow-up

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Abstract A child with a cochlear implant is expected to achieve the successful outcome of facilitated perception of sound and more oral communication. To achieve this goal, ongoing intervention from a variety of professionals is required. These professionals may represent the disciplines of medicine, audiology, social work, education, and speech / language pathology. In India, cochlear implantation is available in only a few large cities. Here, the otolaryngologist will direct the cochlear implant program. Besides determination of medical candidacy, device implantation and medical management, the otolaryngologist is responsible to ensure that other aspects of cochlear implant management are implemented. This paper, the second of two that describe the multidisciplinary, team approach of the Pediatric Cochlear Implant Program of The Children's Hospital of Philadelphia (CHOP), in Pennsylvania, USA, discusses the non-medical aspects of cochlear implant post-implantation follow-up. The first article, previously published, discussed cochlear implant candidacy. The various speech tests used at CHOP for assessment are based on the English language. They may be translated into the regional Indian languages where the assessment and training can be carried out accordingly.

Keywords Cochlear implant · Tuning · Programming · Speech therapy

Introduction

The ideal candidate A child who has been determined to be a candidate for a cochlear implant at the Children's Hospital of Philadelphia (CHOP) will have undergone a number of appointments with a variety of professionals. Cochlear implant candidacy will have been assessed from the perspective of the surgeon and surgical support staff, determining candidacy based on the general health of the child, his/her ability to undergo general anesthesia, and the shape of the cochlea. The child will be older than 12 months of age, unless the child is post meningitis when the cochleae show signs of ossification that would result in profound deafness and would later complicate insertion of the electrode array. An audiologist will have diagnosed severe to profound (>70 dBHL pure tone average) bilateral sensorineural hearing loss. In addition, the child will have demonstrated poor functional benefit from appropriate and consistent amplification (hearing aid). In older children this is quantified as less than a 30% score on the Multisyllabic Lexical Neighborhood Test (MLNT) or the Lexical Neighborhood Test (LNT) (Kirk, et al 1995). In young children, this is quantified as a failure to progress on the Infant Toddler Meaningful Auditory Integration Scale (IT-MAIS) or the Meaningful Auditory Integration Scale (MAIS) [11, 14]. The speech language pathologist will have evaluated the child's expressive and receptive language ages, independent of modality, and found them to be close to the chronological age. The child who is a candidate for the cochlear implant will be able to vocally produce or approximate a stimulus using auditory, visual, or tactile cues, and will have a good attitude toward oral communication. Psychosocial issues that pertain to the cochlear implant candidate's family (or caregivers) will have been assessed by a social worker. The family will communicate effectively and

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will be actively involved with their child. The family's and the child's (when appropriate) expectations for the cochlear implant will have been determined to be realistic. A family will not have geographic and fiscal barriers to follow-up and maintenance of the cochlear implant. An educational consultant will have determined that the child's home and educational environments are consistent and that an academic curriculum is in place that will challenge the child to increase reliance on auditory communication. There will be therapists and teachers familiar or trained to work with cochlear implants, who understand how to troubleshoot the device and are equipped to handle minor problems such as broken cords and drained batteries. In addition, the candidate ideally will have had a short duration of deafness. Any multiply handicapping conditions that exist would not preclude habilitation techniques. The child's behavior will be controllable by the parent.

Not all candidates will fit the profile of an ideal candidate. Children with deviations from each of the previously described candidacy criteria may be candidates for the cochlear implant. In a companion paper, Franck and Mishra [6] discuss the candidacy evaluation process in more detail.

Surgery The discussion of the surgical insertion of the cochlear implant is not considered in this article.

Pre-activation therapy During the time in between determination of cochlear implant candidacy and activation of the device, conditioning therapy can be helpful to maintain or develop skills that will be helpful in device programming. This type of training may involve the use of residual hearing or vibrotactile stimulation to firmly establish conditioned responses.

Orientation An orientation appointment is available to the family and the child approximately one week before activation. At this appointment, the operation of the device is discussed, the device is physically fit on the child, but the implant is not activated. The condition of the healing incision is assessed, and referral to the surgeon is warranted only if there is unusual redness or swelling. Appropriate magnet strength is determined qualitatively as enough to hold the transmitting coil over the receiving coil without exerting excess pressure on the skin. This allows the child to become comfortable with wearing the device, and the family comfortable with fitting the device on the child before stimulation. In addition, the family is instructed to read the device manual before activation. The purpose of this appointment is to reduce the anxiety and increase the confidence of the child and family before activation. This reduces the novelty of the activation appointment only to the presence of electrical stimulation.

Activation At the cochlear implant activation, the plan for programming the device is communicated to the family (and child, if appropriate). Expectations for the child's response to initial stimulation are reiterated. Expectations for activation should be limited to awareness to sounds.

This explicitly excludes initial ability to understand words or identify the speaker.

When the internal device is first interfaced with the speech processor and programming software, patient demographics are entered into the programming file and the processor for identification. Next, electrode impedance values are measured along the array. The impedance values are reported and the audiologist has the ability to deactivate electrodes if necessary. Unusually high electrode impedances ($>150\text{ k}\Omega$ or $>20\text{ k}\Omega$ for Advanced Bionics' Clarion® and Cochlear's Nucleus® implant systems, respectively) may indicate an open circuit. An open circuit may be manifested by a broken electrode lead or an electrode surrounded by a nonconductive medium, such as what would be the case is the electrode were within an air bubble. Unusually low impedances ($<5\text{ k}\Omega$ or $<1.5\text{ k}\Omega$ for Advanced Bionics' Clarion® and Cochlear's Nucleus® implant systems, respectively) may imply that there is a short circuit in the electrode, allowing current to pass through the device rather than the surrounding neural tissue. The rare instances of either unusually high or low impedance values may indicate that an electrode should not be used for mapping or stimulation.

The speech processors offered by each cochlear implant system manufacturer have multiple coding strategies available. These coding strategies exist along a continuum of stimulation waveforms, number of channels of stimulation and stimulation rates. While similarities exist between the processing strategies offered by different manufacturers, device characteristics and strategy implementation make each device unique. Device manufacturers have guidelines to assist the audiologist in selecting the initial speech coding strategy. However, these guidelines are based on average preferences that resulted in a variety of preferred settings [13]. It is important for the clinician to realize that the optimal processor settings cannot be predicted. Typically, the clinician will use manufacturer-suggested default programming parameters for the initial map settings.

Within these parameters and on one electrode at a time, the audiologist will increase the electric current from an imperceptible level to the point where the child responds. This will be repeated on other electrodes for as many as the child will attend, or until all the threshold levels have been set, whichever comes first. Standard audiometric techniques may be used to do this, including Behavioral Observation Audiometry, Visually Reinforced Audiometry, Conditioned Play Audiometry or voluntary responses, depending on the specific abilities of the child.

The audiologist will then increase stimulation level from the threshold of perception to the maximum level where the child consistently responds and shows no sign of discomfort. Like with threshold levels, this is repeated for each stimulating electrode. The child is not required to provide feedback that each stimulus is audible, rather, the child is distracted, being allowed to play or draw. With children

old enough to understand the concept of a stimulus that is comfortable or too loud, the child may point to a happy and sad face, respectively.

Because the number of such psychophysical measurements required to set the threshold and maximum comfortable levels is large, they may not all be measurable within the child's attention span at the initial programming session. For monopolar electrode configurations (where an active intra-scalar electrode is referenced to an extracochlear return electrode), fitting levels can be interpolated in between measured electrodes. If the clinician measures fitting levels in an order that first spans the array, then fills in between, interpolation can be useful. Objective measures have been found to correlate well with fitting levels, allowing non-behavioral means of establishing the range of stimulation [2, 7, 8, 15].

Once threshold and maximum comfortable loudness levels have been established with measurement, interpolation or objective means, stimulation is presented across the array at the maximum comfortable level set previously with the child, typically in a basalward direction. During this sweep of presentations, the child is carefully watched to ensure that no stimulation is uncomfortably loud. Older children may be able to provide feedback to equate loudness levels across the electrode array.

The next step of cochlear implant activation is to set additional signal processing parameters. Some of these parameters may pertain to overall stimulation details, such as electrode firing order, or parameters that pertain to individual channels of stimulation, such as frequency allocation and gain. As with electrode stimulation parameters, manufacturer defaults are typically used, based on average optimal parameters for which there is large variability.

The combination of all stimulation parameters, stimulation levels and signal-processing parameters are defined as a map. The map is saved to the computer and is downloaded to a program in the memory of the speech processor. Cochlear implants have sensitivity and volume controls. The sensitivity control defines the range below which stimulation is not provided, and above which stimulation is compressed. By increasing sensitivity, quieter sounds are stimulated, and by decreasing sensitivity, only louder sounds are presented. Increasing and decreasing the volume control increases and decreases the level of stimulation, respectively. With the sensitivity and volume controls at their minimal positions, the cochlear implant is activated in a live-voice mode. First slowly increasing the sensitivity to the middle of its operational range, then increasing the volume, stimulation is provided while the child is watched closely for signs of discomfort. At the first sign of discomfort, stimulation is decreased and the processor is set so that there is control of the volume, but so that it cannot stimulate above this level without parental manipulation. Initial reaction to this stimulation may include laughing or crying, becoming withdrawn or excited, vocalizing or quieting.

Due to the variety of possible reaction to stimulation, the clinician and the family must be counseled appropriately. The goal at the end of the first day of programming is for the child to leave with nothing more than a map that results in a comfortable, audible percept.

On additional programming positions of the processor, similar maps are saved with increases in threshold or maximal stimulation levels. Often, levels of stimulation that are loud at cochlear implant activation seem soft to the user after they have become acclimated to the new sensation. By switching to programming positions with higher levels of stimulation, the parent is able to increase stimulation levels beyond the level allowed by the volume control.

The child is sent home wearing the processor. The parents are instructed to make their auditory environment as pleasant and enriching as possible. Suggested activities include playing with favorite toys that make pleasant noises, quiet reading and "discovery walks", where environmental sounds are identified. The parents are recommended to slowly increase the level of stimulation using the volume control or by processor programs if the child does not object. At any sign that the stimulation may be too loud, levels should be reduced. Parents are reminded that the goal is to find the right level of stimulation, not to increase the level arbitrarily.

Regular programming The child will return to the clinic for cochlear implant programming each week for the first month, then monthly until the child has had the processor for six months, then approximately two to three times per year. These visits are necessary to optimize the processing parameters for the individual child. Most frequently, manipulations are made to the stimulation levels. However, manipulations should also include changing signal processing parameters and processing strategies. Both new maps and the map that the child used before the tuning appointment can be stored on the processor. This allows the parent or teacher to monitor the child's performance with the new settings, and revert to their previous map if changes were deleterious to performance. As the child becomes a better reporter of sound preference, he or she will become a more active part of the cochlear implant programming experience.

Auditory therapy (auditory training and speech-language therapy) Auditory therapy is a crucial component of providing the best chance for success with the cochlear implant. The amount of therapy recommended for each child will depend on the child's skills, availability of services, and consistency with educational and home environments. Therapy is recommended at least twice per week, until skills have progressed enough that services are no longer necessary. Not all children will progress to the level of open set understanding of spoken communication relying only on auditory cues. It is crucial that each child is considered individually, and appropriate language is provided.

All children learn to listen and talk by passing through developmental stages. The amount of listening and talking

experience that a child with a cochlear implant may have will vary depending on the onset and progression of hearing loss, as well as the amount of therapy provided before the implant. Because of this experience, a deaf child with a cochlear implant may have accomplished some of these developmental stages before a therapy plan is developed. However, when establishing therapy goals for a newly implanted child, it is important to start therapy with beginning skills. This ensures that none of the required skills upon which the child will build are missed. In addition, we assume that the cochlear implant signal is different from any previous auditory experience.

Erber [4] and Estabrooks [5] developed the basic listening skill model that progresses from sound detection, through discrimination and identification to comprehension. This model has served as a framework for auditory therapy at the CHOP cochlear implant program.

The ability of a child to detect the presence of a sound is a fundamental skill required in programming the cochlear implant. Concomitant skill development by the audiologist and therapist helps develop this skill. The child must not only learn to detect sounds of programming, but also detect environmental sounds and components of speech sounds (/m/, /ah/, /oo/, /ee/, /sh/, and /s/). The therapist will assist the child to learn that after detection, sound is associated with meaning. The meaning may be that the child should look at a toy, respond by placing a block in a bucket, or know that someone is knocking at the door.

The next step is for the child to learn how to discriminate stimuli. Discrimination skills exist on a continuum. The easiest auditory skills for a child with an implant to learn to discriminate are differences in length and pattern of stimuli. Then skills of discriminating spectral features of the sound can be developed. For speech stimuli, skills progress from differentiating /aaaaaaaaaaaa/ from /aa/, then /ice cream cone/ from /fish/. Then skills are developed from differentiating /airplane/ from /hotdog/ to /bed/ from /bus/. For non-speech stimuli the child is taught to discriminate the sound of a telephone (regular long stimuli) from sounds of knocking on a door (short stimuli). Sound discrimination ability requires that the cochlear implant is able to differentially stimulate each sound. Once a child has learned to discriminate sounds, differential meaning can be assigned. Identification of environmental sounds is an exciting process for an implanted child. Parents can guide their child around their acoustic environment, identifying the different things that make different sounds. Speech sounds are classified into segmentals and suprasegmentals [10]. Children are taught to identify these components of speech. Segmental skills are defined by the manner they are produced (plosives, nasals, fricative, affricates, liquids and vowels), the place of articulation (bilabial, labiodental, linguadental, alveolar, palatal, velar and glottal), and if the sounds are voiced. Suprasegmentals include the prosodic features of speech superimposed on the speech segments.

This includes speech duration, intensity, pitch, rate and stress.

Comprehension is the final step and the goal in auditory skill development. In response to speech, training is advanced in response complexity. First, a child might be asked to follow simple directions or respond to familiar statements. Tasks are first made more complex by the addition of multiple or sequenced directions. Then, adding noise and distance between the speaker and the listener can increase task complexity.

Family and school participation is crucial to auditory skill development. Consistency between home, classroom and clinical settings ensures that skill development is reinforced in all aspects of the child's life. In any type of intervention, the child's family is positioned to be their best advocate, to assure consistency of care.

Educational consultation The educational environment of a child with a cochlear implant must support oral communication methods. This does not necessarily exclude the use of manual forms of communication. Furthermore, the quality of the acoustic environment must be optimized to reduce background noise. The role of the Educational Consultant is to provide information to the parents of a child with a cochlear about best educational options for that particular child. Once the family has chosen an educational route that is most compatible with their educational philosophy, geographic proximity, etc, the educational consultant can work with the educational agency and the family to develop the Individualized Family Service Plan (IFSP) or Individualized Educational Plan (IEP). The IFSP and the IEP are formal annual outlines of educational goals and objectives that are required by law for any child with special needs. The IFSP services children ages 0–3 yrs, and the IEP services school-age children.

Annual evaluation Yearly, a child with a cochlear implant returns to the clinic for appointments with the cochlear implant team audiologist, speech language pathologist, social worker and surgeon. This is an important set of appointments for each of the involved disciplines.

The audiologist will confirm that the child's processor and accessories are operating and are being used properly. A comprehensive audiologic evaluation will be performed. Sound field warble tone (250–6,000 Hz) and speech reception thresholds should be observed at approximately 30 dBHL. Age and ability-appropriate speech perception scores should be established [6]. Scores can be compared to pre-operative or previous annual evaluations to monitor progress. Modern cochlear implant processors are computers running speech processing software. If available, manufacturer updates to this software are applied. This may include new processing strategies that can be tried on the child's processor. The cochlear implant is re-programmed based on the testing information and the responses of the child.

The speech language pathologist will formally and informally assess the child's progress. A variety of tools may

be used depending of the age and abilities of the child, and the amount of time available in the evaluation. Receptive language vocabulary may be assessed with the Peabody Picture Vocabulary Test – III (American Guidance Service), and the Receptive One-Word Picture Vocabulary Test (Academic Therapy Publications). Similarly, expressive language vocabulary may be assessed with the Receptive One-Word Picture Vocabulary Test (Academic Therapy Publications). Tests have been designed to measure other aspects of language. The Preschool Language Scale-3 (The Psychological Corporation – Harcourt Brace Jovanovich, Inc.) and the Clinical evaluation of Language Fundamentals – Preschool (The Psychological Corporation – Harcourt Brace Jovanovich, Inc.) test a broad range of language abilities in preschool age children. Older children may similarly be assessed using the Clinical evaluation of Language Fundamentals (The Psychological Corporation – Harcourt Brace Jovanovich, Inc.). The Test for Auditory Comprehension of Language (Pro-Ed) focuses on auditory language skills. Formal speech production tests, such as the Goldman Fristoe Articulation test (American Guidance Service). Voice quality and speech intelligibility may be quantified from The Central Institute of the Deaf Picture Speech Intelligibility evaluation test (Central Institute of the Deaf).

The social worker visits with the parents while the child's hearing, speech perception, speech and language abilities are assessed. During this visit, the clinician evaluates the psychosocial aspects of the implanted child's family. The social worker evaluates to what extent device expectations have been realized and if new psychosocial factors may be influencing speech and language development. Parental stress is quantified with The Parent Stress Index (Psychological Assessment Resources, Inc.) and can be compared to preoperative or previous scores. The social worker is able to intervene when appropriate, or refer to other mental health professionals if necessary.

The surgeon annually checks the health status of the patient with the cochlear implant. A routine otologic examination is performed. Particular attention is devoted to examining the health of the skin flap in between the transmitting and receiving antennae. The placement and stability of the internal receiver is also checked. The child is assessed for tinnitus, vestibular problems, any pain associated with the device and facial nerve involvement.

The data in the table I show some general trends from the one hundred forty three children followed at the CHOP cochlear implant program as of November 2000. Twenty percent of the children were implanted between age 1 and 2 yrs. This represents the greatest number of implants at any age. These children were diagnosed earlier than older implanted kids, indicating the utility of early identification of hearing loss. Furthermore, children implanted between age 1–2 yrs have the greatest proportion of aural / oral communicating methods.

The inspiring outcomes achieved by children with cochlear implants at CHOP reflect worldwide trends. Children with cochlear implants are learning language at a normal rate of development, delayed by the time without hearing [1, 13]. Furthermore, the cochlear implant is allowing children more access to mainstream educational environments [3]. Children with cochlear implants have unique needs, because though they may communicate like their hearing peers, they are profoundly hearing impaired. As profoundly hearing impaired individuals, their success depends not only on themselves, but also on their family's, educators' and clinicians' intervention. A cochlear implant clinic must have a long-term multidisciplinary commitment to the children it serves.

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