

# Changing Considerations for Cochlear Implant Candidacy: Age, Hearing Level and Auditory Neuropathy

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## Introduction

The first child fitted with a cochlear implant was a ten year old boy of who received a single channel device from Dr. William House in 1980 (Eisenberg and House 1982; Eisenberg, Berliner, Thielemeir, Kirk and Tiber 1983). At that time the controversy over the safety and general wisdom of providing children with cochlear implants was raging. Prominent physicians and audiologists at the time, felt that cochlear implants were far too experimental and invasive to be used in vulnerable children. Less than 20 years later, the procedure is accepted as standard clinical practice and thousands of children under the age of 18 have received single and multichannel cochlear implants worldwide.

In the 1980's concerns over implantation of children included weighing the benefits of novice devices against the trauma of electrode insertion and issues surrounding head growth and device migration in young children. Many of these fears have been laid to rest and the benefits of cochlear implantation for aiding in speech perception, speech production and language development of children who are pre-lingually deaf, have been extensively documented.

Twenty years after the first child was implanted, a major issue facing cochlear implant teams regarding candidacy is no longer "should we implant young children?" but "how young should we provide this intervention?" Information on neural plasticity and critical periods as well as the success of early intervention programs for remediation of many types of development disabilities has focused our attention on providing intervention for deaf children at the youngest possible age (see Sininger, Doyle and Moore 1999,

for a comprehensive review). World-wide acceptance and implementation of early identification programs for hearing-loss has uncovered a population of deaf infants whose families are now investigating and seeking all possible interventions much earlier than the families of deaf children seen in audiology clinics 5 or 10 years ago. However, implantation of very young children is not without significant concerns. It remains to be seen, what the reasonable lower-limit of age of implantation will be and how well the realities will mesh with our expectations for optimal outcomes. We are again faced with a dilemma that requires careful gathering of data and prudent clinical wisdom.

Implant candidacy guideline borders are shifting. Experience with children using traditional amplification and those using cochlear implants has allowed comparisons of performance across the two groups. As performance of implanted children has approached that of children with amplification who have better hearing thresholds, the "deafness" or degree of hearing loss criterion has been relaxed somewhat. Measures of aided speech perception over time are now used to compare a child's performance while using traditional amplification to the expected growth of ability of a child using a cochlear implant (Miyamoto, Kirk, Todd, Robbins and Osberger 1995). The degree of hearing loss is now a secondary criterion with speech perception development being considered a more important determinant of implant candidacy. If a child using a well-fitted hearing aid is not making progress at the rate expected if he or she were using an implant, then this child may be considered for implantation even if hearing levels are not profound. This has opened the door for implantation of children with severe hearing loss who are otherwise not achieving expected goals with amplification.

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Also, as experience with cochlear implants expands and the fear of trauma and un-repairable damage to the ear subsides, we are gaining experience and knowledge to allow the implantation of patients with a wider variety of auditory disorders and of children with a wider array of disabilities. On an individual basis, children with mental retardation or autism and other disorders who have hearing loss are being considered as cochlear implant candidates (Lenarz 1998; Waltzman, Scalchunes and Cohen 2000). Auditory neuropathy is another recently described disorder that has been shown to respond remarkably well to electrical stimulation (Shallop, Peterson, Facer, Fabry and Driscoll 2001; Trautwein, Shallop, Fabry and Friedman 2001; Trautwein, Sininger and Nelson 2000). More information on auditory neuropathy will follow in a later section. In short, the cochlear implant has become a standard of treatment for children with severe to profound hearing loss. Experience coupled with caution and reality will determine where the expanding boundaries of candidacy will stop.

## **Current Pediatric Candidacy Guidelines**

Terminology and wording of candidacy guidelines vary by manufacturer, but in general, there are some basic areas of attention recommended for cochlear implant teams to use when deciding on cochlear implant candidacy for an individual child. These areas include 1) degree of hearing loss, 2) aided performance, 3) family expectations and 4) physical and developmental limitations. All areas must be considered simultaneously and none is completely sufficient or deficient alone. Cochlear implant teams have discretion to implant or reject a patient based on their combined judgement.

### **Degree of Hearing Loss**

Cochlear implantation for children of 18 months or older is available under standard guidelines. In addition, both Cochlear Corporation and Advanced Bionics Corporation have devices under FDA investigation for infants as young as 12 months of age. Strict adherence to degree of hearing loss guidelines must be maintained when implantation is conducted under FDA investigation for these very young children. When a child has an average, bilateral hearing loss in

the speech range of 90 dB or more, cochlear implant candidacy is not questioned. In some cases children over 18 months of age with better pure-tone hearing sensitivity are considered as implant candidates, especially when speech perception with hearing aids does not proceed as expected.

### **Aided Performance**

Quantification of performance expectations must take into account the age of the child, the duration and concentration of intervention and the expected developmental sequence. In many centers, a minimum of six months use of well-fitted amplification is necessary to determine if expectations for improved speech perception or auditory skills in an individual child will be met. However, the observation/waiting period and trial with amplification is generally waived in the case of acquired deafness due to meningitis when there are indications that imminent ossification of the cochlea will make implantation difficult or impossible.

Advanced Bionics Corporation suggests the following measures to aid in quantification of development of auditory skills. Children under 4 years of age are considered implant candidates if they demonstrate MLNT (Kirk, Pisoni and Osberger 1995) scores of 20% or less or IT-MAIS (Zimmerman-Phillips, Robbins and Osberger 2000) parent questionnaire scores of 2 or less on questions 3, 5 or 6. Criterion for children over 4 years include PBK word lists scores (Haskins 1949) of 12% or less or HINT-C (Nilsson, Soli and Gelnett 1996) sentences in noise scores of 30% or less.

### **Family Expectations**

Guidance regarding judgement of realistic family expectations is less specific. In general, cochlear implant teams look for families who understand that the surgery is just the beginning of a long-term process that mandates a commitment from the whole family. Parents and guardians must have sought out and be prepared to participate in the educational process and auxiliary services to maximize benefit from the implant. They must understand that obvious changes in communication ability can take a year or more to materialize. Children from families who expect that cochlear implant surgery will provide an easy, fast fix for deafness will not make good candidates.

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## Physical and Developmental Considerations

The physical status of the ear and temporal bone must be evaluated by the surgeon to determine whether implantation is possible. Some inner ear deformities such as a common sac or Mondini Dysplasia may not prohibit implantation but should be noted and discussed with parents and audiologists regarding expectations and potential needs for special adjustments to the device. Other malformations, for example complete aplasia of the inner ear or absence of the auditory nerve would contraindicate implantation.

As many as one-third of children with hearing loss may have other handicapping conditions or developmental delays. Clinicians must understand the degree to which a child can participate in the setting of the device and the establishment of basic device functioning. The ability of the child to use the information provided by the device for development of auditory skills and speech perception must also be weighed against the risks and costs of surgery and the stress on the family. There are no hard and fast guidelines, but it is clear that children with visual, motor and developmental disabilities have been able to take advantage of cochlear implantation (Lenarz 1998; Waltzman et al. 2000) and barriers to implantation of children with other disabilities are falling.

## Reasons to Implant Early

Current information suggests that the earliest possible implantation would lead to the most normal developmental patterns of auditory and communicative skills. The healthy human infant is born ready to absorb auditory information. The newborn is developing intricate neural networks to categorize this information and develop a system of communication via speech. We have every reason to believe that critical or at least optimal periods exist during which such development can proceed normally. For a complete review see Sininger, Doyle and Moore (1999).

There is reason to believe that electrical stimulation can aid in neural survival. Leake and colleagues (Leake, Hradek, Rebscher and Snyder 1991) have demonstrated that electrical stimulation to one ear can differentially aid in spiral ganglion cell survival in the deafened kitten when compared to the

unimplanted ear. Other recent evidence of the advantageous aspects of electrical stimulation on neural transmission in the auditory brainstem and midbrain is found in an overall reduction of absolute and inter-peak latencies of electrical ABR during the first year following implantation (Gordon, Papsin and Harrison in press).

There is ample evidence that early implantation results in significant advantages in communication development. Language development growth patterns were found to be normal or even accelerated in children implanted before 18 months of age when compared to older children (Hammes, Novak, Rotz, Willis and Edmondson in press; Novak et al. 2000). In addition, Hammes et al. (in press) found that children in their study implanted prior to 18 months of age were able to successfully transition from manual to oral communication while less than fifty percent of those implanted between 19–30 months were able to do so. Several of the children who received implants after thirty months of age were never able to transition from manual to oral communication.

Increased awareness on the part of families of deaf children regarding the benefits of early intervention will lead to an increase in pressure upon clinicians to provide cochlear implantation for very young children. At the current time, 40 states have laws establishing newborn hearing screening programs. Deafness is being identified in infants during the newborn period rather than at 12–24 months or later as in the recent past. These infants can be fitted with amplification and enrolled in intervention programs within a few months of life and by 8–10 months of age these children may have had a sufficient hearing aid trial and will meet all other standards of CI candidacy. Parents and families have much greater access to information than in the past and are fully aware of the advantages of early treatment. They are speaking to other parents, especially in chat rooms and listserves on the Internet and are fully aware of the significant benefits that can be gained from appropriate implantation. They also have better access to payment for implant services through both private insurance and public payers. These families are, or should be, full partners in the process of decision making regarding cochlear implantation and will demand the best for their children. It is incumbent upon professionals to guide and inform them of the risks and benefits of implantation of infants.

## What are the Obstacles to Implantation Under 12 Months?

There are many issues raised by the suggestion of providing cochlear implants to children under the age of 12 months. The major issues involve 1) the accuracy of audiometric threshold determination in that age group, 2) the accuracy of measures of functional benefit from amplification, 3) the ability to obtain accurate measures for device setting, 4) the physical limitations of the undeveloped ear and mastoid and 5) the risks of anesthesia in infants.

### Determining Thresholds

Assessment of hearing thresholds in infants and young children is challenging but, with current measures emphasizing electrophysiology techniques and a test-battery approach, precise prediction of hearing thresholds is possible in most infants of any age. While current literature warns that accurate assessment of audiometric thresholds using behavioral techniques is not accurate until approximately 6 months of age, it is important to include an observation of auditory behaviors in any pre-implant assessment. This provides two important bits of information: a validation of the assumed thresholds determined electrophysiologically and an assessment of the child's functional ability to recognize and utilize sound in a meaningful way. For the very deaf child, this may require the use of vibro-tactile stimuli. However, behavioral verification provides the clinician with a glimpse of the child's ability to utilize stimulus information and to participate in the assessment following implantation. To some extent, this information also validates developmental/cognitive assessments that should also accompany any evaluation of implant candidacy. A child of 6 months who cannot participate in a stimulus-response-reward paradigm must be suspected of behavioral or developmental complications.

Audiometric thresholds for specific frequencies using tone-burst ABR can be predicted within 10–15 dB of behavioral thresholds obtained at older ages (Sininger, Abdala and Cone-Wesson 1997; Stapells, Gravel and Martin 1995). One must be cautious to avoid using ABR to estimate hearing in the case of children with auditory neuropathy in which case the ABR threshold is not correlated with hearing threshold. Auditory neuropathy should be obvious from the characteristics of the audiometric test results and a

qualified audiologist using a test-battery approach can avoid the mistake of assuming a child with auditory neuropathy has no hearing based on the ABR. The topic of auditory neuropathy will be covered more completely in a later section.

### Functional Benefit from Amplification

Assessment of functional benefit from amplification can be particularly complex in the very young child. In the period before spoken language is expected, more subtle measures of communication must be employed to assess development. Babbling behaviors and overt response to speech and environmental sounds can be assessed directly or by parental report. One such assessment tool, which is suggested for pre-implant evaluation in very young children, is the Infant-toddler Meaningful Auditory Integration Scale (IT-MAIS) developed by Zimmerman-Phillips et al. (2000). This test utilizes an interview-questionnaire format to assess infant vocalization behaviors, alerting to sound and ability to derive meaning from sound. This type of assessments can be used to give general impressions of progress with amplification, if not quantitative proof, and should be used in the context of all other candidacy criteria.

### Device Setting

As with hearing thresholds, one can turn to physiologic measures to supplement and verify behavioral information needed for appropriate setting of the implant thresholds and comfort levels. As stated earlier, the child should be able to give some indication of threshold in a behavioral paradigm but comfort levels are a concept that would not be easily elicited from an infant under the age of 12 months. Neural response telemetry is a technique that allows recording of auditory nerve compound action potentials from the intracochlear electrodes following stimulation of selected electrode pairs. Following the measurement of an amplitude growth curve, the 0 point on the curve can be interpolated and used as a predictor of setting for threshold or comfort level for the specific electrodes under test (Brown, Abbas and Gantz 1990; Shallop, Facer and Peterson 1999). However, while this technique is essentially accurate, some caution must be used in interpreting results in the first few months following implantation when behavioral thresholds can be changing (Thai-Van et al. 2001). The electrically induced stapedial reflex can be used in a similar

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way with visual inspection during the surgical procedure (Shallop et al. 1999) or by post-implant testing in a more traditional manner using a tympanometer (Battmer et al. 1990). In general, at least for the child who is otherwise on-target developmentally and can assist in device setting with predictable behavioral responses, sufficient physiologic measures exist to aid in the determination of reasonable settings for the device. It is not clear if these aids would be sufficient in a child who was developmentally under the age of 6 months.

### Undeveloped Ear

While the cochlea is of adult size and functionally mature at birth, the skull and temporal bone will continue to grow for several years. One question that arises when considering the implantation of infants, is whether the thickness of the skull in the temporal region and the skin overlying this area are sufficient to provide a seat for the receiver-stimulator. A well is drilled into the skull to seat the device and prevent migration over time. The thickness of the skull in the region where the device is placed is only about 2–4 mm in a child aged 1–2 years. The receiver-stimulator itself is 6–7 mm thick. Because the soft tissue covering this area is only 5–9 mm deep, surgeons have resorted to a “deep seating” technique in which the bone is drilled away exposing dura and allowing a soft area with some give for seating the device. To date no complications of this technique have been reported (Hoffman 1997; Lenarz et al. 1999). However, the skull thickness issue has not been explored in children under about 6 months of age and very few children have been implanted at that age. The full range of issues regarding seating of the device in the infant less than 6 months is yet uncertain.

Middle ear growth that would strain the electrode lead is essentially complete in the first year of life. The danger of electrode extrusion has been addressed by building a few millimeters of redundancy into the electrode in the mastoid to allow for growth. The electrode is also anchored in the middle ear near the cochleostomy to avoid extrusion. Hoffman (1997) reports that the incidence of electrode or device migration is the same or less in children than in adults. Roland et al. (1998) also found the electrode array to be stable in a group of 27 children under the age of six years.

In the past, concerns have been raised over the complications of middle ear disease and implantation in this otitis media-prone age group. These fears have

been shown to be unfounded. Clark et al. (1987) demonstrated that a fibrous sheath grows around the electrode and seals the cochleostomy preventing migration of pathogens from the middle ear. Others have found no increase in otitis media following implantation nor any degree of complications when it occurs (Cohen and Hoffman 1993; Dahm, Shepherd and Clark 1993).

### Anesthesia Risks

The most serious concern regarding implantation of infants less than one year of age is the risks of anesthesia in this age group. Nancy Young, M.D. of Children’s Memorial Medical Center in Chicago recently summarized her concerns in this area noting that the incidence of complications from anesthesia is eight times higher in infants under 12 months than in older children (Young in press). She further states that “There is significant evidence that infants are at increased anesthetic risk in comparison to older children and adults” and concludes that “Since infants six months of age and younger are the most likely to experience problems, implantation in this age group in the absence of urgent indication may be ill advised.”

### Auditory Neuropathy

A recent wrinkle in the area of cochlear implant candidacy is the question of whether patients with a disorder of the auditory nerve termed Auditory Neuropathy are appropriate candidates. Many patients with auditory neuropathy have received cochlear implants, some before a clear diagnosis was obtained, and the vast majority has performed quite well.

The term auditory neuropathy was coined by Starr, Picton, Sininger and Berlin (1996). Patients with auditory neuropathy have hearing loss, abnormal or absent auditory brainstem response (ABR), evidence of normal outer hair cell function from either normal otoacoustic emissions or present cochlear microphonics, and very poor speech perception ability (Sininger and Oba 2001). These patients can have any degree of hearing loss from mild to profound but a preponderance show severe-to-profound loss.

Very poor speech perception scores in these patients can be out of proportion to the degree of loss if it is mild or moderate. Another paradox is the

finding that ABR thresholds are not correlated with hearing levels. Both of these findings are explained by very poor neural synchrony and temporal misalignment of neural discharge patterns with the auditory (speech) signal. This in turn is explained by the disease process at the level of the nerve. The consequence is that traditional hearing aids may be able only to help with decreased sensitivity but can do very little to improve degraded speech perception. While some patients are able to obtain limited benefit from hearing aids (Cone-Wesson, Rance and Sininger 2001), alternatives to traditional amplification, such as cochlear implants, are necessary for most of these patients.

Twenty seven percent of all patients with auditory neuropathy, in a series studied by Starr (2001), had evidence of concomitant peripheral sensory and/or motor neuropathy. When data from adult patients are evaluated alone, as many as 80% show evidence of peripheral neuropathy. Both demyelinating and/or axonal neuropathy are the suspected from the pathology evidence from sural nerve biopsy on Starr's patients. Other cases showing evidence of neuropathy of the peripheral auditory nerve at autopsy have been described as well (Nadol, Jr. 2001; Satya-Murti, Cacace and Hanson 1980; Spoendlin 1974; Starr, Picton and Kim 2001).

Some would question how a cochlear implant could provide benefit when the nerve itself is damaged or diseased. In fact, there is significant retrograde deterioration of the auditory nerve following sensory loss and most patients who are deaf due to loss of sensory hair cells, show very poor nerve survival (Spoendlin and Schrott 1989). However, these same patients generally receive significant benefit from cochlear implants. Clearly, a full complement of spiral ganglion cells and peripheral auditory nerve fibers is not needed to achieve success with a cochlear implant. Electrical stimulation of the demyelinated auditory nerve in mice has been shown to be sufficient to produce an ABR (Zhou, Abbas and Assouline 1995). In addition, electrical stimulation itself can help to promote auditory nerve survival (Leake et al. 1991; Leake, Snyder, Hradek and Rebscher 1995).

The real proof lies in the results following cochlear implantation found on many children and adults with clear diagnosis of auditory neuropathy including present OAEs and absent ABR. Many of these children have received cochlear implants and show excellent results that are essentially undiscriminated from

those of peers with sensory loss (Shallop et al. 2001; Trautwein et al. 2001; Trautwein et al. 2000). These patients have been shown to demonstrate auditory nerve action potentials in response to electrical stimulation and excellent performance on speech perception test batteries over time following implantation.

There are, however, some important caveats regarding the implantation of children with auditory neuropathy. One question yet to be answered is: "should a patient with moderate hearing loss and very poor speech discrimination (and presumably a normal complement of hair cells) receive a cochlear implant?" Currently no published data exist on patients with moderate loss due to auditory neuropathy who have been treated with cochlear implants. However, if results in adult patients are promising, then this must be considered for children because of poor prognosis with conventional hearing instruments.

A second issue that is related to the age of implantation, is the characterization of hearing thresholds. Unlike the earlier description of accurate threshold prediction from ABR for infants, the ABR is not a reliable way to predict threshold in patients with auditory neuropathy. Consequently, audiologists must rely on behavioral measures of hearing that are not accurate until around 6 months of age. To further complicate matters, some children with auditory neuropathy lose their otoacoustic emissions for reasons that are unclear (Sininger and Oba 2001) and others may have remission of symptoms or dramatic fluctuation in hearing thresholds. It appears that it would be prudent to be more cautious regarding the implantation of a very young child (under 1 year of age) with auditory neuropathy and to wait until a clear picture of the pathology has emerged, a consensus on neurological status is obtained and until hearing thresholds are stable and reliable. Only then should a young child with auditory neuropathy be considered for cochlear implantation.

## Summary

In summary, there appear to be some insurmountable limitations to the lower limit of age of cochlear implantation at or around 6 months of age. These include physical dimensions of the skull, difficulty with obtaining behavioral information for confirmation of accurate assessments and the increased risk from anesthesia. The actual lower limit for

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children with developmental delays or auditory neuropathy is older and must be determined on a case-by-case basis.

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