ABSTRACT. Objectives. As a result of universal newborn hearing screening and improved evaluation tools, many children with severe to profound hearing loss are being diagnosed as infants. This affords the opportunity to provide these children access to cochlear implantation, although medical and audiologic challenges must be addressed. The purpose of this study was to investigate the safety and efficacy of cochlear implantation in children who are younger than 1 year.

Methods. A prospective study was conducted of 18 children who had confirmed severe to profound sensorineural hearing loss and received cochlear implants at our medical center before 12 months of age. The length of device usage ranged from 6 months to 4 years, 5 months. The main outcomes measured were perioperative and postoperative surgical/medical aspects, the Infant-Toddler Meaningful Auditory Integration Scale and age-appropriate phoneme, and word and sentence recognition tests, when appropriate.

Results. All children had full insertions of the electrode array without surgical complications and are developing age-appropriate auditory perception and oral language skills.

Conclusions. Early implantation is feasible and beneficial in some children who are younger than 12 months and should be considered with attention to variables involved in the decision-making process, including possible increased surgical risk, skull size and scalp thickness, and mastoid development. Pediatrics 2005;116:e487–e493. URL: www.pediatrics.org/cgi/doi/10.1542/peds.2005-0282; cochlear implantation, infants.

ABBREVIATIONS. IT-MAIS, Infant-Toddler Meaningful Auditory Integration Scale; dBHL, db in hearing level; NR, no response; CP, Common Phrases; MLNT, Multisyllabic Lexical Neighborhood Test; LNT, Lexical Neighborhood Test.

Universal newborn hearing screening has allowed for the identification of severe to profound hearing loss at birth or within a few months thereafter. Because universal hearing screening programs have been and continue to be established throughout the United States, between 85% and 99% of newborns are being screened for hearing loss within 1 day of being born, with the remainder being tested within the first few months of life. This new population of hearing-impaired infants, who by virtue of the extent of their hearing losses are possible candidates for cochlear implantation, provides new opportunities and challenges.

Research over time has demonstrated that the early identification of significant hearing loss followed by intervention procedures, including hearing aid usage commencing during the first 6 months of life, significantly increases the level of language development, speech intelligibility, and emotional stability as compared with children with later identification and intervention. On average and over all degrees of hearing loss, children achieve higher levels in linguistic, academic, and social skills when management of hearing loss commences at an early age. Because much achievement in the world is based on linguistic competence, the ability of children to communicate often defines many aspects of their educational and social development. Consequently, the window of opportunity for language learning is critical to overall childhood development. For the most part, those critical years have been determined to be from birth to ~7 years of age, by which time the majority of the tools necessary for effective communicative competence are in place. Congenital severe to profound hearing loss deprives children of the ability to extract those rules and has been shown to limit their ability to develop effective auditory and oral linguistic capabilities and communication. Research has demonstrated that profoundly deaf children have significant delays in their language development skills, including oral and written, as compared with their normal-hearing peers despite the use of appropriate amplification and extensive rehabilitation. Characteristically, pediatric cochlear implant recipients already have significant language and speech delays at the time of implantation considering that, historically, the majority of children were receiving implants at age 2 years and older. Pediatric cochlear implantation, however, has provided these profoundly congenitally/ prelingually deaf children with greater access to sound, which has promoted an increase in auditory skills, speech understanding, and oral linguistic development. Recently, studies with children who were trained in oral communication have shown not only that these delays do not increase after implantation but also that future gains that are realized are often on par with normal-hearing children. Investigators have reported on small groups of severely to profoundly hearing-impaired children who have undergone cochlear implantation and are developing oral language and speech production skills commen-
surate with their normal-hearing peers. To catch up, however, these children often have to learn at a faster-than-normal rate.\textsuperscript{7,8} It stands to reason, then, that if implantation occurred at a younger age, then the gap might be narrower or nonexistent, allowing for language development within the normal range.

In fact, recent reports on early cochlear implantation indeed have revealed greater levels of achievement in some children. In 2002, Hammes et al\textsuperscript{9} found that children who received implants when they were younger than 18 months had considerably better language outcomes than children who received implants when they were older than 18 months. Moreover, the children who received implants early performed better than deaf children who were identified, aided, and provided with appropriate intervention before 6 months of age but received implants at a later age. Waltzman et al\textsuperscript{10} in 1998, Geers et al\textsuperscript{11} in 2003, and Svirsky et al\textsuperscript{12} in 2004 also found that children who received implants when they were younger than 2 years achieved better perception scores and oral language competence than children who received implants when they were older than 2 years. In 2004, Robbins et al\textsuperscript{13} compared 3 groups of children on the basis of age at implantation: 12 to 18 months, 19 to 23 months, and 24 to 36 months. Using the Infant-Toddler Meaningful Auditory Integration Scale (IT-MAIS), a parent questionnaire of communication behaviors, the authors found that at 6 months after implantation, more than half of the youngest group achieved levels equal to that of normal-hearing children, \textasciitilde 25\% in the second group achieved comparable scores, but \textasciitilde 10\% in the oldest group were on par with normal-hearing peers. In addition, some young pediatric recipients have been capable of developing proficiency in \textasciitilde 1 spoken language.\textsuperscript{14} Despite the theoretical positive benefits associated with early implantation, challenges remain.

Cochlear implant surgery in young children may present several additional risks that are not necessarily present in older children and adults. Anesthetic risks, including an increased risk for respiratory failure and bradycardia, have been shown to be higher in infants who are younger than 6 months and in children who are younger than 1 year if other medical illnesses are present. The risk for complications or untoward events has been shown to be lessened when a pediatric anesthesiologist performs anesthesia. Total blood volume is also significantly less in the very young child; therefore, blood loss needs to be kept to a minimum, especially when drilling in the immature mastoid bone marrow.\textsuperscript{15}

In addition to the medical issues, the degree of hearing loss in each ear needs to be established with certainty. Objective testing methods, including auditory evoked potentials and auditory steady-state responses, have been able to provide more accurate thresholds of hearing, thereby increasing the comfort level concerning the amount of hearing loss and residual hearing. The progress in developing age-appropriate speech perception skills assessment tools, however, has been much slower. The majority of these tests are language based, and although versions are being developed for children as young as 18 months, accurate measures for infants is far more challenging, hindering the ability to make final decisions concerning candidacy. In the absence of appropriate measurement tools, how might one judge the efficacy of hearing aids in terms of speech perception in an 8-month-old or in terms of linguistic development? Although investigators are in the process of creating such procedures, clinicians currently rely on less objective measures as well as evidence that cochlear implants are efficacious in older children to decide for implantation in an infant. Postoperative device programming presents additional challenges. Accurate thresholds and comfort levels are the basis for appropriate and effective device programs that are necessary for optimal functioning of a cochlear implant. Although these measures are not expected to be definitive at initial stimulation and are expected to change over time in both older children and adults, a close approximation of accuracy is desired and leads to a better listening environment for all recipients. Although standard pediatric behavioral techniques can be used to elicit responses, advances in neural response telemetry and imaging, which provide guidance as to the exact place of the individual electrodes and henceforth the frequency bands being stimulated, have provided more objective tools of threshold measures to assist in device programming.

Despite that many of the issues surrounding infant implantation have been and continue to be addressed, the basic uncertainty of whether implantation before 12 months of age yields greater auditory and linguistic benefits without increased risk remains unanswered. The purpose of this preliminary study was to examine cochlear implantation in children who were younger than 12 months to determine short-term efficacy and safety.

METHODS

Eighteen children with bilateral severe to profound sensorineural hearing loss were enrolled in this study (Table 1). Seventeen had congenital hearing loss and 1 child, patient 1, was deafened as a result of meningitis at 5 months of age. Thirteen of the 17 children with congenital hearing loss were identified via newborn hearing screening programs, although 5 of those had positive

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family histories and were at high risk for hearing loss. Hearing loss for patients 2, 3, 12, and 15 was diagnosed at 3, 2, 4, and 2 months, respectively. The mean age at implantation was 9.6 months (range: 6–11 months), and the mean length of device usage was 17.5 months (range: 6–53 months). Preoperative amplification usage ranged from 2 months to 8 months: 14 of the children were fit with FM systems, and 4 used traditional hearing aids. All of the children were enrolled in early intervention programs on diagnosis and receiving appropriate rehabilitation services. Seven children received the Nucleus CI24RCS, 9 received the Nucleus CI24RCA, 2 received the Nucleus CI24K, and all who have undergone stimulation are programmed with the Advanced Combination Encoding processing strategy (Cochlear Ltd, Lane Cove, New South Wales, Australia).

Preoperatively, the nature and the extent of the hearing loss was established using frequency-specific auditory evoked potentials, ear-specific behavioral audiograms when possible, and ototympanic movement after implantation. The facial recess (posterior tympano-)skull development may possibly make the device more prone to nonabsorbable sutures because new bone formation and future implantation. The possibility of transected emissary veins must be expeditious to avoid excessive hemorrhage. Although there is generally no anatomic difference in the middle ear dimensions, the semicircular canals and the sigmoid sinus can occur when drilling the trough from the well to the mastoid cavity. The mastoid is often underpneumatized in this population. Using diamond burrs and bone wax provides good bone formation, and future surgery for the mastoid infection is easier. The mastoid becomes thicker in the very young child than in the older child and is handled in an atraumatic and delicate manner. Blood loss is kept to a minimum, with judicious use of the monopolar electrocautery. Attention to and coagulation of transected emissary veins must be expedient to avoid excessive hemorrhage. Although there is generally no anatomic difference in the middle ear dimensions, the semicircular canals and the facial nerve are closer to the mastoid cortex in the very young child. The facial nerve can be just under the skin at the mastoid tip because of the relative underdevelopment of this structure. Therefore, the incision is not taken as inferior as one might do in an older child.

Drilling the well and the tie-down holes to seat the receiver/stimulator requires use of the operating microscope to avoid dural injury in the young child with a thin skull. Exposure of the sigmoid sinus can occur when drilling the trough from the well to the mastoid cavity. The mastoid is often underpneumatized in this population. Using diamond burrs and bone wax provides good hemostasis of the exposed mastoid bone marrow. Attention is paid to areas of slight oozing that might be ignored in an adult or an older child.

Fixing the receiver/stimulator to lower the overall device profile on the skull does require some dural perforation that has not been found to be dangerous. The device is tied down firmly with nonabsorbable sutures because new bone formation and future skull development may possibly make the device more prone to movement after implantation. The facial recess (posterior tympanotomy), cochleostomy, and electrode insertion were no different postoperatively. The facial recess (posterior tympanotomy), cochleostomy, and electrode insertion were no different after implantation. The facial recess (posterior tympanotomy), cochleostomy, and electrode insertion were no different postoperatively.
Postoperative intravenous antibiotics directed toward the cultured microbes (bacteria) were administered for 6 weeks. After cessation of antibiotic therapy, a sinus tract developed and again the same microbe (*Staphylococcus epidermidis*) was cultured.20,21 The decision was made to remove the device but leave the electrode array in the cochlea to preserve a lumen for reimplantation at another time. Three months later, a new device was implanted on the same side. The patient is currently 1 year after reimplantation without incident or deterioration of performance.

**Programming**

The children conditioned well using conditioned orienting response audiometry and subjective and objective measurements including neural response telemetry and electrical stapedius reflex thresholds; responses to electrical stimuli were obtained reliably during initial stimulation and at all follow-up programming sessions. All children adapted well to wearing the device without any adverse reactions.

**Auditory Skills**

Sound field threshold responses to frequencies 250 to 4000 Hz and speech stimuli were in the mild hearing loss range for all stimulated patients by 1 month after stimulation. Postoperative behavioral thresholds were obtained at 6-month intervals and confirmed the preoperative levels obtained in the nonimplanted ear.

**IT-MAIS**

Preoperative (with amplification) and postoperative 6-month IT-MAIS data were obtained on all 18 patients, and 1-year postoperative data are reported for 9 children. Substantial improvement was noted for all 18 children from the preoperative hearing aid condition to the poststimulation 6-month evaluation. The mean preoperative total score was 0.7 (1.75%), and the mean postoperative total point score was 30.4 (76%) of an achievable 40 points. Preoperatively, for category 1 (vocalization behavior), the preoperative mean score was 0.2 of a possible 8 points (2.5%), and postoperatively the mean score was 6.7 (83%), indicating that children consistently increased the use of vocalization after implantation. For category 2 (alerting to sound), the preoperative mean score was 0.4 (2.5%) points, and the poststimulation score was 12.3 of a possible 16 (77%), demonstrating that the children rarely alerted to sound with hearing aids but reliably alerted with the implant. Similar results were noted for category 3 (deriving meaning from sound), for which the preoperative mean score was 0.08 (0.5%), and the postoperative mean score was 11.3 (71%) of a possible 16 (Fig 1).

Continued improvement was noted as length of device usage increased. For patients 1 to 9, when 12-month data are reported, improvement was noted from the 6- to 12-month evaluation interval. The 6-month total score for these 9 patients was 30.6 (76.5%), and the 12-month total score was 34.8 (87%). Category 1 performance improved from 85% to 93.8%, category 2 improved from 75.6% to 87.5%, and category 3 improved from 72.5% to 83.1% (Fig 2).

The mean total scores from the 6-month and 12-month postimplantation intervals were plotted alongside normative data established for the IT-MAIS by Kishon-Rabin et al22 in 2001 and Robbins et al13 in 2004 (Fig 3). The 6-month postoperative mean for the 18 children who received implants is 76.5%, which is ~25% above the mean for normal-hearing 6-month-old children. The 12-month postoperative mean for the 9 children who have reached that evaluation period is 87%, which is ~7% above the mean for normal-hearing 12-month-old children. The higher mean scores of the children who received implants may be reflective of the fact that before implantation, these children wore hearing aids and received some amount of auditory benefit.

**Speech Perception**

Objective postoperative speech perception data for patients 1 to 5 are shown in Fig 4. At the last scheduled evaluation, patients 1 to 4 were administered the MLNT, LNT, and CP. MLNT word scores ranged from 83% to 100%, and phoneme scores ranged from 95% to 100%. LNT word scores ranged from 84% to 97%, and phoneme scores ranged from 93% to 98%. CP test scores ranged from 60% to 100%. Patient 5 obtained a score of 100% on the Glendonald Auditory Screening Procedure word test.

All children are receiving early intervention services. Sixteen of the 18 children either communicate...
orally or are being taught using only oral communication, whereas 2 children use total communication. Of these 2 children, 1 has a deaf mother who uses sign language as her primary means of communication, and the parents of the second child prefer the total communication approach. Preliminary reports from therapists and teachers indicate that all children are developing speech and language skills with a natural-sounding voice. Children who have used the device the longest are progressing at a rate equal to that of their normal-hearing peers, whereas the others have emerging age-appropriate skills in both receptive and expressive language. To date, no differences are evident between children who are using oral communication and those who are using total communication, although it is premature to draw any conclusions at this time on the basis of this limited data set.

In summary, all children had full insertions of the electrode arrays without perioperative or immediate postoperative surgical complications. Patient 3 required revision surgery 20 months after initial implantation as a result of scalp breakdown and persistent infection, which necessitated device removal and reimplantation. Performance reached first implant levels shortly after the second stimulation and has continued to improve. Device programming was accomplished without difficulty and did not pose
any particular age-related problems, and all children are developing auditory and oral language skills.

DISCUSSION

Cochlear implantation was performed successfully in children who were younger than 12 months, and device programming was accomplished without difficulty using a combination of objective and subjective techniques. Auditory competence is emerging, and although receptive and expressive language skills were not the focus of this study, preliminary reports show most of the children developing linguistic behaviors. Because the median age of implantation of this study population was ~10 months and auditory age-appropriate behaviors as measured by the IT-MAIS were either nonexistent or minimal before implantation despite appropriate amplification and intervention, it can be hypothesized that the auditory gains made after stimulation were not attributable solely to maturation, although at this point in time there are limited comparative data available to substantiate this point. These results are encouraging, however, and may be indicative of enhanced communication capabilities. If surgery can be performed without increased risk and access to auditory stimuli can be provided before 12 months of age, then it is possible that severely to profoundly deaf children can avoid the gaps in auditory perception that require learning at a faster-than-normal rate to enable them to reach the age-appropriate levels.

Nevertheless, there remain issues that should be considered with implanting in children who are younger than 12 months. Surgeons need to be cognizant of the areas of increased risk, and programming audiologists need to have appropriate experience with the pediatric population. For instance, scalp and skull thickness is obviously less in the very young child, and it is usually not possible to recess completely the receiver/stimulator. This anatomic issue combined with a thin scalp flap has the potential to increase the incidence of scalp flap complications. Surveillance on magnet strength and scalp flap appearance by the programming audiologists and more frequent visits to the surgeon are indicated. Parents of very young cochlear implant recipients should also receive instructions regarding careful observation of magnet strength and scalp flap appearance. Our 1 case of scalp flap necrosis and breakdown over the receiver/stimulator edge was most likely the result of a lack of careful surveillance and that the child wore glasses. The earpiece of the glasses rested under the ear-level microphone, and both were in contact with the edge of the receiver/stimulator. The device became contaminated with S. epidermidis, a known biofilm-producing organism. Biofilms are responsible for diseases such as otitis media and play a role in numerous other diseases, including bacterial endocarditis. They also may contribute to an assortment of hospital-acquired infections, the starting places of which can include implants, catheters, and other medical devices. In this case, the device could not be sterilized with intravenously administered antibiotics or reoperation with device rotation. Explantation and reimplantation 3 months later became necessary.

Another major area of caution relates to the possibility that the child may have additional disabilities. It is often difficult in an infant to be aware of the presence of and/or diagnose learning disabilities, cognitive deficits, or other variables that potentially can hinder the ability of a child to make use of the auditory information being accessed via the cochlear implant. During preoperative counseling, clinicians need to address this possibility with the parents to ensure that expectations regarding the child’s progress are realistic. Not emphasizing that every child is different and that some children as a result of the potential presence of negative confounding factors may not achieve a high level of performance could lead to great dissatisfaction, frustration, and distress on the part of the parents. Having other disabilities would not preclude implantation but would certainly alter the level of expectation. Such circumstances could speak to the issue of waiting until a child is perhaps 12 to 18 months of age in hopes that other deficits would become more apparent, allowing for a more informed prognosis; long-term studies can be expected to shed light on these issues. There could be other extenuating circumstances. For example, patient 16 to date has shown the least progress of all of the children in this study. The mother passed away, the child is being cared for by relatives, numerous appointments were not kept, and the implant has not been worn on a consistent basis.

In additional, the implant team should be cognizant of the parents’ level of acceptance of the child’s deafness as well as their awareness of other options, including hearing aid use and manual language. The parents should have a level of comfort with their decision to implant and have realistic expectations. Facts regarding the wide range of results with implants and the factors that affect outcomes should be a standard portion of the preoperative information sessions with the parents to ensure informed consent to the best possible degree.

These data are preliminary; many questions remain unanswered and require long-term follow-up. For instance, does implantation in children who are younger than 12 months increase the possibility of short- or long-term postoperative complications? Does this early access to sound increase the chances of age-appropriate oral language and speech production development beyond what can be obtained with later implantation, especially implantation between 1 and 2 years of age? How does the overall performance of these children in academic and social areas compare with the performance of children who received cochlear implants after 12 months of age? Currently, there is no evidence that children who receive implants between the ages of 12 and 18 months are disadvantaged relative to implantation at a younger age; these issues require additional exploration over a long term. For now, however, we can say that implantation in children who are younger than 12 months can be performed safely and provides early and effective access to auditory stimuli,
leading to functional benefit above what can be obtained from conventional amplification.

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REFERENCES

**Cochlear Implantation in Children Younger Than 12 Months**

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