

Surgery and Functional Outcomes in Deaf Children Receiving Cochlear Implants Before Age 2 Years

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Objective: To examine the feasibility of cochlear implantation in children younger than 2 years regarding surgery and functional outcomes.

Design: Prospective study.

Setting: Tertiary pediatric cochlear implant center.

Patients: A consecutive sample of 12 children younger than 2 years at the time of cochlear implantation (8 boys and 4 girls). The cause of hearing loss was meningitis in 6 children and congenital in 6.

Interventions: Multichannel cochlear implantation using the Nucleus C124M (Cochlear Co, Sydney, Australia) device. Functional outcome was assessed using the Listening Progress Profile and the Categories of Auditory Performance.

Main Outcome Measures: Perioperative and postoperative surgical complications and functional outcome.

Results: Eight children had a completely patent cochlea.

Four children required a 3- to 5-mm drilling to reach the scala tympani because of ossification after meningitis. Full insertion was achieved in 11 patients; the other child received 18 electrodes. One patient had temporary facial nerve weakness; 2 others had wound edema and serous discharge that resolved with conservative management. In the longer term, 1 child experienced a single episode of acute otitis media; another had recurrent episodes of otitis media. Mean Listening Progress Profile scores increased from 1 to 42 and median Categories of Auditory Performance scores increased from 0 to 5 at 2 years postsurgery. Comparison with the scores in the 2- to 5-year group showed no significant differences. No significant tuning difficulties were experienced with all children.

Conclusions: Cochlear implantation is feasible in children younger than 2 years without significant surgical complications or particular tuning difficulties. Functional results 2 years after implantation were as good as or better than those of children who underwent implantation between ages 2 and 5 years.

Arch Otolaryngol Head Neck Surg. 2002;128:11-14

IN 1995 the US National Institutes of Health Consensus Conference on Cochlear Implants¹ lowered the age limit to 2 years for cochlear implantation. The major concerns in relation to implantation in younger children include: (1) the relatively small size of the skull and its growth in young children, with potential surgical difficulties of access and a possible increase in surgical risk (later complications of electrode displacement); (2) the high prevalence of otitis media in this age group and its subsequent related complications; and (3) difficulties in rehabilitation.

There is evidence² that insufficient auditory stimulation during critical periods of development leads to linguistic and communicative deficits. Thus, it seems logical that the earlier a child with hearing loss receives an implant the better the outcome. Therefore, the age limit of 2 years

for cochlear implantation may be unnecessarily restrictive. The aim of this study was to assess the feasibility of surgery, the postoperative complications, and the functional outcomes in children who undergo implantation before age 2 years.

RESULTS

SURGERY

The incisions used did not differ from those used in adults: an extended endaural or postauricular incision. The pericranium was raised with the flap, and after performing a cortical mastoidectomy, a bed (bony well) was drilled for the receiver-stimulator unit. There was considerable variation in the thickness of the skull, although exposure of the dura was necessary in 9 of 12 patients for the implant to be comfortably and securely countersunk in a bony well of ad-

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PATIENTS, MATERIALS, AND METHODS

A prospective study was undertaken of 12 children (8 boys and 4 girls) who underwent implantation before age 2 years. All children were profoundly deaf, and radiological assessment was undertaken using magnetic resonance imaging. Age at implantation ranged from 17 to 23 months (mean, 20.5 months). Duration of hearing loss ranged from 8 to 23 months (mean age, 17.9 months). The cause of deafness was meningitis in 6 children and congenital in the other 6. Characteristics of all the children studied are given in the **Table**. Children were assessed prospectively regarding surgical difficulties and postoperative complications, tuning difficulties, and functional outcome. Follow-up ranged from 4.0 months to 4.4 years (mean, 2.2 years).

Functional outcome was assessed using the Listening Progress Profile and the Categories of Auditory Performance. These outcome measures were chosen because they are age appropriate for these children and are sensitive monitors of the progress of audition in young children as early as 6 months after implantation.³⁻⁷ The results were compared with those of children who received an implant between ages 2 and 5 years. Statistical analysis was performed using the *t* test for unpaired observations for normal distributions and the Mann-Whitney test for nonparametric distributions.

equate depth. Posterior tympanotomy and cochleostomy anteroinferior to the round window were performed in the usual way. The facial nerve was not exposed in 11 patients, and in 1 patient the sheath of the nerve was exposed. In all children, the Nucleus CI24M device (Cochlear Co, Sydney, Australia) was used. After insertion of the electrode array, the receiver-stimulator unit was secured in its bed using a nonabsorbable polypropylene (prolene) or synthetic absorbable suture (Vicryl) suture. Additional fixation of the electrode array was achieved using cement at the posterior tympanotomy and synthetic polyester passed through holes drilled at the margin of the cortical mastoidectomy. Small pieces of muscle were then placed around the electrode array in the cochleostomy to effect a seal. Stapedius reflex levels were measured to obtain an approximate estimate of the upper comfort levels for the implant; integrity testing, electrically evoked brainstem responses, and neural response telemetry were also performed. Closure was in 2 layers using subcuticular suture Vicryl to avoid the need for suture removal. A mastoid dressing was applied overnight, and a reverse Stenvers radiograph was taken the following day to check the position of the device. Initial stimulation of the implant was undertaken after roughly 4 weeks.

Eight of the 12 children had a completely patent cochlea. In 4 children, a 3- to 5-mm drilling was needed for access to the scala tympani because of cochlear ossification. Full insertion was accomplished in 11 children; in 1 child, 18 electrodes were inserted. Nine children did not encounter any surgical complications. One

Characteristics of 12 Children Who Underwent Cochlear Implantation Before Age 2 Years

	Mode	Mean	Median	Range
Age at onset of deafness, mo	0	3.0	0.5	0-10
Age at implantation, mo	22	20.5	21.0	17-23
Duration of deafness, mo	22	17.9	19.0	8-23

developed facial nerve weakness after surgery (House-Brackmann scale II to III) that was treated with corticosteroid and antibiotic therapy and subsequently returned to normal (House-Brackmann scale I). Two other patients developed minor complications (edema and oozing from the wound) which were managed conservatively. During follow-up, 10 of 12 children did not encounter problems. One child had an episode of acute otitis media 6 months after surgery that resolved with antibiotic therapy, and the child did not experience any other infections for the following 3 years and 10 months (until the time of the study). The other child experienced recurrent episodes of otitis media that did not resolve with adenoidectomy and prophylactic low-dose antibiotic therapy, so a grommet was inserted 18 months after surgery. The child still wears the cochlear implant, showing substantial progress in auditory perception skills. No children experienced device migration or dislodgment of the electrode array.

POSTOPERATIVE FUNCTIONAL OUTCOME

The mean (median) preoperative Listening Progress Profile score was 1 (0). The mean (median) score increased to 28 (27) at 6 months, 38 (39) at 12 months, and the maximum of 42 (42) at 24 months. The respective scores in the group of children who received an implant between ages 2 and 5 years were 1 (0), 30 (31), 37 (40), and 41 (42).

Regarding the Categories of Auditory Performance (minimum score, 0; maximum score, 7), the median (mean) score before implantation was 0 (0.2). The median (mean) score increased to 4 (3.8) at 6 months, 5 (4.6) at 12 months, and 5 (5.6) at 24 months. The respective scores in the 2- to 5-year-olds were 0 (0.2), 4 (3.4), 4 (4.2), and 5 (5.0). Statistical comparisons between the 2 groups did not reveal any significant differences.

COMMENT

In the United Kingdom, approximately 220 children are born every year with hearing loss in excess of 95 dB, with an additional 80 children losing their hearing later, usually because of bacterial meningitis.⁸ Many of these children cannot derive benefit from hearing aids and therefore are candidates for cochlear implantation, which may help them develop considerable speech perception and speech production abilities.⁹⁻¹⁶ However, several factors affect when these children should undergo implantation.

Of great importance is the issue of neural plasticity. Results of animal studies¹⁷ suggest that initial auditory activation of the brain leads to establishment of an intricate network linking the primary auditory cortex with second-

ary, tertiary, and associated areas of the brain. During the first postnatal week in the white rabbit, dendritic length increases 4-fold, and at the onset of hearing there is substantial proliferation of dendrites and branches that may reach 200% of adult levels, followed by a more gradual loss to 100% of adult levels.¹⁸ Although a vertical orientation of dendrites is present at birth in the white rabbit, the onset of hearing is associated with an explosive elaboration of dendrites toward the pial surface.¹⁸ Deafness may have various effects on neuronal connectivity within the auditory pathway, and the changes in cochlear nucleus cell size in animals were correlated with duration of deafness.^{19,20} Removal of the cochlea in young mice, gerbils, and chicks results in severe transneuronal degeneration, whereas there is little or no change in adult animals.²¹ However, this degeneration does not seem to directly correspond to any features specific to auditory system development, and it was found to be similar in different developmental stages, suggesting that other general systemic factors besides neuroplasticity may cause the differential susceptibility in young animals.²¹ These experiments highlight the complexity of these issues and the fact that the biological mechanisms underlying such phenomena have not yet been defined.

Results of developmental studies²²⁻²⁵ in humans indicate that there are critical or sensitive periods during which the developing central nervous system can most readily use sensory information to form linguistic structure. These periods may vary according to the different elements of language: for phonetic factors, 6 months of fetal life through age 12 months; for syntax, up to age 4 years; and for semantics, up to age 16 years.²

In addition to the possible existence of critical periods for language learning, there is a complementary argument supporting the importance of young age at implantation. The rate of language development in profoundly deaf children with implants exceeds that expected in deaf children without implants,^{13,26} and the gap between chronological age and language age that usually widens over time in deaf children without implants remains constant after implantation.²⁷ Consequently, the earlier the implantation is performed, the smaller the gap will be between chronological and language age. Therefore, it is not surprising that young age at implantation is an important determinant of later functional outcome.²⁸⁻³¹ In these studies, data were limited regarding children who received implants before age 2 years; therefore, there is a compelling need for studies involving such children.

Progressive cochlear ossification after meningitis is a concern that has led many otologists to believe that such children should be "fast-tracked." Ossification can take place as early as 2 months after meningitis.³² When early scanning reveals development of ossification, early cochlear implantation should be considered. In the present study, 4 children had ossification, and in 1 child it prevented full electrode insertion. Age should not be a consideration in patients with rapid development of cochlear ossification; successful implantation has been performed in such cases even in children aged 6 months.

Worries have been raised about the size of the skull and its growth. At birth, the cochlea is already adult sized, but the temporal and parietal bones grow into adulthood.^{33,34} have shown that more than 50% of tem-

poral bone growth is completed in the first 2 years of life, but the skull continues to grow into the late teenage years. The facial recess does not demonstrate postnatal growth. However, the distance between the receiver site and the electrode entry point into the inner ear will be increased by an average of 12 mm; Dahm et al³⁵ recommended that the fixation of the implant in a young child allow up to 25 mm of lead wire lengthening. Electrode migration risk in the present series was reduced by anchoring the electrode array close to the cochleostomy. Data from a study by Hoffman and Cohen³⁶ using the Nucleus device show that electrode migration is the same for adults (1.17% of 3064 cases) and children (1.31% of 1905 cases), whereas the incidence of device migration is lower in children (0.05% of 1905 cases) than in adults (0.26% of 3064 cases). However, most children in the study by Hoffman and Cohen³⁶ were much older than 2 years at the time of implantation.

The issue of the prevalence of acute otitis media or otitis media with effusion in this age group, and subsequent complications related to infection, needs to be addressed. The evidence³⁷ is that the incidence of otitis media with effusion is lower in children with vs without implants, and the nonimplanted ear seems to be more commonly affected. Nonetheless, concerns that middle ear infections may spread to the inner ear have been expressed. Shepherd et al³⁸ reported that clinical experience and animal studies have shown that the implanted cochlea is able to resist the spread of infection nearly as effectively as the cochlea of a nonimplanted ear. There is, however, a vulnerable period during the healing process after implantation. Hoffman and Cohen,³⁶ in a review of 1905 children who received the Nucleus device, found 1 case (0.05%) of meningitis. Lenarz³⁹ reported episodes of acute and serious otitis media in children with implants, but all were successfully treated medically. He suggested that the cortical mastoidectomy and removal of large areas of mucosa in the mastoid reduces the risk of postoperative mastoiditis and inflammation.³⁹ In the present study, only 1 child encountered particular problems with otitis media. However, the recurrent episodes responded well to antibiotic treatment, and there were no episodes of meningitis or other complications of otitis media. The child continues to wear the implant and derives substantial benefit from it.

Surgical complications and the feasibility of surgery need recording. Our results reveal a low rate of surgical complications and provide further evidence that surgery in such young children is safe. However, our implantation center (Nottingham Pediatric Cochlear Implant Programme, Nottingham, England) has considerable experience in pediatric cochlear implantation; the number of children who have received implants approaches 300 (mostly children <5 years). This may have contributed to the low rate of surgical complications because the number of complications decreases when the number of patients who receive implants increases in a cochlear implantation center.⁴⁰

Literature comparing functional outcomes of cochlear implantation in children younger than 2 years vs older children is limited. Waltzman and Cohen⁴¹ reported similar or even better outcomes in children who received implants before age 2 years. However, at least 1 of the 2 tests used to assess children's progress (PBK monosyllabic word test) is designed for much older chil-

dren, and there are doubts about whether these tests are appropriate for such a young population, especially at the early intervals after implantation.

The present study used outcome measures designed for such young children and compared children who received implants before age 2 years with children who received implants between ages 2 and 5 years. However, it is a common weakness for outcome studies not to have the same numbers of children at all intervals, and these numbers usually decrease at subsequent follow-up. The present study was not an exception, although mean follow-up was 2.2 years. The results (means and medians) reveal that children who received implants before age 2 years showed considerable progress in auditory perception after implantation, matching the progress of children in the older age group. Although various difficulties may arise in tuning and rehabilitation regarding such young children, we did not experience any particular problems in these areas, perhaps because the professionals involved had substantial experience with young children. Moreover, the outcomes, at all intervals up to 2 years after implantation, were comparable with those in children who underwent implantation between ages 2 and 5 years. This becomes more important taking into consideration that younger children usually perform worse on auditory tests and scales at the first intervals after implantation and catch up and surpass the older children after 2 years of implantation.²⁸ Therefore, it is encouraging that children who received implants before age 2 years in the present series had no difference from the older children so early after implantation. It will be interesting to see if, in the long term, the younger children will surpass the older children with implants.

In conclusion, this study shows encouraging results in the cochlear implantation of children younger than 2 years. The feasibility of surgery has been demonstrated, with no significant complications and good early results of tuning and rehabilitation. Further long-term follow-up is required to demonstrate improved outcome of these children in comparison to older children undergoing cochlear implantation.

Accepted for publication August 24, 2001.

We thank the professionals (audiologists, scientists, teachers for the deaf, and speech and language therapists) of the Nottingham Cochlear Implant Programme for their valuable contribution in the assessment and management of the children studied.

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