Pediatric Cochlear Implants in Prelingual Deafness: Medium and Long-Term Outcomes

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Cochlear implants have revolutionized the rehabilitation of deafness in adults and in children. These implants include an electrical array that is implanted into the cochlea and directly stimulates the different fibers of the cochlear nerve, thereby bypassing the damaged hair cells. They are indicated in young patients with profound and severe hearing loss who do not benefit from conventional hearing aids, have no medical contraindications and do not harbor unrealistic expectations (neither the patient nor the parents). Cochlear implants in children were first performed in the late 1980s following the earlier positive experience in adults and have exceeded expectations. However, due to a wide range in short-term results, it was essential to define variables contributing to outcomes. During the early 1990s the factors found to be associated with good performance were: a) age at implantation and length of deafness, b) amount of daily use, c) mode of communication, and d) absence of other handicaps.

Deafness in children is commonly *prelingual*, which means that deafness occurred either congenitally or in the first few years of life, before speech had been fully acquired. Non-syndromic genetic deafness is the commonest congenital cause Another common cause is intrauterine infections such as cytomegalovirus. Acquired deafness before speech and language development is less common and includes bacterial meningitis and taking ototoxic medication.

The future of a prelingual deaf child, in terms of hearing, developing speech and integrating into the hearing society, depends not only on optimal amplification with a hearing aid or cochlear implant, but also to a large extent on the type of education he or she receives. In education based on *auditory-verbal* communication, the deaf person learns to use auditory cues optimally, by practicing and perfecting speech understanding and precisely articulating spoken language. This type of education aims at completely integrating the deaf person in the hearing society. In *oral* communication the deaf learn to hear and speak sign language with the aid of lip reading. This type of education also aims at integration in hearing society. *Total* communication teaches both oral and sign language in order to maximize the ability to communicate. *Manual* communication is not based on hearing but rather on manual sign language. Patients who sign commonly do not use amplification, and they typically belong to "deaf society."

When looking at the advances in outcomes of cochlear implant recipients during the last generation, it is important to appreciate the developments in the technical design of the implant. Since the coding strategies of implants have dramatically improved, there has been a significant increase in quantity and quality of auditory information. Upgrading the external part of the implant has enabled the patients who were implanted in earlier years to enjoy most of the new developments in coding strategies.

Since the early 1990s a clear benefit has been shown for cochlear implants in children. Hundreds of studies have reported on outcomes, with follow-up limited to 2–5 years. According to Summerfield and Marshall [1], the benefits of cochlear implants for profoundly deaf children may be defined in three time spans:

- Short term: 1–3 years, in the form of enhanced achievements in audition, speech and language
- Medium term: during the subsequent 5 years, enhanced educational attainments, greater social versatility and robustness, and increased quality of life
- Long-term: spanning at least 20 years from implantation and including greater social independence and quality of life in adulthood.

In this issue of *IMAJ*, Migirov and coauthors [2] publish the results of questionnaires completed by prelingual cochlear implant users on integration into Israeli hearing society when they grew up. Their findings, as well as other research on this issue, are discussed below under the heading Education and Employment. This editorial will focus on the outcome of medium and long-term benefits of prelingually deaf children with cochlear implants.

STUDY DESIGNS

Summerfield and Marshall [1] discuss study designs that would optimally assess medium and long-term outcomes of prelingually deaf children with cochlear

implants. There has been an increasing demand by health organizations/purchasers to adopt the elective procedures with the most rigorous evidence of effectiveness and cost-effectiveness. Randomized controlled studies are the ideal methodology for scientifically studying effectiveness of a medical device, yet no such study has compared cochlear implants with alternative devices (for example, a hearing aid). According to the authors, this may be explained by the very fast technological evolution of implants, which might produce outdated results, and the fact that such studies would span over many years.

Performing retrospective population-based, cross-sectional comparisons may prove a more practical method of assessing outcomes in this group of patients. This methodology is based on comparison of implanted children and non-implanted matched children. Among the many important variables that would have to be included are age, age at onset of deafness, implant duration, degree of hearing, the quality of health and rehabilitation, degree of parent commitment, etc. In such a way material benefits, effectiveness and cost-effectiveness could be examined. Naturally, the strength of the study would depend on the success or failure in performing the matching process.

OUTCOMES

In the next part we will review the medium and long-term results of cochlear implants performed in pediatric prelingual patients, based on available studies, according to the following subjects: auditory and speech development, education and employment, non-usage and device failure, and reimplatation.

AUDITORY AND SPEECH DEVELOPMENT

Open-set word and sentence recognition are the most common tests performed for assessing outcomes of hearing, following cochlear implantation. Presurgery, prelingual children who are implanted in the first few years of life commonly score 0%, a result that reflects their profound hearing loss and lack of ability to recognize speech, even when well fitted with hearing aids.

Spencer et al. [3] reported on a cadre of their first pediatric prelingual implants (1987 to 1995), with a mean 10 years follow-up. Patients scored 70% on word recognition and 68% on sentence test accuracy. Seventy-nine percent of patients correctly recognized phonemes (two syllable-balanced words) with a high correlation between hearing and speech.

Waltzman and collaborators [4] studied 81 prelingually deaf children with a follow-up of 5–13 years. The age of implantation ranged from 13 months to 15 years (mean 4.2 years). Postoperative open-set word and sentence recognition after 5 years were 81% and 94%, respectively, with a statistically significant improvement between post- and preoperative scores.

Beadle et al. (5) studied 30 children at 5 and 10 years after implantation. They used the CAP index (Category of Auditory Performance) with a scale of 0 to 7. The score -0 implies no awareness of environmental sounds, and 7, the ability to talk on the telephone with a familiar speaker. Scores improved from 0 at pre-implantation to a mean category of 6 at 5 years, and 7 at 10 years. The study also examined SIR (Speech Intelligibility Rating), with 1 implying unintelligible speech, usually using manual communication, up to a score of 5 = intelligible speech. Close to half the patients scored 4 and 5 (intelligible speech for an average listener). Median speech intelligibility rating was 3 at 5 years, and 4 at 10 years, also demonstrating that patients' speech continues to improve between 5 and 10 years.

Uziel et al. (6) thoroughly studied the 10 year outcome of 82 children. Seventy-nine percent reported that they could use the phone. The mean open-set word score was 72% and a 44% word recognition in noise. Two-thirds of patients scored a speech intelligibility rating of 4 and 5, while 76% of the group scored significantly lower than their hearing peers.

EDUCATION AND EMPLOYMENT

Ideally, an implanted child would be mainstreamed in regular classrooms and would use *auditory-verbal* and *oral* communication. Reports show that the majority of cochlear-implanted children are in fact educated in mainstream schools. However, this may not necessarily mean that the child or adolescent is attending a normal-hearing class and he or she may require significant additional assistance.

Certification from a mainstream high school is a condition for attending college or university in most countries. Accordingly, attending a school for the deaf, whether the individual underwent a cochlear implant or not, will usually exclude him or her from higher learning and, consequently, from better employment possibilities. Cochlear-implanted children may shift from mainstream education to special education. Such individuals were observed to enroll in apprenticeships and became employed in blue collar jobs [7].

Spencer and team [3] reported the series of their first pediatric prelingual implanted children (1987 to 1995), with a mean 10 year follow-up. All patients attended a regular public school and assisted with a sign language interpreter. Seventy-five percent of patients attended a post-high school institution, and the scholastic achievements in 10th grade were at least as good as their hearing peers.

In the series of Waltzman [4] and Beadle [5], approximately 75% of the implanted children used oral communication. According to Huber and co-researchers [7], 60% of 15 patients who completed a mainstream high school education used oral communication. After 10 years follow-up, of 19 children in compulsory and secondary school, a third attended school for the deaf, a third were mainstreamed in regular classes, and a third were in units for the hearing impaired attached to a mainstream

school [5]. In this study two-thirds of these adolescents managed well with oral communication, with occasional help, and the rest used *total* communication. Some of the patients were engaged in college education, even when using *total* communication, with variable success.

Huber et al. [7] found that 40% of patients who completed high school received advanced education, which was similar to their normal-hearing peers. Although almost all graduates of secondary school and college were employed, when compared to normal-hearing controls, cochlear implant users had significantly less correspondence between career aspiration and their actual occupation. This study also looked at the career prospective from the parents' view. Parent's expectations of their children were significantly less optimistic (26%) than those of normal-hearing children (73%).

In this issue of *IMAJ* Migirov et al. report their results of questionnaires for cochlear implant users. Eighteen patients were studied at a mean of 7 years after receiving their cochlear implants. Fourteen used oral communication and 4 used total communication. Remarkably, all but one were either employed or enrolled in military service, seminary (yeshiva), high school or university. Since the response rates to the questionnaires were 60%, the authors comment that the responders may represent the more successful cochlear implant users.

NON-USAGE

Successful cochlear-implanted patients would be expected to use their implants during all hours of activity. Naturally, non-use is regarded as a waste of an expensive resource and every effort should be made to avoid this situation. Use/non-use is not an all or nothing phenomenon, since when thoroughly questioned, patients may volunteer that they only wear the implant to school or when they want to listen in social situations [5]. Complete non-use was reported by 3–5% of patients [4,5,7]. Interestingly, most of the non-users were older than 10 when implanted [4].

DEVICE FAILURE AND REIMPLATATION

Reimplatation is required in cases of device failure following local infection or device extrusion. Waltzman et al. [4] report a failure rate of 11% after 5–13 years, mostly due to device failure and to device extrusion. Uziel and colleagues [7] reported that 13.4% of their patients required reimplantation after 10 years: one case was due to local infection and the others to device failure. Beadle [6] reported a relatively high rate of 27% device failures after 10 years. Reimplatation is considered highly successful by these authors.

SUMMARY

Unfortunately, no large-scale, well-designed, comprehensive studies on medium and long-term effect of cochlear implants in prelingual children exist. Furthermore, the few studies listed have addressed different aspects of this issue, in a non-standardized manner. With the question of outcomes becoming so important in modern medicine, standardized reporting methods are essential. This would enable a fair comparison between the different commercial devices and between rehabilitation-education methods.

The studies presented above were published between 1999 and 2008. Due to the long follow-up period required for reporting medium and long-term outcomes, they actually represent patients implanted one and two decades ago. At present and in the near future, we may expect better results from patients who were implanted in the last few years for several reasons: a) superior cochlear implants with better coding strategies; b) developments in rehabilitation and higher awareness among parents and staff; c) better health care and universal screening programs, leading to implants being performed in an earlier age; and d) a high rate of performing bilateral cochlear implants (either simultaneously or sequentially) in the last decade, which accomplishes even better results compared to unilateral implants.

From the studies reviewed above, mainstreaming the child who has auditory-verbal and oral communication is the preferred educational setting for maximizing the medium and long-term benefit from a cochlear implant. Hearing and speech skills continue to improve many years after the implant. Non-use and failure rates (as reported by the authors from the medical centers and not solely by the cochlear implant manufacturers) are low, ranging from 1% to 2.7% per year. Overall, patients have a high rate of employment, close to that of the general population. However, they may be less satisfied, as the individual and the parents may feel compromised by their communication skills.

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