

REVIEW ARTICLE

Cochlear Implants in Children—A Review

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Over the past two decades, cochlear implantation has become a widely accepted treatment of deafness in children. Over 20,000 children have received cochlear implants worldwide. Hearing, language and social development outcomes have been positive. We review current issues in cochlear implantation, candidacy, evaluation, surgery, habilitation, ethics and outcomes. *Key words:* cochlear implant, deafness, ear surgery, hearing, rehabilitation.

INTRODUCTION

Historical records describe the treatment of deafness among ancient and primitive tribes through the placement of minerals or plant extracts into the ear canal (1). Although medical interventions for conductive hearing impairment progressed significantly over the years, treatment for sensorineural deafness remained essentially no more effective than these early attempts until the last quarter of the twentieth century. It has only been in the past 20 years that physicians have been able to successfully treat profound sensorineural hearing loss with cochlear implants (CIs). These electronic prosthetic devices are introduced surgically through the mastoid and middle ear into the inner ear, directly stimulating the auditory nerve in response to sound.

Cochlear implant technology has been characterized by rapid and continuous evolution since the development of the first clinical devices in the early 1980s. Single-channel devices quickly gave way to computer-based multichannel devices. For several years emphasis was placed on sound processing and the development of improved speech coding strategies, the paradigms used by the device to reproduce the speech signal as an electrical stimulation pattern. External equipment became smaller and more versatile. By the early 1990s implant devices were able to offer the user options in terms of speech processing strategies. Multiple memory slots allowed the user to judge the efficacy of different strategies in his/her normal environment, and even to select different strategies for different listening situations. Most recently, attention has turned again toward internal electrode design. The goal has been to develop internal arrays that lie closer to the modiolus and cause less insertion trauma. These electrodes run complex programming strategies with less power, opening the way for ear-level and ultimately fully implantable devices.

Reflecting the improved capabilities of the technology, patient candidacy has expanded over time. Most candidacy selection criteria, such as age, degree of benefit received from hearing aids, medical and radiological status, have become less stringent, increasing the potential candidate base. It can be expected that advances in genetic testing for deafness will soon be used not only for diagnostic purposes but also to direct treatment and management options, including candidacy for a CI.

Advances in technology have led to a continuous improvement in cochlear implant performance and, although current-generation implants do not restore normal hearing, recipients are generally able to function at a level comparable to that of less hearing-impaired successful hearing aid users. The majority of adult implantees are able to conduct interactive telephone conversations and prelinguistically deaf children have demonstrated the capacity to develop language at a rate equal to that of their hearing peers (2). However, long-term habilitation continues to be essential for children after cochlear implantation. The degree of success achieved in children is highly correlated with age at implantation and effective use of auditory habilitation. Early training in signed languages, once theorized to enhance later performance with CIs, is now shown to be associated with poorer linguistic outcomes (3).

INDICATIONS FOR CANDIDACY

Indications for cochlear implantation in children have expanded significantly in the past decade as remarkably positive outcomes have been documented. The minimum age for implantation has decreased from 24 to 12 months while the maximum degree of hearing aid benefit acceptable for pediatric implant candidates has increased. Experience has shown that children implanted at a younger age

perform better than children implanted when they are older. Waltzman and Cohen (4) have shown that children implanted below the age of 2 years achieve open-set speech recognition equal to or faster than children implanted above the age of 2 years. Furthermore, children with "aidable" residual hearing often perform better with implants, and those with some measurable open-set speech recognition ability prior to implantation perform better with CIs than children without residual open-set word recognition (5, 6).

Medical and radiological criteria have been expanded to include children with cochlear dysplasia, multiple developmental delays and certain systemic medical conditions (7). Therefore, it is important that each child be considered individually by an experienced cochlear implant team consisting of an otolaryngologist, audiologist, a rehabilitation and educational professional and others as needed.

Criteria for pediatric implantation that have not changed include family commitment and appropriate expectations of the family and the educational setting. In older children and teenagers, especially those who are manual communicators, the child's motivation, in addition to parental desires, should be evaluated.

Current implantation criteria for children are summarized in Table I. Absolute contraindications include agenesis of the inner ear (Michel deformity), absence of the cochlear nerve and systemic illness precluding anesthesia or surgery.

EVALUATION OF CANDIDACY

Medical evaluation

Evaluation should include assessment of the patient's health and ability to undergo general anesthesia. A complete medical history and physical examination should be performed along with appropriate laboratory tests and imaging studies. Ophthalmological referral should be obtained in view of the increased incidence of visual disorders in deaf children (8). If a

family history of sudden cardiac death is elicited then electrocardiography should be carried out in order to exclude cardiac abnormalities associated with the Jervell-Lange-Nielson Syndrome (2). As previously mentioned, selected people with systemic illness, including those with autoimmune disorders and diabetes mellitus and immunosuppressed patients after organ transplantation, may safely be implanted after careful, individualized medical evaluation.

Imaging

CT findings are commonly used to assist in ear selection and to allow the surgeon to plan ahead in unusual cases. CT assessment should include mastoid pneumatization, thickness of parietal bone in the area of the seat, facial nerve, facial recess, cochlear anatomy, round window, vascular anatomy (carotid artery, sigmoid sinus, emissary veins), cochlea and vestibular aqueduct and internal auditory canal.

Adequate imaging allows the surgeon to assess inner ear morphology, including cochlea patency. Cochlear abnormalities should be identified preoperatively as specifically as possible in order to assist in surgical planning and postoperative outcomes counseling.

MRI is indicated when internal auditory canals (IACs) are <1.5 mm, in order to demonstrate the presence of the cochlear nerve, or when there is questionable ossification of the cochlea. In such cases, T2-weighted images will demonstrate loss of the endolymph/perilymph signal.

Audiologic evaluation

Audiologic evaluation is the primary means of determining candidacy for cochlear implantation. It is important to be able to obtain ear-specific auditory information under both aided and unaided conditions. In prelinguistic children, it may not be possible to obtain this information without therapeutic intervention (diagnostic therapy) and repeated visits to the implant audiologists (2). Determining the benefit provided by traditional amplification to preverbal children may require time and expertise beyond the capabilities of many inexperienced or understaffed clinics, but is a necessary component of candidacy evaluation.

Assessment of parental expectations and support, hearing aid history and compliance with the therapy process are all important aspects of the evaluation of the young child. If cochlear implantation is to be maximally successful, auditory-oral, traditional oral or total communication education, with a strong emphasis on oral communication and auditory development, are essential.

Table I. *Current selection criteria for pediatric cochlear implantation*

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|--|
| 1. Age \geq 12 months (unless ossifying) |
| 2. Severe-to-profound bilateral sensorineural hearing loss |
| 3. Benefit from hearing aids less than that expected from cochlear implants |
| 4. No medical contraindications to undergoing general anesthesia |
| 5. Family support, motivation and appropriate expectations |
| 6. (Re)habilitation support for development of oral language, speech and hearing |

Table II. Considerations for selecting the ear to be implanted

Hearing

Choose better ear

- Anacusis opposite ear
- Disuse > 10 years opposite ear
- Neither ear useful

Choose worse ear

- Good residual in both ears

Imaging

Absolute contraindications

- Aplasia
- Absent auditory nerve

Choose better ear

- Ossification
- Dysplasia
- Facial or vascular anomaly
- Pneumatization

Medical

Choose better ear

- Recurrent otitis media
- Prior ear surgery
- Cerebrospinal fluid

Other

Timing

- Most recently deafened

Language

- Choose right ear

Ear selection

Factors used to determine the ear to be implanted are shown in Table II. In general, the more an implanted ear has been used, the better the eventual outcome of implantation. For this reason, it is preferable to implant an ear which has some residual hearing and which has been used with amplification in the recent past.

SPECIAL CONSIDERATIONS IN CHILDREN*Very young children*

Implanting children below the age of 24 months has several potential benefits, but also raises issues of accurate diagnosis, surgical technique and outcomes. During the language-sensitive first 3 years of life, hearing loss has been shown to cause central auditory system abnormalities in experimental animals and auditory perceptual disorders in children (9, 10). In very young children, it is also known to delay the development of language and speech and lead to negative social and educational consequences (2).

Owing to this narrow window of opportunity, timely and accurate hearing evaluation of very young children is another important consideration. While objective measures such as evoked potentials can help determine auditory thresholds, these measures are of limited use in evaluating the benefits provided by

amplification. Careful behavioral testing for functional gain and newer methods such as diagnostic therapy are essential in assessing benefit from amplification. Diagnostic therapy is performed by a therapist well versed in prespeech communicative behaviors and auditory milestones.

The therapist observes the young child's performance with amplification over weeks or even months. An essential element is parental involvement, overseen by the therapist, to ensure constant at-home follow-through of therapy procedures. Absence of appropriate progress while participating in such therapy is a strong indication of inadequate auditory input. Newer instruments, such as the Infant-Toddler Meaningful Auditory Integration Scale (ITMAIS), are also useful for obtaining information from parents regarding the young child's responses to sounds in everyday listening situations (2).

Surgical technique in children 12 months of age is very similar to that in older children. Anesthetic precautions regarding body temperature and fluid management are critical.

Acute otitis media

Children receiving CIs were once considered to be at risk from middle ear infections due to the incidence of acute otitis media (AOM) in this age group. This could theoretically create a problem due to potential spread of infection into the cochlea along the electrode array.

Clinical studies have demonstrated that the prevalence and severity of AOM are not increased by cochlear implantation (11). In potential implant candidates, recurrent AOM should be treated aggressively with antibiotics with or without the use of ventilation tubes to avoid inordinate delay in evaluation and implantation of the otitis-prone child. Once recurrent AOM or otitis media with effusion are controlled, implantation may proceed, even with a ventilation tube in place (11). Oral antibiotics are effective in the treatment of post-implantation AOM and there has been no report of increased incidence of labyrinthitis or meningitis in this group of children (11).

Cochlear dysplasia

Malformation of the inner ear identified by CT was once considered a contraindication to cochlear implantation due to concerns about electrode insertion, electrical stimulation pathways and abnormal tonotopic organization of the spiral ganglion. However, clinical studies have found that patients with minor anomalies (such as an enlarged vestibular aqueduct or Mondini dysplasia) may be implanted with standard techniques and hearing results are ap-

proximately equal to those with normal CT scans (12). Patients with major anomalies (common cavity) require specialized surgical techniques and outcomes can be variable (2, 12). The cochleostomy incision is made in the supero-lateral aspect of common cavities and lumbar drains are not indicated (12).

In general, surgical outcomes are good in the great majority of these cases (12). However, the surgeon must be acutely aware of the potential for anomalous facial nerve position, cerebrospinal fluid gusher and potential misplacement of the electrode into the IAC (13). A facial nerve monitor should always be used as an adjunct, but especially in these cases.

Auditory neuropathy

Children with auditory neuropathy (AN) may have severe-to-profound hearing loss with absent auditory brainstem response, accompanied by the paradoxical presence of otoacoustic emissions and/or cochlear microphonics. Cochlear implantation of children with AN has been efficacious. Implanted children have shown improved listening and communication skills that have enabled them to take advantage of different communication and educational options (14). While the precise pathophysiology of AN is not known, the effectiveness of CIs suggest that it is an isolated inner hair cell disorder.

Post-meningitic deafness and ossified cochlea

Bacterial meningitis is the leading cause of acquired deafness in children. Histopathologic studies on human temporal bones in ears deafened by meningitis reveal a marked reduction in spiral ganglion cells and frequent ossification of the cochlea (15).

In our experience, post-meningitic children with cochlear ossification can be implanted with a variety of techniques and electrically stimulated with generally good outcomes (16). However, these children may require higher stimulation levels and more frequent updates of their programming over time compared to other patients (17).

Revision surgery

The most common indication for reimplantation is device failure. Less frequent indications include upgrading from a single-channel to a multichannel device, infection and flap breakdown (18).

In revision surgery, hearing outcomes are as good as or better than results with the initial CI, with no significant complication rate (13). Following reimplantation, mean length of insertion, number of channels actively programmed and speech recognition scores were at least as good as findings before initial implant failure (18).

Ethical considerations

Organized attempts to restrict the use of CIs in children occurred throughout the 1990s. Certain professionals expressed concerns that CIs did not work well enough to justify the surgical risks. Others, especially those involved with deaf culture, contended that if a large number of deaf children received CIs then deaf society as it currently exists would be diminished (19). However, >90% of deaf children are born to hearing parents (2). For these families there is often a strong desire for the child to hear and speak and to be fully part of the family. Traditional medical ethics gives parents the responsibility to make decisions on behalf of their children. Their decisions should be based on sound research and determination of the best interests of their child rather than the concerns of special interest groups (19). As experience and outcomes with cochlear implantation continue to be safe and successful, many deaf organizations, such as the National Association of the Deaf and Gallaudet University in the US, are moderating their opposition to the implantation of children.

SURGERY

Experience with CI surgery has led to the evolution of surgical techniques that simplify the procedure (20) and reduce complications (13). The physical characteristics of the implantable electronic package and electrode array vary among devices, requiring device-specific surgical techniques in order to minimize complications.

Under general anesthesia, using routine aseptic methods and facial nerve monitoring, cochlear implantation usually takes 1.5–2.5 h and patients are discharged either on the day of surgery or on the following day. Surgery in the pediatric age group requires particular attention to the higher incidence of anomalous structure, the facial nerve exiting the stylomastoid foramen, delicate tissues and small dimensions. Minimally invasive surgery using a 3–4 cm retroauricular incision is performed, an anteriorly based pericranial flap is developed and a skin/subcutaneous tissue pocket is elevated posteriorly (Fig. 1). The site of the template is identified on the skull, providing it is posterior enough to allow use of an ear-level processor, and the appropriate sized seat is drilled. In very young children, it may be necessary to place the receiver-stimulator directly on an “eggshell”-thin layer of bone over the dura. A complete mastoidectomy, posterior tympanotomy and cochleostomy are performed. The electrode array is inserted into the scala tympani through the cochleostomy incision. This incision is packed with muscle

or fascia and the receiver-stimulator is fixed to bone with non-absorbable sutures. The flap is closed over the device.

The first postoperative visit is at 1 week to check wound healing; \approx 2–3 weeks later, the fitting and mapping of the speech processor begins.

Special surgical considerations

Malformed cochlea. In malformations consisting of incomplete partition (Mondini malformation), enlarged vestibule and dilated vestibular aqueduct observed on CT, routine cochleostomy and insertion of the electrode are accomplished. Ears with any form of dysplasia are at risk of cerebrospinal fluid leak through the cochleostomy incision. Adequate packing within the scala is important to prevent complications. Lumbosacral drains are not advised (20).

In common cavity deformities, the modiolus is absent and the facial nerve may run in the lateral wall of the presumptive cochlea. The electrode array may be inserted into an opening created in the lateral recess of the common cavity, approximately corresponding to the expected position of the ampullated limb of the lateral semicircular canal (12). Insertion in this area is performed in order to avoid the abnormal facial nerve. Intraoperative imaging or endoscopy of the cochlea may be used to confirm the electrode position.

Facial nerve monitoring is highly recommended in children with congenital anomalies as atypical routing of the VIIth nerve may be expected. Electrode stabilization with the split-bridge technique is helpful in these cases (21).

Ossified cochlea. If obstruction is limited to the inferior segment of the cochlea (the category of ossification most frequently seen following meningitis), the obstruction may be removed, or a tunnel may be

created through the obstruction to reach an open lumen within the cochlea (22). This technique is successful if obstruction is limited to 8–10 mm as measured from the round window membrane. The new bone formation will be recognized as being less dense and of a lighter color. Following this new bone anteriorly leads the surgeon to a patent scala tympani. In these cases complete electrode insertion is possible and outcomes are equivalent to those of implanted patients with excellent outcomes.

In cases where obstruction extends apically into the ascending segment, the electrode may be inserted in the scala vestibuli by widening the cochleostomy superiorly. Electrode insertion into the scala vestibuli is technically straightforward and is associated with good hearing outcomes. If the scala vestibuli is also obstructed, complete electrode insertion is possible with drill out procedures, either canal wall down as described by Gantz et al. (23) or canal wall up as modified by Balkany et al. (24).

Patients with compromised healing. Patients receiving CIs who have impaired wound-healing capabilities because of immunosuppressive medications or underlying medical conditions are considered to be at increased risk of postoperative infection. However, cochlear implantation of patients with liver and renal transplants, sickle cell disease or reduced healing capabilities has been shown to be safe and effective (7). When considering these patients for cochlear implantation, it is important to consider the specific nature of their health problems as well as the overall medical and social situations of the patients in order to determine if the routine travel, postoperative visits and long-term rehabilitation required are possible.

Habilitation

Children with limited language abilities and limited sound experience present major challenges to pediatric implant teams. Unlike adults, who can report problems with the system, most very young children either do not recognize malfunctions of the equipment or have no way of describing the problem.

As the age of implantees has decreased, the need for more innovative methods of programming and patient management has increased. New technology allows listening through the CI microphone in order to test sound quality; audible alarms indicating low battery power also assist parents and implant teams; telemetry measures provide assurance that no short circuits exist in the array and that the device is working; and objective programming can be performed intraoperatively or as part of the programming process to confirm auditory stimulation and to establish a starting point for behavioral programming (25).

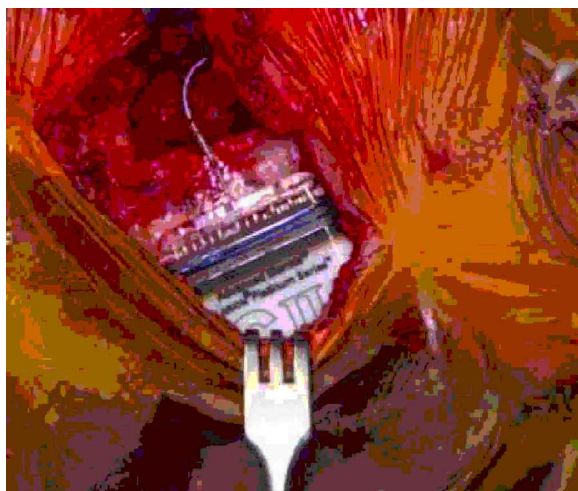


Fig. 1. Minimal incision surgery with implant receiver/stimulator in position.

A recent addition to objective programming is measurement of the electrically evoked whole nerve action potential by means of neural response imaging (NRI) or neural response telemetry (NRT) (26). This measure has the benefit of requiring no external electrodes, is determined directly through the implant system, and can be obtained even if a child is moderately active. NRT responses can be used to confirm auditory stimulation and can be used as a starting point for behavioral programming. Another objective measure that has been shown to be useful in programming young children is the electrically elicited middle ear muscle reflex (EMR), a form of the acoustic or stapedial muscle reflex elicited through a CI, which is highly correlated with desired maximum stimulation levels (27–29). Finally, electrically elicited auditory brainstem responses (EABR) can be recorded either intraoperatively or postoperatively in a cooperative subject, and can be used in a manner similar to NRT (26).

The degree of success in long-term habilitation achieved with CIs is more highly correlated with early age of implantation and habilitation than with the brand of CI used. In children, maximum benefit in both speech perception and language development is achieved when the child participates in a strongly auditory, orally based and family-centered therapy situation (3).

Outcomes

There has been continuous improvement in CI performance over the past 20 years. While younger age at implantation is generally associated with better hearing outcomes, children > 5 years of age have been shown to benefit from cochlear implantation. This benefit is greater than that obtained with hearing aids, but less than that obtained by children implanted at a younger age (2).

Although wide variability exists, in most children CIs and oral therapy promote the development of spoken language beyond what could previously be achieved with hearing aids. Implanted children learn language at the same rate as hearing children of similar hearing age. However, delays in development in place prior to implantation may not be recovered in all cases (3). Such handicaps can be minimized by early implantation. Good speech recognition abilities are positively associated with development of good speech intelligibility 2–5 years after implantation and the speech of implanted children has been found to show continued improvement beyond 5 years (2).

Another measured outcome, and an area of concern often expressed by opponents of pediatric CI, is the long-term psychological effect of cochlear implantation on both the child and family. It has been

argued that CIs are a manifestation of an inability of the parents to accept the child as deaf, thus forming the child's self-image as a defective individual. However, as longer-term global outcomes become more widely available, cochlear implantation has been shown to be associated with long-term psychological benefit, including improved interactive skills with both adults and other children (25).

DISCUSSION

CI technology continues to evolve, resulting in enhanced hearing, speech and cost-effectiveness for children. Indications for implantation of children have also expanded, including consideration of candidates as young as 12 months of age and those who demonstrate limited benefit from traditional amplification.

Several developments are currently being tested clinically and may become widely used in the next few years. Binaural cochlear implantation has been used in children. It is anticipated that in the future the two implants will be integrated and will share a stimulation program to minimize channel interaction and improve hearing in noisy environments and localization of sounds. Development of perimodiolar electrodes, implantable microphones and rechargeable batteries promise fully implanted devices in future.

Implantation of children and adults with residual hearing requires preservation of existing neural elements. One solution may be a hybrid electro/acoustic stimulator (a short, atraumatic electrode coupled to an implantable hearing aid). In these experimental devices, severe-to-profound high tone losses are treated electrically and mild-to-moderate losses are treated acoustically.

Another area of current development is neural preservation in association with implantation. Neurotrophins, molecular genetic techniques and apoptotic pathway blockers, delivered either preoperatively or through CI electrodes, are all under study. If effective, cochlear implantation may become indicated for individuals with moderate-to-severe hearing loss, and assume the role of implantable hearing aids.

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