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Neurosurg Clin N Am 19 (2008) 317–329

NEUROSURGERY
CLINICS
OF NORTH AMERICA

Cochlear and Brainstem Implantation

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Cochlear and auditory brainstem implants offer safe and effective hearing habilitation and rehabilitation for profoundly deafened adults and children. Brainstem implant technology is currently approved for use in patients with neurofibromatosis type 2, who have lost integrity of auditory nerves following vestibular schwannoma removal. An update on implant devices, speech processing strategies, candidacy criteria, and perceptual performance are provided in this article.

Cochlear implants

Cochlear implantation is an established treatment for selected individuals with bilateral severe to profound sensorineural hearing loss (SNHL) who derive limited benefit from conventional hearing aids. The first cochlear implants, developed in the early 1960s, comprised single electrodes that were surgically placed within the scala tympani, in an effort to electrically stimulate the auditory nerve in patients with absent or dysfunctional cochlear hair cells. These early devices restored some degree of sound awareness to recipients and, in many cases, facilitated lip-reading far better than their hearing aids had. The introduction of multichannel devices in the early 1980s, development of advanced speech coding strategies, and refinement of candidacy criteria have led to substantial improvements in postimplant performance, evidenced by improved open-set speech understanding in both children and adults.

This article provides an update on issues related to cochlear implantation, including device design, speech processing strategies, candidate selection, surgical technique, and perceptual performance.

Implant device

Basic components

All cochlear implant systems possess an externally worn device and an implanted internal component (Fig. 1). The external hardware consists of an ear-level microphone, an ear-level or body-worn speech processor, and a transmitter placed behind the ear. The internal component consists of a receiver–stimulator, linked to an intracochlear electrode array via a lead wire. Some implant devices have a second electrode which serves to ground the stimulating electrode. Sound received by the microphone is transduced into electrical signals, which are filtered, analyzed, and digitized by the speech processor and forwarded to the transmitting coil. The encoded signals are then delivered to the implanted receiver–stimulator by radio-frequency electromagnetic induction. This signal is reconverted to an electrical signal, which is then delivered to the implanted electrode within the scala tympani. Current applied to the electrodes radiates into the fluid of the scala tympani, spreads through the habenula perforata of the osseous cochlear modiolus, and stimulates the auditory nerve.

Electrode design

The design of the electrode array differs in the presently available commercial implants. Four implant systems have been FDA approved for

This article originally appeared in Otolaryngologic Clinics of NA: volume 35, issue 2, April 2002; p. 325–42. It has been updated to reflect recent advances in implant technologies.

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1042-3680/08/$ - see front matter
doi:10.1016/j.nec.2008.02.014

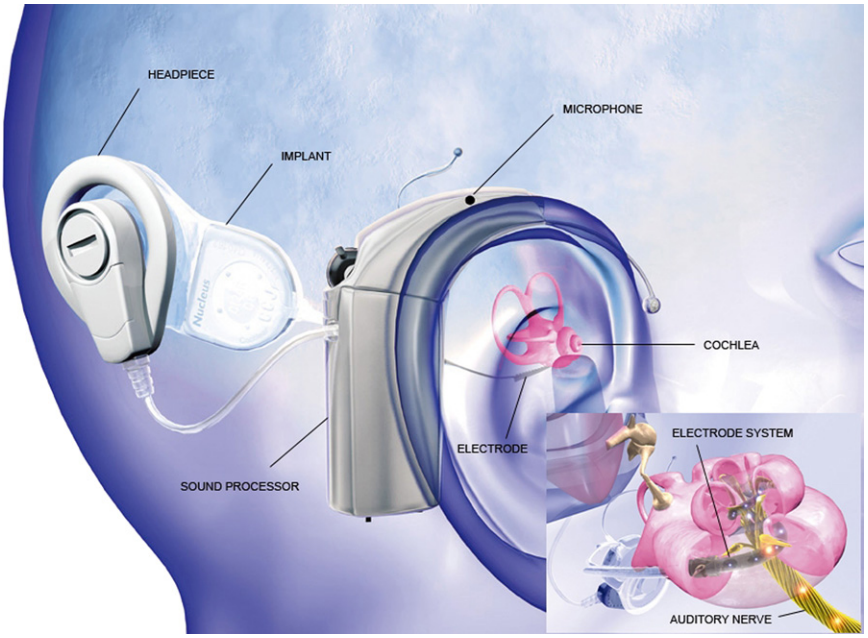

Fig. 1. Essential components of cochlear implant system (Cochlear Corp., Englewood, CO).

use in adults and children in the United States (the Nucleus Freedom Implant [Cochlear Corp., Englewood, Colorado], the HiRes 90K Implant [Advanced Bionics Corp., Sylmar, California], and the PULSAR CI100 and SONATA TI100 Implants [MED-EL Corp., Innsbruck, Austria]). There has been a trend in the past decade toward implantation of modiolar-hugging electrode arrays. The closer proximity of these electrode arrays to spiral ganglion cells offers theoretic advantages of improved sound quality, speech recognition, and power efficiency [1].

The Nucleus Freedom Implant system currently uses the Contour Advance electrode with Softip (Fig. 2). This consists of a 25mm long precurved modiolar-hugging electrode with 22 platinum electrode contact plates held in a straight position with a soft platinum wire stylet. The electrode tip comprises a conical tapered silicone elastomer designed to improve the insertion characteristics of the original Contour electrode and minimize tip fold-over during the insertion process (Fig. 3). The Contour Advance with Softip electrode inserted using the Advance Off Stylet (AOS) technique has been shown to significantly reduce trauma to the intracochlear structures during he insertion process [2]. Using the AOS technique, the electrode is inserted until its tip reaches near the back of the basal turn of the cochlea, then is advanced off the stylet (Fig. 4). A marker on the outer surface of the electrode (11 mm from the tip) delineates the insertion point at which the AOS technique should begin. As with earlier generation Nucleus implant systems, the Freedom Implant includes a ground or reference electrode, allowing monopolar stimulation of all 22 electrodes in the array, thus reducing power consumption.

Similar to the Nucleus Freedom implant, the HiRes 90K implant is housed in a titanium case with a removable magnet and telemetry coil attached and encased in silastic (Fig. 5). The device comes with 2 electrode options. The HiFocus Helix electrode is 24.5 mm long and consists of 16

Fig. 2. Nucleus Freedom implant (Coclear Corp., Englewood, CO).

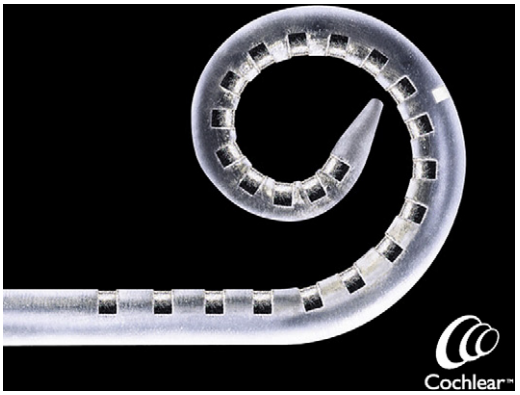

Fig. 3. Advance Contour electrode with Softip (Cochlear Corp., Englewood, CO).

planar platinum-iridium contacts arranged along the medial surface of the silicone electrode array. Dielectric partitions between the electrode contacts prevent channel interaction resulting from longitudinal spread of electrical current toward neighboring groups of nerve fibers. This precurved electrode comes preloaded on a stylet assembly which advances the electrode array off the stylet to an insertion depth of 21.5 mm. This configuration theoretically allows for the electrodes to lie close to the cochlear modiolus, thus providing improved sound fidelity and hearing performance. The HiFocus 1j electrode has the same number of electrode contacts on a slightly longer, narrower, and less curved silicone electrode array, and is designed to be inserted to a depth of 25 mm.

The PULSAR CI100 and SONATA TI100 devices developed by MED-EL Corporation house the same I100 electronics in a ceramic and titanium casing respectively (Fig. 6). The standard electrode array for these 2 devices features 12 pairs of electrode contacts on a soft and flexible straight electrode. The design of this array allows for the deepest insertion depth (approximately 31 mm) which then enables stimulation of a larger number of nerve fibers within the cochlea.

In addition to the standard electrode inventory provided by all the manufacturers, modified electrode configurations including straight, compressed and split electrodes (Fig. 7) are generally available with each device to implant congenitally malformed and ossified cochleae.

In addition to the standard electrode inventory offered with each of these cochlear implant systems, modified electrode designs such as the straight electrode, short electrode and split electrodes, are available for implanting malformed or ossified cochleae.

Speech-coding strategies and speech processors

Speech-coding strategies are software programs stored within the speech processor, which convert pitch, loudness and timing of sound into useful electrical signals [3]. Strategies are typically either non-simultaneous or simultaneous.

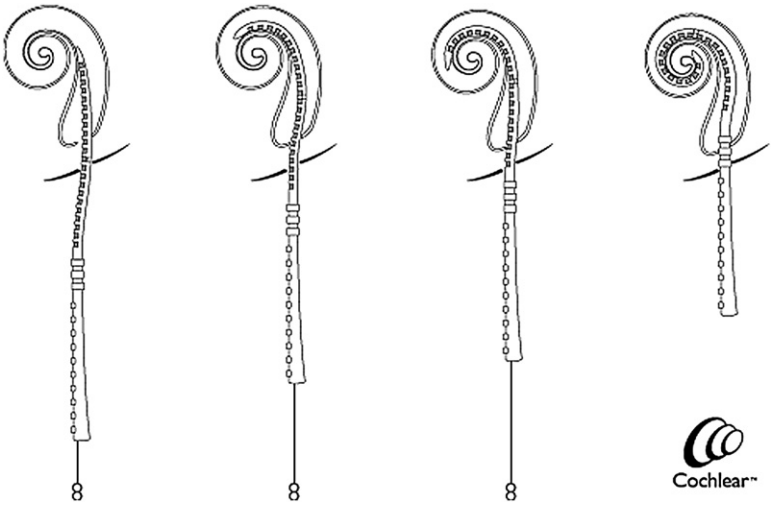

Fig. 4. Advance off stylet technique for insertion of Advance Contour electrode with Softip (Cochlear Corp., Englewood, CO).

Fig. 5. HiRes 90K implant (Advanced Bionics Corp., Sylmar, CA).

The most widely used speech coding strategy, SPEAK, or spectral peak, samples sound approximately every 4 ms and processes this information into 20 different frequency bands [4]. The processor selects an average of six filter bands that have the highest energy in each 4 ms interval and presents pulses sequentially to the six corresponding electrodes. Up to 10 maxima can be sampled in this fashion. This results in stimulation of up to 10 electrodes every 4 ms, representing the spectral energy levels of the sound input during that interval. The ACE, or advanced combined encoder, strategy is similar to the SPEAK strategy but uses a much higher rate of stimulation.

Another popular strategy is the continuous interleaved sampling (CIS) strategy [5,6]. With this strategy, each electrode receives pulses at a rate of 600 to 1000 pulses per second. Speech is divided into several frequency bands, and the amplitude envelope is extracted from each band. This information is then translated into an electrical impulse that drives the electrode representing that frequency band. In the SPEAK and CIS strategies, the electrode pulses are generated sequentially so that no two electrodes are active at the same time. This strategy avoids the problems of electrode interactions.

The Nucleus Freedom processors (Fig. 8) use a combination of the SPEAK, ACE and CIS strategies. The original ACE strategy allows stimulation rates up to 14,400 pps (pulses per second), whereas the more recent ACE (RE) strategy allows for higher stimulation rates of up to 34,000 pps.

HiResolution (HiRes) Sound is available in the HiRes Implant System from Advanced Bionics. HiRes is designed to offer a wide, programmable dynamic range, preserve spectral and temporal details of sound and stimulate at rates of up to 83,000 pps. In the HiRes 90K implants, the number of sites of stimulation can be increased beyond the number of electrode contacts. Through simultaneous delivery of current to pairs of adjacent electrodes, stimulation can be ''steered'' to sites between the contacts by varying

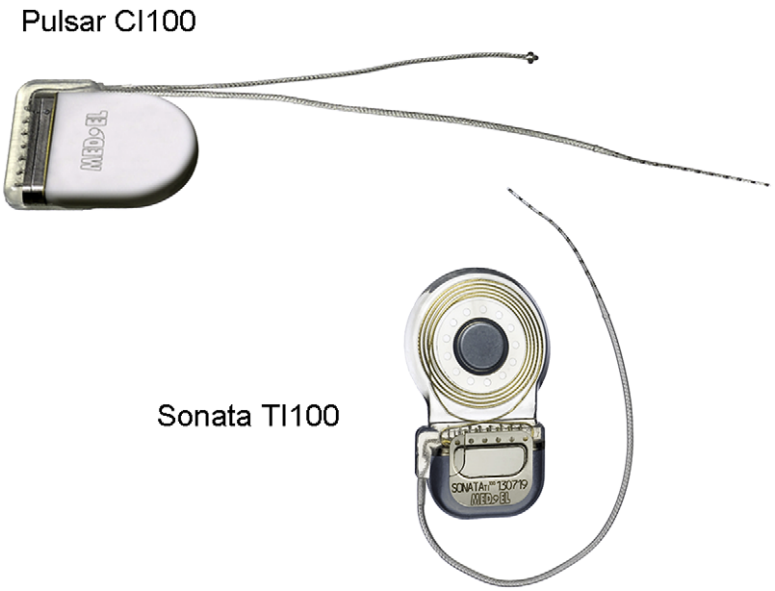

Fig. 6. MED-EL Pulsar CI100 and Sonata TI100 implants (MED-EL Corp., Innsbruck, Austria).

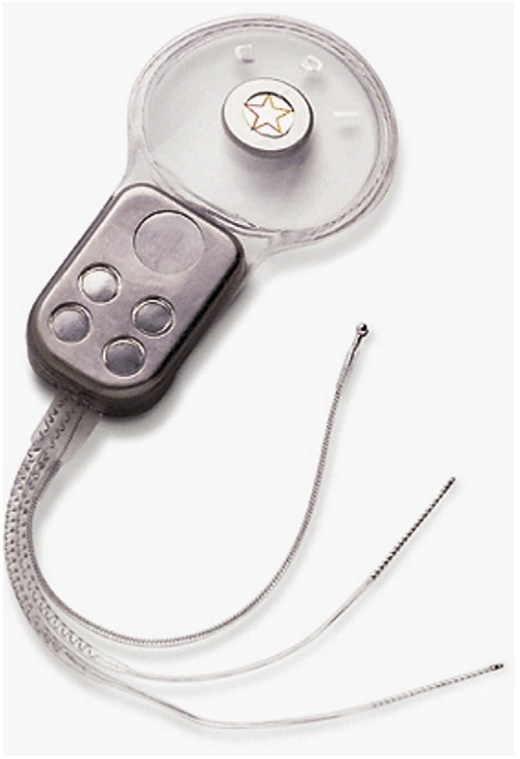

Fig. 7. Nucleus Freedom implant with split electrode (Cochlear Corp., Englewood, CO).

the proportion of current delivered to each electrode of the pairs. The Advanced Bionics strategies currently in use are the Hi-Res-P (Pulsatile), HiRes-S (Simultaneous) and HiRes Fidelity 120.

MED-EL coding strategies provide high stimulation rates up to 50,700 pps and individual current sources for each channel. With the introduction of the I100 electronics platform and the OPUS speech processors, MED-EL developed the FSP (Fixed Place strategy). The timing of stimulation is used to code the temporal structure of the sound signal in the low and mid frequency range by using channel-specific sampling sequences [7].

Telemetry

Neural response telemetry is a method that enables direct measurement of auditory nerve action potentials from cochlear implant patients. This technology is currently available for all four implant devices [7]. Initial recordings are obtained intraoperatively once the implant electrode has been inserted into the scala tympani and the receiver-stimulator has been secured in place.

The information obtained is useful for troubleshooting device failures and optimizing parameters for speech-processing strategies. This is particularly useful in mapping cochlear implants for younger pediatric patients who lack auditory experience.

Patient selection

Candidate selection for cochlear implantation has evolved as the devices and patient performance evolved. In general, adults and children with bilateral severe to profound SNHL, who receive little or no benefit from conventional hearing aids, are in good physical and mental

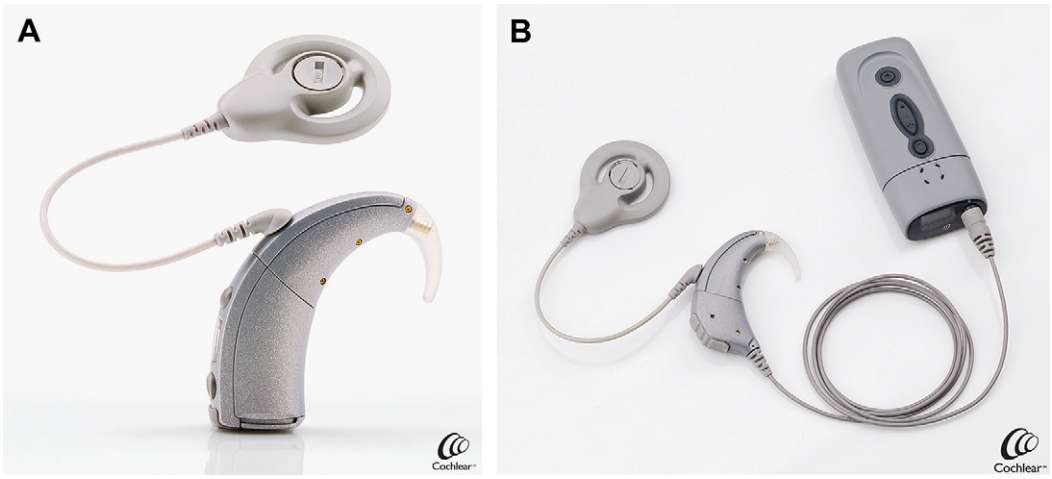

Fig. 8. Nucleus Freedom (*A*) BTE sound processor, (*B*) Bodyworn sound processor (Cochlear Corp., Englewood, CO).

health, and possess the aptitude and motivation to participate meaningfully in the auditory rehabilitation program are potential candidates for cochlear implantation.

Medical evaluation

A complete history and physical examination are necessary to detect problems that may contraindicate surgery or interfere with the patient's ability to complete postimplantation rehabilitation.

The cause of hearing loss is rarely a contraindication to implantation. Profound hearing loss associated with cochlear nerve aplasia is a rare congenital anomaly, in which the lack of auditory innervation obviates the option of cochlear implantation [8]. Prior meningitis with cochlear ossification or fibrosis does not exclude a patient from implantation but may necessitate modification of the surgical technique.

Two crucial factors influencing auditory performance following cochlear implantation include age of onset of deafness and duration of profound hearing loss. The ideal adult candidate has profound acquired SNHL. A period of auditory experience adequate for development of normal speech, speech perception, and language offers a significant advantage in learning to use the implant [9]. These postlingually deafened patients represent the majority of adults undergoing implantation. In these patients, there is a significant correlation between duration of profound hearing loss and performance. Those with prolonged auditory deprivation receive similar auditory information as do other implant patients but are not able to use the information as effectively in the recognition of running speech [10], which is thought to be due to the loss of central auditory processing.

A small number of adult implant recipients are congenitally or prelingually deafened, with prolonged auditory deprivation and little to no experience with sound. These patients typically have greater difficulty assimilating the new auditory information and, in general, have performed less well than those with some degree of auditory memory [11].

For children, the later the onset of deafness, the greater the auditory memory and, generally, the better the speech language skills. Both of these factors contribute to a higher likelihood of successful implant use. For congenitally or prelingually deafened children, increasing data support early implantation for maximal auditory benefit and speech language development [12]. Current FDA guidelines require a child to be at least 1 year old, have profound SNHL, demonstrate no benefit from conventional hearing aids, and be free of medical contraindications. Under exceptional circumstances, infants may be implanted earlier if hearing loss is the result of meningitis and there is concern for cochlear ossification. In addition to these criteria, the commitment of the family and the child's educational setting to postimplantation rehabilitation is a crucial determinant of successful implant use. Bilateral cochlear implantation is currently being recommended in both adults and children where applicable. The additional input from bilateral cochlear implantation has been shown to improve hearing in noisy situations and improve sound localization with the use of intensity cues [13]. The second device may be implanted simultaneously or sequentially.

Audiologic assessment

The basic testing battery includes unaided and aided warble tones and speech-detection thresholds, environmental sound recognition, and speech perception tests, with and without visual cues [14]. In general, a patient is considered a candidate for cochlear implantation when the following criteria are met: bilateral profound SNHL, with a three-frequency pure-tone average (500, 1000, and 2000 Hz), unaided threshold in the better ear of 70 dB hearing loss or poorer, or a speech discrimination score of 50% or less in best-aided conditions in the ear to be implanted and 60% or less in the non-implanted ear or bilaterally.

Audiologic screening in children requires auditory brainstem evoked response testing and otoacoustic emissions testing, in addition to conventional behavioral audiometry. An aggressive 6-month trial period using appropriate amplification and intensive auditory and speech training is an integral component of candidacy assessment in children. The global evaluation of cochlear implant candidacy in children is considerably more challenging than in adults and is best approached by a dedicated team comprising speech and hearing professionals, social workers, psychologists, and educators. The ultimate candidacy of a child is determined by not only a demonstrated physiological need but also the strength of the child's social and educational background.

Imaging

Preoperative imaging serves to complete the candidacy evaluation process and assist in surgical planning. A high-resolution CT scan of the temporal bones using a bone algorithm is the study of choice in most centers. MR imaging is the study of choice in a few centers. These images allow the surgeon to identify cochlear malformations and assess cochlear patency, mastoid pneumatization, and the course of the facial nerve. Observation of a narrow internal auditory canal on CT scans of the temporal bone should alert the surgeon to the possibility of cochlear nerve agenesis, which contraindicates implantation. This may be confirmed on sagittally reconstructed T2-weighted MR images through the internal auditory canal. MR imaging is also superior to CT imaging in assessing soft tissue obliteration of the cochlear lumen.

Electrical testing

Electrical stimulation of the promontory or round window has been used by some implant teams as part of the preoperative candidacy assessment [15]. Patients detecting the stimulus may possess better residual auditory neuronal stimulability. Such testing has not been uniformly adopted by all implant teams because patients with a negative response, particularly at the promontory, may respond to intracochlear stimulation with an implant.

Surgical technique

Selection of the side for implantation is governed by several factors. The most patent cochlea is typically chosen for implantation. It is generally believed that the ear with the shortest duration of deafness may serve as the best ear for implantation; however, if the patient uses a hearing aid in only one ear (the side that is perceived as the better-hearing ear), implanting the contralateral ''worse'' ear does not negatively impact performance [16]. When no specific factors lead to the choice of one ear over the other, the ear on the side of the dominant hand is chosen to facilitate device manipulation [17].

The implant is inserted via a transmastoid facial recess approach to the round window–scala tympani. Placement of a cochlear implants in children is essentially the same as in adults. By age 1 year, the mastoid antrum and facial recess are adequately developed. In patients with mastoid cavities or absent posterior ear canal, obliteration of the mastoid cavity with blind sac closure of the external auditory canal is preferably done at the time of disease removal. Cochlear implant placement is then performed at a second stage approximately 4 to 6 months later.

Surgery is performed with the patient under general anesthesia with the use of continuous intraoperative facial nerve monitoring. Many different incisions have been designed to allow placement of the receiver–stimulator. In general, the skin flap developed must be large enough to cover the receiver–stimulator completely. In adults and older children, temporalis muscle is removed around the receiver–stimulator area to minimize the thickness of the scalp over the internal device. This technique enhances magnetic coupling of the internal receiver and the external transmitter and reduces power consumption. In younger children with thin scalps, the temporalis muscle is elevated with the skin in a single layer. A depressed seat is created in the skull posterosuperior to the pinna, with adequate allowance for placement of a behind-the-ear microphone piece. More recently, minimal access incisions have been developed for placement of cochlear implant devices. In conjunction with minimal access surgery, receiver-stimulators may be tucked under the temporalis muscle without drilling a bony pedestal in the skull to secure the device in.

A complete mastoidectomy is performed, preserving a bony overhang around the margins of the mastoid cavity, to aid stabilization of the carrier coil within the cavity. The short process of the incus and its buttress are then used as bony landmarks to guide development of the facial recess. The chorda tympani is generally left intact unless a narrow recess limits visualization and access to the round window. The lip of bone overhanging the round window niche is then gently removed to allow for visualization of the round-window membrane. The cochleostomy is made anterior and inferior to the round-window membrane, in the basal turn of the cochlea. The electrode array is carefully advanced into the scala tympani. The cochleostomy is then sealed with a small plug of temporalis muscle or fascia.

The postauricular flap is closed in layers without drainage. For purposes of hemostasis, only bipolar electrocautery should be used after insertion of the cochlear implant device.

Surgical complications

The risks of implant surgery are similar to those of routine mastoid surgery and include infection, facial paralysis, dizziness, cerebrospinal fluid (CSF) leak, and meningitis. Skin flap breakdown and wound infection are the most common complications of cochlear implant surgery [18]. Placement of the device too close to the incision, or an excessively thin flap, can lead to device extrusion. It is imperative to seat the device approximately 1 or 2 cm from the wound edge and maintain a flap thickness of at least 6 to 7 mm. Flaps thicker than this can lead to inadequate magnet contact between the outer and inner hardware, with subsequent diminished performance.

During opening of the facial recess, the facial nerve is at particular risk for injury. Heat injury can be avoided by careful dissection and copious irrigation. The use of facial nerve monitoring may reduce the risk for injury, although it is no substitute for knowledge of temporal bone anatomy and good surgical technique.

Cerebrospinal fluid leaks, although unusual, can occur in two locations. The recessed site for the internal receiver often extends down to the dura. Inadvertent injury to the dura can result in CSF leak and should be repaired at the time of surgery with temporalis fascia. In congenitally malformed cochleae, the modiolar base may be deficient, resulting in a gush of CSF subsequent to the cochleostomy. When this situation is anticipated preoperatively based on imaging findings, the eustachian tube should be temporarily obliterated with Surgicel before the cochleostomy. Rarely do patients require further management of this complication, such as placement of a lumbar drain.

In the initial phase of cochlear implant development and insertion, there was a concern about the possibility of bacterial seeding of the CSF through the cochleostomy during bouts of acute otitis media. In 2003, several cases of fatal meningitis were reported in implanted children. Subsequent analysis of these cases demonstrated a significant correlation between the use of silastic intracochlear positioners to enhance perimodiolar electrode placement and post-operative meningitis. The use of these modiolar positioners has since been abandoned. The presence of cochlear malformations was also found to be a significant risk factor for meningitis. Streptococcus pneumoniae was found to be the predominant microbe in these cases. The United States Food and Drug Administration currently recommends preoperative vaccination with Prevnar or Pneumovax, along with the use of perioperative antibiotics for meningitis prophylaxis in all implant patients [19].

In the elderly population, the possible effects of cochlear implantation on the vestibular system should be considered. Transient dizziness and imbalance have been reported in up to 30% of implant recipients [20].

Rehabilitation

The external device is fitted 4 weeks after surgery, allowing for resolution of edema in the postauricular flap. Electrode mapping is also performed at this time. The initial goals of this fitting are to establish thresholds and comfortable loudness levels for each electrode. Most cochlear implant processors assign successive bands of frequencies to each electrode in tonotopic order. In some patients, processor programming may involve adjusting this electrode-to-frequency assignment if the pitch of the electrodes is not tonotopically ordered. All of these parameters are then stored in the speech processor and are collectively referred to as the "MAP."

In very young children with little or no auditory experience, determining these settings can be challenging. Electrically evoked auditory potentials and acoustic reflex thresholds can be useful adjuncts to the mapping process in children [21].

Rehabilitation continues at regular intervals for several years following implantation. Global rehabilitation often includes simple auditory training, speech reading practice, and counseling of the patient and family.

Results

Despite differences in device design and function, postimplant performance has not differed significantly among the devices. Virtually all patients receiving multichannel cochlear implant devices experience substantial benefit. Up to two thirds of adults undergoing implantation obtain open-set speech recognition and comprehend speech to some degree while using the telephone [22–24]. Even poor performers benefit from the awareness of environmental sounds and enhancement of lip-reading abilities.

One of the crucial determinants of performance is memory of previous auditory experience. Postlingually deafened adults with some auditory

experience and a short duration of deafness generally learn to use the sound information provided by the implant more quickly and effectively than do those who were born with profound hearing loss or those who lost their hearing early in life [25].

In children, a supportive family and educational environment, combined with intensive auditory and speech rehabilitation, are crucial for successful device use. Postlingually deafened children often demonstrate rapid improvement in speech perception, usually achieving 100% correct responses on closed-set testing, reaching a maximal benefit within 12 to 18 months [26]. Prelingually deafened children achieve average word recognition scores of 30% to 44% using newer speech-processing strategies [27,28]. Onset of deafness after 2 years of age and duration of deafness of less than 2 years convey the best prognosis for performance [29]. Congenitally deaf children undergoing implantation at a young age demonstrate the most improvement after implantation [12,30,31]. Congenitally deaf adolescents undergoing implantation do less well and have a high nonuser rate [32].

Acquisition of speech and language in children undergoing implantation often parallels speech-perception abilities. Furthermore, speech intelligibility and language skills continue to improve over time and, on average, exceed those of age- and hearing-matched peers using hearing aids [33,34]. Patients in oral-communication settings appear to perform at a higher level than do those in total-communication settings [35–37]. Research efforts are currently focused on developing atraumatic electrodes and insertion techniques to preserve residual hearing in the implanted ear. Investigational devices with shorter electrodes are being implanted in individuals with significant residual low frequency hearing who currently receive limited benefit from conventional hearing aids. The successful implantation of these devices allow for bimodal hearing in the implanted ear with auditory stimulation using a conventional aid to amplify low frequency tones and electrical stimulation using the cochlear implant to restore hearing in the high frequency spectrum [REFERENCE].

Auditory brainstem implants

The auditory brainstem implant (ABI) was designed to restore some hearing to patients lacking functionally intact cochlear nerves and who were, therefore, not candidates for cochlear implantation. These are principally patients with neurofibromatosis type 2 (NF-2), in whom both cochlear nerves are nonfunctional as the result of the vestibular schwannoma or its surgical removal. In such individuals, the cochlear nucleus complex within the brainstem is stimulated directly by placing the ABI electrode into the lateral recess of the fourth ventricle.

The first ball electrode ABI was placed by House and Hitselberger in 1979 in a patient with NF-2 during the removal of her second vestibular schwannoma. Early prototype single-channel devices were replaced by multichannel devices in 1992. The Nucleus multichannel ABI device received FDA approval for implantation in October 2000.

Device design

The current Nucleus multichannel ABI24 device was developed collaboratively by the House Ear Institute and Cochlear Corporation [38]. The device consists of an implanted component and an externally worn component, similar to the cochlear implant device. The implanted receiver–stimulator, modeled after the Nucleus cochlear implant device, is attached to an electrode array composed of 21 platinum disks mounted on a silicone and Dacron mesh carrier (Fig. 9). The external component consists of a microphone, speech processor and a transcutaneous transmitter coil. Unlike the cochlear implant device, however, the receiver magnet is removed at the time of surgery, allowing patients with NF-2 to undergo MRI

Fig. 9. Nucleus multichannel ABI24 device (Cochlear Corp., Englewood, CO).

surveillance of tumors. A small retainer disk is used instead to secure the external transmitter coil in place.

Functionally, the ABI device differs little from the cochlear implant device. A microphone converts sound to an analog electrical signal, which is then modified by a speech processor and delivered to the electrode array via either a percutaneous pedestal or a transcutaneous coil.

Patient selection

The criteria for placement of ABIs include a diagnosis of NF-2, age of at least 12 years, requiring either first or second side surgery, proficiency in the English language, and reasonable expectations.

Initially, the ABI was placed at the second acoustic neuroma surgery. With increased experience, several patients have undergone device placement during surgery for the first tumor, even though they had serviceable hearing in the opposite ear. Earlier implantation allows the patient time to learn to use the device before complete loss of hearing occurs in both ears. Also, waiting to implant the second side often results in placing the device after removal of large tumors, which may have distorted normal landmarks, thus significantly complicating placement of the device. Finally, implanting the first side gives the patient a chance to have the second side implanted should the initial device fail to provide useful auditory sensation [39].

Surgical technique

The cochlear nucleus complex, composed of the dorsal and ventral cochlear nuclei, is the target for placement of the ABI electrode array. The ventral nucleus is the main relay nucleus for nerve VIII input, and its axons form most of the ascending pathway. Both nuclei are not visible from a surgical approach and must be located using surface landmarks. The lateral termination of the fourth ventricle, the foramen of Luschka, is found between the roots of the facial and glossopharyngeal nerves. Usually only a stump of nerve VIII remains, and this may be a useful landmark for the lateral recess as well. Experience has demonstrated that the ideal position for the electrode placement is completely within the lateral recess, adjacent to the dorsal nucleus and the inferior aspect of the ventral nucleus. This position results in optimal auditory stimulation and the least stimulation of adjacent structures, including cranial nerves V, VII, and IX, or the overlying flocculus of the cerebellum. Also, placement completely within the lateral recess aids in the stabilization of the electrode [40].

The approach used for tumor removal in ABI cases is the translabyrinthine craniotomy. This approach offers the most direct access to the lateral recess after tumor removal. Intraoperative monitoring of the trigeminal, facial, and glossopharyngeal nerves is helpful. The translabyrinthine craniotomy skin incision is modified to allow for placement of the internal receiver–stimulator. Once the tumor is removed, a seat is created in the lateral surface of the skull for placement of the receiver, and a groove is created as a path for the electrode lead. Meticulous hemostasis is obtained in the cerebellopontine angle, and then the landmarks leading to the surface of the cochlear nuclei are identified. Normally, the opening of foramen of Luschka is marked by intact choroid plexus and the taenia traverses the roof of the lateral recess. After removal of larger tumors, however, these landmarks may be obscured. In such cases, the stump of nerve VIII can usually be followed to the opening of the lateral recess, and nerve IX reliably leads to the floor of the recess. Once identified, the location of the lateral recess can usually be confirmed by observing the egress of spinal fluid during a Valsalva maneuver.

The electrode array is then placed completely within the lateral recess as noted earlier. This placement situates the array immediately adjacent to the dorsal cochlear nucleus and the posterior tip of the ventral cochlear nucleus. Orienting the electrode array such that the electrodes face medially and superiorly maximizes stimulation of the cochlear nuclei.

After placement of the electrode, tests for electrical auditory responses are performed, as are stimulation of adjacent cranial nerve nuclei and vital sign changes. The position of the electrode array is adjusted to maximize auditory stimulation while nonauditory stimulation is minimized. The electrical ABR is very useful in confirming placement of the array, especially in cases in which the tumor has distorted normal anatomy. The electrode array is secured by placing a small piece of fat or polytetrafluroethylene (Teflon) felt within the opening of the lateral recess. The electrode lead is then positioned in the mastoid cavity, while the ground electrode is inserted under the temporalis muscle. The magnet in the receiver–stimulator is removed

and replaced with a silicone plug to allow for postoperative MR imaging. The mastoid defect is packed with fat and the wound closed in layers.

Surgical complications

The most common complications related to ABI placement include CSF leak, electrode migration, and nonauditory side effects. CSF leak in these cases may be related to tracking of CSF along the electrode lead, from the subarachnoid space to the subcutaneous plane. These leaks may be managed in a similar fashion to leaks occurring after conventional neuro-otologic surgery. Rarely is re-exploration necessary for control of the leak [39].

Migration of the electrode may occur as a result of unstable positioning or changes in shape and position of the brainstem after tumor removal. Electrode position may be confirmed on high-resolution CT scans in which soft tissue windows of the posterior fossa are overlaid with images enhanced for high-contrast objects [41].

Nonauditory side effects have occurred in 42% of multichannel implant users and seem to be related to electrode position [38]. In patients with a low electrode placement, as demonstrated on CT scans, symptoms related to glossopharyngeal nerve stimulation have occurred and are typically a sense of tingling or constriction in the throat. One patient with an exceptionally low placement had nausea and shoulder contraction related to vagal and accessory nerve stimulation respectively [40]. High electrode placement has caused facial twitching due to stimulation of the intact facial nerve. A mild sense of jittering of the visual field also has been reported, possibly related to activation of the flocculus of the cerebellum. Nonauditory side effects in the multichannel device generally occur with stimulation of the more medial or lateral electrodes. They can usually be reduced by switching reference electrodes, increasing the duration of the stimulus pulse, or turning off the electrode. The severity of the nonauditory sensations often decreases over time, sometimes allowing for reactivation of electrodes previously turned off.

Rehabilitation

Initial device stimulation is usually carried out 4 to 8 weeks after implantation and consists of measuring threshold and comfort levels and detecting nonauditory side effects, which may be controlled with modification of stimulus parameters.

Results

With some notable exceptions, the overall performance of patients with ABIs have paralleled that of patients with earlier single-channel cochlear implants. Eighty-five percent of patients undergoing implantation receive auditory sensations [42]. Combined with lip-reading cues, 93% of patients demonstrate improved sentence understanding at 3 to 6 months. Most patients have environmental-sound awareness and understanding of closed-set words, consonants, and vowels. Open-set speech recognition in patients receiving ABIs is the exception rather than the norm. Sound recognition and speech perception generally continue to improve for up to 8 years following implantation [43], although overall progress tends to be slower and of a lesser magnitude than that seen with multichannel cochlear implants. The ABI device is currently being implanted in non-NF2 patients in Europe who are not candidates for cochlear implantation. These include patients with extensive cochlear ossification, cochlear nerve aplasia, or cochlear nerve injuries. Early results indicate better hearing outcomes in these patients compared to NF2 patients. A significant number of these non-tumor patients are able to understand speech at a level comparable to that of the most successful cochlear implant users, including the ability to converse on telephones [44]. Clinical trials are currently underway in the United States for this clinical indication.

Summary

Cochlear implantation is an established habilitative and rehabilitative option for profoundly deafened individuals over 1 year of age who derive limited benefit from conventional hearing aids. Auditory performance varies among individuals and is determined primarily by age at implantation, preexistence of speech and language skills, and the time interval between onset of deafness and implantation. Successful implant users generally demonstrate improved auditory abilities and speech production skills beyond those achieved with hearing aids.

Multichannel ABIs can provide useful auditory information to patients with NF-2 who have lost integrity of auditory nerves following removal of vestibular schwannomas. The implant allows for awareness of environmental sounds and, potentially, speech recognition. Most patients undergoing implantation demonstrate improved

lip-reading skills, and exceptional performers achieve understanding of open-set speech.

Acknowledgments

The authors thank Karen Berliner and Laurel Fisher for their assistance in the preparation of the original manuscript.

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