Chapter 129 – Cochlear Implantation

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Cochlear implants are implantable electronic prostheses designed to convert mechanical sound energy into electrical signals that directly stimulate the auditory nerve in severely to profoundly deaf individuals. They have become the standard of care for rehabilitating deafened individuals who no longer benefit from the use of conventional hearing aids. The Food and Drug Administration (FDA) approved the use of cochlear implant devices in the United States for adults in 1985 and for children in 1990. Since then, more than 80,000 devices have been implanted in this country.

The functional anatomy of the inner ear as it applies to cochlear implantation is briefly reviewed here. The cochlea consists of a central bony skeleton, the modiolus, and the surrounding bony otic capsule. Within the modiolus lies the auditory nerve with bipolar primary afferent neurons, their cell bodies (spiral ganglia), and efferent nerve fibers. The presence of at least 10,000 surviving cochlear neurons is vital for successful hearing outcomes using a cochlear implant device. The spiral ganglion neurons follow the spiral arrangement of the cochlear duct with maximal ganglion cell density located between the upper basal turn and lower middle turn of the cochlear duct. This has clinical significance in cochlear implantation for placing the electrode array close to the greatest density of cochlear neurons.

All cochlear implant systems have an externally worn device and an implanted internal component. 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. 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.

Three implant systems (Nucleus Freedom system, Harmony HiResolution Bionic Ear system, and Med-El Sonata^{TI100} and PulsarCI^{100} systems) are currently FDA approved for use in adults and children in the United States. All are multichannel systems with 16 to 24 electrodes designed to take advantage of the tonotopic organization of the cochlea. Incoming speech signals are filtered into a number of frequency bands, each corresponding to a given electrode in the array. The Nucleus Freedom and HiResolution Bionic Ear systems use precurved modiolar-hugging electrode arrays. The relative proximity of these precurved electrode arrays to spiral ganglion cells offers theoretical advantages of improved sound quality, speech recognition, and power efficiency. By contrast, the longer tapered straight electrode of the Med-El PulsarCI^{100} and Sonata^{TI100} systems are designed for deep intracochlear insertion and electrical stimulation of the full sound frequency range. Overall hearing outcomes as reflected by standard audiologic test scores have been comparable using devices from all three manufacturers. The selection of device to be implanted is, in most practices, left to the patient.

PATIENT SELECTION

Candidacy criteria for cochlear implantation in adults and children continue to evolve with advances in electrode design, surgical techniques for atraumatic electrode insertion, and speech processing strategies. Current devices use either precurved perimodiolar electrode arrays or slim-profile tapered arrays to minimize trauma to surviving neural elements within the cochlea. With the possibility of preserving residual hearing in the implanted ear, candidacy criteria have expanded to include individuals with more residual hearing.

Adults

Current candidacy criteria for cochlear implantation in adults are listed in Table 129-1. It is well established that individuals who lose hearing after acquiring speech and language skills (postlingually deafened) and those with a shorter duration of deafness before implantation perform better with cochlear implants. In fact, duration of deafness has been found to be the only reliable predictor of auditory performance.[1] Other variables, including age at implantation, age at onset of hearing loss, age of deafness, hearing aid use, side implanted, and preoperative audiologic test scores, have not consistently correlated with postimplantation performance. Prelingually deafened

adults who have been habilitated using aural and oral education may receive some benefit from a cochlear implant.[2] There is no upper age limit for implantation in adults with good health.

Table 129-1 -- CANDIDACY CRITERIA FOR COCHLEAR IMPLANTATION IN ADULTS OLDER THAN 18 YEARS

- Bilateral severe to profound hearing loss: a three-frequency pure-tone average (500, 1000, 2000 Hz) unaided threshold in the better ear of greater than 70 dB
- Less than 20% word recognition score with consonant nucleus consonant (CNC) words bilaterally
- Minimal benefit from conventional hearing aids, as defined by hearing in noise sentence testing (HINT) recognition scores of less than 50% correct in the best aided condition
- No medical contraindications

Children

The selection procedure for children is more complex than for adults. In contrast to adults, both prelingually and postlingually deafened children are candidates for cochlear implantation. Research has shown that early auditory experience is critical for the development of central and peripheral auditory neural pathways. There exists an early period of language development in humans between ages 2 and 5, during which auditory input is paramount. Therefore, early hearing habilitation using a cochlear implant in children with congenital deafness and early profound acquired hearing loss is critical. Early implantation significantly improves the chance for development of normal speech and language in children.

Since the legislation of universal newborn hearing screening across the United States, many more infants with congenital hearing loss are being identified within the first few months of life. Current FDA guidelines permit implantation of children as young as 12 months of age. Exceptions to this lower age limit may be made in children with deafness resulting from meningitis. Earlier implantation in these children may allow for successful electrode insertion before the development of intracochlear ossification. Criteria for the pediatric age group are listed in Table 129-2. Appropriate family motivation, expectations, and support are also important factors in determining suitability of the child for implantation.

Table 129-2 -- CANDIDACY CRITERIA FOR COCHLEAR IMPLANTATION IN CHILDREN

Children age 12 to 24 months:

- Bilateral profound hearing loss
- Lack of auditory skills development and minimal hearing aid benefit
- No medical contraindications
- Enrollment in an education program emphasizing auditory development

Children age 25 months to 18 years:

- Bilateral severe to profound hearing loss
- Lack of auditory skills development and minimal hearing aid benefit (open set word recognition scores of less than 30% correct)
- No medical contraindications
- Enrollment in an education program emphasizing auditory development

PREOPERATIVE 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 post-implantation rehabilitation. Etiology 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 for cochlear implantation. Prior meningitis with cochlear ossification or fibrosis does not exclude a patient from implantation, but may necessitate modification of the surgical technique itself. The ear selected for implantation must be free of infection and ideally have an intact tympanic membrane.

An audiologic evaluation that characterizes hearing loss in both unaided conditions and in best binaurally aided conditions is performed. This includes measurement of pure tone thresholds, word, and sentence recognition testing. Aided speech recognition scores are the primary audiologic determinants of cochlear implant candidacy. Audiologic screening in children requires auditory brain stem evoked response testing, and otoacoustic emissions testing, in addition to conventional behavioral audiometry. A 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. The ultimate candidacy of a child is determined not only by a demonstrated physiologic need but also by the strength of the child's social and educational background.

Preoperative imaging necessarily includes a high-resolution, thin-cut computed tomography (CT) scan of the temporal bone in both the axial and coronal planes. This is used to determine the presence and patency of the cochlea, identify any congenital inner ear malformations, assess the caliber of the internal auditory canal, and assess the anatomy of the mastoid, middle ear, and facial nerve. Congenital malformations of the inner ear and cochlear ossification per se are not contraindications to cochlear implantation, although modifications in surgical technique or the use of nonstandard implant electrodes may be necessary and should be anticipated preoperatively. More recently, some implant teams have advocated the routine use of magnetic resonance imaging (MRI) of the internal auditory canals as the preoperative imaging modality of choice. In a retrospective review by Parry and colleagues, MRI was found to be more sensitive and specific in diagnosing soft tissue abnormalities in the inner ear than high-resolution computed tomography in cochlear implant candidates.[3] Using high-resolution T2-weighted sequences, MRI offers the capability for visualizing intracochlear fluid and is therefore more sensitive for detecting both bony and soft tissue obliteration of the cochlea. MRI can also be used to identify neural components within the internal auditory canal on sagittal reconstructions of the internal auditory canal to confirm the presence of a cochlear nerve.

In children, a comprehensive psychosocial evaluation is performed to identify factors that may affect subsequent adjustment to or benefit from the implant. Reasonable implant expectations and acceptance of the child or adult recipient by family members are addressed.

Several factors influence the selection of ear to be implanted. Generally, the ear with the shortest duration of deafness, the more consistent use of a hearing aid, and the most radiographically favorable anatomy (well-pneumatized mastoid, normal facial nerve anatomy, normal inner ear development, and patent cochlea) is selected for implantation. More recently, Friedland and colleagues have suggested that cochlear implantation of the poorer hearing ear does not compromise postoperative performance as measured using standard audiologic tests.[1] While duration of deafness was still the most consistent and significant determinant of postoperative speech recognition, the authors concluded that the overall auditory experience of the individual, rather than ear-specific criteria, was the best predicator of postoperative cochlear implant performance in the postlingually deafened adult. When both ears are equally suitable for implantation, the ear on the side of dominant hand is selected to facilitate usage of the device.

Of the more than 80,000 patients worldwide who have received cochlear implants, approximately 3500 have received bilateral cochlear implants.[4] Given the advantages of conventional binaural amplification for bilateral sensorineural hearing loss over monaural amplification, we should expect similar auditory performance with bilateral cochlear implantation. The reported benefits of bilateral implantation include significant decrease in head-shadow effect, improved speech understanding in noise, binaural summation, and sound localization. Implants may be placed sequentially at separate surgeries or simultaneously.

Children with cochlear implants have an increased risk for *Streptococcus pneumoniae* meningitis compared with children without cochlear implants. Routine preoperative pneumococcal vaccination is now recommended for all patients.[5] Guidelines for vaccination vary by patient age and are listed in Table 129-3.

Table 129-3 -- GUIDELINES FOR MENINGITIS PROPHYLAXIS IN COCHLEAR IMPLANT CANDIDATES

- Children who have completed the pneumococcal conjugate vaccine (Prevnar) series should receive one dose of the pneumococcal polysaccharide vaccine (Pneumovax 23). If they have just received the pneumococcal conjugate vaccine, they should wait at least 2 months before receiving the pneumococcal polysaccharide vaccine.
- Children between 24 and 59 months of age who have never received either the pneumococcal conjugate vaccine or pneumococcal polysaccharide vaccine should receive two doses of pneumococcal conjugate vaccine 2 or more months apart and then receive one dose of pneumococcal polysaccharide vaccine at least 2 months later.
- Individuals age 5 years and older with cochlear implants should receive one dose of pneumococcal polysaccharide vaccine.

Source: *http://www.cdc.gov/vaccines/vpd-vac/mening/cochlear/dis-cochlear-hcp.htm*

SURGICAL APPROACHES

Surgery is performed under general anesthesia with the patient in the supine position. Continuous intraoperative facial nerve monitoring is routinely employed. The use of long-acting muscle relaxants is avoided for this reason. A single dose of a broad-spectrum antibiotic such as cefazolin (Ancef), 2 g, is administered at the time of anesthesia induction.

Device templates provided by the manufacturer are used to outline the position of the behind-the-ear (BTE) processor, internal receiver, and surgical incision. The internal receiver is placed above and behind the BTE processor, avoiding any overlap between the two components. The position of the internal receiver may be marked at this point using methylene blue and an 18-gauge needle if desired. A variety of skin flaps have been described, most of which are some variation of the standard postauricular incision used for routine mastoid surgery with a posterosuperior extension to provide surgical exposure for seating the internal receiver (Fig. 129-1). Minimal access approaches have been described for cochlear implant insertion if cosmesis is of concern to the patient. In this case, an oblique incision no greater than 3 cm in length, approximately 0.5 cm behind the postauricular crease, is made above and behind the pinna (Fig. 129-2).[6] After infiltration of the planned incision with 1% lidocaine with 1:100,000 epinephrine solution, the skin and subcutaneous tissue are incised down to the level of the temporalis fascia superiorly and mastoid periosteum inferiorly (Fig. 129-3). Skin flaps are then developed anteriorly to the ear canal and posteriorly to allow for placement of the internal receiver. An anteroinferiorly based musculoperiosteal flap is then created by incising the temporalis fascia, muscle, and periosteum. Posterosuperiorly, this flap should be long enough to completely cover the electrodes as they emerge from the internal receiver. The mastoid cortex and retromastoid cranium are exposed by elevating this musculoperiosteal flap anteriorly until the spine of Henle is visualized (Fig. 129-4).

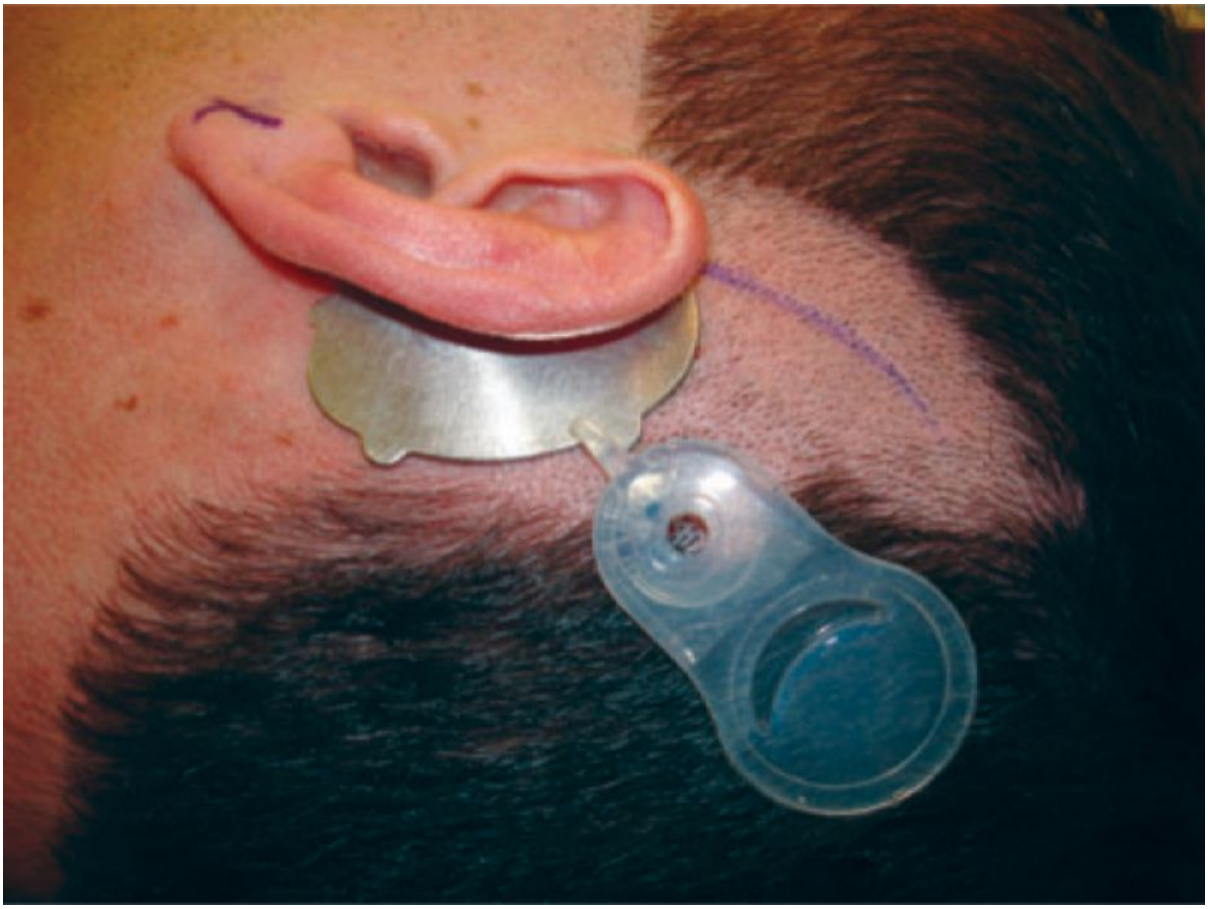

Figure 129-1 Implant template placement and surgical incision marking (Nucleus Freedom device).

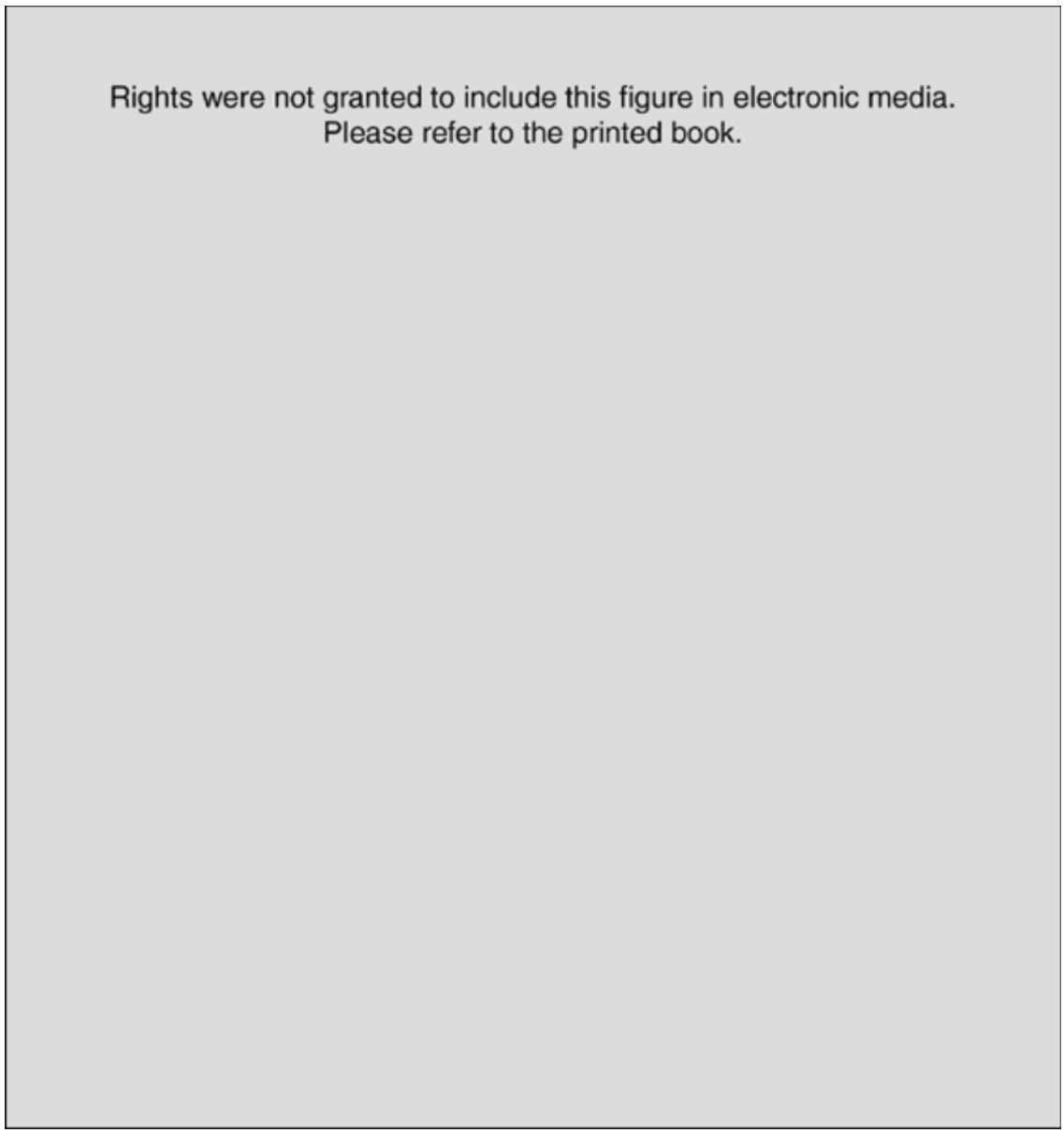

Figure 129-2 Minimal access incision for cochlear implantation.
(From O'Donoghue GM, Nikolopoulos TP: Minimal access surgery for pediatric cochlear implantation. Oto Neurotol 23:891-894, 2002. Reprinted with permission from Lippincott Williams & Wilkins, Optometry and Vision Science.)

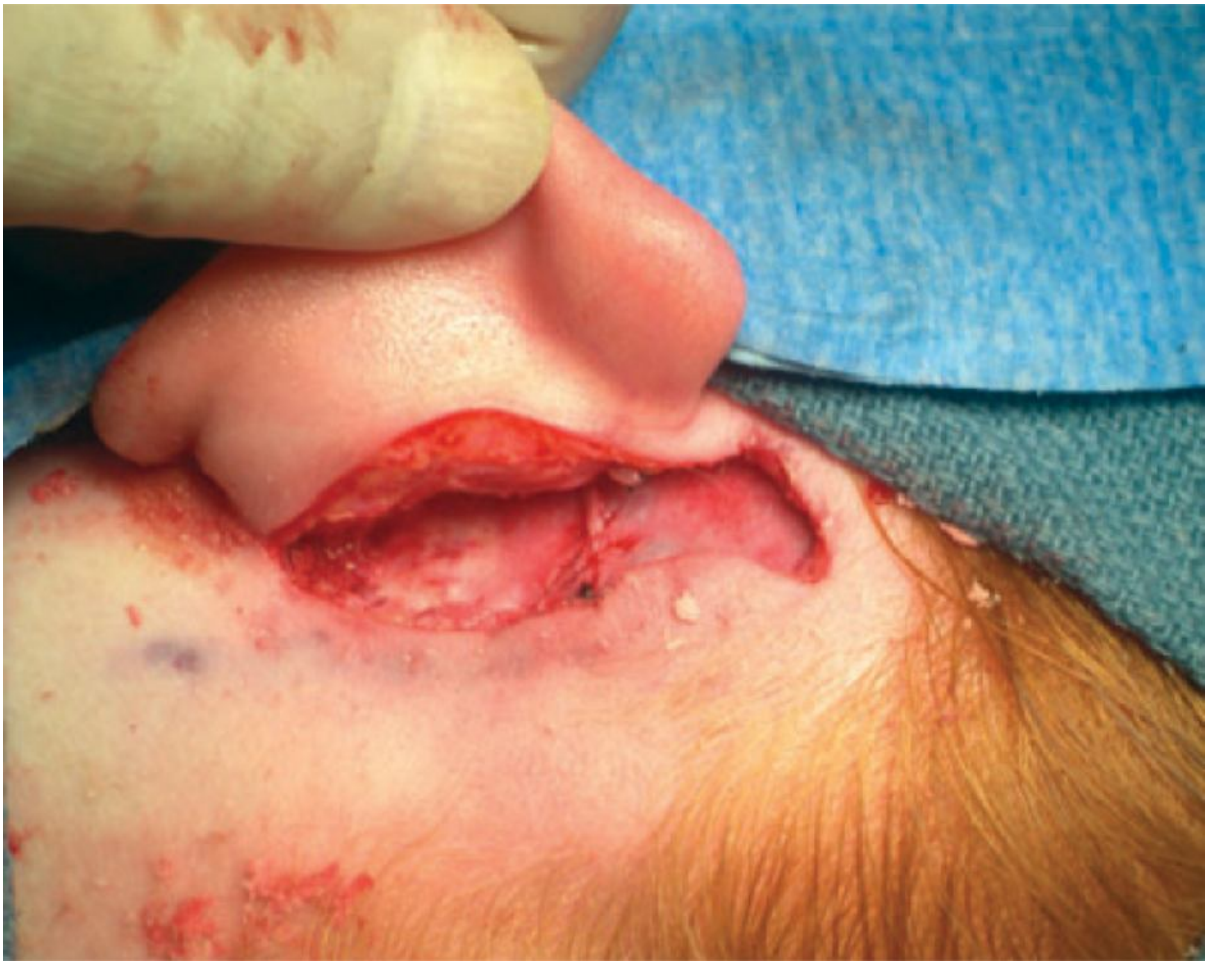

Figure 129-3 Skin incision and flap elevation.

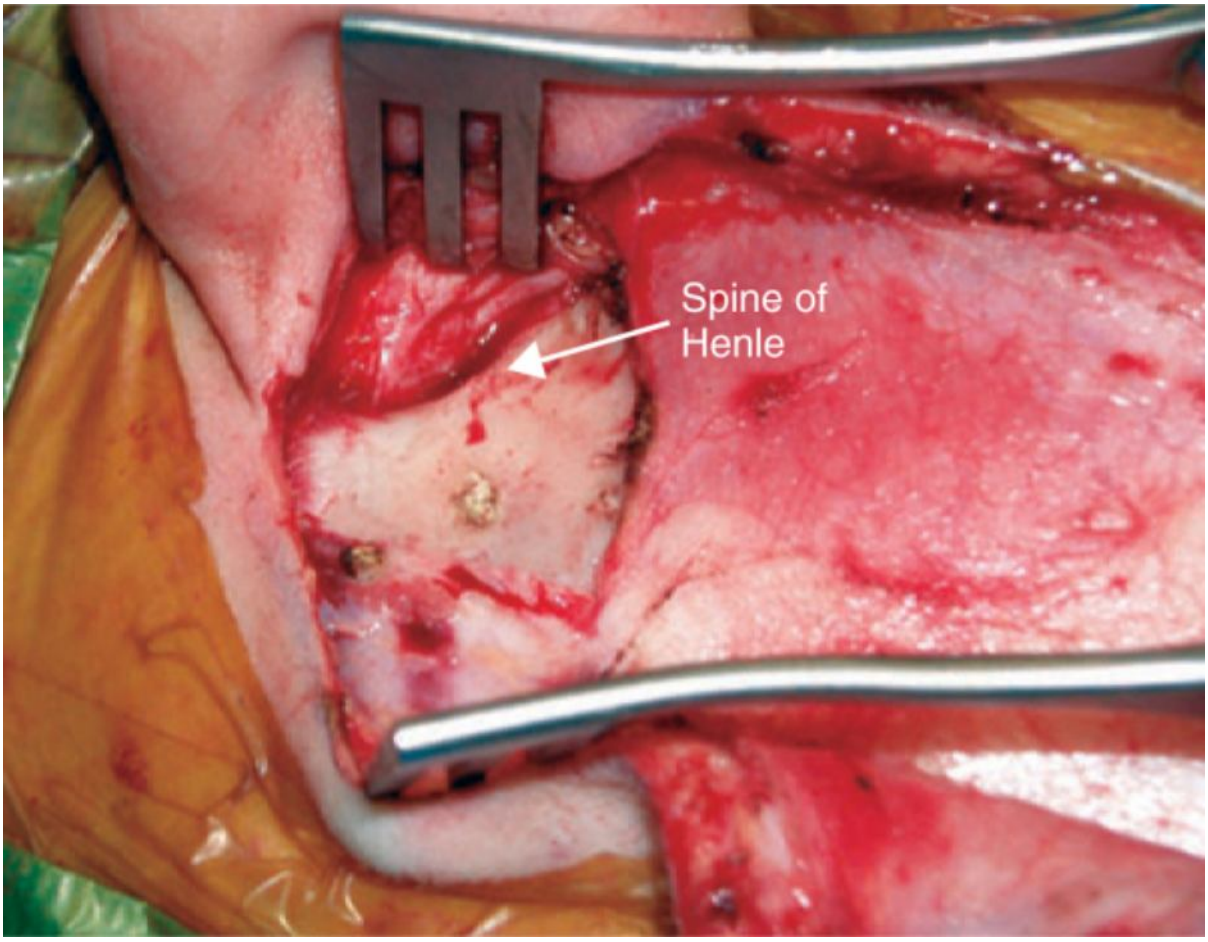

Figure 129-4 Musculoperiosteal flap elevated.

A complete mastoidectomy is performed using a combination of cutting and diamond drill burrs and continuous suction irrigation, without saucerization of the mastoid cavity. A bony overhang along the superior and posterior margins of the mastoid cavity is preserved for securing the electrode array within the cavity. Once the mastoid antrum is entered, the lateral semicircular canal is identified and drilling continues anterosuperiorly until the body of the incus is identified. The latter is used to define the level of the facial recess. Using 1- to 3-mm diamond burrs, the facial recess, bound by the chorda tympani nerve, facial nerve, and incus buttress, is carefully saucerized with copious irrigation to avoid thermal injury to the facial nerve (Fig. 129-5). Removal of bone anteromedial to the facial nerve in the facial recess is sometimes necessary to visualize the round window niche. When the facial recess is narrow, the chorda tympani nerve may need to be sacrificed to gain adequate exposure to the cochlea. In this situation, extreme caution should be exercised when removing bone lateral to the chorda tympani nerve to avoid violating the external auditory canal. Where a prominent round window lip is evident and obscures visualization of the round window membrane, the bony lip may be carefully drilled down using a 1- to 1.5-mm diamond burr. The mastoid cavity is then thoroughly irrigated with bacitracin solution (50,000 units bacitracin powder dissolved in 1000 mL normal saline solution) to remove any bone dust. The epitympanum and facial recess are plugged with Gelfoam or cotton prior to irrigation to avoid dispersing bone dust into the middle ear.

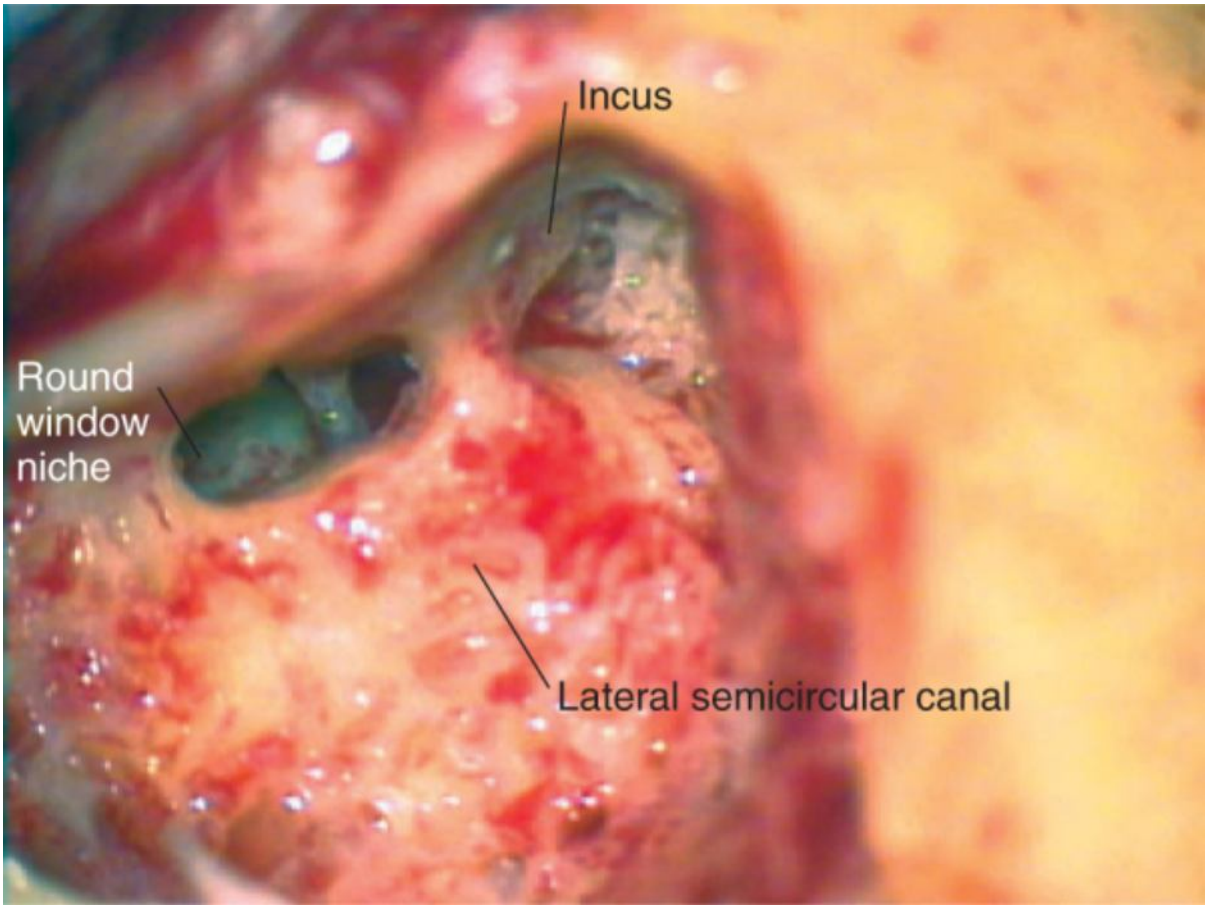

Figure 129-5 Facial recess opened for surgical access to round window.

The position of the internal receiver is then confirmed at this point using the BTE and device templates. A dummy internal receiver is used to mark out the dimensions of the bony recess necessary to seat the internal receiver. Cutting and diamond drill burrs are used to develop this recess and the tunnel connecting this recess to the posterosuperior limit of the mastoid cavity. In pediatric patients, the limited thickness of the skull may necessitate removal of bone down to dura, leaving a central island of bone, to allow for adequate recessing of the device. Several techniques are available to secure the device within the bony recess, including the use of nonabsorbable sutures with or without screws, Gore-Tex strips, and microplates. We use four 4-0 silk sutures placed through suture tunnels created alongside the bony recess (Fig. 129-6).

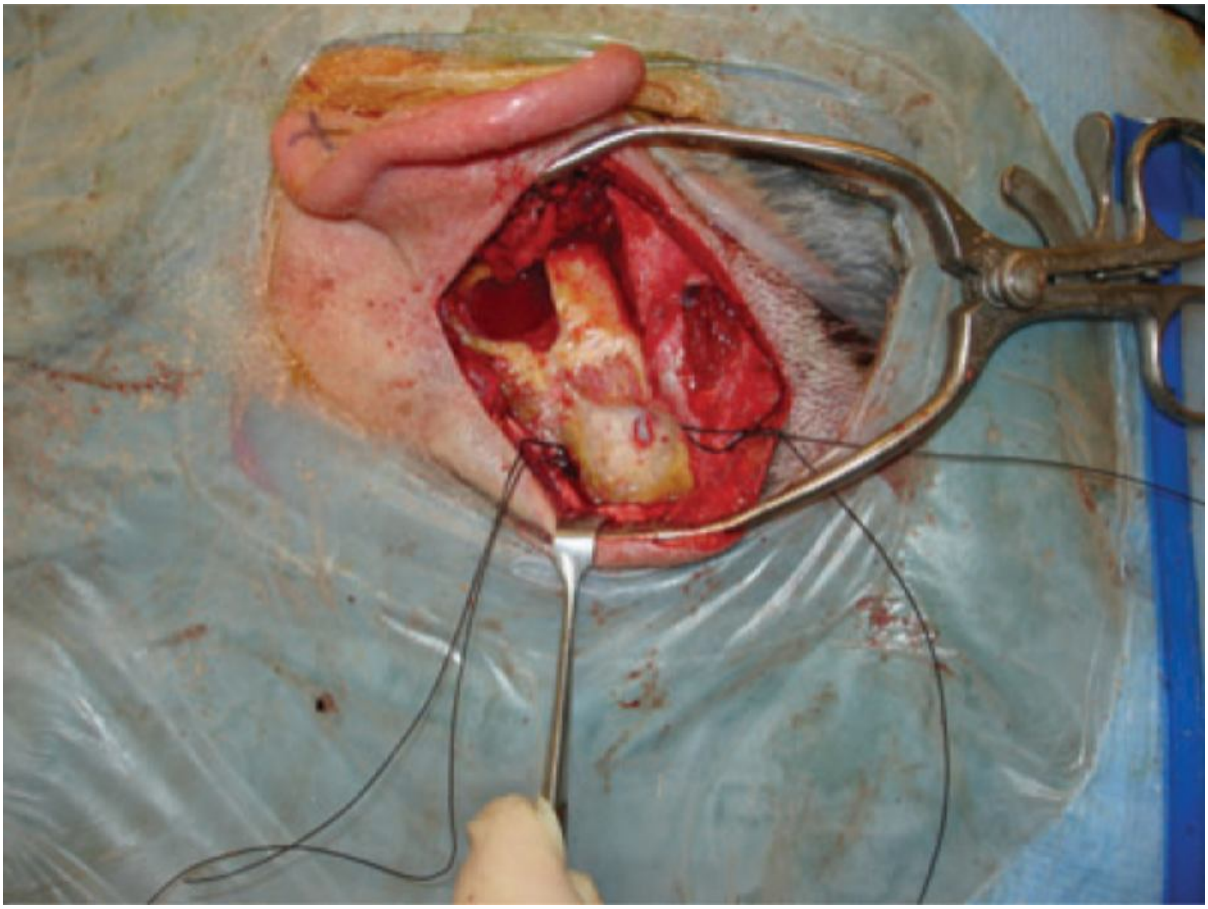

Figure 129-6 Bony recess and tunnel drilled for internal receiver and electrodes. Silk ties in place to secure internal receiver within recess.

Next, the cochleostomy is performed. Using a 1-mm diamond burr, a small cochleostomy is made immediately anterior and inferior to the round window membrane (Fig. 129-7). This allows surgical access directly into the scala tympani of the cochlear basal turn. Placement of the electrode array into this location is also possible directly through the round window membrane.

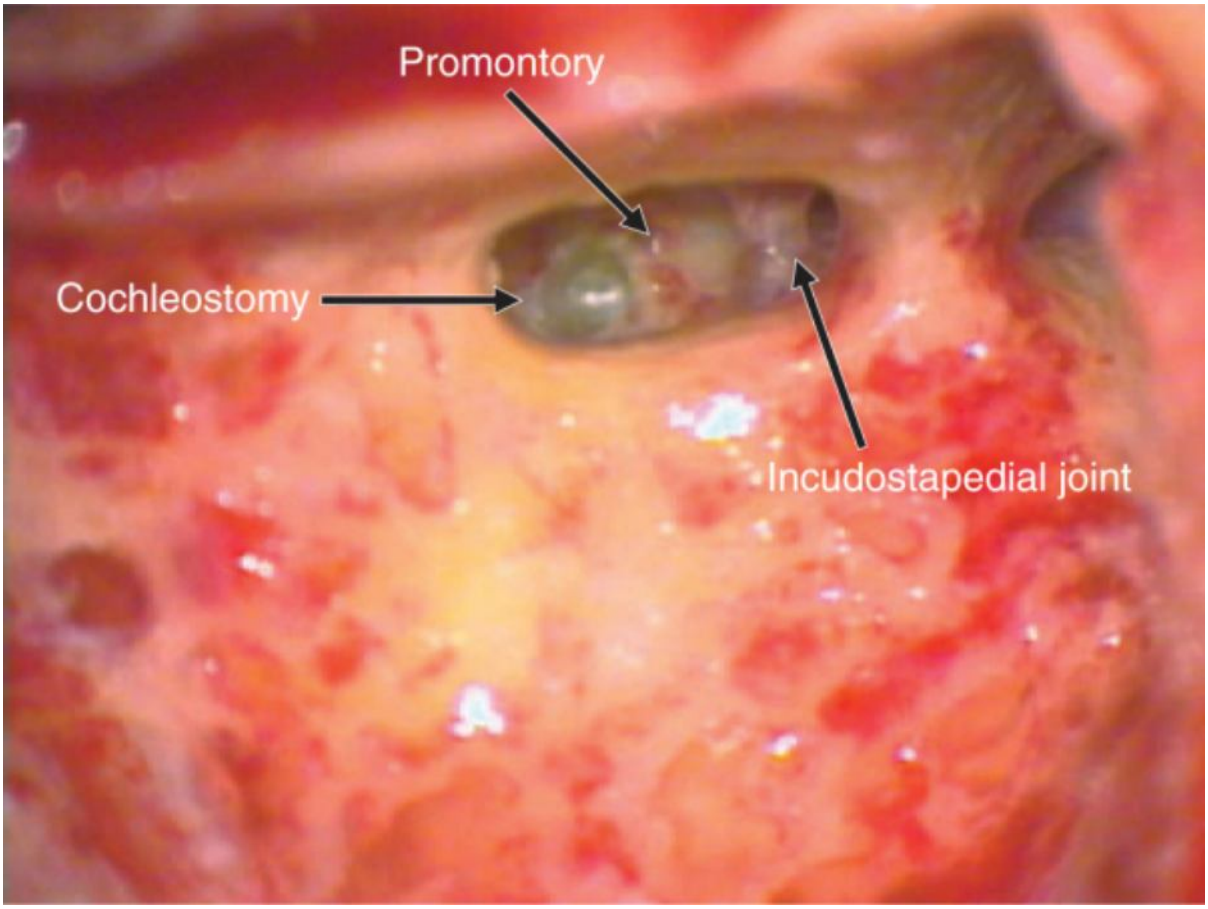

Figure 129-7 Placement of cochleostomy anterior and inferior to round window membrane.

The internal receiver is then placed in the bony recess previously created. The magnet of the internal device may be placed medial or lateral to the temporalis muscle (Fig. 129-8). Once the receiver has been secured in place, the electrode array is inserted into the scala tympani of the cochlea (Fig. 129-9). A variety of tools are provided by the implant manufacturers to facilitate electrode insertion. Undue force should not be used during electrode insertion to avoid electrode kinking within the cochlea and minimize insertional trauma to surviving neural elements within the cochlea. The cochleostomy is then plugged with small pieces of temporalis muscle or fascia. The ground electrode is then buried under the temporalis muscle anterosuperiorly. Once the implant is in place, only bipolar electrocautery should be used in order to minimize the risk of electrical current conduction through the device or into the cochlea. Intraoperative measurements (neural-response telemetry and impedence testing) are obtained to confirm proper functioning of the device before or during wound closure. A plain anteroposterior view radiograph of the skull may be routinely or selectively obtained intraoperatively to confirm correct placement of the electrode array within the cochlea (Fig. 129-10). The wound is closed in layers without drainage. Care should be taken to ensure that the musculoperiosteal flap completely covers the mastoid cavity containing the electrode array. A mastoid dressing is applied to the operated ear for 24 hours.

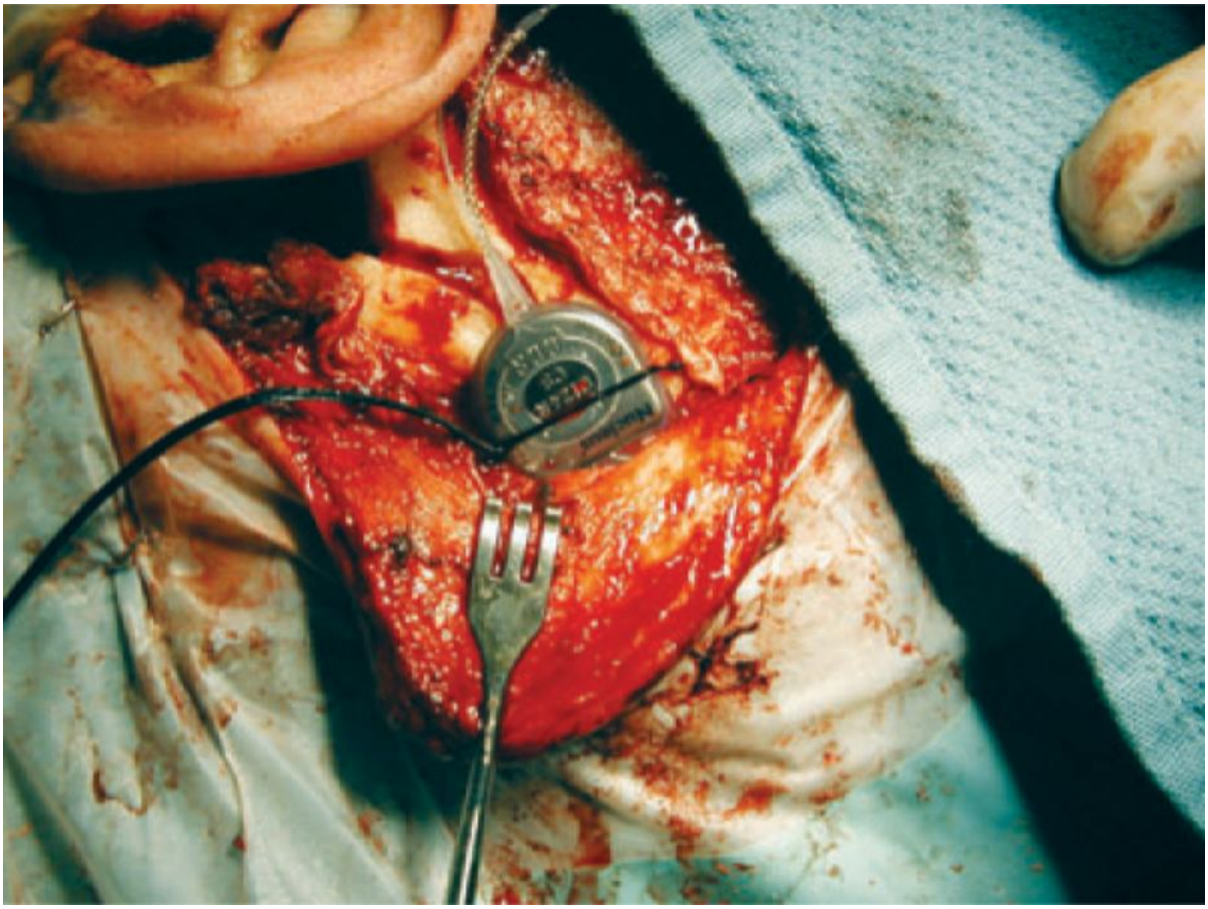

Figure 129-8 Device secured within bony recess and under temporalis muscle.

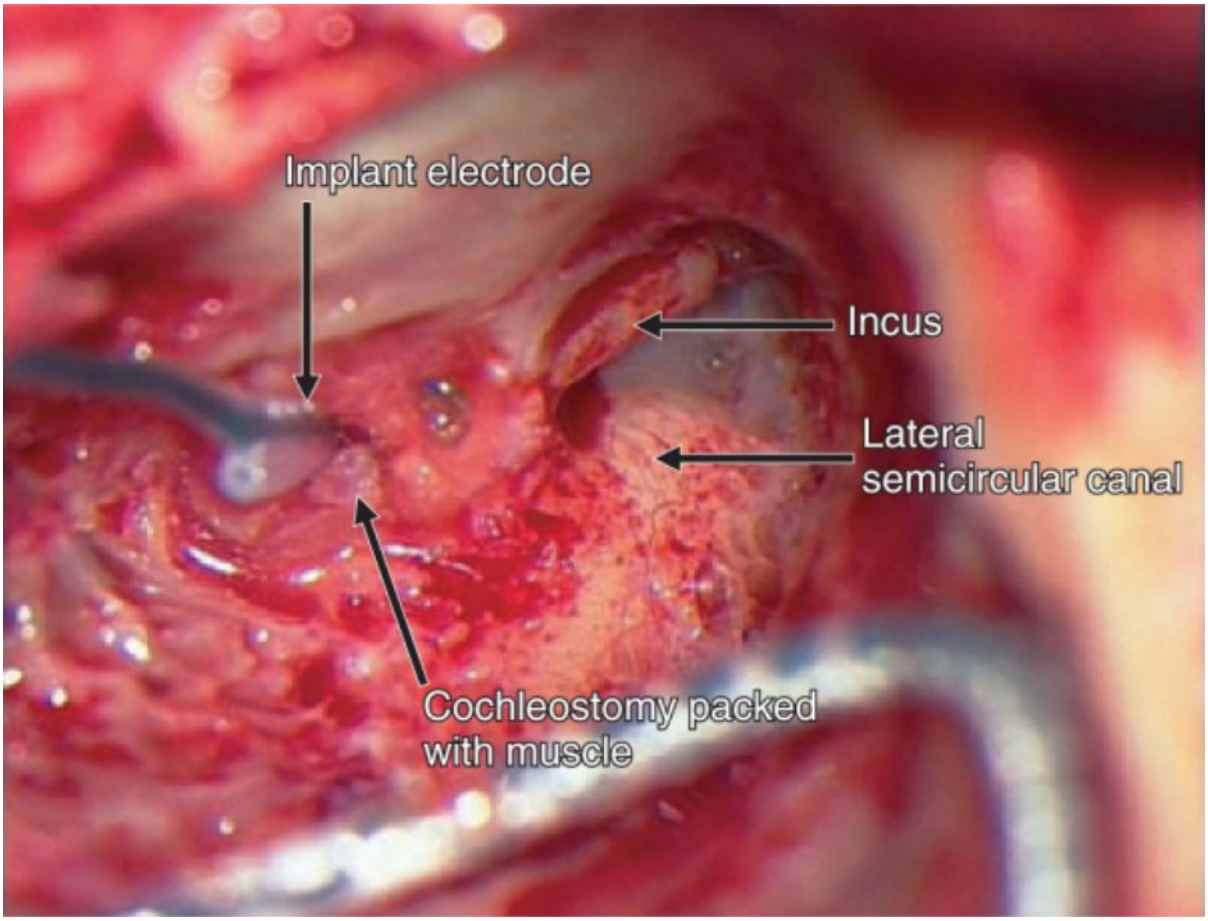

Figure 129-9 Electrode placed in scala tympani through facial recess.

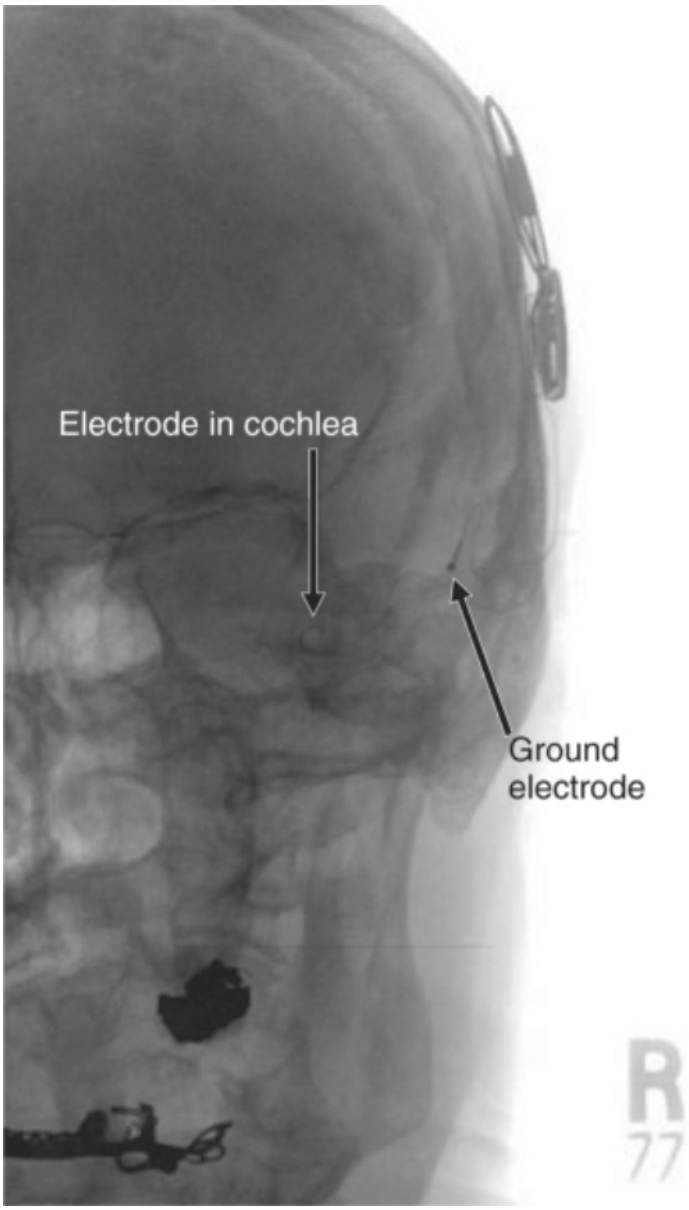

Figure 129-10 Anteroposterior radiograph of the skull showing good electrode placement within the cochlea.

POSTOPERATIVE MANAGEMENT

The patient is discharged home on the day of surgery with a 7-day course of oral antibiotics with antistreptococcal coverage. The patient is instructed to remove the mastoid dressing on postoperative day 1. The initial follow-up visit is in 1 week. The patient is fitted with a signal processor 4 weeks after surgery, and individual programming/rehabilitation begins. Regular follow-up visits with the implant physician and audiologist for the first several years with objective assessment of performance are planned.

COMPLICATIONS

Potential risks of the implant procedure are similar for chronic ear surgery: infection, facial nerve paralysis, dysgeusia, dizziness, cerebrospinal fluid (CSF) drainage, meningitis, risks of general anesthesia, and bleeding.

Wound infection that leads to failure of healing at the incision site is the most common problem associated with implant surgery. Breakdown of the skin flap with potential extrusion of the device can be prevented by keeping the skin incision at least 1 cm away from the edge of the internal receiver and maintaining a 6- to 7-mm thick skin flap overlying the internal receiver and magnet.[7] Any sign of cellulitis warrants aggressive treatment with broad-

spectrum intravenous antibiotics. If implant exposure results from necrosis of the overlying skin flap, local rotational flaps may be necessary to cover the device (Fig. 129-11).

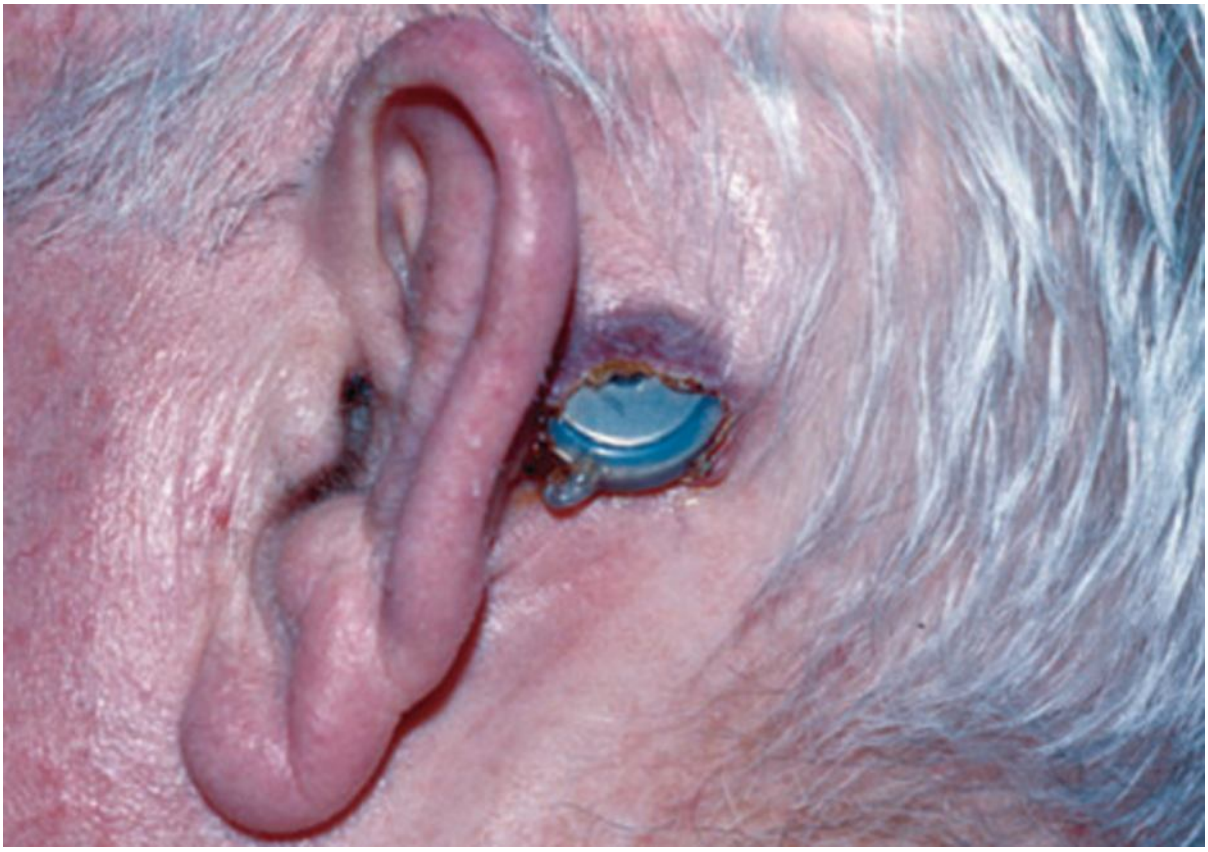

Figure 129-11 Wound breakdown over internal receiver with early extrusion of the device.

Facial nerve injury can occur as a result of surgery. Although intraoperative facial nerve monitoring may reduce this risk, a thorough knowledge of temporal bone anatomy and meticulous surgical technique, along with copious irrigation while drilling in and through the facial recess, remain critical.

CSF leak has occurred when drilling the bony well for the internal receiver in patients with thin temporal squama. A small dural tear can be covered with temporalis fascia or repaired primarily. Firm packing of the cochleostomy site with connective tissue prevents perilymphatic fistulae following electrode array insertion. In patients with congenitally malformed inner ears, a postoperative CSF leak is anticipated based on the preoperative imaging findings. The eustachian tube should be temporarily obliterated before the cochleostomy. The elective placement of a lumbar drain in these cases is optional. If a significant postoperative CSF leak develops despite these measures, the eustachian tube and middle ear may be obliterated.

Worldwide meningitis cases in children and adult cochlear implant recipients reported in 2002 led the FDA and Centers for Disease Control and Prevention to develop recommendations for preoperative vaccination for all implant recipients. Studies have reemphasized the importance of packing the cochleostomy site with soft tissue to prevent bacteria from entering the inner ear and intracranial space.[8,9]

Intracochlear ossification can be anticipated preoperatively on CT imaging of the temporal bone. Surgical management options include drilling through the basal turn of the cochlea with partial electrode insertion,[10] insertion of a split electrode array through two separate cochleostomies in the basal and apical cochlear turns, respectively, to maximize the number of electrodes implanted.[11] The first cochleostomy is created at the standard site, just anterior to the round window membrane. The second cochleostomy is created approximately 2 mm anterior to the oval window.[12] An extensive drillout procedure to gain access to the upper basal turn has also been described by Gantz and colleagues for the ossified cochlea.[13]

PEARLS

- A limited cortical mastoidectomy with inferior exposure down to the level of the inferior external auditory canal is usually adequate for cochlear implantation.

- A combined transmastoid/transcanal approach, leaving the ear canal skin intact, may be helpful for added surgical exposure of the cochlea when a contracted mastoid is encountered.
- Removal of bone medial and anterior to the facial nerve within the facial recess may be necessary for exposure of the round window niche.
- Attention should be directed to inserting the cochlear implant electrode into the scala tympani with the electrode plates directed toward the modiolus.
- The musculoperiosteal flap should be reapproximated such that it completely covers the mastoid cavity with the carrier and ground electrodes within it.

PITFALLS

- Drilling lateral to the chorda tympani nerve when opening the facial recess may result in violation of the tympanic membrane or external auditory canal.
- Thermal injury to the facial nerve may occur while drilling in the facial recess if continuous irrigation is not applied.
- Inadequate surgical exposure and visualization of the round window may result in erroneous placement of the implant electrode within hypotympanic air cells.
- Suctioning at or within the cochleostomy may result in trauma to the surviving intracochlear neural elements, thus limiting postoperative auditory function.
- The presence of congenital inner ear malformations may increase the risk of postoperative CSF leak and meningitis.