

Continuing Education Independent Study Series



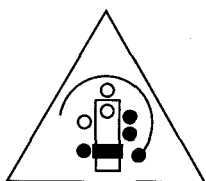
COCHLEAR IMPLANTS AND SENSORINEURAL DEAFNESS

MARY KENNEDY, CST/CFA

**ENT-HEAD AND NECK SURGERY
SHANDS HOSPITAL AT THE UNIVERSITY OF FLORIDA,
GAINESVILLE, FLORIDA**

Association of Surgical Technologists

*Publication made possible by an educational grant provided by
Kimberly-Clark Corporation*



ASSOCIATION
OF SURGICAL
TECHNOLOGISTS

*AEGER PRIMO -
THE PATIENT FIRST®*

Association of Surgical Technologists, Inc.

7108-C S. Alton Way, Suite 100

Englewood, CO 80112-2106

303-694-9130

ISBN 0-926805-15-0

Copyright© 1996 by the Association of Surgical Technologists, Inc. All rights reserved. Printed in the United States of America.
No part of this publication may be reproduced, stored in a retrieval system, or transmitted, in any form or by any means, electronic,
mechanical, photocopying, recording, or otherwise, without the prior written permission of the publisher.



PREFACE

“Cochlear Implants and Sensorineural Deafness” is part of the AST Continuing Education Independent Study Series. The series has been specifically designed for surgical technologists to provide independent study opportunities that are relevant to the field and to support the educational goals of the profession and the Association.

Acknowledgments

AST gratefully acknowledges the generous support of Kimberly-Clark Corporation, Roswell, Georgia, without whom this project could not have been undertaken.

INTRODUCTION

Purpose

The purpose of this module is to (1) refresh the learner's understanding of the anatomy of the ear, the physiology of hearing, and the various otologic disorders linked with sensorineural deafness, and (2) provide a thorough discussion of the history of cochlear implant technology as well as the aspects of patient selection, surgical technique, and expected benefits involved in implantation. Upon completing this module, the learner will receive 2 continuing education (CE) credits in category 3 (applicable to CFA certification).

Objectives

Upon completing this module, the learner will be able to do the following:

1. Describe the anatomy of the ear.
2. Discuss the hearing process and how the anatomic structures of the ear create the sense of hearing.
3. Define sensorineural hearing loss and describe the disorders that cause sensorineural hearing loss, their associated symptoms, and the treatment measures employed.
4. Discuss the history of cochlear implants and how researchers and surgeons developed the technology used in current implant models.
5. Describe the criteria involved in selecting candidates for cochlear implantation and the tests and studies that candidates must undergo.
7. Discuss the successive steps involved in the implantation procedure, including the surgical instruments used; describe the role of special picks and support rings in placing the implant; and discuss the steps following implantation, including placement of the receiver-stimulator.
8. Discuss how the cochlear implant works and how it is programmed during the postsurgical rehabilitation period.
9. Discuss the rehabilitation process for cochlear implant patients, particularly for children.

Using the Module

1. Read the information provided, referring to the appropriate figures.
2. Complete the enclosed exam without referring back to the text. The questions are in a multiple choice format. Select the best answer from the alternatives given.
3. Mail the completed exam to AST, CEIS Series, 7108-C S. Alton Way, Suite 100, Englewood, CO 80112-2106. Please keep a copy of your answers before mailing the exam. You must return the original copy of the answer sheet; this exam may not be copied and distributed to others.
4. Your exam will be graded, and you will be awarded continuing education credit upon achieving a minimum passing score of 70%. If you are an AST member, your credits will be automatically recorded and you do not need to submit the credits with your yearly CE report form.
5. You will be sent the correct answers to the exam. Compare your answers with the correct answers to evaluate your level of knowledge and to determine what areas you need to review.

Studying Technical Material

To study technical material, find a quiet place where you can work uninterrupted. Sitting at a desk or work table will be most conducive to studying.

Having a medical dictionary available as you study is very helpful so you can look up any words with which you are unfamiliar. Make notes in the margins of any new definitions so that you can review them.

The ultimate test of how well you learn this material is your ability to relate your knowledge to what is happening in the surgical field. Apply your knowledge to the progress of the surgery and the postoperative results for the patient.

Acknowledgments

The author would like to thank the Cochlear Corporation of Englewood, Colorado, for all of the information that was provided to her.

COCHLEAR IMPLANTS AND SENSORINEURAL DEAFNESS

The Anatomy of the Ear

The human ear consists of three anatomic regions: the external ear, the middle ear, and the inner ear. The **external ear** is composed of the outer appendage, called the auricle or pinna, and the external auditory canal (meatus). The **middle ear** begins at the tympanic membrane or eardrum, which lies across the base of the external auditory canal. The three bones (auditory ossicles) within the middle ear are the malleus, the incus, and the stapes. The handle of the malleus rests against the tympanic membrane; its head is attached by ligaments to the body of the incus. The incus articulates with the head of the stapes, the base or footplate of which rests in the oval window of the cochlea (Figure 1). The **inner ear** has two

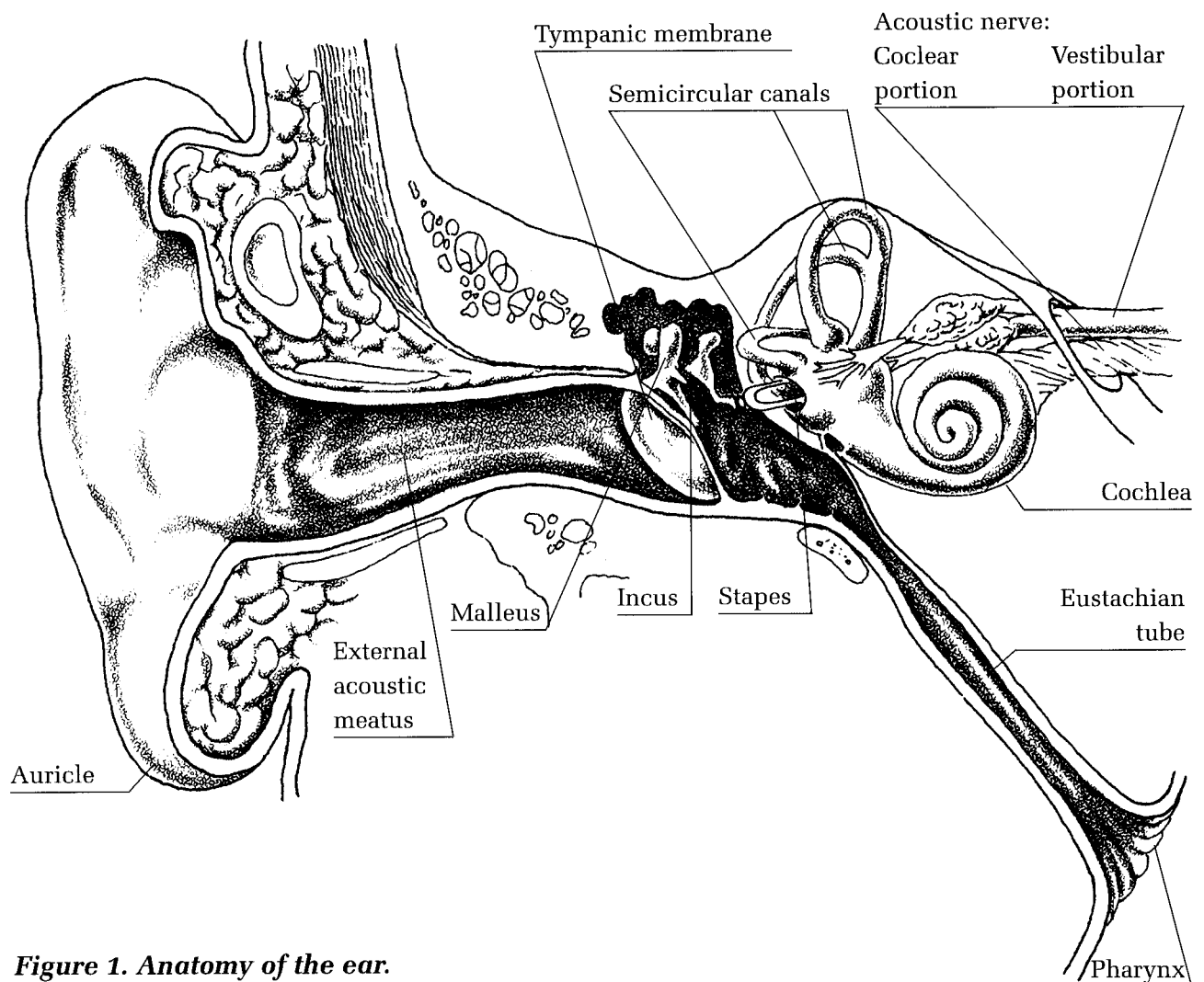


Figure 1. Anatomy of the ear.

divisions, the osseus labyrinth and the membranous labyrinth. The osseus labyrinth consists of the semicircular canals, which control bodily balance, and the cochlea, which contains the organ of hearing. The membranous labyrinth lies within the osseus labyrinth; these structures will be discussed in greater detail later.

The inner and middle ear reside in the temporal bone. Behind the external ear (postauricular area) lies the mastoid. In its normal state, this portion of the temporal bone is filled with holes or “air cells.” Below the mastoid is the sigmoid sinus, which is filled with blood and drains into the internal jugular vein. The facial nerve (cranial nerve VII) travels through the temporal bone and exits in front of the ear to innervate the face. The facial nerve has two branches in the middle ear: one innervates the stapedius muscle that moves the stapes; the other is the chorda tympani, which contains taste fibers from the anterior two-thirds of the tongue. For any major ear surgery, knowing where the facial nerve travels in the temporal bone as well as the location of the sigmoid sinus is important.

The Physiology of Hearing

Hearing, which is a mechanoreceptive sense, is made possible by the ear’s response to the mechanical vibrations of sound waves entering it. The funnel-shaped auricle acts to channel the sound waves through the external auditory canal toward the tympanic membrane. The tympanic membrane is cone-shaped, its concavity facing downward and outward toward the auditory canal.¹ Sound waves strike the tympanic membrane, causing it to vibrate, thereby effecting vibration of the middle-ear ossicles.

The ossicles are suspended by ligaments so that the malleus and incus together act as a single lever, having its fulcrum approximately at the border of the tympanic membrane. The head of the malleus, which is opposite its handle, balances the other end of the lever so that changes in body position are prevented from increasing or decreasing tension on the tympanic membrane.¹

The handle of the malleus is held in a constant state of being pulled inward by ligaments and by the tensor tympani muscle, which keeps the tympanic membrane tense. Such conditions allow sound vibrations acting on any portion of the tympanic membrane to be transmitted to the malleus. The incus moves in concert with the malleus because of their being tightly bound to one another by ligaments. The articulation of the ossicles with one another is such that the stapes is caused to press on the cochlear fluid each time the incus moves inward and to pull on the fluid when the malleus moves outward.

The amplitude of movement of the stapes footplate with each sound vibration is only three-fourths as great as that of the handle of the malleus. The ossicular system serves to reduce the amplitude of this movement while increasing its force at the stapes footplate by approximately 1.3 times. Furthermore, the surface area of the tympanic membrane measures approximately 55 square mm, whereas the surface area of the stapes footplate is approximately 3.2 square mm. This 17-fold difference in surface area, when multiplied by the extent of force (1.3 times) achieved by the ossicular system, allows the energy of a sound wave impinging on the tympanic membrane to be applied—through the pressing of the stapes footplate on the oval window—with approximately 22 times as much pressure on the cochlear fluid as was originally exerted by the sound wave against the tympanic membrane.¹ In the absence of the ossicular system and tympanic membrane, sound waves would travel directly through the middle ear and enter the cochlea at the oval window, resulting in a 30-decibel hearing loss.

Cochlear Function in Hearing

The cochlea is comprised of three coiled tubes lying side by side: the scala vestibuli, the scala media, and the scala tympani. The scala vestibuli and scala media are separated by a thin membrane called Reissner's (or vestibular) membrane, which serves to maintain a special fluid in the scala media that is required for normal function of the "hair cells" (Figure 2).

The basilar membrane separates the scala tympani and the scala media; on its surface lies the **Organ of Corti**. This "organ of hearing" contains mechanically sensitive hair cells, which are the receptor-end organs that generate nerve impulses in response to sound vibrations. Two types of hair cells are present: one row of **internal hair cells** contains nearly 3,500 cells, each cell having a diameter of 12 microns; three or four rows of **external hair cells** contain approximately 20,000 cells, each cell measuring 8 microns in diameter.¹ Both the base and sides of the hair cell synapse with a network of cochlear nerve endings that lead to the spiral ganglion of Corti, which lies in the modiolus (center) of the cochlea. The spiral ganglion

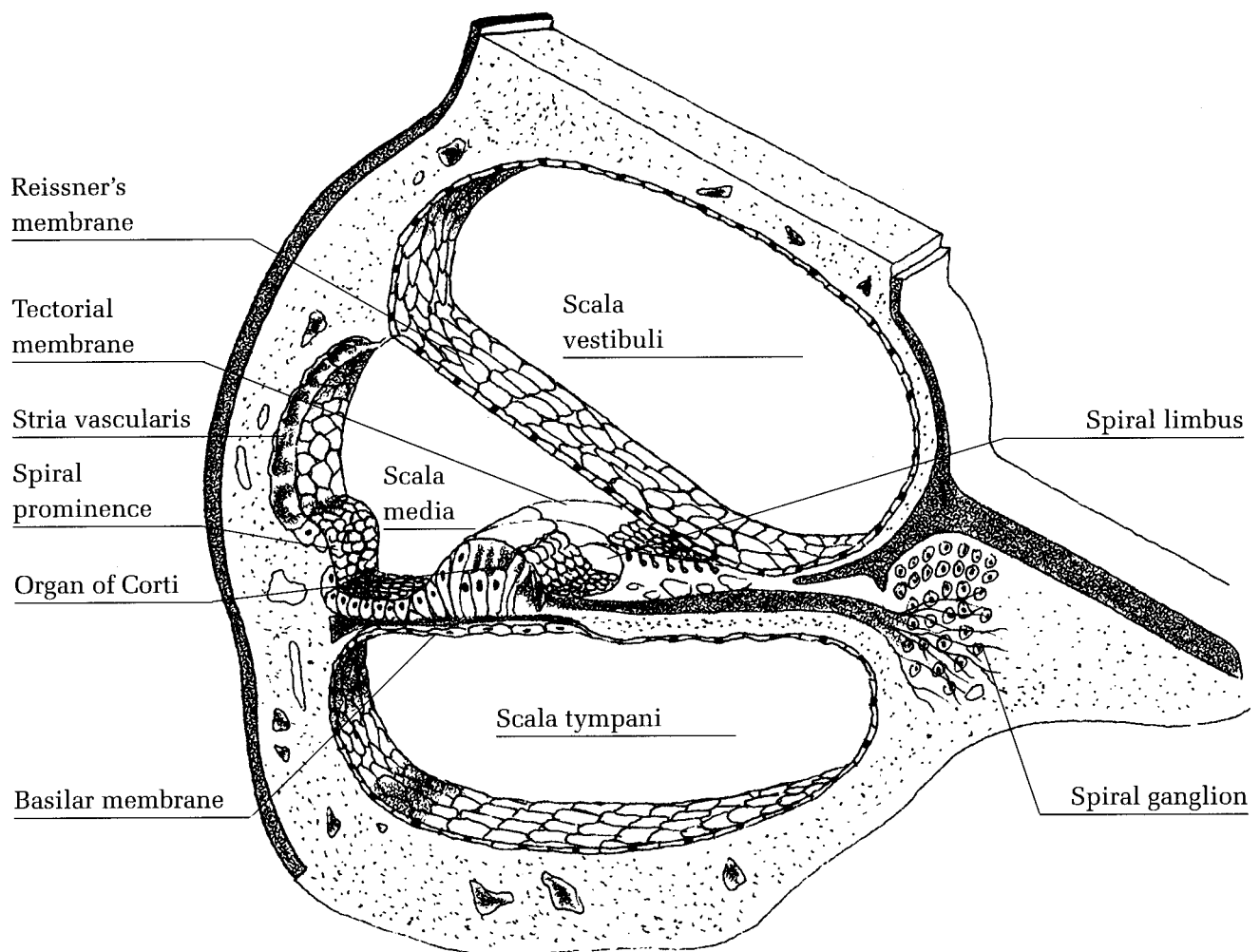


Figure 2. Cochlear anatomy.

sends axons into the cochlear nerve and thus into the central nervous system via the auditory nerve (cranial nerve VIII) at the upper level of the medulla oblongata of the brain.

Hairs project upward from the hair cells and either touch or are embedded in the gelatinous surface coating of the tectorial membrane that lies above the cilia in the scala media. The bending of the hairs in one direction depolarizes the hair cells, thus exciting the nerve fibers and culminating in the sensation of hearing.

Two types of hearing loss are conductive and sensorineural. **Conductive hearing loss** usually results from an interruption in the ossicular system and is caused by conditions such as otosclerosis or cholesteatoma. A variety of treatments exist for conductive hearing loss; their selection is dependent on the medical condition involved. This discussion will explore **sensorineural deafness**, which is the result of some form of breakdown in the neural pathway. A discussion of a variety of otologic disorders that can lead to sensorineural hearing loss, as well as their treatment, follows.

Sensorineural Hearing Loss

Sensorineural hearing loss in children can be attributed to many etiologic factors. It is estimated that one in every 750 infants born will have a potentially disabling sensorineural hearing loss.² The following risk factors may contribute to placing certain infants at high risk of hearing impairment:

1. family history of impaired hearing in childhood.
2. congenital or prenatal infections eg; toxoplasmosis, rubella, cytomegalovirus, herpes, or syphilis.
3. anatomic malformations of the head or neck.
4. low birth weight (less than 1500 grams).
5. unconjugated hyperbilirubinemia at levels exceeding indications for exchange transfusion.
6. bacterial meningitis.
7. severe asphyxia.²

Approximately half of all instances of hearing loss can be attributed to genetic factors; for the majority of these cases, no effective treatment exists.² Because so few of the physical defects associated with hearing loss are visible at birth, congenital hearing impairment often escapes diagnosis. A delay in speech development may be the first indication that a hearing problem exists. For patients afflicted with hearing loss, particularly in the case of pediatric patients, a thorough history-taking forms the basis for diagnosis and treatment. If the patient is an infant, the mother's pregnancy history is an important factor to be considered: many events that occur during the mother's pregnancy may have a deleterious effect on her infant's hearing ability. The patient's physical examination should include a microscopic ear examination as well as a neurologic examination. The auditory evaluation includes measuring air and bone conduction in addition to speech discrimination. Electrocochleography (ECoG) measures the electrical activity of the cochlea in response to sound stimulus. This test can be used to determine whether nerve loss originates in the cochlea, the auditory nerve, or higher structures. Magnetic resonance imaging (MRI) and computed tomography (CT) scans as well as various laboratory tests assist in determining the cause of hearing loss.

Several forms of inner ear disorders that cause sensorineural deafness are subject to medical or surgical treatment and most of these (Meniere's disease, for example) have a better success rate with early

intervention. **Meniere's disease** is characterized by fluctuating hearing loss, a sensation of fullness in the ear, tinnitus (ringing in the ear), and episodic vertigo (dizziness).

Meniere's disease is noted for its distinctive signature on ECoG.³ Early treatment consists of restricting intake of salt and caffeine; smoking cessation; and instigating the use of diuretics, vasodilators, or vestibular suppressants to treat vertigo. If such treatment measures prove ineffective, several surgical options are available that include endolymphatic sac-mastoid shunt, vestibular nerve section, or labyrinthectomy, as indicated by the severity of the disease.

Syphilis can cause inner-ear problems in the form of sensorineural hearing loss and symptoms similar to those in Meniere's disease. Syphilitic hearing impairment is treated with steroids and antibiotics, and prognosis is very good.

Autoimmune-related sensorineural hearing loss is a treatable, yet severe form of hearing loss that occurs when the body's immune system attacks and progressively destroys the inner ear.³ This disorder is diagnosed through an immune screen and is treated with steroids, cyclophosphamide, and plasma phoresis. With early intervention, 95% of the potential destruction to one's hearing caused by this disease can be controlled.

Cochlear otosclerosis is a disorder characterized by flat hearing loss, although the patient's level of speech understanding remains good to excellent. This condition afflicts individuals in their 30s or 40s, and a familial history of stapedial otosclerosis may be identified in some patients. This type of sensorineural hearing loss can be arrested in nearly 85% of patients when treatment consists of fluoride, vitamin D, and calcium supplementation. Fluoride therapy is administered with caution, particularly when treating children and pregnant women; improper dosage can cause premature closure of the epiphyseal plates of developing bones.

Some medications have potentially toxic effects on hearing and balance. These include aminoglycosides (for example, streptomycin), chemotherapy agents, aspirin, vancomycin, erythromycin, loop diuretics (lasix), and certain combinations of these drugs. The extent of hearing damage that can be caused by these drugs varies depending on the inherent ototoxicity of the drug. Streptomycin, for example, is extremely ototoxic; conversely, aspirin is considered not ototoxic and any adverse impact it may have on hearing is easily reversed.

Perilymph fistula, defined as leakage of perilymph fluid from the inner ear into the middle ear through the round and/or oval windows, involves such symptoms as hearing loss and dizziness. This condition can result from an individual's being subjected to a sudden, loud popping noise or to extreme pressure change in the ear, and heavy lifting, straining, or coughing by the patient can exacerbate the symptoms. Conservative treatment consisting of bed rest, stool softeners, and cough suppressants is effective for most patients. If symptoms persist, or if they progress to either positional vertigo or ataxia, surgery may be necessary. A tympanotomy (entailing tissue sealing of the oval and round windows) eliminates the dizziness symptom in 95% of patients and will improve or stabilize hearing ability in 50% of patients.³

Bacterial meningitis causes profound sensorineural hearing loss in many of the patients diagnosed with this infection. Total bilateral nerve deafness is the most usual outcome; unfortunately, the deafness is untreatable even when the most powerful hearing amplification aid is provided. These patients may be considered candidates for cochlear implantation.

Various types of congenital hearing loss are characterized by agenesis or dysgenesis of the components of the inner ear, and most of these anomalies can not be treated.

Michel's aplasia is characterized by the total lack of development of the inner ear. Spaces may be found in the petrous portion of the temporal bone, but no resemblance to normal anatomy exists. The external and middle ears may be formed normally and capable of functioning.

Mondini's aplasia is described as a malformed cochlea that is flattened, and only the basal coil is fully developed: only 1.5 turns of cochlear coil may exist instead of the normal 2.5 turns. This condition rarely is bilateral, but the unaffected ear still bears evidence of malformation. The semicircular canals are underdeveloped, and in some cases the membranous labyrinth is absent. Mondini's aplasia may also be associated with the absence of the oval and round windows as well as other aplastic lesions of the middle ear. Auditory functions in patients with this disorder range from normal hearing to marked deafness, depending on the degree of dysgenesis involved.

In **Scheibe's aplasia**, although the bony labyrinth is fully formed, dysgenesis occurs in the membranous labyrinth. Scheibe's aplasia is the most common malformation associated with a congenital hearing loss, especially one caused by diseases having autosomal recessive gene inheritance.² Because a few hair cells may still be functional, a hearing aid may be useful to these patients.

Aplasia of the cochlear duct, which is an inherited disorder, is called **Alexander's aplasia**. The Organ of Corti (spiral organ) and the ganglion cells of the basal coil are affected, resulting in an inability to hear high-frequency sound waves. Because it is still possible to hear at lower frequencies, a hearing aid may be useful to patients with Alexander's aplasia.

Congenital sensorineural hearing loss is associated with other abnormalities. In the United States, **Waardenberg syndrome** accounts for approximately 2% to 3% of all instances of hearing loss identified as congenital.² Two distinct types of this syndrome have been described. Type 1 features lateral displacement of the inner canthi (dystopia canthorum) and lacrimal points of the eye, a flat nasal root, hyperplasia of the eyebrow, partial or total heterochromia of the irides, and partial albinism revealed in a white forelock. Hearing loss occurs in nearly 25% of these patients. In type 2, dystopia canthi is absent and hearing loss occurs in approximately 50% of these patients.² Such patients have atrophy of the Organ of Corti and a reduced number of nerve cells in the spiral ganglion, which cause their hearing loss.

Pendred's syndrome accounts for up to 10% of the cases of congenital sensorineural hearing loss diagnosed.² This type of hearing loss generally is bilateral and symmetrical; the level of hearing impairment is noted to be most severe when the patient is exposed to sound waves of higher frequency. Goiter is one feature of this disease that usually appears in early childhood and results from defective synthesis of the hormone thyroxine. These patients are known to have malformations similar to those in Mondini's aplasia.

Usher's syndrome is estimated to account for 6% to 10% of all instances of congenital sensorineural hearing loss.² This disorder is characterized by progressive retinitis pigmentosa and a level of hearing loss ranging from moderate to severe. The hearing impairment, which is caused by dysfunction of the cochlear hair cells, usually precedes the onset of visual symptoms.

Down's syndrome is a congenital disorder of chromosomal etiology in which hearing loss is estimated to occur in 40% to 77% of cases.² The hearing loss may be sensorineural, conductive, or mixed. These patients usually have stenosis of the external auditory canal and experience a higher incidence of serous otitis media.

Once the cause and extent of sensorineural deafness in children have been determined, the ultimate goal is to maximize their speech and language development. To achieve this goal, numerous methods of treating hearing loss are available, including the use of vibrotactile devices (usually indicated for blind-deaf patients), hearing aids, and cochlear implants for those who qualify. The child may also be taught manual sign language and finger spelling to improve communications skills. Some experts (representing

the various fields relating to hearing loss) suggest that whenever possible, hearing-impaired children be integrated with normal-hearing children in kindergarten or grade school.

History of Cochlear Implants

More than 200 years ago, electrical stimulation of the ear was discovered to be effective in producing the sensation of hearing. In 1790, Volta inserted metal rods into each of his ears and connected the rods to a circuit that produced approximately 50 volts. On closing the circuit, he reported a sensation resembling a blow to the head followed by a sound described as that which might be produced by the boiling of a viscous fluid.⁴ In the late 1800s, physicians Politzer and Gradenigo advocated the use of electrical stimulation in the diagnosis and treatment of various ear diseases. This form of therapy lost popularity when it was used inappropriately to treat diseases in which the treatment had no benefit.

In 1925, radio engineers discovered that auditory sensations could be produced when electrodes placed near the ear were stimulated by a modulated, alternating current. In 1930, Weaver and Bray discovered the electrical potential of the cochlea when it is triggered by acoustic stimulation. This discovery led to the 1937 studies of Stevens, Jones, Lurie, and Flottorp that pertained to electrophonic hearing: an alternating current established in the audible-frequency range was sent through an electrode to the skin near the ear. The electrode and skin act as two plates of a condenser microphone and the resulting vibrations are transmitted to the cochlea by means of air and bone conduction, thus producing auditory sensations.⁴ In all types of electrophonic hearing, a normal or near-normal cochlea is required. Therefore, stimulation of hearing by electrophonic means had no application in hearing-impaired patients with a malformed cochlea. However, the revelation that the cochlea acts essentially as a transducer of acoustical energy to electrical energy led scientists to investigate the role of direct stimulation of the cochlear nerve in achieving artificial hearing.

In 1957, Djourno and Eyries were the first to stimulate the acoustic nerve in a profoundly deaf person by direct application of an electrode. The individual reported hearing sounds resembling those made by "crickets" or by a spinning "roulette wheel." The patient was also aware of background sounds and could distinguish a few simple words, but was unable to develop any significant powers of speech discrimination.

In 1958, Galambos and Rupert were the first to insert cochlear implants in the United States. In an experiment conducted at Walter Reed Hospital, they placed electrodes in the round windows of the ears of domestic cats. In 1963, the first scientific article on human cochlear implants was published in the United States by Doyle, who was the engineer who work with House. The early electrodes used were insulated with silicone rubber that contained toxic agents, and patients in whom the electrodes had been implanted developed redness and swelling around the surgical wound within 3 weeks postoperatively, prompting electrode removal.

Initially, physicians were hesitant to insert electrodes directly into the inner ear in fear of causing further damage. Subsequent studies proved that direct stimulation of the cochlear nerve within the inner ear was the treatment of choice. In addition, scientists learned that electrical stimulation of the acoustic nerve by means of a single contact electrode probably can not be expected to provide unaided understanding of speech.⁵

The implants currently on the market are both single-channel and multichannel, although the former rarely are used. Through participation in symposiums, physicians have agreed that intracochlear implants should (1) be able to access large numbers of acoustic nerve fibers within the cochlea, (2) allow delivery of safe levels of electrical stimulation without destroying auditory neurons, (3) be designed with

handling properties that allow safe insertion of the electrode by surgeons, and (4) be equipped with electrodes that will function for a lifetime.⁵

In 1985, the FDA approved the clinical use of cochlear implants in adults; in 1990, such approval was granted for children aged 2 years and older, although use was limited to the Nucleus 22 Channel™ (Figure 3).

Cochlear Implant Technology

The cochlear implant is an electronic device that uses minute electrical currents to excite auditory nerve fibers so that signals reach the brainstem's auditory nuclei and ultimately the auditory cortex. Such electrical stimulation results in perception of sound, though differing from normal hearing.⁶

The early cochlear implant model had a single short electrode that served to stimulate available auditory nerve fibers. Variations in pitch were received with marginal distinction by the implant so that sounds were perceived as buzzes. Although both single-channel and multichannel implants are available, the multichannel type is known to provide better speech discrimination.

Cochlear implants have both internal (implanted) and external components. The internal components include the receiver-stimulator and the electrode array; the latter is inserted into the cochlea, while the receiver-stimulator lies in a shallow bed in the mastoid behind the auricle. The external components are placed outside of the patient's body and include a microphone, speech processor, and transmitter. The transmitter is in contact with the receiver-stimulator via magnets. The microphone receives sounds from the environment; these are coded by the speech processor and sent by the transmitter to the receiver-stimulator. The receiver-stimulator directs the

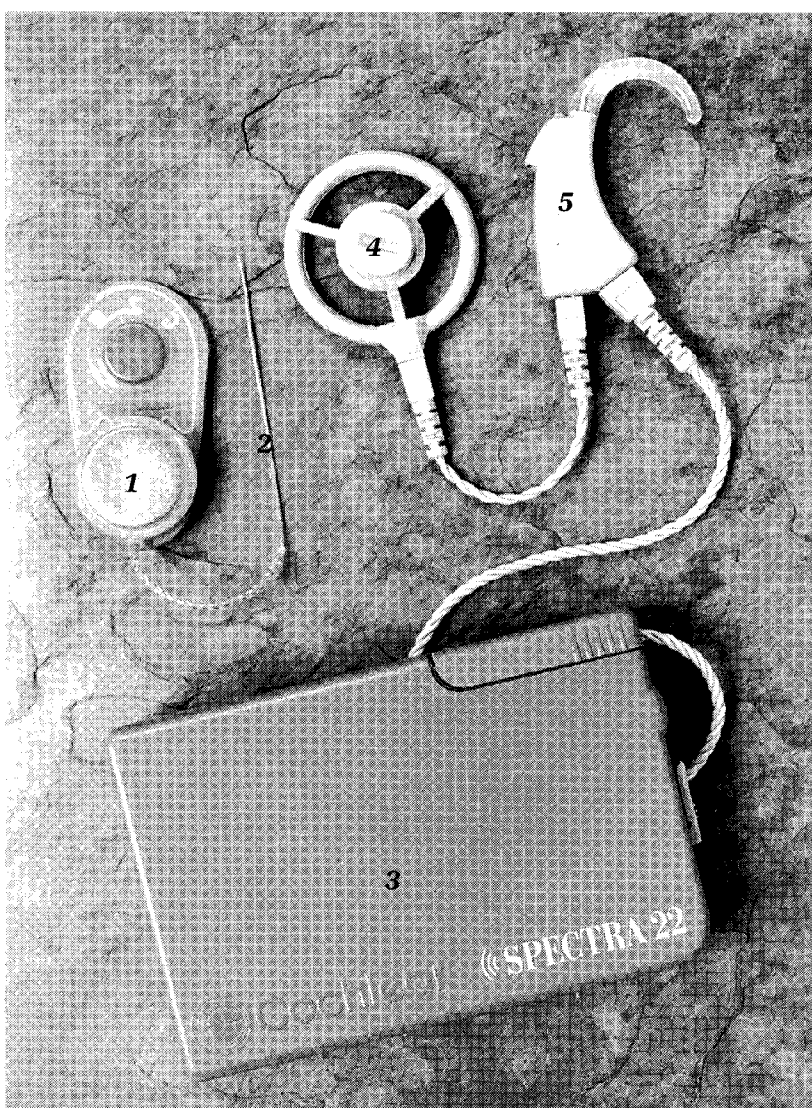


Figure 3. The Nucleus 22 Channel™ cochlear implant. 1, Receiver/stimulator; 2, Electrode array; 3, Speech processor; 4, Transmitter; 5, Microphone. (Photo provided courtesy of the Cochlear Corporation, Englewood, Colorado)

activation of selected electrodes at appropriate energy levels. The current emanating from the electrodes of the cochlear implant directly stimulate the nerve fibers while effectively bypassing damaged receptors. Cochlear implants currently are approved for unilateral use; however, when approved, bilateral use may improve the patient's quality of hearing.

Patient Criteria

Candidates for cochlear implantation are carefully selected on the basis of auditory, medical, psychological, social, and in some instances educational criteria.⁶ The basic auditory profile of such patients includes a diagnosis of sensorineural, bilateral hearing loss (classified as severe to profound) and an inability to recognize speech through hearing aid amplification. Adult patients must have been deafened postlingually (ie, deafened after having learned oral speech and language). If an adult patient was deafened before he/she could speak or recognize language, or was born deaf, he/she generally is not considered a candidate for cochlear implantation. Such persons are considered to have become members of the deaf community, as signified by communicating through American Sign Language (ASL), and these individuals usually have little need for or interest in auditory information. Similarly, deaf children born to deaf parents who use ASL and have strong ties to the deaf community rarely are considered to be candidates for cochlear implantation.⁶ In fact, some members of the deaf community oppose the use of cochlear implants in deaf children of hearing parents because they believe that hearing parents deprive the deaf child of a "birthright to silence" and therefore should not be permitted to make a decision in favor of implantation. Others fear that in providing functional hearing to deaf children, cochlear implants may reduce the number of those choosing to be associated with the "deaf culture."⁶

Medically, the implantation candidate must be able to undergo surgery that is conducted under general anesthesia. Even those patients who have serious conditions (such as leukemia, multiple sclerosis, or cerebral palsy, or who have undergone organ transplantation) are known to have had a successful outcome as cochlear implant recipients. In terms of psychological preparedness, the candidate must harbor realistic expectations of the implant's effect on his or her life. If a patient holds unrealistic beliefs about the implant's ability to restore normal hearing, he/she will be required to undergo counseling and must be denied candidacy if an exaggerated level of expectation persists. In addition, the patient must express a desire to join the hearing world.

An important factor in patient selection for cochlear implantation is the existence of familial and social support, particularly in the case of pediatric patients who are family-dependent. Evidence of strong family support and the family's holding appropriate expectations is crucial. Pediatric patients also require an educational setting that is conducive to maximizing the benefits of the implant: this is achieved when auditory skills training is emphasized in the school curriculum.

Finally, the financial aspect of implantation must be considered: implants range in cost from \$14,000 to \$18,000; surgery and rehabilitation expenses are additional concerns. Recently, the federal Medicare and Medicaid programs accepted cochlear implantation as a procedure for which funding would be provided. Patients must investigate whether sufficient insurance coverage is available to them or obtain adequate funding prior to undergoing the procedure.

Once approved for cochlear implantation, the patient completes the usual preoperative tests that are required in cases involving general anesthesia. A CT scan of the head is performed in order to determine which ear will receive the implant.

Surgical Technique

In most cases, cochlear implantation is a 2- to 3-hour procedure. The patient is placed in the supine position on the operating room table with the head turned so that the ear to be implanted is exposed. The table is positioned according to the surgeon's wishes, and the patient is appropriately padded. The head is shaved in the temporal region of the implanted side; the amount shaved is specified by the surgeon. The surgeon and anesthesiologist together must determine if a Foley catheter should be placed: a urinary tract infection resulting from catheterization may cause the implant site to become infected, potentially undermining the proper functioning of the implant. The patient is appropriately prepped and draped according to the surgeon's preference.

A postauricular flap is made in the area immediately behind the auricle and extends outward along the temporal bone; this incision is another aspect of the surgery that is dependent on the surgeon's preference. The skin flap is dissected away from the mastoid and temporal bone, and a portion of the temporal fascia or muscle may be harvested for later use. The flap is retracted, and the surgeon performs a mastoidectomy with a facial recess. It is important to locate the facial nerve, passing as it does through the mastoid, so that the nerve will not be injured while the surgeon locates the round window of the cochlea. Most surgeons use a facial nerve monitor that assists in locating and preventing damage to the nerve.

The round window of the cochlea is located and, prior to the cochleostomy itself, several tasks must be completed in preparation for the implant. First, a shallow, round bed is made in the mastoid to seat the implant, requiring that the sigmoid sinus be carefully avoided. A special milling burr is used, and an implant dummy is fitted into the bed to ensure proper fit. Some surgeons may place the implant under the muscle layer overlying the temporal bone in such a manner that would permit the patient to continue wearing glasses comfortably; in this instance, an implant bed would be unnecessary. Next, a groove is made for the "tail" of the implant to sit comfortably in the mastoid, a configuration that lessens the potential for electrode displacement. Once the drilling is complete, the entire area is irrigated to remove any bone dust. The cochleostomy is performed with a small drill (such as a stapes drill) using a 1-mm bit. Following this, the implant is brought to the field and the surgeon prepares it for implantation. Special cochlear implant picks are used to help guide the electrodes into the cochlea, and the surgeon turns the implant as needed to ensure that all of the electrodes are properly guided into the cochlea. At the implant end of the electrode array are support rings that keep the electrodes from being pulled out of the cochlea. Ideally, the surgeon is able to maneuver all or most of these support rings into the cochlea.

Hospitals are required to provide documentation to the Cochlear Company (on special forms that accompany the implant) stating the number of support rings that were actually placed in the cochlea by surgeons during the implantation surgery, and records containing such data are maintained in the event that implant malfunction occurs at a later time (original documents are sent to the Cochlear Company; copies are retained in the patient's medical record.) The surgeon also documents this information on the operative record.

Once the implant is placed in the cochlea, it is important that the bovie must be either turned off or taken away from the operative field because the electric current may travel up the electrodes and potentially cause damage to them, to the auditory nerve, or to the brain. Thereafter, bipolar cautery is used to maintain hemostasis. Prior to implantation, some surgeons may inspect the flap as well as the operative site in general to ascertain whether use of a bovie is indicated. After the implantation, surgeons may choose to place small plugs of muscle or fascia around the cochleostomy site to prevent leakage of perilymph fluid. The receiver-stimulator normally is sutured in place using nonabsorbable sutures with a

figure-of-eight stitch to immobilize it and to prevent the electrodes from being pulled out of the cochlea. Finally, the flap is closed and a large mastoid dressing is placed over the site to provide pressure. Most surgeons employ a small drain that is removed the following day. Patients generally are admitted for 23-hour observation, although many surgeons are choosing to perform cochlear implantation on an outpatient basis.

Postsurgical Rehabilitation

The surgical site normally requires a healing period of 4 to 6 weeks before fitting of the external component can be attempted. The patient then begins the period of rehabilitation during which the implant is programmed and the patient is assisted in learning to use the sounds that he/she now is able to hear. In the first programming session, the internal components are connected to a desktop computer, and the amount of electrical current sent to individual electrodes is controlled by means of a computer program provided by the implant manufacturer. An audiologist begins the session by stimulating one electrode at a time with a small amount of current, the level of which is raised gradually until the patient perceives a sound, typically a beep. Because of the placement of the electrodes within the cochlea, the stimulation of each results in the patient's perceiving different pitches. Stimulation of the apical electrodes results in sounds of lower pitch; the basally positioned electrodes convey sounds of higher pitch.⁶ Thresholds for sound perception are established so that the amounts of current that result in loud but not painful sounds can be determined. Such data allow the computer to map an acceptable range of current for each electrode. This programming procedure can be completed in several hours' time for adults, whereas children may require a period of several days in which to complete this process.

A final stage entails disconnection of the implant from the computer and its subsequent connection to a microphone, allowing the patient finally to hear sounds from the environment. The microphone detects a sound; the speech processor analyzes its pitch and loudness and stimulates the appropriate electrodes. Rehabilitation for adults involves speech training exercises consisting of increasingly difficult listening tasks (for example, distinguishing similar-sounding words).

Successful rehabilitation in pediatric patients depends on the individual patient. Children who lost their hearing postlingually hold an advantage over those who were congenitally deafened. Congenitally deafened children have no point of reference on which to base the perception of sound: when first enabled to perceive sound, their initial reaction may be that of surprise or fright; some exhibit marginal overt awareness of the sounds to which they are subjected. For these children, the process of learning language and how to distinguish sounds requires daily training in which the child's attention is drawn to spoken words; subsequently, the meaning of those words is provided. Most younger children generally require 1 to 2 years in which to realize the full potential of their cochlear implants.⁶ Although congenitally deafened children tend to require a lengthier period of adjustment, they may receive substantial benefit from implantation. Additionally, the length of time that a child was deaf (prior to his/her receiving the implant) may influence the rate of progress, because the ability to learn language peaks early in life.

Most recipients of cochlear implants can expect improvement in their speech-reading ability and are able to hear and recognize a variety of environmental sounds such as the ringing of a doorbell or telephone. Nearly half of all adult patients become capable of understanding some speech without the assistance of visual information.⁶ A few patients can even conduct telephone conversations.

Because children who have received cochlear implants are growing and maturing while learning to use their implants, determining the extent to which the child's progress can be attributed to the presence

of the implant as opposed to normal development is difficult for physicians. However, studies have shown that pediatric patients treated with cochlear implants improve more rapidly in their receptive and expressive language skills than do those using hearing aids. Outcomes vary according to the individual patient.

Conclusion

Many patients with sensorineural deafness may not be eligible for treatment because of the cause of their hearing loss. Patients whose disorders have not affected the anatomy of the inner ear, however, can receive treatment with a good prognosis in most cases. Those patients who have profound bilateral deafness—and for whom treatment with hearing aids is ineffective—may be candidates for cochlear implantation. Such candidates must undergo a series of critical studies before implantation can be authorized. In these patients, the auditory receptors are impaired by the disease process, although the cochlea itself is normal.

The electrodes of the cochlear implant are placed directly into the cochlea to stimulate the cochlear nerve, which is a branch of the acoustic nerve. Early implants were placed near the cochlea in the fear that structures of the inner ear may otherwise become damaged. Later studies showed that direct stimulation of the cochlear nerve produced better sound perception; therefore, intracochlear implants became the treatment of choice.

Both single-channel and multichannel cochlear implants are available, although studies have shown that better sound discrimination and pitch perception can be achieved with multichannel implants. These studies also show that little appreciable difference exists among the different types of multichannel implants (ie, models having 4, 8, or 22 channels). The single-channel implants rarely are used.

The FDA has authorized the use of cochlear implants in adult patients for more than a decade, and since 1990, only one device, the Nucleus 22 Channel™, has been approved for use in children. Patients who have undergone cochlear implantation must undergo an extensive rehabilitation program. Congenitally deafened children who receive implants seem to require a longer period of time to demonstrate progress, having had no prior experience in hearing. Expense is an important consideration in cochlear implantation because the cumulative costs of the implant device, the surgical implantation, and the rehabilitation process may exceed \$25,000.⁶ The most important component of the cochlear implant is the speech processor, the device that processes environmental sounds and speech and directs the receiver-stimulator to stimulate certain electrodes; thus, when considering the attributes of a given implant, the speech processor component and its particular abilities must be evaluated carefully.

Physicians and engineers continue to contribute their expertise toward improving the cochlear implant and the speech processor. Also under development is a brainstem implant (functioning in nearly the same manner as the cochlear implant) for the benefit of patients who have undergone acoustic nerve resection in the treatment of an acoustic neuroma. In addition, studies are being conducted to determine whether a benefit exists, in the areas of speech understanding and discrimination, for patients who receive bilateral cochlear implants: a better quality of life is anticipated for such persons.

Most patients who have received cochlear implants declare satisfaction with their devices, stating that quality of life has improved for them through hearing restoration; an enhanced enjoyment of life is made possible. Children appear to receive the greatest benefit from cochlear implants because in these patients, normal development evolves concurrently with acclimation to the implant. Cochlear implant technology, unlike that of many other medical implants and devices, is considered still to be in its

infancy; therefore, many significant advances in the field of cochlear implantation are anticipated in the years to come.

References

1. Guyton AC. *Textbook of Medical Physiology*. 6th ed. Philadelphia, Penn: W. B. Saunders Co; 1981.
2. Paparella MM, Fox RY, Schachern PA. Diagnosis and treatment of sensorineural hearing loss in children. *Otolaryng Clin North Am*. 1989;22:51-71.
3. Hicks GW, Wright JW. Treatment of sensorineural hearing loss. *Indiana Med*. August 1991:540-544.
4. Gray R, ed. *Cochlear Implants*. San Diego, Calif: College Hill; 1985.
5. Schindler R, Merzenich M, eds. *Cochlear Implants*. New York, NY: Raven Press; 1985.
6. Telischi FF, Hodges AV, Balkany TJ. When to refer cochlear implants for deafness. *Hospital Practice*. May 15, 1994:55-66.

Suggested Readings

1. House JR, Luxford WM. Facial injury in cochlear implantation. *Otolaryngol Head Neck Surg*. 1993;109:1078-1082.
2. Hoffman RA, Cohen NL. Surgical pitfalls in cochlear implantation. *Laryngoscope*. 1993;103:741-744.
3. Doyle PJ, Sipke P, Noel FJ. The cochlear implant: a comparison of single-channel and multichannel results. *J Otolaryngol*. 1991;20:204-208.