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Hearing Loss

Hope Through Research



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Cover:
A wax mold ensures a custom fit for a new hearing aid.

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Hearing Loss— Hope Through Research

Some years ago pollsters asked a sample of adults, "If you had to choose between becoming blind or becoming deaf, which would you choose?" A surprisingly large number chose blindness. As terrifying as a world of darkness may have seemed, a world of silence seemed even worse.

When you think about it, the reasons are not hard to fathom. Human beings are talkers, social creatures who seek out their fellow beings for conversation and contact. From birth on we cry and coo, look and listen, in our first attempts to communicate. But for over 200,000 newborns and young infants who are born deaf or suffer severe hearing loss in the first years of life, that vital communications link never gets forged. Not only are deaf children denied the wealth of experience that comes from listening to the sounds of nature, of music, and of the human voice, but they must also struggle hard to master speech and language.

For those who become profoundly hard of hearing later in life, the impact is hardly less tragic. At present nearly 2 million Americans are either totally deaf or suffer such significant hearing loss in both ears that they cannot hear conversation, a phone ringing, traffic noises, or a fire alarm. Another 14-15 million are moderately to severely impaired.

All these people know the loneliness, isolation, and frustration that comes from hearing loss. They know what it's like to sit in company and miss the joke or the gist of conversation. Worse, they soon realize that society can be cruel. People are impatient. They don't like repeating their words or raising their voices. They show by facial expression and gesture how annoyed they are when talking to someone "who doesn't understand what I'm saying." It is only a short step from that attitude to the assumption that people who are hard of hearing are soft in intelligence. Ages ago that assumption led to the vulgar use of the word *dumb* to mean stupid. *Dumb* derives from Old English roots that mean mute, unable to speak; once the common fate of those born deaf.

Fortunately, many elements in society—including the hearing impaired themselves—have rallied to fight ignorance and prejudice. Such well-known figures as Henry Fonda, Nanette Fabray, Lou Ferrigno (TV's Incredible Hulk), and *New Yorker* cartoonist and children's book author William Steig have publicly acknowledged their hearing problems. Their example, along with individuals as celebrated as Beethoven, Thomas Edison, Helen Keller and Winston Churchill, has helped dispel the embarrassment that many hearing-impaired people feel, an embarrassment that often makes them deny they have a problem and avoid seeking treatment or wearing a hearing aid.

Pediatricians, family physicians, ear specialists and parents are increasingly aware of the need to diagnose hearing impairments early in life, so that remedial



Rare photograph of Helen Keller, her teacher Annie Sullivan, and Alexander Graham Bell.

The sooner hearing loss in
infancy is diagnosed, the
better.

measures and language and speech training can begin. Many lay and professional organizations have formed to aid those with hearing impairments.

In 1975 one of the 11 research Institutes of the National Institutes of Health in Bethesda, Md., added *communicative disorders* to its name to emphasize its increasing attention to research on hearing, speech and language problems. The National Institute of Neurological and Communicative Disorders and Stroke (NINCDS) is the principal Federal agency supporting research on the many causes of hearing loss, as well as on prevention, treatment, and rehabilitation. Support includes the development of better hearing aids and other ways to augment hearing; research on hearing *gains*, one might say, to counter hearing *losses*.

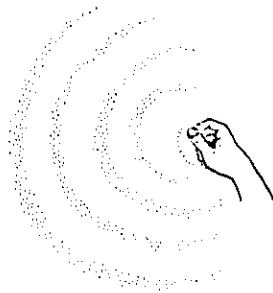
Through basic studies of normal hearing and studies targeted on specific impairments, the primary goal of the NINCDS research program is to develop improved methods of *prevention*. But until this goal of prevention is achieved, research attention will continue to be directed toward better therapies for the hearing impaired.

Sound and hearing

Human hearing depends on a series of mechanical and electrical events that enable sound waves in air to be converted to electrical impulses carried by nerves to the brain.

Sound itself is a form of energy. Suppose you snap your fingers. The snap generates a force that presses against the molecules of air surrounding your fingertips. The molecules are pushed out a short distance in all directions, crowding into space occupied by other air molecules so that a densely packed shell of air molecules forms. That shell—sound experts call it a “shell of compression”—in turn presses against other air molecules nearby so that they, too, are pushed out to form a second, slightly larger shell of compression, which nudges a third layer of air, and so on.

Meanwhile, the air around your fingertips has become less dense as a result of those first molecules being



When you snap your fingers the sound energy creates shells of compression and rarefaction in air.

pushed out. A partial vacuum is created by this "rarefied" air, and the molecules that moved out now rush back to fill that vacuum. Their return creates a second partial vacuum in their wake, which the molecules of the second shell rush back and fill. And so it goes. Thus, the original sound energy generated by your fingersnap moves through the air on a "wave" which is really a succession of shells of compression and rarefaction created by molecules moving back and forth—vibrating.

The number of shells of compression that pass a given point every second determines the frequency of the sound, measured in cycles per second (cps) or Hertz (Hz). Human beings interpret frequency as pitch; the greater the frequency, the higher the pitch. How far the molecules move back and forth as they vibrate is a measure of the energy or intensity of sound. Human beings interpret sound intensity as loudness.

Our ears are sensitive to only certain ranges of frequency and intensity. Healthy young adults can hear notes as low as 20 Hz—lower than the lowest notes of a bass fiddle—as well as sounds at 20,000 Hz, beyond the

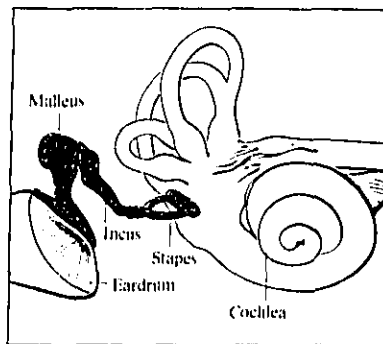
upper reaches of a flute. The intensity range to which our ears respond is enormous. When sound is just audible—the threshold level of hearing—the force of sound waves acting on the ear is about 140 million times smaller than the force needed to lift a 1-ounce weight. At the other extreme, human ears can respond—painfully—to sonic booms, explosions, or the noise of jackhammers breaking up city streets.

Because the ear can respond to such enormous ranges of sound energy, intensity is measured in ratios. That is the basis of the decibel (dB) scale. A sound 10 times more intense than another at the same frequency differs from it by 10 dB; a sound 100 times more intense differs by 20 dB. The decibel scale is usually set at an arbitrary zero level (0 dB), which does not mean the absence of sound, but the average threshold level of hearing of healthy young adults. On that basis, a whisper is about 20 dB, and normal conversation about 60 dB. The noise of a jet taking off is on the order of 160 dB—10 quadrillion times the zero level!

Tuning in

Understanding these fundamentals of sound explains a lot of what goes on—and what can go wrong—with the hearing process. Initially the job of the ears is to pick up sound waves and conduct them accurately to the inner ear. That the ears can manage this task with great skill and efficiency is due in part to the design of the outer and middle ear.

When sound waves enter the ear they travel for an inch or so down a narrow tube, the external auditory canal, before striking the delicate, skin-covered tympanic membrane, or eardrum. The drum is shaped like a broad flat cone about 1/2-inch across and less than 1/50-inch thick. The drum vibrates in tune with the sound waves striking it and transmits the vibrations accurately to three tiny bones in the middle ear, the ossicles. These bones—the malleus, the incus, and the stapes—amplify the vibrations so that the waves can pass on to the inner ear.

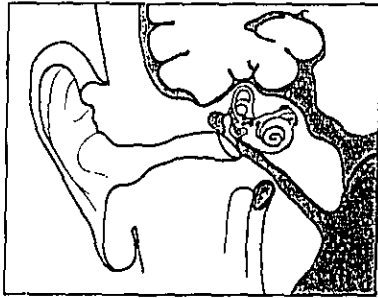


The bones of the middle ear shown in position in relation to the eardrum and inner ear.

It's not hard to understand why specialists find the ear a stunning example of design as well as a challenge for study. For the ear's high-fidelity equipment is miniaturized. The ossicles are the smallest bones in the human body; they fit into a stringbean-seed sized cavity that has been carved out of the temporal bone of the human skull.

Two other features of the middle ear are important: One is that the compartment connects to the throat by a narrow canal with collapsible walls, the eustachian tube. When you swallow, the eustachian tube opens so that air pressure in the middle ear and throat is equalized. That mechanism protects your middle ear from harmful pressure differences that can occur in a fast-rising elevator, for example, or on takeoff and landing in an airplane. The second important middle ear feature is also protective. Muscles attached to the ossicles automatically contract in response to loud noises. These automatic reflexes may prevent strong sound pressures from damaging the delicate structures of the inner ear.

To summarize, when the eardrum and middle ear bones are working properly, sound waves striking the



The organ of Corti, containing the auditory receptors and nerve fibers embedded in a snail-shaped bony canal, collectively called the cochlea.

drum are faithfully conducted across the middle ear and boosted in energy. The energy boost helps prepare sound waves that have been traveling in air for the more resistant watery medium of their next stop: a fluid-filled bony shell in the inner ear called the cochlea.

Conductive problems

A variety of problems can affect hearing before sound reaches the cochlea. Because these early stages in the hearing process are concerned with picking up and conducting sound signals, specialists refer to the hearing impairments involved as *conductive* problems. Among the most common problems are:

- *External blockage.* Sometimes there is a buildup of wax in the ears. Sometimes children put things in their ears. Sometimes a bug crawls in. These are obvious plugs that partially block sound. The removal of impacted wax and foreign objects is best left to experts to avoid the possibility of damaging the eardrum.
- *Perforated eardrum.* A hole or a tear in the eardrum can occur as a result of injury, sudden pressure

Otitis media is the leading cause of conductive impairment.

change, or infection. Ear specialists can repair or completely rebuild the eardrum using the latest techniques of microsurgery.

- *Genetic and congenital abnormalities.* Malformations of the outer and middle ear sometimes occur in connection with hereditary disease or as a result of injuries and illnesses that affect a baby before or around the time of birth. Surgery can sometimes correct these problems.

- *Otitis media.* By far the most prevalent cause of conductive impairments is a common middle ear disease, otitis media. The problem can occur at any age but is particularly prevalent in children. An estimated two-thirds of preschoolers have at least one episode. The reason that children are so vulnerable may be that their eustachian tubes are shorter and positioned more horizontally than in adults. Infectious agents causing colds or other upper respiratory disease can easily spread to the middle ear. At the same time, mucus, pus, or other fluids accumulating in the middle ear tend not to drain off. Thus the middle ear can become inflamed, swollen, fluid-filled and painful—the classic symptoms of otitis media that can result in temporary and sometimes permanent hearing impairment. Thanks to today's medications, most middle ear infections can be cleared up with no lasting damage.

Some children are particularly prone to middle ear disease, however, suffering five or six bouts a year. When the condition recurs that often and is accompanied by fluid in the ear, it is called chronic otitis media with effusion (also serous otitis media). NINCDS-supported scientists at the University of Minnesota are currently studying chronic otitis media to determine if changes in the body's immune system are involved. One possibility is that the body's immune defenses successfully fight off the initial infection, but that some residue of the virus or infectious agent remains to stimulate an immune reaction, leading to the accumulation of fluid. NINCDS is also supporting a

major Otitis Media Research Center in Pittsburgh, Pa., in association with Children's Hospital of the University Health Center. The center will study all aspects of otitis media in animals and in human patients.

Patients with chronic middle ear disease with effusion need careful supervision and treatment by ear specialists (otologists) or ear, nose, and throat specialists (otolaryngologists). Not only is there danger of permanent hearing impairment, but frequent bouts of temporary hearing loss may in themselves be serious. The auditory system, especially in the developing years, needs the regular stimulation of sound for healthy growth. Periodic episodes of hearing loss may starve the auditory cells in the brain and contribute to impairments in speech and language skills. Studies of such "auditory deprivation" are under way by NINCDS grantees at Louisiana State University in New Orleans.

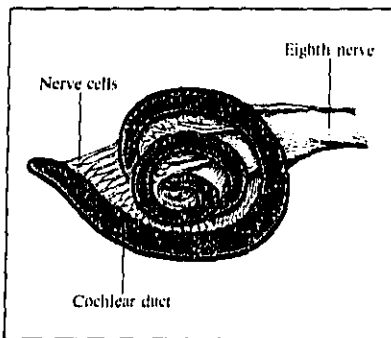
Physicians treating otitis media with effusion can drain the fluid by making a small incision in the eardrum. The incision will heal and the problem may disappear. If fluids continue to build up, however, the physician may insert a small drainage tube in the eardrum in an operation called a tympanostomy. Tympanostomy tubes are sometimes dislodged accidentally



A close look inside the ear with an otoscope is an essential part of any ear examination.

and may be rejected by the body, but often they may stay in place for as long as a year and effectively control the problem. Because there is a small risk that the tubes can give rise to scarring or thickening of the drum after removal, some doctors prefer not to use them, and choose instead to wait and see if the ear problem will improve in time.

• *Otosclerosis.* An example of a hereditary hearing problem that develops in adults is otosclerosis, a condition in which there is an overgrowth of bone in the middle ear. Usually the tiny stirrup-shaped stapes bone is the most affected and becomes fixed in place, impeding sound conduction. Otosclerosis can often be remedied by surgery to remove the excess bone and replace all or part of the stapes with an artificial part. Those who have undergone successful surgery describe the results as miraculous. "I was completely deaf before the operation," one woman said. "As soon as I woke up I could hear again!"



The snail-shaped cochlea has been cut away to show the center compartment, the soft cochlear duct that contains the organ of Corti. The nerve fibers stimulated by hair cells spiral around the cochlea and come together in the eighth nerve.

Delicate “hair cells” in the ear translate sound to electricity.

• *Presbycusis*. Specialists have coined the word *presbycusis*—literally, old hearing—to describe hearing impairments that occur in aging. While presbycusis is primarily associated with changes in the inner ear and brain (discussed later), conductive impairments may also occur. The bones of the middle ear may become stiff, for example, or the eardrum thicker and less flexible. Both those changes may reflect a less rich blood supply to the ear as a result of heart disease, high blood pressure, or other circulatory problems in older people.

Conductive impairments can be detected in the course of an ear examination that includes a variety of diagnostic tests. Before we describe the tests, let us pick up the story of what happens to sound when it reaches the cochlea.

From ear to brain

When sound vibrates the three middle ear bones, the last in line, the stapes, presses against a membrane called the oval window. This membrane is fitted into a thin shell of bone that encloses all the inner ear structures. About an inch down from the oval window, the bone spirals to form the snail-shaped cochlea, another ministructure less than 1/2-inch across at its base, rising a mere 1/4-inch to its tip.

The cochlea is composed of three fluid-filled compartments. The center and smallest compartment is a duct of soft tissue that contains the organ of hearing, called the organ of Corti, after the Italian scientist who first described it. Like the retina of the eye, the organ of Corti contains special cells called sensory receptors that take incoming energy—light in case of the eye, sound for the ear—and transform that energy into electrical signals. The ear cells that do the transforming are called hair cells because the cell tops are fringed with fine hairs that stick up into the fluid filling the duct. The hair cells are sandwiched between two membranes: one membrane rests lightly on the hair tips; the other forms the floor or base of the duct, and so is called the basilar membrane.



Microscopic clusters of hairs fringe the tops of auditory cells in the inner ear.

Research had led to greater understanding of how the organ of Corti works and to a Nobel prize for the investigator who contributed significantly to that understanding, Georg von Békésy. Put very simply, when the stapes kicks in the oval window, the fluid in the cochlea is stirred and sets the basilar membrane moving in a very special way: Sounds of high frequency cause the greatest movements of the membrane under hair cells at the base of the cochlea, agitating the tips of the cells' protruding hairs. Sounds of middle frequency cause maximum movements of the membrane further toward the center of the cochlea, while sounds of lowest frequencies cause peak membrane movements near the top of the cochlea.

The movements of the hairs cause changes inside the cells that lead to the production of electrical signals. These signals excite nearby nerve cells whose long fibers—some 30,000 in each ear—spiral out from the cochlea to form the eighth, or auditory nerve, which goes from the ear to the brain.

Soon after entering the brain, eighth nerve fibers contact nerve cells in the first of many nerve centers concerned with hearing. Ultimately the auditory signals reach the cortex, the outermost covering of the brain.

The cortex contains centers associated with interpreting speech and music, with thinking, memory, learning, and other higher mental faculties.

There are a great many details and subtleties about human hearing that scientists are continuing to work out. How do we manage to block out unwanted sounds, for example, enjoying an intimate conversation in the midst of a noisy party? How do we detect subtle differences in loudness as well as pitch? What enables some of us to hear a tune for the first time and repeat it perfectly? How do we locate the source of sound and judge how near or far it is?

Much of this research requires a detailed analysis of what happens at the cochlea and at auditory centers in the brain. In recent years NINCDS-supported scientists have been able to remove embryonic cochlear tissue and grow it in the laboratory. Small animals like guinea pigs or chinchillas are often used in these studies because their cochleas are relatively large and their brains not very complex.

Work with human volunteers is also essential. Persons with normal hearing are often studied in experiments in which different messages are piped into each ear or where computers are used to generate garbled or synthetic speech. These studies are aimed at analyzing how people recognize speech and how they detect a meaningful message amidst noise.

Also of great interest are studies of how the two ears and the two halves of the brain work individually and together in the perception of speech, music, and other complex sounds. Experiments with hearing-impaired individuals are equally important, leading to a better understanding of the cues hearing-impaired people use to recognize speech—and how noise affects them.

Hearing problems higher up

Hearing impairments that result from damage to the sensory apparatus (the hair cells and other parts of the inner ear) or to the eighth nerve and auditory centers higher up in the brain (the neural apparatus) are often

lumped together as "sensorineural" problems. These include:

- *Hearing loss at birth.* Some 4,000 infants are born deaf every year in the United States. Close to half those cases are due to hereditary disorders.

A larger group of babies is born deaf or with major hearing impairments as a result of congenital disorders or difficult labor and delivery. Mothers who contract certain infections during pregnancy or who take certain drugs may give birth to hearing-impaired infants. Sometimes the hearing loss accompanies other prenatal or birth-related problems that result in cerebral palsy, seizures (epilepsy), or mental retardation.

Before the advent of a vaccine for German measles (rubella), this viral disease in pregnant women was a notorious cause of hearing impairment at birth. Common measles (rubeola) in pregnant women also imposed a threat to the unborn child. Fortunately, there are now vaccines for both these viral diseases so that women can (and should) be immunized well before becoming pregnant.

There are other virus infections for which no effective treatments or vaccines exist as yet. Of these, cytomegalovirus and herpes simplex type 2 virus (which causes a genital infection) can seriously affect the nervous system of infected newborns. Cytomegalovirus infection is estimated to affect 1 out of every 100 children born in the U.S. Between 5 percent and 10 percent of those children develop hearing impairments, especially for high frequencies, and have IQ scores less than 80 later on in school.

- *Hereditary hearing loss.* It is important to realize that hereditary conditions not only can result in deafness at birth, but also account for a variety of hearing impairments occurring later. Hereditary disorders can affect the outer and middle ear, as in otosclerosis, but generally involve damage to the cochlea or higher nerve centers. Because there are so many kinds of hereditary disorders, with different risks of inheritance, couples with a history of deafness on either side of the family

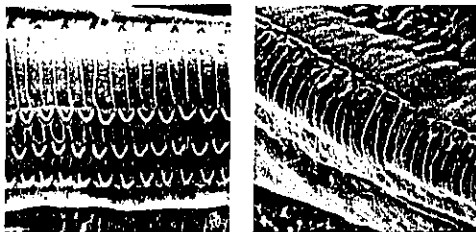
should consult genetic counselors for information.

- *Trauma-induced problems.* A severe blow to the head, an accident, stroke, brain hemorrhage or other trauma that affects the ear or any of the auditory pathways and brain centers will obviously take its toll on hearing ability.

- *Tumors.* Patients with eighth nerve tumors—called acoustic neuromas—may complain of hearing loss in one or both ears, headaches, dizziness, ringing in the ear (called tinnitus), or numbness over the face. Such symptoms deserve prompt attention. If an eighth nerve tumor is detected early, surgery to remove the tumor can be completely successful, leaving no hearing or other impairment. Tumors diagnosed at later stages may have grown large enough to be life-threatening, or their surgical removal may result in hearing loss, disturbances in the sense of balance (also located in the inner ear), loss of sensation in the face, or facial paralysis.

Acoustic neuromas can occur for no known reason, but can also arise as a result of a hereditary disease called neurofibromatosis.

- *Noise damage.* Brief exposures to high intensity sound can cause a temporary but reversible hearing loss. But continued exposure to loud noise means trouble: Eventually the hair cells sustain permanent damage, resulting in gradual hearing loss.



The hair cells in the left photo are normal; right photo shows that exposure to noise leads to total destruction of hair cells (lower layers) or damage (upper layer).

Concern about noise damage has inspired major research.

During the early days of industrialization nobody doubted that the din surrounding boilermakers, hydraulic press operators, or steel mill workers rendered their hearing less than perfect. Nowadays specialists are concerned that the everyday sounds of our highly technological society are also wilting our hair cells. Think of the power mowers and chain saws, the disposals, stereo sets, dishwashers, and food processors we live with... and the sounds of airplanes, motorcycles, city and highway traffic, fire and emergency trucks. Think, too, of joggers wearing earphones or young people at rock concerts or disco clubs, and you have the reason so many hearing specialists are worried.

Concern about noise has inspired research at several major hearing laboratories supported by NINCDS. Scientists at Washington University Medical School in St. Louis, Mo., for example, are systematically changing the frequency, intensity and duration of noise to see how each of these factors affects the structures of the inner ear. Investigators at the Central Institute for the Deaf, also in St. Louis, are analyzing the effects of noise on the inner ears of animals and also observing how such stress affects the animals' behavior. Other NINCDS-supported investigators are studying how noise damage may lead to degeneration of eighth nerve fibers, why some people are more affected by noise than others, and why the ear, once impaired by noise, often becomes hypersensitive—even more vulnerable to noise damage.

- *Drug-induced hearing loss.* Drugs as common as aspirin, the antibiotics streptomycin or neomycin, and certain of the water pills (diuretics) used to treat high blood pressure can damage the hair cells or other vital parts of the inner ear. Anyone who, while under medication, has a sudden change in hearing, or experiences dizziness or ringing in the ears (tinnitus), or has other problems with hearing or balance should report the symptoms to a physician at once. Often changes in the prescription can eliminate the symptoms and prevent permanent damage to the ear.

Certain powerful anticancer drugs may also damage hearing. NINCDS is currently conducting research studies of patients undergoing cancer chemotherapy at the Clinical Center, the research hospital of the National Institutes of Health. Audiologists measure the patient's hearing before treatment and then at periodic intervals thereafter to determine if the drugs have affected hearing.

- *Tinnitus.* Many people have experienced one or more occasions when they felt a ringing or buzzing in the ears or inside the head. But a surprising number of people, especially in middle age or later years, complain of a constant ringing in the head for no known reason. In some cases the symptoms may be unnoticed if a person is busy at work, talking, or otherwise distracted. For other people, however, the unpleasant sounds are present during every waking hour, interfering with all activities. In the most severe cases tinnitus even interrupts sleep. The hapless victim is tormented by an incessant internal siren sounding off. The psychological effects on a person can be devastating.

Tinnitus is considered a sensorineural disorder, but what causes it and where in the ear or brain the trouble lies are unknown. Because excessive amounts of aspirin and related drugs produce temporary tinnitus in human beings, NINCDS-supported investigators are studying the effects of these drugs in animals. The scientists can detect whether the animal is experiencing tinnitus by first training the animal to behave in specific ways in response to typical tinnitus sounds. They then observe whether the animal behaves the same way after drug treatment. Other NINCDS grantees are studying sounds that may be generated within the cochlea itself. Still a third NINCDS-supported group at the University of Oregon is developing an ear device designed to mask the tinnitus sounds, a method of treatment that appears to help some tinnitus sufferers. A variety of tinnitus maskers can now be obtained commercially.

- *Presbycusis.* Changes associated with aging are responsible for the majority of hearing impairments in

Six million Americans 65 or over have moderate to severe hearing losses.

adults. Many people in their forties and fifties experience a decline in sensitivity to high frequencies. The decline is gradual and progressive so that by their sixties and seventies as many as 25 percent of the elderly are noticeably impaired. However, investigators are beginning to question whether "aging factors" *per se* are at fault. There are cultures in the world—the Mabaan people of the African Sudan, for example—where presbycusis doesn't exist. Both men and women have excellent hearing in old age. The environment of the Mabaans is exceptionally quiet by Western standards. Further, the Mabaans do not suffer from heart disease, high blood pressure, ulcers or asthma. They lead relatively stress-free lives. No conclusions can yet be drawn, except that presbycusis is clearly not an inevitable result of aging.

Many experts now think that lifelong exposure to noise, as well as the high prevalence of heart disease, high blood pressure, and other blood vessel disorders increase the odds of hearing loss in later years.

In addition, some hereditary predisposition may be involved: As one investigator puts it, "Some of us may simply be programmed to suffer a decline and fall of our hair cells or our auditory neurons starting at a particular age—as young as the twenties and thirties in some people." Further, the decline may be selective: The hair cells and inner ear structures may be healthy in some older individuals so that they can pass a hearing test for pure tones with flying colors. Yet those same people may have trouble understanding speech, especially under trying conditions. Experts suspect that the listener's confusion is associated with tissue damage or loss of nerve cells in the brain where centers for speech perception and discrimination are located.

With the realization that people are living longer, the problem of presbycusis has become an area of growing concern to the National Institute on Aging (NIA) as well as NINCDS. At present 6 million Americans 65 or over have moderate to severe hearing losses. By the year 2000 an estimated 32 million Americans will be 65 or

older. If 25 percent continue to be affected by presbycusis that means that 8 million Americans may know only too well the limitations on activities, the lack of enjoyment, the isolation and boredom that severe hearing impairments can impose. To deal with this problem, cooperative research ventures are being initiated by the NINCDS with its sister organization, the NIA.

Currently NINCDS-supported research on presbycusis includes programs at the University of Michigan, focusing on the causes of presbycusis and microscopic changes in the inner ear, and a program at the University of California at Los Angeles concerned with how age affects the transmission of nerve signals along the auditory pathways in the brain.

Detecting hearing loss

In the case of a simple infection or impacted wax, the diagnosis and treatment of a hearing problem may begin and end in the family doctor's office. More complicated cases call for the expertise of the otologist. He or she will conduct a thorough ear examination, note the patient's medical history, and inquire about hearing problems affecting other members of the family. Certain blood tests or other laboratory analyses may be necessary, as well as standard hearing tests. The specialist may also want X-rays of the head or the computerized X-ray images of the brain called CT scans.

Hearing tests are usually conducted by audiologists, professionals educated in the science of hearing and in



An NINCDS-supported investigator at the University of California, Los Angeles, tests the hearing of a patient participating in research studies.

the battery of tests used to assess and analyze hearing impairment. Audiologists also provide counseling and nonmedical rehabilitation for the hearing impaired, such as lipreading and hearing aid evaluation.

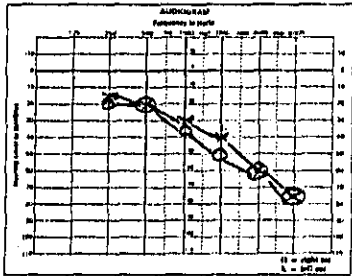
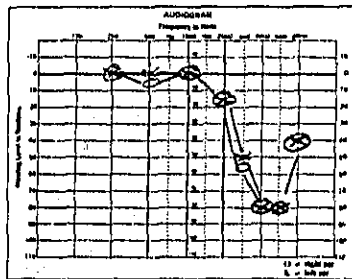
Persons undergoing audiological testing sit in a small soundproof room. The examination usually includes tests to determine how well the eardrum and middle ear bones conduct sound. These tests depend on inserting a snug-fitting probe with wires attached into the external canal of the ear. Then air pressure between the probe tip and the eardrum is varied at the same time that a tone is sounded through the probe tip. A machine analyzes the movements of the drum and middle ear bones, printing out the results on a graph—the "tympanogram."

The probe can also be used to check the acoustic reflex to loud noise. The tympanogram and acoustic reflex tests take only a few minutes. Since the tests depend on automatic responses of the auditory system, they can be used to test hearing in infants and others who cannot respond voluntarily.



The baby doesn't have to do a thing. The machine measures sound conduction in the middle ear and acoustic reflexes. The tests are simple and painless.

PHOTO COURTESY OF THE NATIONAL HEARING SOCIETY



Audiograms measure how well you hear pure tones with each ear. Upper audiogram shows a typical noise-induced hearing loss—worst at around 4000 Hz. Lower audiogram shows a typical presbycusis pattern: hearing in both ears gradually worsens as frequency increases.

The audiologist then measures the patient's thresholds for two-syllable words and for pure tones in a range from 250 Hz to 8000 Hz. The patient wears headphones and indicates when he or she can just barely detect sounds as the decibel level is varied. The graph that plots sound frequency against decibel level is the audiogram. The audiologist also measures the ability to discriminate speech by having the individual repeat one-syllable words. The audiologist may conduct further

tests to determine the nature of the hearing loss. These tests may involve manipulations of pure tones and noises or the use of tape recordings that introduce distortions into voice or sound signals.

An EEG for hearing

In the past decade investigators have developed ways of recording the electrical activity of brain centers associated with hearing. Electrodes are attached to the top of the head and at each side, near the ear. The individual wears earphones and sits quietly in a soundproof room listening to clicks at different intensities. A computer analyzes the nerve cell activity in response to the clicks and displays the brain wave pattern on a video screen. Audiologists know the normal shape of the waves and the time it takes for nerve signals to move from center to center along the auditory pathways. Delays in the appearance of certain waves or changes in their pattern help localize the problem. Because the brain cells recorded lie in the brainstem—a core of brain tissue located below the cortex—the brain wave recording is called the auditory brainstem response. Like tympanometry, the auditory brainstem response test is automatic and so can be used to study hearing in infants. However, the test takes up to an hour and the subject must remain stationary and quiet, so testing in young children usually requires mild sedation.

Many people think that you can't measure children's hearing in the first months or years of life. Even without the newer automatic tests, however, there have always been ways of measuring infants' hearing. Parents can approach a baby from behind and sound a bell or rattle. The child who hears will be startled, and, depending on the stage of brain development, may turn toward the direction of the sound. Audiologists can also test an infant's hearing with standard pure tone tests. The infant wears earphones and sits on its mother's lap in the test room. The audiologist trains the child to associate a certain sound with the appearance of a toy or other interesting object displayed on a



The auditory brainstem response test measures brain wave activity picked up by electrodes on the scalp in response to clicks transmitted to the earphones. The test is automatic and the boy can relax, doze, or sleep while the computer whirs.

screen. By observing the child's behavior in response to speech or noises at various intensities, the audiologist can measure how well the child can hear.

The results of medical and audiological examinations may indicate a problem that can be helped by surgery, medication, or a hearing aid. Sometimes preventive measures are urged: Adults with middle ear infections are cautioned to avoid flying; workers who are beginning to show noise-induced hearing losses are advised to transfer to less noisy departments or at least wear ear protection. Often, however, the hearing loss is longstanding and irreparable. In those instances there is no instant remedy or miraculous cure. But there *are* important things that can be done.

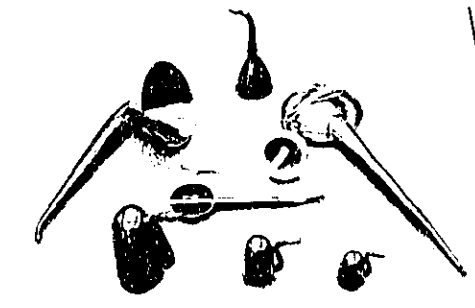
What hearing aids do

A hearing aid amplifies sound. The aid provides the extra power to boost sound so that it can stimulate the cochlear cells. Hearing aids can benefit anyone, as long as some hearing remains. How well hearing aids work is another matter. Their effectiveness depends not only on the design of the aid (is it a quiet, high-quality, easy-to-maintain instrument?), but also on how well the aid matches the individual's needs. Present day aids are a far cry from the ear trumpets used generations ago. A modern aid is lightweight, battery-operated, and miniaturized. It can be molded to fit inside the ear, worn behind the ear, or fitted into eyeglasses.

Many individuals with hearing impairments become sensitive to amplified sound. This does not mean they cannot wear a hearing aid; it does mean that the aid must accommodate their sensitivities. Some investigators now suspect that hard-of-hearing people occasionally retain "islands of hearing"—frequency ranges where sound is still perceived at near normal levels. Such individuals might find the amplification provided by hearing aids uncomfortable. Scientists at Louisiana State University in New Orleans are currently investigating islands of hearing in hearing-impaired people to see how common the phenomenon is, and whether special aids making use of these frequencies could be designed to enable individuals to understand speech.

But no matter how well designed and appropriate to the wearer's hearing impairment, the chances of the aid benefiting the user largely depend on attitude and motivation. It is important to realize that adjustments in the aid have to be made and all wearers go through a period of learning and adaptation. In short, the recommendation and fitting of a hearing aid is not the end of an audiological examination, but the beginning of a new way of life. Followup in the first few weeks, proper maintenance, and periodic checkups to see how the human ear and aid are both doing are a necessary part of the process.

At the same time, a person can learn simple skills to



Hearing aids have come a long way—from yesterday's trumpets to today's sensitive, powerful, and all but hidden instruments.

enhance the usefulness of an aid. Speechreading (lip-reading) is one. Most people already possess this skill to a remarkable degree. If you think you are not a speech reader, consider the times you have watched a TV movie where sound was not quite synchronized with lip movement.

Hearing handicaps before age 3

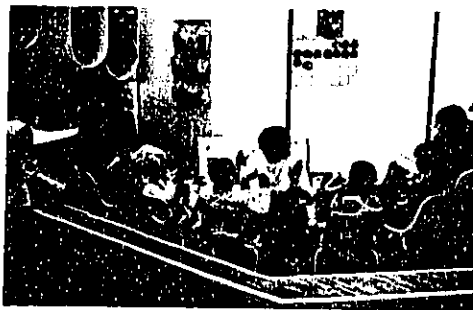
The problems of communication are vastly more complicated for the child who has never heard speech than for those whose hearing problems develop later.

Normally we learn to speak by imitating others and listening to the sounds we make. That instant replay—auditory feedback—allows us to correct our speech and





The dancers at this school for the deaf can barely hear—but they're rehearsing a number for a school musical. All wear hearing aids and listen to the beat amplified by loudspeakers on stage.



Hearing-impaired and learning-disabled children play rhythm games in a class taught by a speech therapist and two aides.

“Rehabilitation begins with parent education...”

continuously adjust the tone, rhythm, and loudness of our voices. Children who suffer a hearing loss in infancy must overcome a double hurdle: They are cut off from a major source of learning about the people, places, and things of this world. And they cannot benefit from the natural feedback system that makes speaking one's native tongue the inevitable event in development that it is for most of us.

Children who become moderately to severely impaired before age 3 (the prelingual years) can be helped by hearing aids, but as one specialist put it, “Rehabilitation begins with parent education...the audiologist and hearing therapist must use considerable skill in helping parents (and children) adjust to the benefits and the limitations of a hearing aid.” How much progress can be made also depends on what other handicaps a child may have. Visual problems, movement disorders or mental retardation compound the basic communication and learning problems.

It is testimony to human resourcefulness and to progress in hearing research that people no longer despair of overcoming the problems of hearing loss in infancy. In times past those afflicted often remained mute or chose not to make vocal sounds. They communicated with the speaking world through notes, gestures, and signs—not unlike the tourist struggling in a foreign land. Society tended to enforce the communications barrier by ignoring the problem or else relegating deaf people to special schools or institutions. One result has been that deaf people have tended to form a separate culture among themselves, employing their own means of communication and seeking each other's company. Even today 95 percent of deaf people marry other deaf people. Many have the native intelligence and creativity to succeed at professional careers, but communication and social barriers continue to work against their achieving their full potential.

At the end of the 19th century, authorities urged reforms in education aimed at solving the problem of the isolation of the deaf: Enforce a strictly oral mode of



Hearing-impaired adults learn American Sign Language (Ameslan) in government-sponsored classes.

speech training, they declared. Forbid children to use any form of sign language, finger spelling, or other manual communication. Instead, teach speechreading and vocal training exercises so that the deaf will have to master the sounds of speech and the principles of spoken language. This strictly oral approach has been effective in many cases and remains the guiding philosophy of many training centers and schools for the deaf throughout the world.

In the United States, however, opposition to the oral-only tradition remained active, in part due to the efforts of such teachers as Thomas Gallaudet, for whom Gallaudet College for the Deaf in Washington, D.C., is named. Gallaudet believed that the all-important goal for the deaf individual was to be able to communicate with *anyone*. He strongly advocated sign language. Others agreed, urging that whatever means aided communication—speechreading, vocal training, gestures, or sign language—should be encouraged. Today this "total communication" approach, as it has come to be

called, coexists with the oral-only school, each with strong adherents and notable successes.

Important in the development of the total communication approach has been the growing use of American Sign Language, *Ameslan*. Ameslan uses subtle combinations of hand, face, and body movements to comprise a vocabulary and grammar that are distinct from English. Ameslan "signers" now make it possible for profoundly hearing-impaired people who have learned the language to watch the television news or attend some public meetings. In the hands of the creative actors and writers of the National Theater of the Deaf, Ameslan has also developed as a vehicle for imaginative expression in poetry and drama.

Hearing horizons today...

The hearing-impaired individual has more learning opportunities and communications aids available now than at any other time in history. Legislation enacted in 1973 prohibits discrimination against the handicapped



At this federally sponsored job fair for the handicapped, interviewer and applicant exchange information.

in employment or education by any organization receiving any amount of Federal funds. This has meant new opportunities for the profoundly hard of hearing in the job market and at all levels of education.

In addition, variants on the training methods followed by either oralist or total communication practitioners have been developed. "Cued speech" is an outgrowth of the desire to make speechreading more effective. Speakers learn to augment their lip movements by finger signs to indicate particular consonantal or vowel sounds. The signs clarify whether the lips are saying "bad" or "pad," "road" or "load." (Only about a third of spoken English can be understood by interpreting lip movements alone.)

There has also been renewed interest in teaching the deaf to understand speech by having the deaf person feel the muscles of the throat and neck as the speaker pronounces words. This was the method Helen Keller's teacher used, and NINCDS-supported investigators at Massachusetts Institute of Technology (Cambridge, Mass.) are currently exploring its effectiveness.

An increasing variety of amplifying devices and signals are now available for the hearing impaired. Tele-



Feeling vibrations through the jaw during normal speech helps the deaf child learn how to talk.

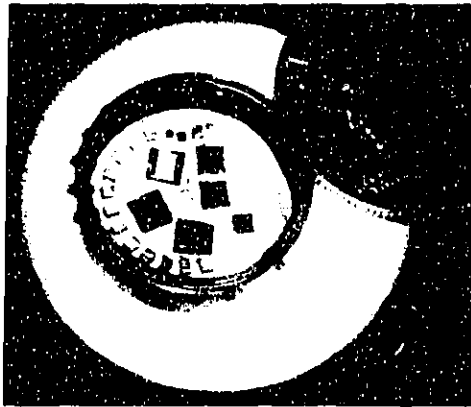
The most ambitious aid to hearing is the cochlear implant.

phones can be equipped with lights to signal when the phone is ringing and amplifiers can be built into the receiver. Other communication equipment includes teleprinters which enable a sender to type a message that is then coded electronically for transmission over the phone. A decoder at the other end reconverts the signals to a printed message.

...and tomorrow

Meanwhile scientists are exploring how to use the body's other sense systems as a means of communication when hearing fails. The skin is sensitive to vibrations over a range of frequencies. By attaching a set of vibrators around the waist or chest, the wearer can learn to interpret a sequence of vibrations as words and sentences. Such "vibrotactile aids" are currently being investigated by NINCDS-supported scientists at the University of Washington in Seattle and the Callier Center for Communication, Dallas, Texas.

Perhaps the most ambitious scientific program on the design boards today is the cochlear implant, a device to aid individuals with sensorineural deafness. The implant uses tiny electrodes to apply electrical stimulation directly to auditory nerve fibers. NINCDS currently supports implant research at Stanford University, Palo Alto, Calif., the University of California at San Francisco, and the University of Washington in Seattle. In some designs the electrodes are implanted in the cochlea itself, positioned to stimulate selected nerve fibers as they spiral around the shell. In other designs the electrodes are applied to nerve fibers after they have been bundled together in the eighth nerve. Either way, the trick is to stimulate fibers associated with a selection of different frequencies so that the brain can distinguish different tones. So far the implants use only a few electrodes, too crude to enable even the rudiments of speech to be encoded and deciphered. But in the few instances where profoundly deaf individuals have volunteered to have the devices surgically implanted, the hardware seems to be well tolerated and the wearers



NINCDS-sponsored scientists at Stanford have developed a 12-channel radio receiver-stimulator that is smaller in diameter than the U.S. quarter in the background. The device is attached to a cochlear electrode array to form a cochlear implant.

generally report pleasure in being able to hear any sound at all.

The state of the art of cochlear implants is in its infancy. A dozen or so microelectrodes cannot be expected to replace the 30,000 nerve fibers we are born with in each ear. But advances in electronics, in computers, in audio engineering, and in hearing science have been great and continue to develop at an impressive rate. Tomorrow's cochlear implants may be even more miniaturized, allowing greater numbers of fibers to be stimulated with less risk that the implant itself may damage tissue. And if, in the end, the cochlear implant is not the ideal device, the same kind of thinking that led imaginative men and women to try it in the first place will stimulate other research directions, ultimately more successful solutions for the problems of the hearing impaired. That is the hope through research.

Voluntary health organizations

There are many organizations concerned with the problems of the hearing impaired. One of the largest groups is the American Speech-Language-Hearing Association, which is both a professional and lay organization. Another important professional organization is the American Academy of Otolaryngology/Head and Neck Surgery, which provides information to the general public.

Some groups espouse particular philosophies in speech education and training. The Alexander Graham Bell Association for the Deaf, for example, adopts the oralist approach. All the groups listed provide literature and advice, and can generally indicate schools and other resources available in local communities. Some organizations, like the National Easter Seal Society, have local chapters providing information and rehabilitation for a variety of handicapping conditions. One organization, the Better Hearing Institute, provides information on a toll-free Hearing HelpLine: (800) 424-8576. In addition, some organizations also support research. Most can supply practical information for individuals interested in donating temporal bone tissue to Temporal Bone Banks for research study. The temporal bone of the skull, collected at autopsy, contains the organs of hearing and balance and is valuable source material for scientists studying the tissue changes that occur in communicative disorders.

Organizations:

Alexander Graham Bell Association for the Deaf, Inc.
3417 Volta Place N.W.
Washington, D.C. 20007
(202) 337-5220

American Academy of Otolaryngology/Head and Neck Surgery
1101 Vermont Avenue, N.W., Suite 302
Washington, D.C. 20005
(202) 289-4607

American Speech-Language-Hearing Association
10801 Rockville Pike
Rockville, Md. 20852
(301) 897-5700

American Tinnitus Association
P. O. Box #5
Portland, Oreg. 97207
(503) 248-9985

Better Hearing Institute
1430 K St., N.W.
Washington, D.C. 20005
Toll-free Hearing HelpLine: (800) 424-8576

The Deafness Research Foundation
55 East 34th Street
New York, N.Y. 10016
(212) 684-6556

National Association for Hearing and Speech Action
10801 Rockville Pike
Rockville, Md. 20852
(301) 897-8682 (Call Collect)

National Association of the Deaf
Suite 301
814 Thayer Avenue
Silver Spring, Md. 20910
(301) 587-1788

*National Black Association for Speech, Language,
and Hearing*
P. O. Box 50214
Washington, D.C. 20004

National Easter Seal Society, Inc.
2023 West Ogden Avenue
Chicago, Ill. 60612
(312) 243-8400

National Hearing Association

Suite 308
1010 Jorie Boulevard
Oak Brook, Ill. 60521
(312) 323-7200

Self-Help for Hard of Hearing People, Inc.

P. O. Box 34889
Bethesda, Md. 20034
(301) 365-3548

Publications

The following publications are published for people with hearing impairment and others concerned with the problem. (Subscription prices are not given since they are subject to change.)

Silent News
193 Main Street
Lincoln Park, N.J. 07035

Deaf American
(Published by the National Association of the Deaf,
listed above)

NINCDS Information

Additional information concerning hearing research supported by the Communicative Disorders Program of the National Institute of Neurological and Communicative Disorders and Stroke can be obtained from:

Office of Scientific and Health Reports
National Institute of Neurological and
Communicative Disorders and Stroke
Building 31, Room 8A-06
National Institutes of Health
Bethesda, Md. 20205
(301) 496-5751

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Bob Guthrie/The Boys Town Institute for Communication Disorders in Children—pages 9, 23, 26 (lower)

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President's Committee on Employment of the Handicapped—pages 28, 29

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