

Hearing Loss

Making a Difference Today

Of the five classical senses (sight, hearing, taste, smell, and touch), hearing is the one that people are most likely to lose. Approximately 28 million Americans have some form of hearing loss, ranging from mild to profound. Loss of hearing can occur at any age. Of every 1,000 infants in the United States, 1 to 3 are born deaf or hard of hearing. By age 18, approximately 17 in 1,000 children have a hearing problem. The incidence increases with age. More than 30 percent of people over the age of 65 and 40 percent to 50 percent of people over the age of 75 have difficulty hearing.

Although people with a hearing loss can lead full, productive lives, the impairment often can delay a child's ability to learn and can hinder his or her social and emotional development. Among older adults, loss of hearing can lead to social isolation and depression. Because people with hearing loss have a long-term need for supportive care and services, the economic costs of the impairment are also considerable. The Centers for Disease Control recently estimated the lifetime costs for all people with hearing loss who were born in 2000 at \$2.1 billion (in 2003 dollars). These costs include direct medical costs such as doctor visits, direct nonmedical expenses such as special education, and indirect costs such as lost wages when a person cannot work as a result of hearing loss.

Research Breakthroughs

Most hearing loss occurs when the 15,000 or so tiny sensory hair cells in the inner ear (cochlea) become damaged, usually as the result of overexposure to loud sounds, an infection, a genetic disorder, or aging. These hair cells are needed to convert sound waves into electrical impulses that are then sent via the auditory nerve to the brain. More than 30 years ago, the National Institutes of Health (NIH) began funding a concerted research effort that eventually led to the development of the cochlear implant—a remarkable device that bypasses damaged hair cells and directly stimulates the auditory nerve, thus enabling people with certain types of hearing loss to understand speech and other sounds again. During the past 20 years, more than 59,000 people around the world have experienced the “miracle” of these devices and, as a result, are now functioning much more fluently in the hearing world. Experts estimate that as many as 250,000 Americans who are currently hearing impaired would benefit from cochlear implants. Traditional hearing aids also benefit the hearing-impaired. Unlike cochlear implants, hearing aids amplify sounds and then send them through the ear canal up to the brain, resulting in hearing.

Until recently, scientists thought that damaged cochlear hair cells could not regenerate in people and other mammals. In 2003, however, NIH-funded researchers used gene transfer technology to grow new hair cells in the cochlea of laboratory animals—the first time that such cells have been regenerated in adult mammals. Such findings suggest exciting new possibilities for hearing loss treatments by retaining the very cells that transform and send sound waves as electrical signals to the brain, resulting in the sensation of hearing.

Gene Discoveries

Within the past decade, researchers also have made great strides in identifying genes with mutations that result in hearing loss. Many such gene mutations have been found, including one linked to the most common cause of hearing loss among Americans—a mutation in the Gap Junction Beta 2 (connexin 26) gene. Researchers also have recently identified a gene mutation responsible for Usher syndrome type 1, a devastating disorder that causes both deafness and loss of vision. This mutation is particularly prevalent among Jewish people of Eastern European descent. The discovery of these and other gene mutations will help scientists develop more effective diagnostic and intervention strategies for inherited hearing losses—strategies that can improve the lives of millions of Americans.

For more information please email brss@sfn.org.

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The snail-shaped cochlea is the hearing organ of the ear. It contains about 15,000 hair cells that convert sound waves into electrical impulses that travel to the brain via the auditory nerve. Most hearing loss occurs when these hair cells are destroyed, usually due to noise, infections, aging, or genetic disorders.

Continued funding for research could lead to:

- Smaller and more effective cochlear implants, digital hearing aids, and other technologies that can help remediate hearing loss.
- Methods for regenerating cochlear hair cells; the destruction of these cells is a major factor in hearing loss.
- More precise gene-based diagnosis to aid in the treatment of hereditary hearing impairment.
- New technologies for diagnosing hearing loss in infants, thus enabling hearing-impaired children to receive early interventions that can help them develop language skills comparable to their peers.



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Hearing Loss

Making a Difference Tomorrow

A combination of factors—noise exposure, infections, genetics, and aging—make hearing loss a common and significant problem in the United States. Did you know that:

- About 28 million Americans have a hearing loss, ranging from mild to profound.
- Noise is responsible for at least one-third of hearing loss cases in the United States.
- Hearing loss is more common in men than in women.
- Ninety percent of children who are born deaf in the United States are born to parents who can hear.

Research Brings Hope for the Future

With funding from the National Institutes of Health (NIH), scientists have made tremendous progress during the past decade in understanding the basic biology that underlies hearing loss. Researchers now have valuable new knowledge about how hearing “works”—how the auditory nerve encodes and transmits complex signals to the brain, for example, and how the brain creates maps of auditory space. They are also exploring the possibility of regenerating cochlear hair cells in humans; the destruction of these hair cells is the primary factor in most cases of hearing loss.

Other exciting developments have occurred in the field of genetics. Scientists have identified dozens of genes with mutations that can cause hearing loss. Such findings may lead to mutation-specific treatments that can delay or prevent certain forms of genetic hearing loss.

A New Generation of Hearing Devices

With NIH support, researchers are studying the precise mechanism by which electrical stimulation activates the auditory nerve—and thus how to build even more effective cochlear implants. Scientists are also trying to determine if implants actually help slow or halt progressive hearing loss in some people by protecting fibers in the auditory nerve from further degeneration. In addition, new types of implantable hearing devices are being explored. NIH-funded grants, for example, are enabling researchers to determine how nanotechnology—the science of building highly miniaturized devices that work on the molecular and atomic level—can be used to create a new generation of implants that would be even smaller and more powerful than those currently available.

Earlier Intervention

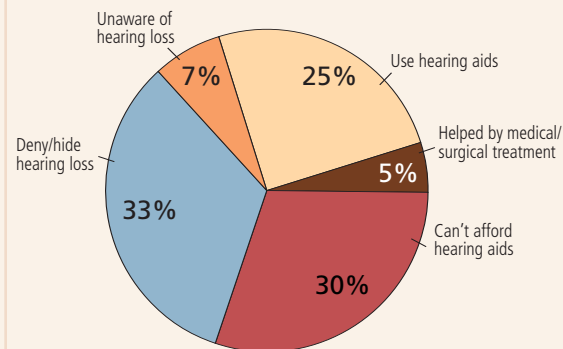
Research to identify the best ways to diagnose and treat very young infants with hearing loss is also underway. Researchers have found that early (within the first few months of life) detection of hearing loss in children and subsequent intervention strategies can make a huge difference in the development of a child’s language skills. New technological advances, along with genetic screening, are making it possible to more accurately evaluate and diagnose the hearing of newborn infants. The result: Children born with a hearing impairment will have the opportunity to develop language skills comparable to their hearing peers.

Hope for Other Diseases

Only with continued funding will researchers be able to bring about the scientific breakthroughs needed to help prevent and, in some cases, reverse hearing loss. Such research also promises to benefit people with other genetically complex communications disorders, ranging from stuttering to autism—disorders that affect millions of additional Americans.

For more information please email brss@sfn.org.

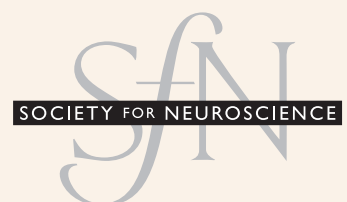
Hearing Loss in Americans



Only 5 percent of hearing loss in adults can be improved through medical or surgical treatment. Although almost 95 percent of Americans with hearing loss could have their hearing treated with hearing aids, many cannot or do not take advantage of these devices.

Already research has led to:

- The development of the cochlear implant, which can help people with certain types of hearing loss understand speech and other sounds.
- The discovery that cochlear hair cells, which play a crucial role in hearing, can be regenerated in adult mammals—a finding that suggests new and exciting treatment possibilities for hearing loss.
- The identification of dozens of gene mutations linked to hearing loss.



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