

**THE COCHLEAR IMPLANT HAS CONNECTED TENS OF THOUSANDS OF DEAF ADULTS AND CHILDREN TO THE WORLD OF SOUND. THANKS TO CONTINUING RESEARCH INTO THE NEUROSCIENCE OF HEARING, BIOMEDICAL ENGINEERS ARE CREATING NEW GENERATIONS OF HEARING DEVICES THAT WILL MORE CLOSELY MIMIC THE COMPLEX WORKINGS OF THE INNER EAR.**

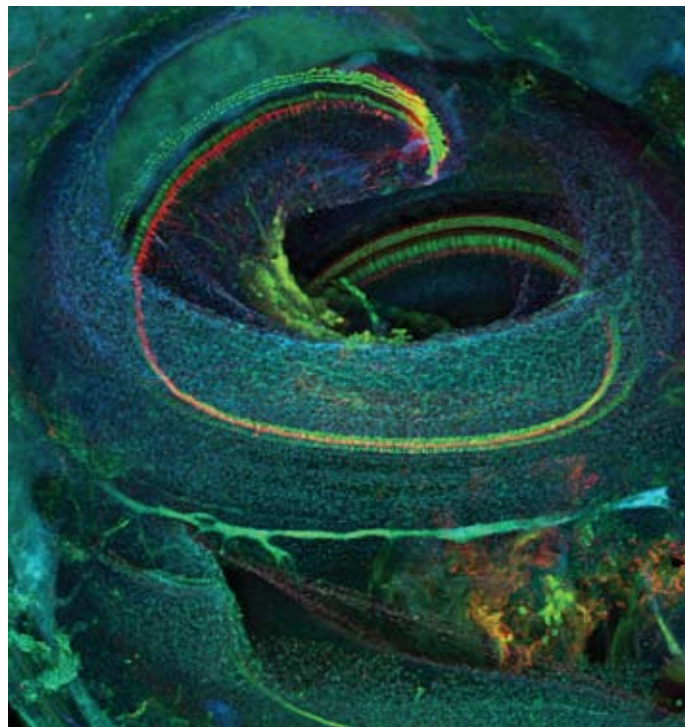
## COCHLEAR IMPLANTS

The small electronic device known as the cochlear implant has made it possible for thousands of individuals who are profoundly deaf or severely hard-of-hearing to understand the speech of others, even over the telephone.

Hearing loss is a major disability in the United States, affecting some 32 million people. Of those, about 650,000 are functionally deaf. Most are adults, although each year an estimated 3 in 1,000 U.S. children are born deaf or with a partial hearing loss.

Profound hearing loss usually occurs when hair cells in the snail-shaped cochlea, or inner ear, are missing or damaged due to a genetic factor, an infection, exposure to loud sound, or other cause. Without these hair cells, the ear is unable to convert sound vibrations into electrical impulses to send to the brain.

Hearing aids, which mainly amplify sound, can help hard-of-hearing individuals who have a significant number of still-healthy cochlear hair cells. Such aids don't work, however, for the profoundly deaf. Enter the cochlear implant. It converts the acoustic vibrations of sound into electrical impulses and shoots them directly to the brain via the auditory nerve, bypassing receptor cells in the cochlea altogether. The device enacts this neurological end run by stimulating



▲ INNER EAR ANATOMY IS SHAPED INTO A VERY COMPLEX SPIRAL STRUCTURE. THIS MOUSE INNER EAR WAS FLUORESCENTLY LABELED AND IMAGED BY CONFOCAL MICROSCOPY. THE SINGLE ROW OF INNER HAIR CELLS AND THREE ROWS OF OUTER HAIRS ARE LABELED GREEN. NERVE FIBERS CONDUCTING SIGNALS TO THE BRAIN ARE SHOWN AS RED BENEATH THE INNER HAIR CELLS. CELL NUCLEI ARE BLUE. THIS VIEW LOOKS FROM THE TOP TOWARD THE BASE TO SHOW THE HIGHEST TURN. THE LOWEST TURN IS A THIN GREEN STRIPE AT THE BOTTOM OF THE IMAGE.

COURTESY OF GEN MACDONALD AND EDWIN RUEBEL, VIRGINIA HERBELL BICEDEL HEARING RESEARCH CENTER, UNIVERSITY OF WASHINGTON.

nerve endings that would normally contact the hair cells.

The cochlear implant is not a miracle cure for deafness. Hearing with the device is not the same as normal hearing, and people must learn how to interpret the sounds it creates. Even with extensive training, music and nuances of speech, such as emotion and emphasis, elude most cochlear implant users.

Recent discoveries in the neuroscience of hearing coupled with more advanced technologies are helping

scientists overcome these limitations and design increasingly sophisticated cochlear implants as well as other types of devices that restore or enhance hearing. This exciting research is also leading to:

- Better methods of diagnosing and treating hearing loss, particularly in young children.
- More knowledge of how sounds are processed in the brain.
- Greater understanding of the molecular and genetic causes of hereditary deafness.

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■ The development of devices that can help people with other types of sensory problems, such as a severe loss of balance.

Many hard-of-hearing people can hear low-frequency sounds, including musical tones, but not the high-frequency sounds produced by human speech.

One of the newer developments in cochlear implant design is a device that stimulates only the part of the cochlea where high-frequency sounds are picked up, leaving low-frequency areas of the cochlea untouched—and undamaged by the insertion of the implant. Users are thus able to continue to hear low-pitched sounds, from Brahms' bass notes to a basset hound's bark. Sometimes a conventional hearing aid is also worn to amplify these lower sounds.

Cochlear implants don't work for people whose auditory nerves have been destroyed—by a tumor, for example. In 2000, the FDA approved another device, the auditory brainstem implant (ABI), which bypasses both the cochlea and the auditory nerve and connects directly to the brainstem. Most ABI users

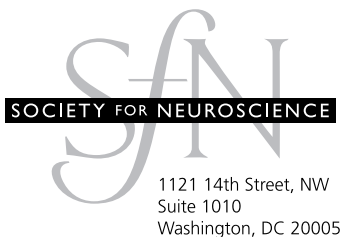
can hear car horns and ringing telephones, as well as more general sound cues, which, when combined with lip reading, can help them participate in conversations and connect again to the world of sound.

Recent research has uncovered the importance of two nerve-protecting proteins in the transmission of both low- and high-frequency sounds. Future cochlear implants may pump therapeutic doses of these proteins into specific locations within the inner ear to enhance hearing. Scientists are also exploring other drug-delivery possibilities for cochlear implants, such as the release of medications that prevent further hair cell loss.

If scientists can discover how to keep cochlear hair cells from dying or even how to grow new ones, cochlear implants won't be needed in the future. Since the early 1990s, scientists have identified dozens of genes related to hearing loss, including the GJB2 gene, which encodes a protein that plays an important role in the functioning of the cochlea. Mutations

in the GJB2 gene are one of the most common causes of inherited deafness. Scientists are now actively searching for gene therapies as well as other strategies that may one day protect cochlear hair cells from damage or degeneration or perhaps even encourage new cells to grow.

Finally, the cochlear implant offers a window on the power and potential of the human brain. Engineering advances now offer cochlear implants containing up to 22 channels that send electrical impulses to the auditory nerve—but this still pales in comparison to thousands of hair cells used to process signals in the normally functioning ear. Nonetheless, with training, the brains of implant users turn this limited auditory information into remarkably robust comprehension of sound and speech. This discovery provides a powerful example of what scientists are learning about the human brain's amazing adaptability and how this knowledge can ultimately be applied to improve quality of life.



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