Advances in Genetics and Devices Are Helping People with Communication Disorders

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During the past decade, scientists have made astonishing advances in the NIDCD's mission areas of hearing, balance, smell, taste, voice, speech and language. Numerous discoveries have expanded our knowledge base amid one of the most exciting periods in the history of communication research.

Genetics ranks high on the list of areas in which we've made significant progress. Before the Decade of the Brain, we knew that deafness could be inherited, but we knew little about the genes involved. Twenty years later, we've identified hundreds of genetic mutations linked to inherited hearing loss, with more than 80 genes mapped just in the past 10 years. Further study has shown us the functions of many of the proteins that these genes encode and has revealed molecular pathways essential for normal hearing.

Similar explorations in speech and language have turned up genetic mutations that are responsible for delayed language development in young children and that also play a supporting role in dyslexia and some cases of autism. This kind of discovery, which reveals common neural pathways in speech, reading and language development, could be the key to freeing thousands of children now locked inside their own worlds.

Another exciting gene discovery, the result of a collaboration across the National Institutes of Health and internationally, recently identified the first genetic mutations responsible for stuttering, which places this speech disorder squarely in the medical world. Researchers are currently working with animal models to understand how this gene influences the neural circuits that control expressive language.

Combating hearing loss by regenerating hair cells, small sensory cells in the inner ear, also is showing promise. Our ability to hear relies on these hair cells, and defects in them or damage to them cause hearing loss. Although fish, amphibians, and birds are able to grow new hair cells, humans and other mammals can't. Scientists are trying to understand the molecules and genes involved in hair cell regeneration in animals, with hopes of learning how to mimic the process in humans. Research in hair cell regeneration could one day offer a powerful treatment option, if not a cure, for hearing loss.

Beyond genetic discoveries, we continue to focus on the development of devices that bring sound into the worlds of people who are profoundly deaf or hard of hearing. The cochlear implant, one of the most groundbreaking biomedical achievements of the past 30 years, uses direct electrical stimulation of the auditory nerve via implanted electrodes to bypass inner ear damage and provide a sense of sound. Although cochlear implants have helped close to 200,000 people worldwide, most still have problems clearly hearing conversations in noisy environments. Scientists are currently looking at how to better localize sound by using advanced signal processing techniques and improved electrode design.

Hearing aid users have similar problems in noisy environments. An ingenious solution has emerged from the study of the ears of a parasitic fly, Ormia ochracea, which is extraordinarily successful at localizing sound. Using the lessons from this research, scientists are developing a miniature directional microphone that can zero in on a single voice and make communication in noisy places a more effective.

As this new decade begins, we're applying the technology of cochlear implants to the development of other potential neural prostheses for hearing,

balance and speech. These include auditory brainstem implants, which reconnect the ear to the brain in people whose auditory nerves have been surgically removed; vestibular implants to normalize balance by electrically stimulating the vestibular nerve; and brain-computer interfaces to help patients with locked-in syndrome translate thought into synthesized speech.

In smell and taste research, we'll focus less on the nose and the tongue and more on the brain, tackling questions about how the brain interprets sensory data and mapping the functional organization of the neural circuits that mediate these senses. We are just beginning to understand the complicated neural networks that turn objects and words into speech, but newer imaging techniques, such as voxel-based morphometry, will allow us to localize brain function at a much finer spatial resolution than fMRI and will become a powerful tool for researchers to see which areas of the brain are active during speech and word retrieval.

I am certain that we will end this new decade with a far better understanding of how language and speech are processed in the brain. We'll also have more sensitive, individually tailored and effective technologies for people with hearing loss. Finally, our continued studies in genetics, and the rapid accumulation of knowledge about genes and their functions, mean that the era of precise genotype-based diagnosis may be at hand for many of the communication disorders we study.

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