DEPARTMENTS OF LABOR, HEALTH AND HUMAN SERVICES, EDUCATION, AND RELATED AGENCIES APPROPRIATIONS FOR 2001

HEARINGS

BEFORE A

SUBCOMMITTEE OF THE

COMMITTEE ON APPROPRIATIONS

HOUSE OF REPRESENTATIVES

ONE HUNDRED SIXTH CONGRESS

SECOND SESSION

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VOLUME 4A

WEDNESDAY, FEBRUARY 16, 2000.

NATIONAL INSTITUTE ON DEAFNESS AND OTHER COMMUNICATION DISORDERS

WITNESSES

DR. JAMES F. BATTEY, JR., DIRECTOR, NATIONAL INSTITUTE ON DEAFNESS AND OTHER COMMUNICATION DISORDERS

DR. DONALD H. LUECKE, DEPUTY DIRECTOR, NATIONAL INSTITUTE ON DEAFNESS AND OTHER COMMUNICATION DISORDERS

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DR. RUTH L. KIRSCHSTEIN, ACTING DIRECTOR, NIH

DENNIS P. WILLIAMS, DEPUTY ASSISTANT SECRETARY, BUDGET, DHHS

Mr. PORTER. The subcommittee will come to order and continue our hearing with the National Institute on Deafness and Other Communication Disorders. We are pleased to welcome Dr. Battey. Please proceed.

INTRODUCTION OF WITNESSES

Dr. BATTEY. Thank you, Mr. Chairman. Joining me at the table are Ms. Sparks, Budget Officer; Mr. Kerr, Executive Officer; Dr. Luecke, Deputy Director for the Institute; and you know Dr. Kirschstein and Mr. Williams.

Before I begin my formal remarks, I would like to add my thanks to that of Dr. Alexander for your remarkable leadership on this subcommittee and for the subcommittee's remarkable support of the National Institutes of Health and the National Institute on Deafness and Other Communication Disorders. I think after you heard what Dr. Alexander told you and what I will tell you and what all of the other directors will tell you, you will realize that you made a great difference for the Nation's health and that difference will be felt for many, many years; and I give you my sincere thanks for that support.

OPENING STATEMENT

I am honored to appear before you as the Director of the National Institute on Deafness and Other Communication Disorders or NIDCD. Within the last year, we have witnessed outstanding research progress in hearing, balance, smell, taste, voice, speech, and language by NIDCD-supported scientists and clinicians, progress further accelerated by our collaborations with other NIH institutes. Today, I would like to highlight a few of NIDCD's many science ad-

vances that are relevant to hearing and hearing impairment throughout the human life span.

In humans, auditory sensory cells, hair cells, and other internal parts of the ear form as early as the third month of development. These hair cells establish connections with the central nervous system and are essential for the hearing process. Loss of, or damage to, these hair cells following injury is permanent and results in deafness or hearing impairment.

If I can direct your attention to my poster, what you see is three parallel rows, which are the tops of cylindrical hair cells that you see down below. You see the beautiful symmetry and order, those cells are responsible for the fine tuning of the auditory response. On the right side they are wiped out or grossly distorted; that ear on the right has been exposed to excessive noise; and you can see that the hair cells have been completely damaged.

Mr. HOYER. Is that from a rock concert?

Dr. BATTEY. A teenager going once to one rock concert will not damage their hearing. What is required to get noise-induced hearing loss would be to be the guitar player sitting next to the speaker every night.

NIDCD investigators have recently provided direct evidence of the mechanism that regulates the number of progenitor cells that develop as hair cells, providing important clues about the molecular processes critical for hair cell development and the potential for hair cell regeneration. NIDCD-supported investigators have also shown that in the mouse, the *Math1* gene is essential for regulating the development of hair cells and progenitor cells. Mice deficient in this *Math1* gene develop no hair cells in their inner ear.

These findings provide insight into the molecular mechanisms regulating hair cell differentiation and specification and provide the initial clues as to how we might ultimately regenerate these hair cells which are so often the cause of hearing loss. Immediately after birth, the ears of a newborn are filled with the new sounds of his or her surrounding environment. Unfortunately, not every newborn will be able to hear these sounds. It is estimated that as many as 12,000 babies each year are born in the U.S. with significant hearing loss, making it a far more common congenital disorder. NIDCD-supported research has shown that the detection of hearing impairment and intervention within the first 6 months after birth are very important for optimizing language development in young children.

In a 5-year multi-center study, NIDCD-supported scientists determined the optimal test procedures for neonatal hearing screening. This study was the first controlled comparison of normal hearing and hearing-impaired infants assessing multiple physiological responses to sound. The development of precise and timely diagnostic screening procedures is the first important step in providing early intervention strategies that will optimize the development of either spoken or signed language skills. Not only is hearing screening becoming available for all newborns, but breakthroughs in medical genetics will enable clinicians and scientists to determine precisely the genetic changes which cause hearing impairment.

About one child in 2,000 is born with hereditary hearing impairment which compromises the development of normal spoken lan-

guage skills. The first gene where mutations result in a regrettably common form of hereditary hearing impairment was mapped to a region of chromosome 13 in 1994. Three years later, scientists discovered that mutations in a gene called GJB2 were the cause of deafness in families with DFNB1.

Since this discovery, scientists studying the nature of mutations in this deafness gene have learned that about one-third of all recessive hereditary hearing impairment in the United States is caused by mutations in GJB2. This is a major player among the tens to hundreds of genes where mutations can cause hearing impairment. With the identification of mutant genes, genetic testing becomes possible, but the technical capability does not necessarily mean the genetic testing is appropriate for widespread clinical application.

The NIDCD is particularly interested in research to develop and validate diagnostic genetic tests to assess the impact of genetic testing on various cultural groups and individuals and to provide education and counseling to facilitate informed decisionmaking about reproductive issues and treatment strategies. Last year, I had the opportunity to describe NIDCD's research on the cochlear implant. In fact, I brought a cochlear implant for you to see.

This is a bioengineering advance that restores the perception of sound to both children and adults with profound hearing impairment. The cochlear implant is an array of roughly 22 electrodes inserted into the inner ear that converts sound into electrical impulses that directly stimulate the acoustic nerve thereby bypassing the damaged hair cells that I told you about a few minutes ago. If I can direct your attention to my second poster you will see a 4-year-old girl. She is wearing a cochlear implant, although you can barely tell. You have to look closely to see the wire. She received her implant at 2, she is now 4 and her caregiver reports to me that she is on age level for language development and, in fact, describes her as chatty which is remarkable given the fact that she has never heard a sound in her life other than the sounds through the cochlear implant that she received at 2 years of age.

Scientists supported by the NIDCD conducted a study to measure language achievement in children with cochlear implants. The study compared a group of children who received cochlear implants and a second group who were using hearing aids and saw significant differences in language achievement favoring the children with cochlear implants.

As research moves forward to reduce the burden of disease in America, the NIDCD is committed to the idea that all segments of American people should benefit from this progress. In comparison to the general U.S. population, Native American children have one of the highest rates of otitis media. NIDCD is continuing its support of a study on the epidemiology of this disorder and hearing loss among Native American infants from birth to age 2 at the White Earth Reservation in Minnesota.

Recent assessment shows that intervention programs should focus on parental smoking as a significant risk factor for otitis media in Native American infants. The study also includes prevention strategies to reduce the burden of otitis media such as promoting breast feeding. Understanding the natural history and risk

factors is vital to gaining insight into the epidemiology of otitis media in both majority and minority populations.

Loss of hearing can occur in childhood, but also later in life. The NIDCD is conducting research on neurofibromatosis type 2, a genetic disorder that often leads to tumors on both acoustic nerves causing deafness in children and adults. Scientists supported by the NIDCD have determined that specific mutations in the NF2 gene result in different levels of severity of the disease. There is a correlation between the severity of the mutation in the gene and the progression and severity of the disease. This finding will facilitate earlier DNA-based diagnoses and improve disease management and increase the preservation of hearing in NF2 patients.

Regrettably, the surgery required to manage this disease often results in resection of both acoustic nerves so that sound perception cannot be restored with cochlear implants. NIDCD-supported scientists have developed a specialized auditory prosthesis that is implanted directly into the ventral cochlear nucleus of the animals, that is the portion of the central auditory system where the acoustic nerve fibers make their initial connections. These animal studies have demonstrated the safety of this technique and deaf NF2 patients are scheduled to be fitted with these devices within the next few years with the hope of restoring auditory perception.

A recent NIDCD-supported study has demonstrated a clear genetic component for age-related hearing loss or presbycusis. It is likely that different mutations in the same genes that cause profound hereditary hearing impairment in children also cause age-related hearing loss later on. With the ability to predict who is at risk for age-related hearing loss, better strategies to minimize or delay hearing loss within the aging population can be developed.

The older population of the U.S. is large and growing. As they live longer, they have a greater chance of developing hearing impairment. Hearing aids are the most common means for assistance for persons with hearing loss. Roughly 20 million Americans have hearing aids right now. The Department of Veterans Affairs and the NIDCD conducted a multi-center trial which included elderly volunteers to compare the efficacy of three commonly used hearing aid circuits. Data from the trial showed that performance differences among the three hearing aid circuits were minimal. Of greater importance, the trial demonstrated that each circuit improved speech recognition with improvements observed under both quiet and noisy listening conditions. We remain committed to supporting research leading to smaller and better hearing aids, capitalizing on bioengineering advances in micro electronics.

My colleagues and I will be happy to respond as best we can to your questions.

Mr. PORTER. Thank you, Dr. Battey.

[The written statement of Dr. Battey follows:]