

**DEPARTMENTS OF LABOR, HEALTH AND HUMAN SERVICES, EDUCATION, AND RELATED AGENCIES
APPROPRIATIONS FOR 1994**

**HEARINGS
BEFORE A
SUBCOMMITTEE OF THE
COMMITTEE ON APPROPRIATIONS
HOUSE OF REPRESENTATIVES**

ONE HUNDRED THIRD CONGRESS

FIRST SESSION

SUBCOMMITTEE ON THE DEPARTMENTS OF LABOR, HEALTH AND HUMAN SERVICES, EDUCATION, AND RELATED AGENCIES

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NATIONAL INSTITUTES OF HEALTH

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**DEPARTMENTS OF LABOR, HEALTH AND
HUMAN SERVICES, EDUCATION, AND RELAT-
ED AGENCIES APPROPRIATIONS FOR 1994**

TUESDAY, MARCH 23, 1993.

**NATIONAL INSTITUTE OF NEUROLOGICAL DISORDERS
AND STROKE, NIH**

WITNESSES

**MURRAY GOLDSTEIN, D.O., DIRECTOR, NATIONAL INSTITUTE OF NEURO-
LOGICAL DISORDERS AND STROKE**

**JAY MOSKOWITZ, PH.D., ASSOCIATE DIRECTOR FOR SCIENCE POLICY
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**ANDREW C. BALDUS, BUDGET OFFICER, NATIONAL INSTITUTE OF NEU-
ROLOGICAL DISORDERS AND STROKE, NIH**

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

Mr. NATCHER. The Committee will come to order.

At this time, we start with the hearings of the National Insti-
tutes of Health.

A great many of you ladies and gentlemen who are in this Com-
mittee room have been with the National Institutes of Health for
many years.

You have heard me say on more than one occasion that one of
the great achievements we claim on our subcommittee and the full
Committee, is the fact that when I was elected a Member of this
Committee, we had \$73 million for the National Institutes of
Health. That is what the budget was, \$73 million. We are up to
\$10.327 billion.

Dr. Goldstein, if I had my way, it would be \$15 billion, I don't
mind telling you.

Dr. GOLDSTEIN. I would agree with you.

Mr. NATCHER. We maintain on this Committee that as long as
you take care of the health of your people and continue educating
your children, you will continue to live in the strongest country in
the world.

This morning we begin two weeks of hearings on the National
Institutes of Health. We would normally wait until the fiscal year
1994 budget request is submitted to the Committee before begin-
ning our hearings, but we will not have enough time once the

TUESDAY, MARCH 30, 1993.

**NATIONAL INSTITUTE ON DEAFNESS AND OTHER
COMMUNICATION DISORDERS**

WITNESSES

**DR. JAMES B. SNOW, JR., DIRECTOR, NATIONAL INSTITUTE ON DEAFNESS
AND OTHER COMMUNICATION DISORDERS, NIH**
**DR. JAY MOSKOWITZ, DEPUTY DIRECTOR FOR SCIENCE POLICY AND
LEGISLATION, NIH**
**DR. LEAMON LEE, DIRECTOR OF DIVISION OF FINANCIAL MANAGEMENT,
NIH**
**W. DAVID KERR, EXECUTIVE OFFICER, NATIONAL INSTITUTE ON DEAF-
NESS AND OTHER COMMUNICATION DISORDERS, NIH**
**PATIENCE T. SPARKS, BUDGET OFFICER, NATIONAL INSTITUTE ON DEAF-
NESS AND OTHER COMMUNICATION DISORDERS, NIH**
DENNIS P. WILLIAMS, DEPUTY ASSISTANT SECRETARY, BUDGET, DHHS

Mr. NATCHER. At this time we have before the subcommittee, Dr. James Snow, the Director of the National Institute on Deafness and Other Communication Disorders.

Doctor, before you give us your statement tell us who you have with you there at the table.

Dr. SNOW. Thank you, Mr. Natcher.

INTRODUCTION OF WITNESSES

To my right is Dr. Jay Moskowitz, the Deputy Director for Science Policy and Legislation of NIH; Dr. Leamon Lee, the Director of the Division of Financial Management of NIH; and Mr. Dennis P. Williams, Deputy Assistant Secretary, Budget, Department of Health and Human Services.

On my immediate right, Ms. Patience T. Sparks, Budget Officer, NIDCD; and on my left is Mr. W. David Kerr, the Executive Officer of NIDCD; and Ms. Sandra Mitchell, Sign Language Interpreter of NIDCD.

Mr. NATCHER. We are delighted to have you all.

OPENING STATEMENT

Dr. SNOW. Mr. Chairman, it is a privilege to appear before this subcommittee to discuss the programs of the National Institute on Deafness and Other Communication Disorders.

Forty-six million Americans have disorders of hearing, balance, smell, taste, voice, speech or language. I am pleased to bring to you good news about progress that will benefit many of these individuals.

The first three years of life are crucial in the development of speech and language. Impairment of hearing during this period se-

verely limits speech and language development. When hearing impairment is identified early, rehabilitation can start, as early as four to six months of age, with hearing aids and special language training. Unfortunately, the average age for identification of hearing impairment in the United States is close to three years.

I am pleased to report that at the end of the NIDCD/NIH-supported consensus development conference held this month an historic event occurred. For the first time, a scientific advisory panel has recommended universal screening for hearing impairment at birth. Dissemination of this information and development of the best possible screening methods will allow for much better outcomes for children with hearing loss. This will allow them to acquire language, whether spoken or signed, at the earliest possible opportunity. This will also assure them greater success in life and save billions of dollars in too-little, too-late rehabilitation.

In current hearing research, NIDCD scientists are at work on a wide variety of elusive and confounding diseases. The NIDCD continues the search for the causes of hereditary hearing impairment. One in 1,000 infants is born deaf, and the hearing impairment is genetic in 50 percent.

When the Institute was formed in 1988, no genes for hereditary forms of deafness had been found. Now we have located 22, but untangling the scheme of mutations is a complex task. The state of the art in molecular genetics has given us an opportunity to make substitution therapy and gene-transfer therapy a possibility by the end of the century.

We are also at work on prevention of noise-induced hearing loss. It affects 10 million Americans, and NIDCD scientists are determining why and how some people are predisposed to greater hearing loss from noise exposure.

The development of a vaccine against otitis media or middle ear infections has been selected for critically important intramural and extramural research. Otitis media is the most common infection in children for which antibiotic therapy is required. This disease damages the emerging communication skills for many of the nation's children.

For years, older Americans have complained about hearing aids because the aids failed to help them understand speech in background noise. The NIDCD, with the cooperation of the Department of Veterans Affairs, has taken an aggressive approach to solving this long-standing problem.

As part of a new initiative that uses the triple approach of grants mechanisms, contracts and clinical trials, the Institute is supporting the development and evaluation of hearing aid technology designed to meet the needs of hearing-impaired people. This research will benefit the five million people who are currently hearing-aid users and the 15 million people in the United States who could benefit from these devices but have not been able to do so.

Under energetic investigation by our Institute are ototoxicity, the adverse effect of drugs on hearing; tinnitus, a ringing or roaring in the ear; otosclerosis, a condition where there is an overgrowth of bone in the ear; and AIDS-related hearing impairment, caused by the most common secondary infection among AIDS patients—striking when communication is crucial.

Loss or reduction of the sense of smell can create life-threatening situations, especially for the elderly. This past year, there was a breakthrough in understanding how the olfactory system is able to discriminate such a vast array of odors. A large family of genes unique to the receptor neurons in the olfactory neuroepithelium was found, opening many new research approaches.

Several distinct proteins have recently been found in taste receptor cells known to mediate the conversion of chemical signals to nerve impulses.

In studies of early diet on later food preferences, findings indicate that components of breast milk, such as sodium, may have lasting effects on later nutritional choices.

Recent voice research includes progress in understanding the causes of nodules, polyps and contact ulcers and the treatment of spasmodic dysphonia, vocal tremor and vocal cord paralysis.

Stuttering, developmental speech disorders and the speech of persons with hearing impairment are being investigated, and practical approaches to these problems are developing.

Recent research on aphasia, the loss of language function that is the result of stroke, revealed that improvements can be made throughout the first full year following a stroke, not just for the first three months as was previously believed. Interestingly, the greatest improvements were made by an older patient group, 65 to 80 years old.

In research on language of deaf children and adults, results indicate that the timing of acquisition of language greatly influences comprehension skill later in life. The effects are more pronounced for first than for second language learning—whether spoken or signed.

In order to track disease and disorder incidence and prevalence, we have created the first epidemiology branch to deal exclusively with communication disorders. It maintains close liaison with the National Center for Health Statistics in the collection and analysis of data about disorders of human communication across the life span.

The NIDCD has held four working group meetings of important constituencies historically underrepresented in scientific planning. Groups advised the Institute about the needs of the deaf community, the needs of oral, auditory hearing-impaired persons, the unique needs of women and women's health issues as well as the needs of minority persons and minority health issues. Because of the usefulness of the recommendations we received, we will be holding three additional working groups.

The NIDCD is now four years old. We are proud to be building a long-needed infrastructure for research in human communication.

Mr. Chairman, I look forward to another year of progress, and I would be happy to answer any questions.

Mr. NATCHER. Dr. Snow, thank you very much.

[The statement of Dr. James Snow follows:]

DEPARTMENT OF HEALTH AND HUMAN SERVICES

National Institutes of Health

Statement of the Director

National Institute on Deafness and Other Communication Disorders

It is a privilege to appear before this committee to discuss the programs of the National Institute on Deafness and Other Communication Disorders (NIDCD).

Forty-six million Americans are directly affected by a communication disorder. Twenty-eight million have hearing impairments; 14 million have disorders of voice, speech, or language; 2 million have problems with balance; and 2 million have disorders of smell and taste. Family, friends, colleagues and, finally, all citizens are affected by the costs of these conditions. There is much to be hopeful about from what is being discovered today and what is planned for tomorrow in the basic and applied science underway extramurally and intramurally at the NIDCD.

NIDCD was created in October 1988 by The National Deafness and Other Communication Disorders Act as one of the institutes of the NIH to fund and conduct research in the normal and disordered processes of human communication.

In hearing research, NIDCD scientists are battling a wide variety of elusive and confounding diseases. The NIDCD is searching for the causes of hereditary hearing impairment because 1 in 1000 infants is born deaf and 50 percent of those cases are genetic. The state of the art of molecular genetics has given us the ideal opportunity to make gene therapy a real possibility by the end of the century.

When the institute was formed there were no known genes for hereditary forms of deafness, but untangling the scheme of mutations and alterations has yielded discovery of 22 of these genes. We are also at work on the preventable and costly cause of hearing loss that affects ten million Americans, noise-induced hearing loss. We are determining why and how some people, perhaps genetically, are predisposed to higher sensitivity to sound-- and greater hearing loss. Selected for intense investigation through both intramural and extramural resources is the development of a vaccine for otitis media, a disease familiar to almost every parent of an infant or toddler. Otitis media is the cause of most visits of young children to physicians. This disease has the potential for damaging the emerging communication skills of the nation's children and affecting their language and literacy for a lifetime. Presbycusis, the hearing loss we have long assumed to be part of the process of growing older, in fact may be preventable or delayable. We have found that there appears to be a genetic predisposition to this kind of hearing loss.

For years Americans have complained about the inability of currently available hearing aids to deal with background noise. The NIDCD has taken an aggressive approach to solving this long-term problem, with the cooperation of the Department of Veterans Affairs.

As part of a new initiative using the triple approach of grant mechanisms, contracts, and clinical trials, the institute is supporting the evaluation and development of hearing aid technology designed to meet the individual needs of subgroups of consumers. This research will not only benefit the 5 million people who are currently hearing-aid users, but also the 15 million people in the U.S. who could benefit from these devices but have not been able to adjust to them.

Under energetic investigation by our institute are ototoxicity, the adverse effect of drugs on hearing, tinnitus, and otosclerosis, a condition in which there is an overgrowth of bone in the middle ear, and AIDS-related hearing impairment, caused by the most common secondary infection among AIDS patients--striking when communication is crucial.

I am eager to report to the committee a recent historic event: For the first time in the world a group of scientific advisors called for the identification of infants with hearing loss--at birth! The announcement was made at the end of an NIDCD/NIH-supported consensus development conference. The average age for identification of this loss in children in the U. S. had been close to 3 years. Dissemination of this information and development of the best possible screening methods will allow for much better outcomes for children with hearing loss, allowing them to acquire language, whether spoken or signed, at the earliest possible opportunity, assuring them greater success in life and saving billions of dollars in too-little too-late habilitation.

In research on balance, scientists are at work on Ménière's disease and acoustic neurinoma. We have all experienced what is called "dizziness" associated with flu or loss of orientation in space. Imagine trying to function at home or in the workplace in a perpetual or intermittent state of dizziness--the results are devastating. Eighty percent of the astronauts working in space suffer from motion-sickness that limits their ability to function fully, making missions less cost-effective. With a new NIDCD-NASA collaboration we have created an opportunity for NIDCD scientists to study causes and develop management strategies for motion-sickness on earth as well as in space.

Loss or reduction of the sense of smell can create life-threatening situations, especially for the elderly. This past year there was a breakthrough in understanding how the olfactory system is able to discriminate such a vast array of odors. A large family of genes that is unique to the receptor neurons in the olfactory epithelium were found. This advance has had a tremendous impact on research efforts, sparking a wave of studies long needed in this field.

In taste, a distinct protein, gustducin, has recently been found in taste receptor cells and joins the growing family of G proteins known to mediate signal transduction. In studies of early diets on