

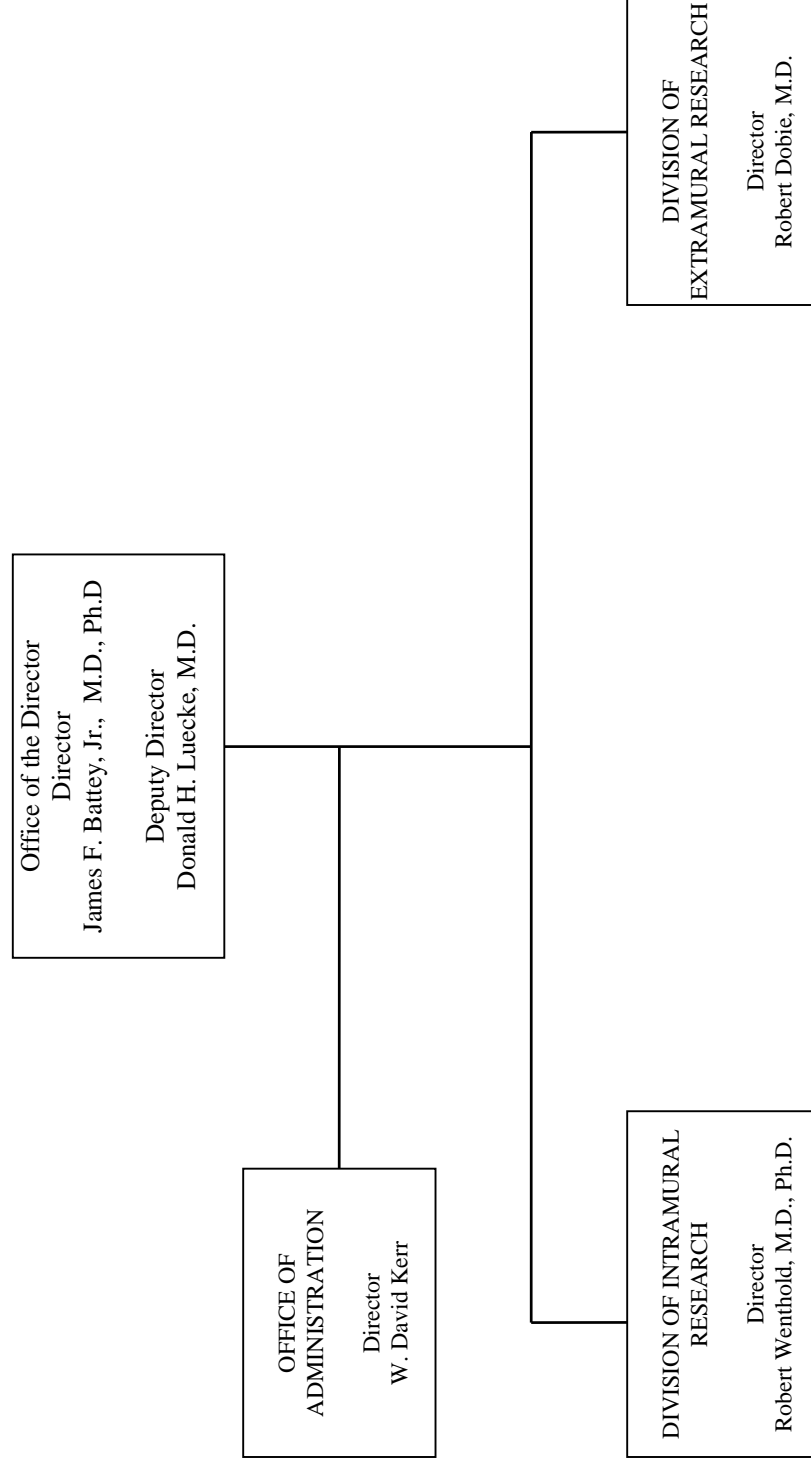
DEPARTMENT OF HEALTH AND HUMAN SERVICES

NATIONAL INSTITUTES OF HEALTH

National Institute on Deafness and Other Communication Disorders

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NATIONAL INSTITUTES OF HEALTH
National Institute on Deafness and Other Communication Disorders



NATIONAL INSTITUTES OF HEALTH

National Institute on Deafness and Other Communication Disorders

For carrying out Section 301 and Title IV of the Public Health Service Act with respect to deafness and other communication disorders, [\$342,072,000] *\$364,186,000*.

[Departments of Labor, Health and Human Services, Education, and Related Agencies
Appropriation Act for FY 2002, (P.L. 107-116)]

National Institutes of Health

National Institute on Deafness and Other Communication Disorders
Amounts Available for Obligation 1/

Source of Funding	FY 2001 Actual	FY 2002 Estimate	FY 2003 Estimate
Appropriation	\$300,581,000	\$342,072,000	\$363,040,000
Enacted Rescission	(100,000)	(107,000)	---
Subtotal, Adjusted Appropriation	300,481,000	341,965,000	363,040,000
Comparable adjustment for legislative proposal for accrued retirement costs	1,024,000	1,106,000	1,146,000
Real transfer to:			
Other HHS Agencies through Secretary's one-percent transfer authority	(57,000)	---	---
Real transfer to HHS for the Office of Human Research Protection	(63,000)	---	---
Comparative transfer from:			
Office of the Director for the Academic Research Enhancement Award program	708,000	0	
National Cancer Institute for research activities	---	---	7,765,000
Subtotal	302,093,000	343,071,000	371,951,000
Unobligated balance, lapsing	(79,000)	---	---
Subtotal, adjusted budget authority	302,014,000	343,071,000	371,951,000
Total obligations	302,014,000	343,071,000	371,951,000

1/ Excludes the following amounts for reimbursable activities carried out by this account:

FY 2001 - \$1,844,000; FY 2002 - \$1,844,000; FY 2003 - \$1,844,000

Excludes \$42,767 in FY 2001 and \$50,800 in FY 2002 for royalties

Justification

National Institute on Deafness and Other Communication Disorders

Authorizing Legislation: Section 301 of the Public Health Service Act, as amended.
Reauthorizing legislation will be submitted.

Budget Authority:

	2001 <u>Actual</u>	2002 <u>Appropriation</u>	2002 Current <u>Estimate</u>	2003 <u>Estimate</u>	Increase or <u>Decrease</u>
Current Law BA	\$301,069,000	\$342,072,000	\$341,965,000	\$370,805,000	\$28,840,000
Accrued Costs	\$1,024,000	\$1,106,000	\$1,106,000	\$1,146,000	\$40,000
Proposed Law BA	\$302,093,000	\$343,178,000	\$343,071,000	\$371,951,000	\$28,880,000
FTE	150	155	155	155	---

This document provides justification for the Fiscal Year 2003 activities of the National Institute on Deafness and Other Communication Disorders (NIDCD), including HIV/AIDS activities. A more detailed description of NIH-wide Fiscal Year 2003 HIV/AIDS activities can be found in the NIH section entitled "Office of AIDS Research (OAR)."

The President's appropriations request of \$371,951,000 for this account includes current law adjusted by assuming Congressional action on the proposed Managerial Flexibility Act of 2001.

INTRODUCTION

Excessive noise has long been recognized as an occupational hazard among adults, and hearing conservation programs have been implemented in the workplace. However, the resiliency of a child's auditory system following noise exposure needs further research. Chronic exposure to loud music, fireworks, lawn mowers or toys can accumulate over a lifetime to gradually produce irreversible damage to the sensory cells of the inner ear. The results of a recent survey conducted by the Centers for Disease Control and Prevention revealed that approximately 5.2 million American youths have some degree of hearing loss due to exposure to noise at hazardous levels.

Disorders of hearing, balance, smell, taste, voice, speech, and language exact a significant economic, social, and personal cost for many individuals. The National Institute on Deafness and Other Communication Disorders (NIDCD) supports and conducts research and research training in the normal processes and the disorders of human communication that affect many

millions of Americans. Human communication research now has more potential for productive exploration than at any time in history. With substantive investigations conducted over the past decades and the advent of exciting new research tools, the NIDCD is pursuing a more complete understanding of the scientific mechanisms underlying normal communication and the etiology of human communication disorders. Results of this research investment will foster the development of more precise diagnostic techniques, novel intervention and prevention strategies, and more effective treatment methods.

Science Advances: The Genetics of Human Communication

Identification of Genes Causing Deafness in Humans

Hearing loss occurs with a frequency of about 1 in 1,000 newborns and is also a prevalent, but not necessarily inevitable, feature of the aging process.¹ Causes of hearing loss in children and the elderly include viral and bacterial infections, loud noise, head trauma, drugs or other chemicals that are toxic to the sensory cells of the inner ear or mutations in genes critical for normal auditory function and development. At present, the underlying biological processes responsible for hearing loss are not well understood for any of these various etiologies. NIDCD scientists are identifying the genes whose mutations result in hearing loss.

Recently, NIDCD Intramural scientists identified a gene located on chromosome 10 that is involved in Usher syndrome type 1D (USH1D). Individuals that inherit two copies of this mutated gene are born profoundly deaf, have severe balance problems and gradually lose their sight beginning in adolescence. Less profound mutations of USH1D gene cause severe deafness but do not cause the loss of sight. The scientists discovered that USH1D gene encodes a protein called cadherin-23. Studies are underway to determine the function of cadherin-23 in the ear and eye. Knowledge of the function of cadherin-23 will provide new insight into cellular processes essential for normal auditory function, which may ultimately guide the development of improved diagnosis and treatment methods.

NIDCD scientists also identified a gene located on chromosome 21 whose mutation caused recessively inherited hearing loss (DFNB29). This gene encodes a protein called claudin-14, which is believed to help seal cells together in the inner ear thus preventing the leakage of a fluid (endolymph) between cells. The endolymph bathes the sound transduction cells and is essential for conversion of the mechanical energy of sound into an electrical signal that is sent to the brain. Studies are underway in a mouse model to advance the understanding of the function of claudin-14.

¹NIH Consensus Development Conference: Early Identification of Hearing Impairment in Infants and Young Children, 1993.

Exploiting Mouse Models of Disease: Discovery of Novel Deafness Genes and Genetic Characterization of Hearing Impairment

NIDCD has developed a substantial research portfolio to study existing mouse mutants as well as creating new mouse models to facilitate the discovery and analysis of genes whose mutation causes hereditary hearing impairment in humans. In a recent study utilizing the mouse mutant Waltzer, NIDCD Intramural scientists showed that mutations in members of the human cadherin gene family cause Usher Syndrome type 1D. The function of cadherins include cell and tissue polarization, cell sorting, cell migration and cell rearrangements. This mouse model is a critical tool for research to determine the identification of the mechanisms by which cadherin mutations cause this devastating deafness and blindness syndrome.

In other studies, a mouse nuclear gene previously identified by NIDCD-supported researchers has now been shown to interact with mutated genes in the mitochondria to significantly alter the severity of age-related hearing loss. This model system should provide important information regarding age-related hearing loss in humans, a relatively common and debilitating health problem within the aging U.S. population. These findings underscore the power of mouse genetics and the value of mouse models of deafness for the identification and detailed molecular characterization of human hearing impairment.

Hearing Parents of Deaf Children Favor Genetic Testing for Deafness

Genetic testing has become an option for deaf individuals and their families. However, little attention has been given to the public's perception on the value and impact of such testing. To investigate this issue, parents with normal hearing, who have one or more deaf children, were surveyed about their attitudes toward diagnostic and prenatal testing for deafness. This population was chosen because over 90% of deaf children are born to two parents with normal hearing.

The study showed that 96% of the respondents had a positive attitude toward genetic testing for deafness, including prenatal testing. None of these parents stated that they would use this information to terminate a pregnancy. The most common reason given for wanting testing (93%) was to identify the cause of deafness. Other reasons included determining the recurrence risk for future children and refining the affected child's future medical management and/or treatment. The results of this study contrast sharply with previous surveys of deaf adults (who may or may not have deaf and/or hearing children) who had a predominantly negative attitude toward genetic testing for deafness, with the majority stating that they believed such tests would do more harm than good.

Importantly, questioning of the respondents in the current study revealed a lack of understanding of the implications of the information provided by genetic tests. The study concludes that genetic testing should be combined with appropriate genetic counseling to assist parents of deaf children in making informed decisions concerning medical management and appropriate intervention strategies for their children.

Gene Therapy and Aminoglycoside Protection

Aminoglycosides are a class of antibiotics sometimes prescribed for a variety of severe bacterial infections. Unfortunately, aminoglycoside treatment can cause degeneration of inner ear hair cells (ototoxicity) resulting in hearing loss. At present there is no clinical treatment for the irreversible ototoxic effects of aminoglycosides. One potentially promising treatment to prevent or reverse ototoxicity involves gene therapy. However, efficient delivery and expression of a gene is critical to the success of gene therapy, and delivery to the closed environment of the inner ear poses unique challenges. NIDCD-supported scientists are currently working to elucidate many of the cellular and molecular mechanisms important for inner ear gene delivery. One such study in a mouse model has demonstrated significant protective levels against ototoxic damage following the delivery of the gene encoding glial cell line-derived neurotrophic factor (GDNF), a protein that affects the development and survival of neurons in the central and peripheral nervous system. Using viruses to carry the gene encoding GDNF, the scientists achieved successful delivery and expression of GDNF in the inner ear and subsequent preservation of hearing upon exposure to aminoglycosides in a mouse model system. While using viruses to deliver genes shows promise, NIDCD-supported scientists are exploring other avenues of gene delivery systems to the inner ear, including the use of a gelatin sponge. In this case, the desired gene is soaked into the sponge and directly placed on the round window membrane opening of the inner ear. Subsequently, the researchers detected the expression and presence of the targeted gene throughout the inner ear. These studies of gene delivery are the first steps towards development of gene-based treatment of sensory hearing loss.

An Essential Gene in Development of Hearing and Balance

A team of NIDCD-supported scientists have used a mouse model to discover a gene that regulates sensory and nerve cell development within both the auditory and vestibular systems. This gene, *BETA2/NeuroD1*, is critical for the formation of nerves in the cochlear-vestibular ganglion (CVG), the nerve bundle carrying signals from the inner ear to the brainstem auditory and vestibular centers. At a very early stage of development, the absence of the *BETA2/NeuroD1* protein results in slow development and differentiation of the CVG neuronal precursors (neuroblasts). Lack of *BETA2/NeuroD1* also causes alterations in the sensory cells and supporting structures of the inner ear. In addition, mutation in the *BETA2/NeuroD1* gene prevents development of the dorsal cochlear nucleus neurons and the granule cells, brain structures critical to hearing and balance. Mice lacking a functional *BETA2/NeuroD1* gene are completely deaf and suffer from severe imbalance and lack of coordination, head tilting, inability to right themselves when laid on their sides or backs, circling behavior and loss of muscle control.

How Sweet It Is! Scientists Identify Sweet Taste Receptor Gene

The sense of taste plays a critical role in the ingestion of nutrients and as a warning system to avoid spoiled food and poisonous substances. Understanding the molecular and cellular events that occur at the early stages of taste perception at the level of the taste receptor cell will provide important insight into how we taste different sweet, bitter, salty and sour substances.

A variety of distinct signaling pathways are activated by the basic taste qualities of salty, sour (acid taste), sweet, and bitter. Salty- and sour-tasting compounds activate ion channels that are located at taste receptor cells clustered within taste buds of the tongue and palate. Bitter and sweet compounds bind to G protein-coupled receptors that are similarly located. Considerable progress has been made in identification of receptors and other intracellular pathways that mediate bitter taste. In contrast, the genes and G protein-coupled receptors involved in sweet taste have been more elusive. Recently, four NIDCD-supported laboratories independently identified a sweet taste receptor gene, *T1R3*, at the mouse *Sac* locus. Differences in sweetener intake among inbred strains of mice are partially determined by variation in genes at the saccharin preference (*Sac*) locus. As predicted, the encoded receptor, T1R3, differs in amino acid sequence in "sweet preferring" versus "sweet indifferent" mouse strains and appears to represent one subunit of sweet receptor. Both human and mouse T1R3 are G protein-coupled receptors, and are selectively expressed in subsets of taste receptor cells. The mechanisms underlying bitter- and sweet-tasting quality coding are being studied in genetically altered mice where key components of the signal transduction cascade (such as candidate receptors and G-protein subunits) are modified or deleted in an attempt to determine the molecular basis for normal taste function.

Abilities in Auditory Pitch Recognition are Largely Inherited

Auditory pitch recognition is a complex process that allows us to determine the pitch or tone of a sound. This process relies on the ears, which receive the sound signal traveling through the air, and the brain, which interprets this signal to produce the pitch we perceive. Individuals with problems in pitch recognition are sometimes referred to as "tone deaf." Severe deficits in pitch recognition may be associated with speech and language disorders. It was long known that tone deafness can run in families. However, it was not known whether this disorder was due to genes inherited in these families, or to a common environment shared by family members. To answer this question, NIDCD Intramural scientists performed a large study on twins. They administered the Distorted Tunes Test, which measures people's ability to recognize incorrect notes in popular melodies, together with the American Academy of Otolaryngology's Five Minute Hearing Test questionnaire. Over 600 twins were tested, and the results show that identical twins scored much more alike than fraternal twins on the Distorted Tunes Test. The data revealed that approximately 70-80% of an individual's score is due to their genes and 20-30% due to other factors. The discovery that individual differences in pitch recognition are mostly genetic opens up the possibility of using genetic methods and information from the Human Genome Project to find the genes essential for pitch recognition. Identifying such genes and how they function will provide new insight into how the brain processes sound.

The Genetic and Environmental Etiology of Stuttering

Stuttering is a disorder in the production of fluent speech. Within the last two decades, NIDCD-supported scientists have shown that genetic factors may predispose individuals to stutter. A current twin study screened a large population-based twin sample from the Australian Twin Registry. Telephone interview-based diagnoses were obtained for 457 of these individuals. The data revealed that approximately 70% of the variance in risk for stuttering was found to be attributable to genetic effects, with the remainder due to unshared environmental effects (e.g.,

birth events, traumas or illnesses, peer influences, etc). There is preliminary evidence to suggest that two subgroups of affected cases may exist: stutterers whose etiology is primarily “genetic” in origin (those with a positive family history) and non-familial (sporadic) cases whose stuttering may have been the result of early brain damage. This study provides additional support for research to identify the genes whose mutation predisposes individuals to stutter in families with a history of stuttering.

Story of Discovery: Hearing Aids - How Basic Biology Translates into Technology to Help the Hearing Impaired

Over the past decade, Cornell University neuroscientists knew they had one amazing fly on their hands when they tested *Ormia ochracea*, a tiny insect parasite with such acute directional hearing that it has inspired a new generation of hearing aids and nanoscale listening devices. But it wasn't until the scientists ran an experiment on a fly-sized treadmill that they fully appreciated *Ormia's* talent for sound localization. Not only can the fly match the species thought to have the best directional hearing--*Homo sapiens*--it does so with a fraction of the distance between the two ears, suggesting new strategies for miniaturization of man-made devices. *Ormia* can detect changes in sound-source position as small as two degrees: humans trying to detect who is speaking in a crowded room cannot do better than that. These latest findings have led to collaborations between neurobiologists and engineers to make a directional hearing aid that would be smaller, simpler and cost less than currently available devices. Nanoscale listening devices based on the *Ormia* ear are under development at several industrial and university laboratories.

Directional hearing is critical for reproductive behavior of *Ormia ochracea*. In order to reproduce, female flies must find a host, such as chirping crickets, to climb aboard and deposit tiny larvae. First the flies must locate and home in on the chirping cricket by landing close by, then tiptoe the last few steps to their unsuspecting host. This stealth maneuver gave the neuroscientists an idea for an experiment. They placed a speaker on a movable arm to play recorded cricket sounds. On a ping-pong ball, they painted hundred of dots so that the ball's position could be tracked by a computer. The ball was floated on a jet of air so that it can be easily turned by a walking fly on a tether. The scientists discovered that no matter where the cricket sound was detected, to the right or left of center, the fly altered its path to walk directly toward the cricket chirps.

Since human ears are about 6 inches apart, it takes about 10 microseconds to make the same calculation that the *Ormia* fly, with its half-millimeter head, makes in about 50 nanoseconds – two hundred times faster. This accomplishment is due to the unique anatomy of the eardrums of *Ormia*. Located behind the fly's head, the eardrums are connected internally by a cuticle-based bridge that functions as a flexible lever. This unusual structure allows the membranes of the eardrum to vibrate in response to sound in two distinct ways, with different resonant frequencies. Trying to mimic the *Ormia* ear in silicon, engineering groups so far have developed prototype "microphone eardrums" that function "*Ormia* -like" as predicted but at ultrasonic frequencies. Additional research will be needed to generate prototypes that detect sound in the range of normal human hearing, that will be highly directional, fit inside the ear canal, and be affordable. Other applications of the *Ormia*-inspired silicon ear might include robotic listening devices.

Individuals who use hearing aids often struggle to understand conversation because of competing sounds and noises that come from other directions. The biological lessons provided by a parasitic fly's abilities in hyper acute time-coding and localization of sound provide strategies for improved nano/micro-scale directional microphones in hearing aids that would focus sound amplification on speech. Applications of these new principles may improve the quality of life for individuals with hearing loss who depend upon hearing aids to understand spoken language.

Science Advances: Increasing Knowledge on Disorders of Human Communication

Functional Brain Imaging as a Tool to Understand Cochlear Implant Performance

The cochlear implant is the first clinically useful neural sensory prosthesis to replace a human sense. It converts sound into electrical impulses on an array of electrodes that is surgically inserted into the inner ear, bypassing the inner ear hair cells and stimulating the auditory nerve directly, restoring the perception of sound to persons who are totally, or almost totally, deaf. This device has allowed adults who lost their hearing to recover an ability to understand speech. Although speech perception performance of adults has steadily increased with new advances in cochlear implantation, wide performance variations exist among cochlear implant recipients. Differences in structural and functional abnormalities of the auditory system may play a role in this variability. However, little is known about the reorganization of the auditory system following deafness, or on the preservation or recovery of auditory function following cochlear implantation.

Recently, functional neuroimaging techniques have been used to assess the brain activity associated with auditory performance. These studies have now been extended to individuals with cochlear implants. To examine the possibility that variation in auditory cortex activity might contribute to the wide range in perceptual performance found in cochlear implant users, NIDCD- supported scientists completed preliminary studies examining functional brain imaging using single photon emission computed tomography (SPECT) in individuals before and after cochlear implantation. The data suggest that preoperative to postoperative changes in auditory cortex responsiveness as measured by SPECT imaging are related to improvements in speech perception scores. Also, despite relatively similar hearing losses in each ear, significant differences in preoperative auditory cortex activation were observed between ears, which may help guide selection of the more appropriate ear for implantation. These data suggest that functional brain imaging may be a useful tool for exploring the responsiveness of the auditory cortex in cochlear implant candidates and for understanding individual performance variability.

Language Development in Profoundly Deaf Children with Cochlear Implants

Cochlear implants have been shown to be a useful aid to communication in deaf adults. Many can have fluent conversations with some lipreading and some can communicate fluently over the telephone, a difficult task in the absence of visual cues. However, approximately one-half of current cochlear implant recipients are children. Parental choice of a cochlear implant implies a desire on behalf of the parents to have their child fully participate in the hearing world, with spoken (oral) language skills. One of the expected benefits of cochlear implantation in children is the acquisition of spoken language. Recent data have shown that cochlear implants have a significant beneficial effect on the development of English language in profoundly deaf children. After receiving the implants, deaf children start developing their English language skills (on average) at a rate similar to that of normal hearing children, and it exceeds the rate expected from deaf children without implants. These findings suggest that earlier implantation in deaf children would result in shorter delays in language development. Further comparisons were made between cochlear implant children in oral communication programs (which excludes the use of manual signs) and total communication programs (which simultaneously uses oral and

manual sign language). Even when language skills and knowledge of the English language rule systems are similar in oral and total communication users, children who use oral communication typically have more intelligible speech and higher levels of speech perception than those children using total communication.

Phase I Clinical Trial of an Otitis Media Vaccine Candidate

Otitis media is the most common reason for a sick child to be evaluated by a physician, a public health burden estimated to cost approximately five billion dollars a year in the United States.² In addition to the cost savings, prevention of otitis media is particularly important because repeated antibiotic treatment of otitis media often results in the appearance of drug-resistant strains of bacteria which can no longer be eradicated with first-line antibiotics.

NIDCD Intramural scientists have developed candidate vaccines that would protect infants from otitis media caused by two major bacterial pathogens: nontypeable *Haemophilus influenzae* and *Moraxella catarrhalis*. These two pathogens account for two-thirds of otitis media cases in children, and there is no vaccine available for prevention of the disease. The scientists used a bacterial surface antigen, lipooligosaccharide, as a component to make conjugate vaccines. Pre-clinical testing with such vaccines from nontypeable *H. influenzae* demonstrated that the vaccines could generate specific immunity against the bacteria and reduce bacterial colonization in nose and throat, and reduce the incidence of otitis media in animal models. In a Phase I clinical trial involving forty normal human adult volunteers, one such vaccine directed against *H. influenzae* proved to be both safe and effective, eliciting a significant immune response against the bacteria. This candidate vaccine will soon be tested in a second trial for safety and effectiveness in children. For *Moraxella catarrhalis*, similar approaches were taken, resulting in several candidate vaccines. Pre-clinical testing in animal models with vaccines for *Moraxella catarrhalis* demonstrated that the vaccines were safe and effective, eliciting a significant immune response that inhibited bacterial growth. Phase I clinical trials are planned to test these candidate vaccines for safety and efficacy in humans. These studies are significant advances towards the long-term goal of developing a multivalent vaccine that reduces the incidence of otitis media caused by all major bacterial pathogens in children.

Symptomatic Congenital CMV Infection in Infants of Women with Preconceptional Immunity

Congenital cytomegalovirus (CMV) is one of the most common opportunistic pathogens in individuals with AIDS. It is also among the leading causes of sensorineural hearing loss and brain damage in children in the United States. NIDCD-supported investigators continue to study the significant contribution of CMV infection to infant morbidity, which includes progressive onset of hearing impairment during early childhood. A recent study shows that women with

²National Institutes of Health: Table - Cost of illness and NIH support for selected diseases and conditions. Disease-Specific Estimates of Direct and Indirect Costs of Illness and NIH Support, 1997.

preconceptional immunity to one strain of CMV can transmit a totally different CMV strain to their developing babies, causing symptomatic congenital infection. This result is crucial for understanding the components of protective immunity and for developing effective vaccines that prevent congenital CMV infections. These findings suggest that a vaccine against only one strain of CMV is unlikely to provide adequate protection against CMV reinfection in pregnant women. This research will guide the development of public health approaches (e.g., neonatal CMV screening, vaccine development, drug development, etc.) that can be effective in reducing and eventually eliminating this health problem.

Development of Stereocilia Orientation in Hair Cells

In humans, sounds are detected in the cochlea, the snail-shaped organ within the inner ear. The cochlea contains thousands of cells called sensory hair cells. These hair cells are arranged in four rows, which travel along the cochlear spiral. Each of these cells has numerous specialized, finger-like stereocilia bundles that project into a fluid-filled space within the cochlea. As sound waves pass into the ear, they are converted into pressure waves in the fluid-filled cochlea that induce vibrations of the hair cell stereocilia bundles. Vibrations of the stereocilia bundles lead to the release of chemical neurotransmitter signals that stimulate the auditory nerve carrying information into the brain. Thus, the vibration of the stereocilia bundle is essential for the perception of sound. Stereocilia bundles are only sensitive to vibrations in a single direction; therefore, the orientation of the bundle is crucial for normal function. In a normal cochlea, stereocilia bundles of all hair cells point in the same direction. However, recent studies suggest that developmental defects can lead to abnormal stereocilia bundle orientation and to hearing loss. However, the cellular and genetic factors that play a role in the specification of bundle orientation are unknown.

Recent experiments conducted by NIDCD-supported scientists indicate that a secreted signaling protein, Wnt-7a, is critical for orienting stereocilia bundles of cochlear hair cells. Specifically, the *Wnt-7a* gene is expressed in cells located adjacent to the inner row of the hair cells. Secreted Wnt-7a protein establishes a concentration gradient across the hair cell region. Disruption of this gradient, either through the addition of excess Wnt-7a protein or by blocking the signaling pathway, leads to the development of disoriented bundles in the embryonic ear. Developing hair cells detect the gradient of Wnt-7a protein and use this gradient as a mechanism to orient all stereocilia bundles in the same direction. This research is providing new information on the physiology of the developing inner ear and causes of hearing impairment.

A Prosthesis Providing Motion Cues to the Nervous System in the Absence of Balance

The vestibular system provides the nervous system with information on the motion and orientation of the head. This information is used by the nervous system to maintain stable images seen through the eyes even when the head is in motion, to stabilize the head and body during motion, and to provide an accurate percept of three-dimensional space during daily living. Loss of vestibular function can cause unpleasant or debilitating sensations of dizziness, vertigo (the illusion of motion) and spatial disorientation, as well as blurred vision and imbalance. Neural prostheses have been developed to restore function to the auditory system, which restores

the perception of sound to profoundly hearing impaired individuals who have lost the function of their inner ear hair cells. However, no such device has been developed for the vestibular system.

Recently, a prototype vestibular neural prosthesis was developed by a team of NIDCD-supported scientists. This device provides electrical stimulation directly to the vestibular nerve in response to head motion. Preliminary investigations on animals demonstrate that the nervous system adapts to electrical stimulation and that such stimulation can provide rotational cues to the nervous system in the absence of vestibular sensation. Over a twenty-four hour period, the central nervous system readily adapted to constant-rate electrical stimulation and even retained a “memory” of the adapted state when electrical stimulation was removed. The animals responded to head rotations in a horizontal plane with electrically-evoked, compensatory eye movements, although of lower magnitude than normal. This demonstrates that function of the vestibular organs was partially restored through electrical stimulation of the vestibular nerve. Research is progressing to refine the vestibular prosthesis and to determine its viability for application in humans with vestibular disorders.

Effects of Socioeconomic Status on Aphasia Severity and Recovery

Low levels of educational attainment and low occupational status have been found to be associated with poor health, increased incidence of disease and shorter life span. However, the specific correlation of these two socioeconomic factors with both initial severity of aphasia (the language problems associated with stroke or other types of brain damage) and subsequent recovery from aphasia has not been examined. Attempts have been made to correlate aphasia severity with educational level and literacy, yielding varied and often conflicting conclusions.

NIDCD-supported scientists examined the histories of a group of persons with aphasia due to a single, unilateral left-hemisphere lesion, at two points in time: approximately four months post-onset, and at eight to nine years post-onset. Severity of aphasia at these two points in time was determined using the Boston Diagnostic Aphasia Examination. Education and occupation correlated significantly with aphasia severity. The lower the level of educational attainment and occupational status, the more severe was the aphasia, both at four months as well as at eight to nine years post-onset. However, the rate of recovery was the same, regardless of level of education or occupation. Lesion size did not explain the differences in the severity ratings for the high and low education and occupation groups. The influence of education and occupation on aphasia in the early stages is consistent with reports from other studies demonstrating links between socioeconomic status (SES) and severity of illness in many organ systems. These results address the need for ensuring access to care and rehabilitation, since the potential for recovery exists for individuals regardless of education and occupation.

Spoken and Written Language Disabilities in School-Aged Children

Many previous studies of young children with language impairments have focused on errors of grammar in spoken language. However, much less is known about the persistence of these grammatical errors or about the occurrence of errors in written language. NIDCD-supported scientists are examining the speaking and writing skills within a group of sixty school-aged

children. Areas examined included use of the smallest grammatical elements of words that mark verb finiteness (regular past tense, third person singular present tense, and forms of the verb “to be”), as well as noun morphology (such as regular plural, possessive, and articles). School-aged children with normal language skills mastered the verb and noun morphology in spoken and written language. In contrast, children with language learning disabilities showed substantial difficulty in the written samples. These results indicate that particular aspects of written grammar remain an area of relative difficulty for children with language learning difficulties. Due to the visibility and importance of written language, the consequences of grammatical errors that persist in writing in the upper elementary school years and beyond should not be underestimated. Researchers found that these errors cluster in a limited number of types, increasing the chance that intervention can be targeted and effective. The results of this study emphasize the need to evaluate children’s written language skills, and provide intervention when needed.

New Therapies for Individuals with Head and Neck Cancer

Over 280,000 Americans suffer partial or complete loss of voice and speech as a result of cancer of the head and neck, and 12,000 of these individuals die each year.³ Intramural scientists from NIDCD and the National Cancer Institute (NCI) have collaborated to develop new therapy alternatives to surgery for patients with head and neck cancer which result in remission and preservation of the organs involved in voice and speech. As part of the collaboration, NIDCD scientists completed a Phase I clinical trial to determine the tolerance and response of individuals with advanced head and neck cancer to combined treatment with the chemotherapy agent Paclitaxel (Taxol®) and radiation. The study was based on laboratory observations that taxol can sensitize and increase response of cancer cells to radiation. All of the individuals entering the study had advanced or inoperable cancer of the head and neck. The study resulted in 70% of the patients with advanced cancers attaining a complete remission and preserving their voice and speech. Fifty-one percent remain in complete remission and 56% are alive three years after treatment, which is similar or better than results obtained with surgery in patients at the same stage. Treatment as an outpatient was well tolerated due to a low incidence of acute toxicity from chemotherapy, but side effects of the combined therapy included a several month delay of recovery of swallowing, which was relieved by nutritional supplements. Follow-up studies are likely to include the addition of a drug to reduce the side effects experienced in this trial.

In addition, new drugs that target the specific molecular abnormalities that cause cancers involving the vocal tract are being studied. NIDCD and NCI are collaborating to conduct a two-year, Phase I trial of a new drug to be given in combination with radiation for treatment of patients with cancers of the vocal tract (head and neck). The scientists discovered that a signal, called Nuclear Factor kappaB, is permanently switched on in head and neck cancer cells, activating other genes that cause these cancers to grow. Using cancer cells grown in the laboratory or in mice, the scientists were successful in blocking the activation of Nuclear Factor kappaB with an investigational drug, PS-341, and inhibiting survival and growth of cancers cells.

³Fast Stats, 1998 SEER Data for Larynx, Oral Cavity and Pharynx, National Cancer Institute.

These studies were supported by a Cooperative Research and Development Agreement between the NIH and Millennium Pharmaceuticals, the company that produces PS-341. Studies to identify the genes activated by Nuclear factor kappaB which cause these cancers are also underway, so that new tests may be developed for diagnosing and selecting the best therapy for individuals with head and neck cancer.

New Initiatives in Human Communication Research

Genetic Testing and the Clinical Management of Nonsyndromic Hereditary Hearing Impairment

In the last decade, approximately 20 genes whose mutations result in nonsyndromic hearing impairment have been identified and isolated. With the identification of genes that contribute to hearing function, genetic testing becomes technically possible but not necessarily suitable for widespread clinical application at present. There are many unresolved issues regarding the prevalence of genetic mutations in various populations, the clinical significance of these mutations, and the short and long-term impact of genetic testing on individuals and their families. There are unique issues to consider before implementing genetic testing for hearing impairment, since these conditions are not universally considered to be disabling or even undesirable.

Thirty-five states have now enacted some type of legislation that requires universal hearing screening for newborns. Not only are infants with severe hearing impairment identified much earlier in life but infants with lesser degrees of hearing impairment are now also being identified. Many unresolved issues remain for clinicians as they characterize auditory performance in a newborn who fails hearing screening, design intervention strategies to optimize communicative success and ensure that a “medical home” exists for the infant with hearing impairment.

The advances in the genetics of hereditary hearing impairment and in the early identification of hearing impairment have now converged. These advances have led some to suggest genetic testing/evaluation for all infants who are identified with a hearing loss at birth. In consideration of these developments, the NIDCD and the National Human Genome Research Institute are collaborating on an initiative to address the clinical relationship between genetic and audiologic/otologic information, as well as to address the clinical validity and utility of genetic testing in the diagnosis, treatment and management of nonsyndromic hereditary hearing impairment, as well as its psychosocial effects.

Mechanisms Underlying the Innervation of Specific Taste Receptor Cells

The process of how taste is perceived involves a dynamic interaction between individual taste receptor cells on the tongue and taste nerves that project to the brain. Taste receptor cells are continually being regenerated, and the new cells need to receive the proper neural connection to provide meaningful input into the brain. The factors that regulate and promote the patterns of connection between individual taste ganglion cells with specific types of taste receptor cells are still not known. Advances in cellular and molecular biology and genetics provide new

opportunities to understand these factors. The NIDCD has developed a new initiative for research on how nerve connections are established between specific types of taste receptor cells by peripheral gustatory ganglion neurons. These studies will contribute to knowledge on the mechanisms involved in nerve cells migration, process outgrowth from peripheral taste ganglion cells, axon guidance, selection of synaptic targets and the innervation of specific taste receptor cells.

Innovative Treatment Approaches to Autism

Autism is a developmental disability that is associated with problems in communication and social interaction. This disorder typically appears during infancy and lasts throughout a person's life. Currently, treatments for autism are limited in their effectiveness, providing only partial alleviation of the symptoms that characterize this disorder. Available pharmacological treatments are especially limited in the scope, duration and degree of their efficacy in lessening the problems created by this disorder. In addition, there is a lack of research data on the effectiveness of existing pharmacological, psychotherapeutic or other interventions. Thus, further characterization of existing treatments and the development of novel approaches are needed, as well as better measures of treatment. Following recommendations made at a 1999 workshop held by the NIH Autism Coordinating Committee, the National Institute of Child Health and Human Development, National Institute of Mental Health, National Institute of Neurological Disorders and Stroke, and the NIDCD have collaborated on an initiative for further research into the areas that could provide a foundation for new and innovative treatment for autism. The NIDCD is particularly interested in research on methods to teach speech to nonverbal children with autism.

Fast Track Grants for Parkinson's Disease Research

Parkinson's Disease (PD) is the most common neurodegenerative movement disorder. It is caused by selective degeneration of the nerve cells in the area of the brain that is critical for purposeful control of movement. As the disease progresses, further complications can arise such as cognitive impairment or dementia, depression, and disturbances of the autonomic nervous system. To promote more research relevant to this devastating disease, the NIH has entered into a unique partnership with several voluntary organizations. The NIDCD is cosponsoring an initiative with the National Institute of Neurological Disorders and Stroke, the National Institute of Environmental Health Sciences and the National Institute of Mental Health, that utilizes an expedited, fast track, award process to support new research for the cure, cause, prevention or improved treatment of PD and its complications. Also collaborating in this initiative is the Michael J. Fox Foundation for Parkinson's Research, the Parkinson's Disease Foundation/National Parkinson's Foundation and the Parkinson's Alliance. The NIDCD is specifically interested in research on speech and swallowing difficulties experienced by individuals with PD.

Innovations in Management and Administration

NIDCD research administrators are using innovative ways to stimulate research, research training and career development opportunities for individuals interested in research on human

communication. To attract new scientists to work in fields of basic, clinical, and health services research, the NIDCD is implementing several programs that will encourage new scientists to pursue careers in communication sciences and its disorders.

NIDCD is encouraging fellowship applications from new scientists by implementing a faster review process. Following the recommendations made by the Work Group on Research Training and Career Development (a meeting of scientists convened to discuss research training/career development relevant to the mission areas of the NIDCD), the Institute now conducts the initial peer review of National Research Service Awards Fellowship applications within its own Scientific Review Branch using an expedited four-month process from submission to award. Not only will these new scientists receive funding for their research sooner, but this expedited process provides enough time for those applicants who did not receive an award to reapply for the next application cycle. Applications for individual pre- and postdoctoral fellows, as well as dual degree (M.D./Ph.D.) fellowships for training physician-scientists are managed using this new process.

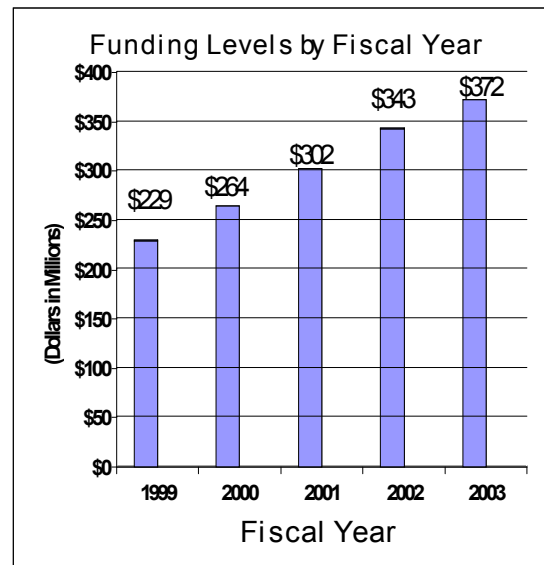
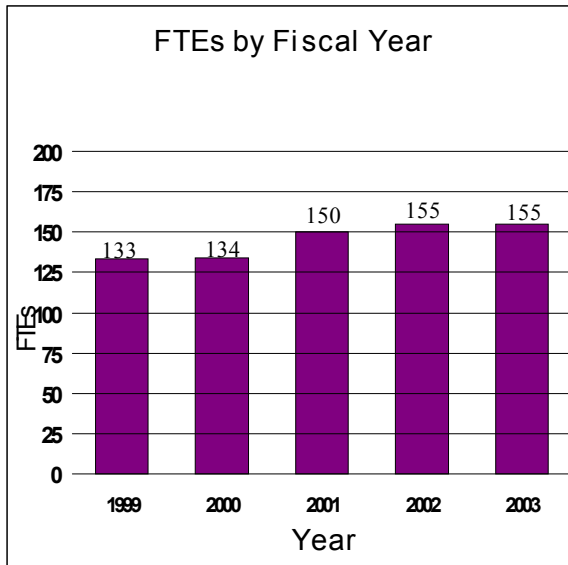
The NIDCD has also targeted its Small Grant Program (R03) for supporting scientists who are in the early stages of establishing an independent research career in the basic and clinical sciences of human communication. The NIDCD R03 may also be used to support individuals transitioning from postdoctoral status to their first independent research position. In addition, the R03 may be used by clinician-scientists planning to make future application for an NIDCD mentored research career development award.

NIDCD is giving high priority to applications from new independent research scientists. Applications from these scientists are given special consideration by the National Deafness and Other Communication Disorders Advisory Council. The applicant is given the unique opportunity to respond to concerns of the proposed research applications raised during initial peer review. These responses are reviewed by NIDCD program staff and Advisory Council members for their merit. This process provides scientists at the beginning stages of their research career with the opportunity to avoid the need for extensive revision of their application by allowing clarification of concerns about research plans raised by initial peer review.

Budget Policy

The Fiscal Year 2003 budget request for the NIDCD is \$371,951,000, including AIDS, an increase of \$28,880,000 and 8.4 percent over the FY 2002 level.

A five year history of FTEs and Funding Levels for NIDCD are shown in the graphs below. Note that Fiscal Years 2000 and 1999 are not comparable for the Managerial Flexibility Act of 2001 legislative proposal.



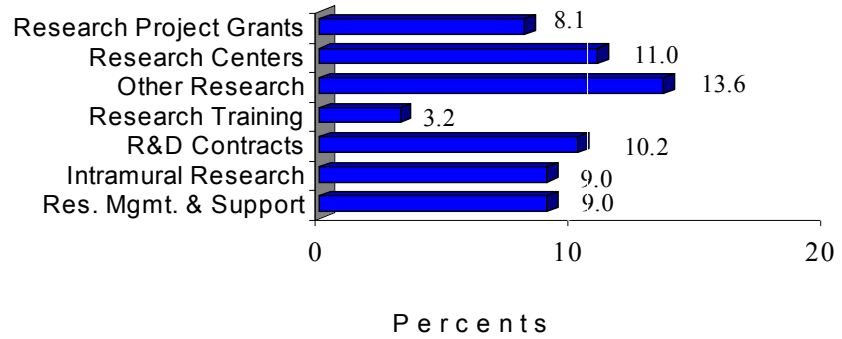
One of NIH's highest priorities is the funding of medical research through research project grants (RPGs). Support for RPGs allows NIH to sustain the scientific momentum of investigator-initiated research while providing new research opportunities. The Fiscal Year 2003 request provides average cost increases for competing RPGs equal to the Biomedical Research and Development Price Index (BRDPI), estimated at 4.0 percent. Noncompeting RPGs will be funded at committed levels which include increases of 3 percent on average for recurring direct costs.

Future promises for advancement in medical research rest in part with new investigators with new ideas. In the Fiscal Year 2003 request, NIDCD will support 309 pre- and postdoctoral trainees in full-time training positions, the same number as in FY 2002. Stipend levels for NRSA trainees will increase by 4 percent over Fiscal Year 2002 levels.

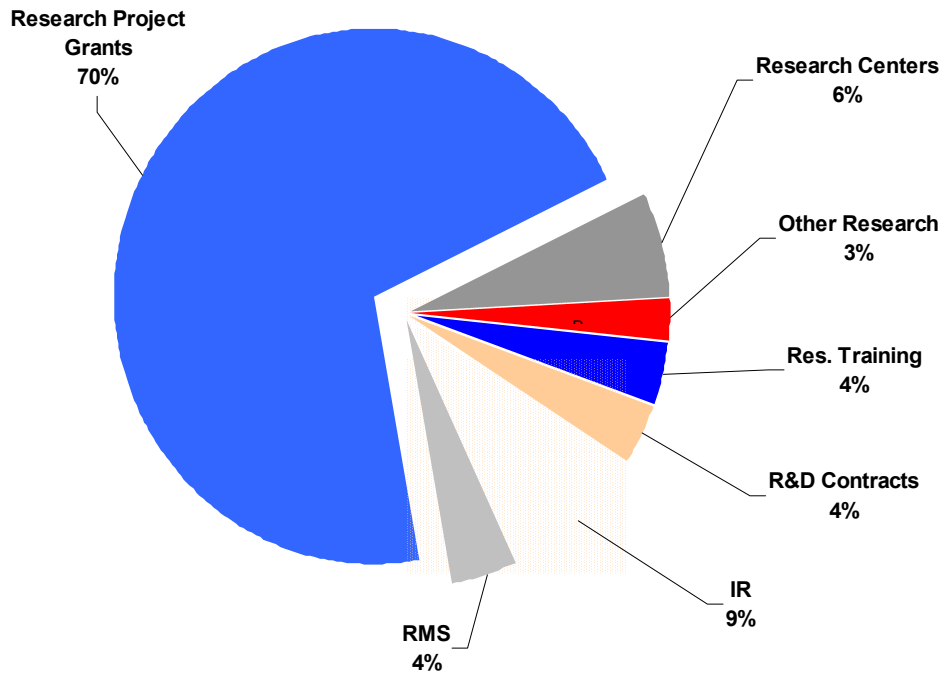
The Fiscal Year 2003 request includes funding for 26 research centers, 74 other research grants, including 43 clinical career awards, and 35 R&D contracts. The R&D contracts mechanism also includes support for 5 contracts for the Extramural Clinical and Pediatric Loan Repayment Programs. Intramural Research and Research Management and Support receive increases of 9 percent over FY 2002.

The mechanism distribution by dollars and percent change are displayed below.

FY 2003 Estimate
Percent Change
from
FY 2002 Mechanism



FY 2003 Budget Mechanism
(Dollars in Millions)



NATIONAL INSTITUTES OF HEALTH

National Institute on Deafness and Other Communication Disorders
**TOTAL - Current Law
 Budget Mechanism**

MECHANISM	FY 2001 Actual		FY 2002 Appropriation		FY 2002 Current Estimate		FY 2003 Estimate	
	No.	Amount	No.	Amount	No.	Amount	No.	Amount
Research Grants:								
<u>Research Projects:</u>								
Noncompeting	553	\$152,432,000	622	\$172,957,000	622	\$172,957,000	645	\$182,368,000
Administrative supplements	(30)	1,562,000	(30)	1,100,000	(30)	1,100,000	(35)	1,500,000
<u>Competing:</u>								
Renewal	69	24,336,000	73	26,498,000	73	26,498,000	82	30,904,000
New	160	32,102,000	163	34,183,000	163	34,183,000	178	38,721,000
Supplements	2	281,000	2	306,000	2	306,000	2	357,000
Subtotal, competing	231	56,719,000	238	60,987,000	238	60,987,000	262	69,982,000
Subtotal, RPGs	784	210,713,000	860	235,044,000	860	235,044,000	907	253,850,000
SBIR/STTR	30	6,890,000	30	7,932,000	30	7,932,000	35	8,862,000
Subtotal, RPGs	814	217,603,000	890	242,976,000	890	242,976,000	942	262,712,000
<u>Research Centers:</u>								
Specialized/comprehensive	19	16,963,000	24	21,547,000	24	21,547,000	26	23,919,000
Clinical research	0	0	0	0	0	0	0	0
Biotechnology	0	0	0	0	0	0	0	0
Comparative medicine	0	0	0	0	0	0	0	0
Research Centers in Minority Institution	0	0	0	0	0	0	0	0
Subtotal, Centers	19	16,963,000	24	21,547,000	24	21,547,000	26	23,919,000
<u>Other Research:</u>								
Research careers	43	5,091,000	41	5,397,000	41	5,397,000	43	6,281,000
Cancer education	0	0	0	0	0	0	0	0
Cooperative clinical research	0	0	0	0	0	0	0	0
Biomedical research support	0	0	0	0	0	0	0	0
Minority biomedical research support	0	0	0	0	0	0	0	0
Other	23	2,444,000	22	2,812,000	22	2,812,000	31	3,045,000
Subtotal, Other Research	66	7,535,000	63	8,209,000	63	8,209,000	74	9,326,000
Total Research Grants	899	242,101,000	977	272,732,000	977	272,732,000	1042	295,957,000
<u>Training:</u>								
Individual awards	101	3,550,000	129	4,406,000	129	4,406,000	134	4,785,000
Institutional awards	200	6,736,000	180	8,791,000	180	8,791,000	175	8,833,000
Total, Training	301	10,286,000	309	13,197,000	309	13,197,000	309	13,618,000
Research & development contracts (SBIR/STTR)	31 (0)	10,424,000 (0)	32 (0)	12,720,000 (0)	32 (0)	12,720,000 (0)	35 (0)	14,019,000 (0)
Intramural research	68	26,398,000	74	29,962,000	74	29,889,000	74	32,576,000
Research management and support	82	11,860,000	81	13,461,000	81	13,427,000	81	14,635,000
Cancer prevention & control	0	0	0	0	0	0	0	0
Construction		0		0		0		0
Total, NIDCD	150	301,069,000	155	342,072,000	155	341,965,000	155	370,805,000
(Clinical Trials)		(2,705,000)		(3,031,000)		(3,031,000)		(3,278,000)

National Institute on Deafness and Other Communication Disorders
TOTAL - Accrued Costs for Retirement and Health Benefits
Budget Mechanism

MECHANISM	FY 2001 Actual		FY 2002 Appropriation		FY 2002 Current Estimate		FY 2003 Estimate	
	No.	Amount	No.	Amount	No.	Amount	No.	Amount
Research Grants:								
<u>Research Projects:</u>								
Noncompeting								
Administrative supplements								
Competing:								
Renewal								
New								
Supplements								
Subtotal, competing								
Subtotal, RPGs								
SBIR/STTR								
Subtotal, RPGs								
<u>Research Centers:</u>								
Specialized/comprehensive								
Clinical research								
Biotechnology								
Comparative medicine								
Research Centers in Minority Institutions								
Subtotal, Centers								
<u>Other Research:</u>								
Research careers								
Cancer education								
Cooperative clinical research								
Biomedical research support								
Minority biomedical research support								
Other								
Subtotal, Other Research								
Total Research Grants								
<u>Training:</u>	<u>FTEs</u>		<u>FTEs</u>		<u>FTEs</u>		<u>FTEs</u>	
Individual awards								
Institutional awards								
Total, Training								
Research & development contracts (SBIR/STTR)								
<u>Intramural research</u>	<u>FTEs</u>		<u>FTEs</u>		<u>FTEs</u>		<u>FTEs</u>	
0	256,000	0	276,000	0	276,000	0	286,000	
Research management and support	0	768,000	0	830,000	0	830,000	0	860,000
Cancer prevention & control	0	0	0	0	0	0	0	0
Construction								
Total, NIDCD	0	1,024,000	0	1,106,000	0	1,106,000	0	1,146,000
(Clinical Trials)		(2,705,000)		(3,031,000)		(3,031,000)		(3,278,000)

NATIONAL INSTITUTES OF HEALTH

National Institute on Deafness and Other Communication Disorders
**TOTAL - Proposed Law
 Budget Mechanism**

MECHANISM	FY 2001 Actual		FY 2002 Appropriation		FY 2002 Current Estimate		FY 2003 Estimate	
	No.	Amount	No.	Amount	No.	Amount	No.	Amount
Research Grants:								
<u>Research Projects:</u>								
Noncompeting	553	\$152,432,000	622	\$172,957,000	622	\$172,957,000	645	\$182,368,000
Administrative supplements	(30)	1,562,000	(30)	1,100,000	(30)	1,100,000	(35)	1,500,000
<u>Competing:</u>								
Renewal	69	24,336,000	73	26,498,000	73	26,498,000	82	30,904,000
New	160	32,102,000	163	34,183,000	163	34,183,000	178	38,721,000
Supplements	2	281,000	2	306,000	2	306,000	2	357,000
Subtotal, competing	231	56,719,000	238	60,987,000	238	60,987,000	262	69,982,000
Subtotal, RPGs	784	210,713,000	860	235,044,000	860	235,044,000	907	253,850,000
SBIR/STTR	30	6,890,000	30	7,932,000	30	7,932,000	35	8,862,000
Subtotal, RPGs	814	217,603,000	890	242,976,000	890	242,976,000	942	262,712,000
<u>Research Centers:</u>								
Specialized/comprehensive	19	16,963,000	24	21,547,000	24	21,547,000	26	23,919,000
Clinical research	0	0	0	0	0	0	0	0
Biotechnology	0	0	0	0	0	0	0	0
Comparative medicine	0	0	0	0	0	0	0	0
Research Centers in Minority Institution	0	0	0	0	0	0	0	0
Subtotal, Centers	19	16,963,000	24	21,547,000	24	21,547,000	26	23,919,000
<u>Other Research:</u>								
Research careers	43	5,091,000	41	5,397,000	41	5,397,000	43	6,281,000
Cancer education	0	0	0	0	0	0	0	0
Cooperative clinical research	0	0	0	0	0	0	0	0
Biomedical research support	0	0	0	0	0	0	0	0
Minority biomedical research support	0	0	0	0	0	0	0	0
Other	23	2,444,000	22	2,812,000	22	2,812,000	31	3,045,000
Subtotal, Other Research	66	7,535,000	63	8,209,000	63	8,209,000	74	9,326,000
Total Research Grants	899	242,101,000	977	272,732,000	977	272,732,000	1042	295,957,000
<u>Training:</u>	<u>FTEPs</u>		<u>FTEPs</u>		<u>FTEPs</u>		<u>FTEPs</u>	
Individual awards	101	3,550,000	129	4,406,000	129	4,406,000	134	4,785,000
Institutional awards	200	6,736,000	180	8,791,000	180	8,791,000	175	8,833,000
Total, Training	301	10,286,000	309	13,197,000	309	13,197,000	309	13,618,000
Research & development contracts (SBIR/STTR)	31 (0)	10,424,000 (0)	32 (0)	12,720,000 (0)	32 (0)	12,720,000 (0)	35 (0)	14,019,000 (0)
Intramural research	<u>FTEs</u> 68	26,654,000	<u>FTEs</u> 74	30,238,000	<u>FTEs</u> 74	30,165,000	<u>FTEs</u> 74	32,862,000
Research management and support	82	12,628,000	81	14,291,000	81	14,257,000	81	15,495,000
Cancer prevention & control	0	0	0	0	0	0	0	0
Construction		0		0		0		0
Total, NIDCD	150	302,093,000	155	343,178,000	155	343,071,000	155	371,951,000
(Clinical Trials)		(5,410,000)		(6,062,000)		(6,062,000)		(6,556,000)

NATIONAL INSTITUTES OF HEALTH

National Institute on Deafness and Other Communication Disorders
Budget Authority by Activity ^{1/}
(dollars in thousands)

ACTIVITY	FY 2001 Actual		FY 2002 Estimate		FY 2003 Estimate		Change	
	FTEs	Amount	FTEs	Amount	FTEs	Amount	FTEs	Amount
Extramural Research: Deafness and Other Communication Disorders		\$262,811		\$298,649		\$323,594		\$24,945
Subtotal, extramural research		262,811		298,649		323,594		24,945
Intramural research	68	26,654	74	30,165	74	32,862	0	2,697
Research management and support	82	12,628	81	14,257	81	15,495	0	1,238
Total	150	302,093	155	343,071	155	371,951	0	28,880

^{1/} Please see the following tables for the crosswalk from current law to proposed law to reflect the administration's proposal for full accrued retirement and health benefits.

National Institutes of Health

National Institute on Deafness and Other Communication Disorders

2001 Crosswalk for Accrued Retirement and Health Benefit Costs
(Dollars in thousands)

	<u>2001 Actual Current Law</u>	<u>2001 Additional Accrual Costs</u>	<u>2001 Actual Proposed Law</u>
Extramural Research: Deafness and Other Communication Disorders	\$262,811	\$0	\$262,811
Subtotal, extramural resarch	262,811	0	262,811
Intramural Research	26,398	256	26,654
Research management and support	11,860	768	12,628
Total	301,069	1,024	302,093

National Institutes of Health

National Institute on Deafness and Other Communication Disorders

2002 Crosswalk for Accrued Retirement and Health Benefit Costs
(Dollars in thousands)

	2002 Current Estimate <u>Current Law</u>	2002 Additional <u>Accrual Costs</u>	2002 Appropriation <u>Proposed Law</u>
Extramural Research: Deafness and Other Communication Disorders	\$298,649	\$0	\$298,649
Subtotal, extramural research	298,649	0	298,649
Intramural Research	29,889	276	30,165
Research management and support	13,427	830	14,257
Total	341,965	1,106	343,071

National Institutes of Health

National Institute on Deafness and Other Communication Disorders

2003 Crosswalk for Accrued Retirement and Health Benefit Costs
(Dollars in thousands)

	2003 Estimate <u>Current Law</u>	2003 Additional <u>Accrual Costs</u>	2003 Estimate <u>Proposed Law</u>
Extramural Research: Deafness and Other Communication Disorders	\$324,594	\$0	\$323,594
Subtotal, extramural research	324,594	0	323,594
Intramural Research	32,576	286	32,862
Research management and support	14,635	860	15,495
Total	371,805	1,146	371,951

NATIONAL INSTITUTES OF HEALTH

National Institute on Deafness and Other Communication Disorders
Summary of Changes

2002 Estimated budget authority		\$343,071,000	
2003 Estimated budget authority		371,951,000	
Net change		28,880,000	
CHANGES	2002 Current Estimate Base		Change from Base
	FTEs	Budget Authority	FTEs Budget Authority
A. Built-in:			
1. Intramural research:			
a. Within grade increase		\$7,940,000	\$107,000
b. Annualization of January 2002 pay increase		7,940,000	97,000
c. January 2003 pay increase		7,940,000	157,000
d. Payment for centrally furnished services		5,143,000	463,000
e. Increased cost of laboratory supplies, materials, and other expenses		16,806,000	430,000
f. Accrued costs for retirement and health benefits		276,000	10,000
Subtotal			1,264,000
2. Research Management and Support:			
a. Within grade increase		7,332,000	125,000
b. Annualization of January 2002 pay increase		7,332,000	89,000
c. January 2003 pay increase		7,332,000	145,000
d. FTE reduction		7,332,000	(50,000)
e. Payment for centrally furnished services		1,569,000	141,000
f. Increased cost of laboratory supplies, materials, and other expenses		4,526,000	149,000
g. Accrued costs for retirement and health benefits		830,000	30,000
Subtotal			629,000
Subtotal, Built-in			1,893,000

NATIONAL INSTITUTES OF HEALTH

National Institute on Deafness and Other Communication Disorders
Summary of Changes--continued

CHANGES	2002 Current Estimate Base		Change from Base	
	No.	Amount	No.	Amount
B. Program:				
1. Research project grants:				
a. Noncompeting	622	174,057,000	23	9,811,000
b. Competing	238	60,987,000	24	8,995,000
c. SBIR/STTR	30	7,932,000	5	930,000
Total	890	242,976,000	52	19,736,000
2. Centers	24	21,547,000	2	2,372,000
3. Other research	63	8,209,000	11	1,117,000
4. Research training	309	13,197,000	0	421,000
5. Research and development contracts	32	12,720,000	3	1,299,000
Subtotal, extramural				24,945,000
6. Intramural research	<u>FTEs</u> 74	30,165,000	<u>FTEs</u> 0	1,433,000
7. Research management and support	81	14,257,000	0	609,000
8. Construction		0	0	0
Subtotal, program		343,071,000		26,987,000
Total changes	155			28,880,000

NATIONAL INSTITUTES OF HEALTH

National Institute on Deafness and Other Communication Disorders
Budget Authority by Object

	FY 2002 Appropriation	FY 2002 Current Estimate	FY 2003 Estimate	Increase or Decrease
Total compensable workyears:				
Full-time employment	155	155	155	0
Full-time equivalent of overtime and holiday hours	0	0	0	0
Average ES salary	\$137,901	\$137,901	\$141,486	\$3,585
Average GM/GS grade	10.5	10.5	10.5	0.0
Average GM/GS salary	\$62,874	\$62,874	\$64,508	\$1,634
Average salary, grades established by act of July 1, 1944 (42 U.S.C. 207)	\$99,360	\$99,360	\$101,943	\$2,583
Average salary of ungraded positions	\$67,275	\$67,275	\$69,024	\$1,749
OBJECT CLASSES	FY 2002 Appropriation	FY 2002 Estimate	FY 2003 Estimate	Increase or Decrease
Personnel Compensation:				
11.1 Full-Time Permanent	\$7,346,000	\$7,346,000	\$7,730,000	\$384,000
11.3 Other than Full-Time Permanent	3,283,000	3,283,000	3,445,000	162,000
11.5 Other Personnel Compensation	372,000	372,000	393,000	21,000
11.8 Special Personnel Services Payments	1,515,000	1,515,000	1,605,000	90,000
11.9 Total Personnel Compensation	12,516,000	12,516,000	13,173,000	657,000
12.1 Personnel Benefits	2,678,000	2,678,000	2,807,000	129,000
12.1 Personnel Benefits, Accrued Retirement Costs	724,000	724,000	752,000	28,000
13.0 Benefits for Former Personnel	2,000	2,000	2,000	0
Subtotal, Pay Cost, Current Law	15,196,000	15,196,000	15,982,000	786,000
Subtotal, Pay Cost, Proposed Law	15,920,000	15,920,000	16,734,000	814,000
21.0 Travel and Transportation of Persons	420,000	420,000	455,000	35,000
22.0 Transportation of Things	46,000	46,000	50,000	4,000
23.1 Rental Payments to GSA	0	0	0	0
23.2 Rental Payments to Others	650,000	650,000	690,000	40,000
23.3 Communications, Utilities and Miscellaneous Charges	580,000	580,000	650,000	70,000
24.0 Printing and Reproduction	216,000	216,000	227,000	11,000
25.1 Consulting Services	168,000	168,000	177,000	9,000
25.2 Other Services	3,535,000	3,535,000	3,713,000	178,000
25.3 Purchase of Goods and Services from Government Accounts	19,894,000	19,787,000	22,628,000	2,841,000
25.3 Accrued Retirement Costs	382,000	382,000	394,000	12,000
25.4 Operation and Maintenance of Facilities	1,203,000	1,203,000	1,258,000	55,000
25.5 Research and Development Contracts	6,524,000	6,524,000	7,267,000	743,000
25.6 Medical Care	233,000	233,000	242,000	9,000
25.7 Operation and Maintenance of Equipment	1,872,000	1,872,000	1,937,000	65,000
25.8 Subsistence and Support of Persons	0	0	0	0
25.0 Subtotal, Other Contractual Services, Current Law	33,429,000	33,322,000	37,222,000	3,900,000
25.0 Subtotal, Other Contractual Services, Proposed Law	33,811,000	33,704,000	37,616,000	3,912,000
26.0 Supplies and Materials	3,384,000	3,384,000	3,644,000	260,000
31.0 Equipment	2,222,000	2,222,000	2,310,000	88,000
32.0 Land and Structures	0	0	0	0
33.0 Investments and Loans	0	0	0	0
41.0 Grants, Subsidies and Contributions	285,929,000	285,929,000	309,575,000	23,646,000
42.0 Insurance Claims and Indemnities	0	0	0	0
43.0 Interest and Dividends	0	0	0	0
44.0 Refunds	0	0	0	0
Subtotal, Non-Pay Costs, Current Law	326,876,000	326,769,000	354,823,000	28,054,000
Subtotal, Non-Pay Costs, Proposed Law	327,258,000	327,151,000	355,217,000	28,066,000
Total Budget Authority by Object, Current	342,072,000	341,965,000	370,805,000	28,840,000
Total Budget Authority by Object, Proposed	343,178,000	343,071,000	371,951,000	28,880,000
Total Accrued Retirement Costs	1,106,000	1,106,000	1,146,000	40,000

NATIONAL INSTITUTES OF HEALTH

National Institute on Deafness and Other Communication Disorders
Salaries and Expenses

OBJECT CLASSES	FY 2002 Appropriation	FY 2002 Current Estimate	FY 2003 Estimate	Increase or Decrease
Personnel Compensation:				
Full-Time Permanent (11.1)	\$7,346,000	\$7,346,000	\$7,730,000	\$384,000
Other Than Full-Time Permanent (11.3)	3,283,000	3,283,000	3,445,000	162,000
Other Personnel Compensation (11.5)	372,000	372,000	393,000	21,000
Special Personnel Services Payments (11.8)	1,515,000	1,515,000	1,605,000	90,000
Total Personnel Compensation (11.9)	12,516,000	12,516,000	13,173,000	657,000
Civilian Personnel Benefits (12.1)	2,678,000	2,678,000	2,807,000	129,000
Accrued Costs of Retirement Benefits (12.1)	724,000	724,000	752,000	28,000
Benefits to Former Personnel (13.0)	2,000	2,000	2,000	0
Subtotal, Pay Costs, Current Law	15,196,000	15,196,000	15,982,000	786,000
Subtotal, Pay Costs, Proposed Law	15,920,000	15,920,000	16,734,000	814,000
Travel (21.0)	420,000	420,000	455,000	35,000
Transportation of Things (22.0)	46,000	46,000	50,000	4,000
Rental Payments to Others (23.2)	650,000	650,000	690,000	40,000
Communications, Utilities and Miscellaneous Charges (23.3)	580,000	580,000	650,000	70,000
Printing and Reproduction (24.0)	216,000	216,000	227,000	11,000
Other Contractual Services:				
Advisory and Assistance Services (25.1)	111,000	111,000	117,000	6,000
Other Services (25.2)	3,535,000	3,535,000	3,713,000	178,000
Purchases from Govt. Accounts (25.3)	13,132,000	13,132,000	15,374,000	2,242,000
Accrued Retirement Costs (25.3)	382,000	382,000	394,000	12,000
Operation & Maintenance of Facilities (25.4)	1,203,000	1,203,000	1,258,000	55,000
Operation & Maintenance of Equipment (25.7)	1,872,000	1,872,000	1,937,000	65,000
Subsistence & Support of Persons (25.8)	0	0	0	0
Subtotal, Other Contractual Services, Current Law	19,853,000	19,853,000	22,399,000	2,546,000
Subtotal, Other Contractual Services, Proposed Law	20,235,000	20,235,000	22,793,000	2,558,000
Supplies and Materials (26.0)	2,996,000	2,996,000	3,221,000	225,000
Subtotal, Non-Pay Costs, Current Law	22,849,000	22,849,000	27,532,000	4,683,000
Subtotal, Non-Pay Costs, Proposed Law	23,231,000	23,231,000	27,926,000	4,695,000
Total, Administrative Costs, Current Law	38,045,000	38,045,000	43,514,000	5,469,000
Total, Accrued Costs	1,106,000	1,106,000	1,146,000	40,000
Total, Administrative Costs, Proposed Law	39,151,000	39,151,000	44,660,000	5,509,000

NATIONAL INSTITUTES OF HEALTH

National Institute on Deafness and Other Communication Disorders

SIGNIFICANT ITEMS IN HOUSE, SENATE, AND CONFERENCE APPROPRIATIONS COMMITTEE REPORTS

FY 2002 House Appropriations Committee Report Language (H. Rpt.107-229)

Item

Dysphonia - The Committee continues to be pleased with NIDCD's expanding intramural research program with respect to dysphonia. The Committee encourages NIDCD to explore possibilities for a more active extramural research effort on dysphonia, and for collaboration with other NIH Institutes on this important disorder. (p. 158)

Action taken or to be taken

The NIDCD Extramural Research Program in Voice currently supports projects that focus on spasmodic dysphonia, which may be a form of dystonia. NIDCD-supported projects are providing physiological and neurological insights into the etiology of spasmodic dysphonia and methods for assessment and treatment. Examples include projects that examine methods for quantifying the irregular vocal fold oscillations in spasmodic dysphonia for the objective assessment of vocal hyperfunction as well as the perceptual qualities of the impaired voice.

Item

Noise-induced hearing loss - The Committee continues to be concerned by the number of Americans who suffer from noise-induced hearing loss. Thirty million Americans are exposed to dangerous levels of noise that can permanently impair their hearing. Ten million Americans have suffered irreversible noise-induced hearing loss. The Committee has been pleased by the Institute's efforts to tackle this preventable health problem. The Wise Ears campaign has the potential to make significant inroads towards educating Americans of all ages. The Committee urges the Institute to provide sufficient funds to expand this promising new initiative. (p. 158)

Action taken or to be taken

Ten million Americans have already suffered irreversible damage from noise, and 30 million are exposed to dangerous levels of noise each day, as reported in the NIH Consensus Development Conference: Consensus Statement - Noise and Hearing Loss, Vol. 8, No. 1, January 1990. Exposure to harmful sounds causes damage to the sensitive hair cells of the inner ear, eventually affecting hearing. These structures can be injured by noise in two different ways: from an intense brief impulse, such as an explosion from a firecracker; or from continuous exposure to noise, such as in a woodworking shop.

The WISE EARS! Campaign was initiated on July 4, 1999 and now includes a Coalition of more than 80 organizations of workers, employers, health and medical professionals, advocates for children and older Americans, teachers, parents, children, unions, industry, federal, regional and local government agencies and institutes, and the general public. The Wise Ears! Campaign has been published nationwide in over 1,000 newspapers with an estimated readership of 91 million. The campaign is designed to reach minority individuals as identified by specific occupational or recreational risk. Future plans include increasing the Coalition membership and expanding the campaign to all 50 states and territories.

In addition, a special outreach effort is underway to reach industrial workers, Hispanic/Latino/Latina individuals, and a special effort to reach Native American teenagers in work and recreational environments that are damaging to their hearing. Many materials are available in Spanish. This is a collaborative effort with the NIH Office of Hispanic Communication.

FY 2002 Senate Appropriations Committee Report Language (S. Rpt.107-84)

Item

Neurofibromatosis [NF] - The Committee encourages NIDCD to enhance its NF research and to coordinate its efforts with other Institutes conducting NF research. (p. 89)

Action taken or to be taken

The NIDCD continues to support studies to determine whether specific mutations in the NF2 gene result in different levels of disease severity. In addition, NIDCD continues to support the development of several new technologies to enhance the successful treatment of NF2 patients, such as development of a Doppler ultrasound cochlear blood flow monitor that will provide benefits in intra-operative monitoring during acoustic neuromasurgery, and as a diagnostic aid for sudden deafness. The NIDCD is also supporting the development of specialized auditory prosthesis for NF2 patients which stimulates the auditory brain stem directly.

NIDCD was one of several Institutes that provided major support for the National Neurofibromatosis Foundation (NNFF)-sponsored meeting of the International Consortium for the Molecular Biology of NF1 and NF2 held in May 2001 in Aspen, Colorado. At this gathering of the world's leading scientists working on NF, new and exciting results were reported by a number of different investigators in studies ranging from animal models to tumors to learning disabilities. The meeting was also structured to attract exceptional new investigators to the field of NF research. NIDCD also participated in a workshop in May 2000 to assess the status of NF research that included many other NIH Institutes, the DoD and the VA, and advocacy groups. The workshop identified research needs and opportunities in NF research that has guided NIDCD's activities in this area.

FY 2002 Conference Committee Report Language (H. Rpt.107-116)

Item

Hearing screening technologies - The conferees continue to support the expansion of NIDCD's research on the efficacy of new hearing screening technologies through all available mechanisms, as appropriate, including clinical studies on screening methodologies and studies on the efficacy of intervention and follow-up, and related research.

Action taken or to be taken

NIDCD continues to support research focusing on the early identification of hearing and has recently completed an initiative in early ID calling for clinical research grants.

The NIDCD has convened the Working Group on Early Identification of Hearing Impairment to provide advice on the most pressing research questions regarding diagnostic and intervention strategies following neonatal hearing screening. Based on these research recommendations, a Program Announcement was published requesting grant applications focusing on intervention strategies following identification of neonatal hearing impairment. Applications were encouraged that address relevant issues including, but not limited to: hardware (hearing aids, cochlear implants and other sensory aids); behavioral treatment programs; development of outcome measures to determine the benefit of intervention strategies; and, studies on the efficacy of intervention. The NIDCD made available approximately \$1.5 million in direct costs for the first year of support. Four awards have now been made. These applications address issues such as the optimization of fitting hearing aids in infants, auditory development in early amplified children, evaluating speech therapy in toddlers, and studying the effects of very early cochlear implantation on language. Not only is this initiative important to the research field, but it also has provided a funding opportunity to which the research audiology community has responded. As neonatal hearing screening advances, new questions are emerging and diligence in the training of an adequate pool of research clinicians remains critical to future endeavors.

NATIONAL INSTITUTES OF HEALTH

National Institute on Deafness and Other Communication Disorders
Authorizing Legislation

	PHS Act/ Other Citation	U.S. Code Citation	2001 Amount Authorized	2002 Estimate	2003 Amount Authorized	2003 Budget Estimate 1/
Research and Investigation	Section 301	42§241	Indefinite		Indefinite	
Deafness & Communication	Section 417B	42§285	Indefinite	\$329,874,000	Indefinite	\$358,333,000
National Research Service Awards	Section 487(d)	42§288	a/	13,197,000	b/	13,618,000
Total, Budget Authority				343,071,000		371,951,000

a/ Funding provided under the Departments of Labor, Health and Human Services, Education, and Related Agencies Appropriations Act, 2002 (P.L. 107-116).

b/ Reauthorizing legislation will be submitted.

1/ Reflects proposed transfer from the National Cancer Institute

NATIONAL INSTITUTES OF HEALTH

National Institute on Deafness and Other Communication Disorders
Appropriation History

Fiscal Year	Budget Estimate to Congress	House Allowance	Senate Allowance	Appropriation 1/
1994	\$153,088,000	\$162,823,000	\$162,813,000	\$162,823,000
1995	<u>2/</u> 167,129,000	166,155,000	167,129,000	166,761,000 <u>3/</u>
Rescission	0	0	0	(101,000)
1996	172,399,000 <u>2/</u>	174,852,000	170,540,000 <u>2/</u>	174,852,000
Rescission	0	0	0	(119,000)
1997	179,090,000 <u>2/</u>	189,243,000	182,693,000	188,422,000 <u>4/</u>
1998	192,477,000 <u>2/</u>	198,373,000	198,583,000	198,857,000
1999	213,184,000 <u>2/</u> <u>5/</u>	216,995,000	229,887,000	229,887,000
Rescission				(152,000)
2000	235,297,000 <u>2/</u>	251,218,000	261,962,000	265,185,000
Rescission				(1,414,000)
2001	276,418,000	301,787,000	303,541,000	300,581,000
Rescission				(100,000)
2002	336,757,000	334,161,000	349,983,000	342,072,000
Rescission				(107,000)
2003	370,805,000			

1/ Reflects enacted supplementals, rescissions and reappropriations.

2/ Excludes funds for HIV/AIDS research activities consolidated in the NIH Office of AIDS Research

3/ Excludes enacted administrative reductions of \$125,000

4/ Excludes enacted administrative reductions of \$77,000

5/ Reflects a decrease of \$650,000 for the budget amendment for bioterrorism.

NATIONAL INSTITUTES OF HEALTH

National Institute on Deafness and Other Communication Disorders
Detail of Full-Time Equivalent Employment (FTEs)

OFFICE/DIVISION	FY 2001 Actual	FY 2002 Estimate	FY 2003 Estimate
Office of the Director	8	8	8
Office of Administration	40	40	40
Division of Intramural Research	68	72	72
Division of Extramural Research	34	35	35
Total, NIDCD	150	155	155
Statutorily-ceiling exempt FTEs not included above	(0)	(0)	(0)
Funds to support these FTEs are provided by Cooperative Research and Development			
FISCAL YEAR	Average GM/GS Grade		
1999	10.5		
2000	10.5		
2001	10.5		
2002	10.5		
2003	10.5		

NATIONAL INSTITUTES OF HEALTH

National Institute on Deafness and Other Communication Disorders
Detail of Positions

GRADE	FY 2001 Actual	FY 2002 Estimate	FY 2003 Estimate
ES-6	0	0	0
ES-5	0	0	0
ES-4	0	0	0
ES-3	0	1	1
ES-2	1	0	0
ES-1	0	0	0
Subtotal	1	1	1
Total - ES Salary	\$125,883	\$137,901	\$141,486
GM/GS-15	19	19	19
GM/GS-14	9	10	10
GM/GS-13	13	13	13
GS-12	15	15	15
GS-11	14	14	14
GS-10	2	2	2
GS-9	16	16	16
GS-8	10	10	10
GS-7	5	6	6
GS-6	1	2	2
GS-5	6	6	6
GS-4	2	2	2
GS-3	3	3	3
GS-2	2	2	2
GS-1	0	0	0
Subtotal	117	120	120
Grades established by Act of July 1, 1944 (42 U.S.C. 207):			
Assistant Surgeon General	1	1	1
Director Grade			
Senior Grade			
Full Grade			
Senior Assistant Grade			
Subtotal	1	1	1
Ungraded	47	49	49
Total permanent positions	104	111	111
Total positions, end of year	166	173	173
Total full-time equivalent (FTE) employment, end of year	150	155	155
Average ES level	ES-2	ES-3	ES-3
Average ES salary	\$125,883	\$137,901	\$141,486
Average GM/GS grade	10.5	10.5	10.5
Average GM/GS salary	\$59,994	\$62,874	\$64,508