Deafness Research Foundation has once again increased its grant giving. Meet the 25 outstanding scientists whose groundbreaking research is devoted to hearing health.

Each year Deafness Research Foundation’s National Hearing Health Grants Center awards research grants to young investigators exploring new avenues in hearing and balance science. We are excited to announce that it has awarded more than $600,000 to 25 outstanding research scientists in 2011. For the second year in a row, this is an increase in our grant giving.

These funds support research in the following areas:
- Fundamental Auditory Research: development, genetics, molecular biology, physiology, anatomy, and regeneration biology
- Hearing and Balance Restoration: cochlear implants, sensory hair cell regeneration, and auditory nerve regeneration
- Hearing Loss: age-related, noise-induced, drug-induced, otosclerosis, and otitis media
- Central Auditory Processing Disorders
- Congenital Deafness: Usher syndrome, Connexin 26
- Vestibular and Balance Disorders: dizziness and vertigo, Ménière’s disease

Deafness Research Foundation (DRF) is the leading national source of private funding for research in hearing and balance science. Research made possible by DRF grants has resulted in dramatic innovations that have increased options for those living with hearing and balance disorders, as well as protected those at risk. Since our inception in 1958, we have awarded over $26.5 million through more than 2,000 scientific research grants to researchers who are dedicated to exploring new avenues of hearing and balance science. With the potential of hearing restoration through regeneration biology, the scope of DRF-funded research has expanded enormously. Since 1972, DRF has funded nearly 40 research grants that have been instrumental in the development, evaluation and improvement of cochlear implants. Approximately 188,000 implant procedures have been completed worldwide with beneficial results, especially in infants receiving cochlear implants.

DRF’s Scientific Review Committee and Council of Scientific Trustees reviewed applications from scientists at renowned research institutions throughout the United States. The selected research projects underwent competitive peer review for scientific merit and program relevance.

FIRST-YEAR DRF GRANT RECIPIENTS

Keith E. Bryan, Ph.D., University of Iowa Carver College of Medicine
Bryan earned his B.A. in biology from Simpson College in Iowa and his M.S. and Ph.D. in biochemistry at the University of Iowa, where he is now a postdoctoral fellow. During his Ph.D. work, Bryan studied how mutations in actin, an important structural protein in the specialized hair cells of the inner ear, may cause deafness. He credits “Biology of the Inner Ear” at the Marine Biological Laboratory in Woods Hole, Mass., for furthering his research interest.

**Research area:** fundamental auditory research

**Investigating the role of CaBP1 in KCNQ4 channel modulation:** KCNQ4 potassium channels play an important role in controlling the responsiveness of auditory hair cells to sound stimulation. Mutation of the gene encoding this channel causes deafness in humans, which may be due to the improper functioning of these channels in the ear. Bryan has identified a novel interaction between Ca2+ binding protein 1 (CaBP1), which is highly expressed in auditory hair cells, and KCNQ4. The goals of this research are to evaluate the functional consequences of this interaction on the cellular localization and biophysical properties of KCNQ4 channels in auditory hair cells.

**Long-term goal:** to understand at the molecular level how hair cells function normally in sound detection and to develop new therapeutic strategies for treating patients with genetic forms of hearing loss.

Brenton G. Cooper, Ph.D., Texas Christian University
Cooper received his B.S. in psychology from the University of New Mexico in 1993. In 2003, he completed his Ph.D. in psychology at the University of Utah. He then went on to a National Institutes of Health-funded postdoctoral position and

Presenting the 2011 DRF Grantees
a research faculty appointment in the biology department at the University of Utah. In 2007, he joined the faculty at Texas Christian University as an assistant professor of psychology in the College of Science and Engineering.

**Research area:** auditory processing/CAPD

**Lateralization of acoustic perception in Bengalese finches:** Cooper's research aims to further our understanding of how different sides of the brain are specialized for processing different frequencies of sounds. Auditory processing of speech and language is lateralized to the left hemisphere of the human brain. Cooper's specific aims are to determine whether auditory processing in the Bengalese finch is lateralized to specific sides of the brain, in humans, and to determine whether the lateralization is learned or genetically determined.

**Long-term goal:** to develop this animal model for testing and refining treatments for hearing loss and lateralized frequency processing deficits in humans, including CAPD.

**Elizabeth Dinces, M.D., M.S., Albert Einstein College of Medicine**

Dinces received her B.A. in chemistry from Amherst College in 1987. She went on to earn an M.D. in 1991 and a M.S. in clinical research in 2004, both from the Albert Einstein College of Medicine in the Bronx, N.Y. Dinces is board-certified in otolaryngology—head and neck surgery and has a subspecialty certification in neurotology. She is active in resident education, clinical otology, and neurotology, and in research at the Albert Einstein College of Medicine. Her clinical activities include an academic practice in the Bronx dealing with ear disease and skull base tumors.

**Research areas:** auditory research, CAPD

**Effects of aging on selective attention in complex, multisource sound environments:** Dinces’ basic science research focuses on understanding how the brain processes sounds into meaningful language and includes auditory scene analysis in the elderly, sound intensity processing in children, and development of auditory processing after cochlear implantation. The value of learning the role of attention and understanding the active and passive processes of stream segregation in aging populations will be to help develop therapeutic strategies to improve listening and understanding in noisy sound environments of aging adults.

**Long-term goal:** to explain mechanisms of auditory scene analysis that break down in aging.

**Sung-Ho Huh, Ph.D., Washington University School of Medicine**

Huh received his B.S. and M.S. from Korea University in South Korea in 1999 and 2001, respectively, and a Ph.D. from Washington University in St. Louis, Missouri, in 2009. Since then he has been working as a postdoctoral fellow at Washington University School of Medicine.

**Research area:** sensory cell regeneration

**Role of fgfs in cochlear sensory epithelium:** Congenital hearing loss, derived from inner-ear defects, is one of the most common hereditary disabilities, affecting one in 1000 children. Fgf20-null mice have congenital hearing loss associated with loss of sensory cells, and inactivation of both fgf9 and fgf20 result in a shortened cochlea. Huh's goal is to understand the cellular and molecular functions of fgf9 and fgf20 in inner-ear development in vivo.

**Long-term goal:** to direct the regeneration of damaged sensory tissue to restore or improve hearing.

**Albena Kantardzhieva, Ph.D., Massachusetts Eye and Ear Infirmary**

Kantardzhieva received her B.S. and M.Sc. in biotechnology from the Sofia University “St. Kliment Ohridski” in Bulgaria in 2000, and a Ph.D. in neurobiology from the University of Amsterdam/Netherlands Institute for Neuroscience in the Netherlands in 2006. Currently she is a postdoctoral fellow in the Eaton-Peabody Laboratory at the Massachusetts Eye and Ear Infirmary, Harvard Medical School, Boston.

**Research area:** fundamental auditory research

**Defining the interaction partners of major proteins in the hair cell’s synaptic ribbon:** Kantardzhieva’s research will investigate what proteins make up the synaptic ribbon, which is a structure specific to hair cells in the inner ear and photoreceptors in the retina. She developed a procedure to purify these ribbons and directly identify various proteins. Knowing the building blocks of the ribbon led to a hypothesis about the ribbon’s function. She believes that the ribbon breaks large neurotransmitter-filled cisterns into small vesicles and thus contributes to the ability of the ribbon synapses to encode graded responses to various signal intensities. The importance of the synaptic ribbon has been long recognized, but its exact function is still unknown.

**Long-term goal:** to explore how the proteins located at the synaptic ribbon work together to achieve signal propagation. This will add knowledge about the molecular mechanisms behind hearing, and in general will help us better understand communication among neurons elsewhere in the nervous system.

**Shuh-Yow Lin, Ph.D., University of California, San Diego, School of Medicine**

Lin received his Ph.D. in neurobiology from Yale University. He completed his postdoctoral training in zebrafish genetics at the Massachusetts Institute of Technology and in sensory neurobiology
from Harvard Medical School. He is currently an assistant professor of surgery in the Division of Otolaryngology—Head and Neck Surgery at the University of Florida.

**Research areas:** hearing loss, ototoxicity

**Prevention of aminoglycoside-induced hearing loss with the mitochondria-targeted antioxidant MitoQ:** Aminoglycoside antibiotics, such as gentamicin, are commonly used to treat serious bacterial infections due to bacteria. However, these drugs can cause hearing loss. At present, there is no solution to prevent hearing loss caused by aminoglycoside antibiotics. This research will determine if the antioxidant MitoQ will prevent hearing loss induced by aminoglycoside antibiotics.

**Long-term goal:** to develop and apply practicable intervention strategies to prevent hearing loss induced by drugs that are toxic to the inner ear.

---

**Debashree Mukherjea, Ph.D., Southern Illinois University School of Medicine**

Mukherjea received her M.S. in medical biochemistry from Kasturba Medical College, Manipal University, India, in 1999, and a Ph.D. in molecular biology, microbiology, and biochemistry from Southern Illinois University (SIU) School of Medicine in 2004. She did six years of postdoctoral training in the departments of pharmacology and surgery at SIU School of Medicine. She is currently a research assistant professor in the Department of Surgery, Division of Otolaryngology, at SIU School of Medicine.

**Research area:** noise-induced hearing loss (NIHL)

**Long-term goal:** to provide a tangible treatment option and a window of opportunity for treatment for NIHL.

**Carolyn P. Ojano-Dirain, Ph.D., University of Florida College of Medicine**

Ojano-Dirain received a B.S. in animal nutrition from the University of the Philippines in Los Baños, Laguna, Philippines, in 1997. In 2002, she received a M.S. in poultry nutrition and in 2006, a Ph.D. in cell and molecular biology, both from the University of Arkansas. Her postdoctoral training in mitochondrial metabolism and gene therapy was at the Department of Medicine at the University of Florida and she is currently a faculty member in the Department of Otolaryngology—Head and Neck Surgery at the University of Florida.

**Research areas:** deafness genes, hair cell synaptic transmission, acoustic trauma, auditory neuropathy

**Molecular mechanisms of synaptic transmission in hair cells:** Lin’s research is focused on understanding hair cells that detect sound and balance information in the ears. The research will test several candidate genes that may be critical for sending information by using hair cells in zebrafish, whose function can be effectively modulated and whose behavior can easily be observed.

**Long-term goal:** to provide important clues about how hair cells deliver information to the brain.

**Regie Lyn P. Santos-Cortez, M.D., Ph.D., Baylor College of Medicine**

Santos-Cortez graduated from the University of the Philippines Manila College of Medicine, Philippine General Hospital, for both her medical education and residency in otorhinolaryngology. She studied genetic epidemiology at the Erasmus Medical Centre in the Netherlands and did most of her Ph.D. work on the genetics of nonsyndromic hearing impairment at the Leal Lab at Baylor College of Medicine in Houston. Currently she serves as assistant professor at the Center for Statistical Genetics, Department of Molecular and Human Genetics, at Baylor College of Medicine.

---

**Erin K. Purcell, Ph.D., Kresge Hearing Research Institute, University of Michigan**

Purcell received her Ph.D. in biomedical engineering from the University of Michigan in 2008. She is a research fellow in the Kresge Hearing Research Institute at the university and is currently training in sensory neurophysiology. Her expertise includes neurophysiology, neural electrode interfaces, stem cell biology, and regenerative scaffolding.

**Research areas:** sensory neurobiology, auditory nerve regeneration, CAPD

**A stem cell-seeded nanofibrous scaffold for auditory nerve regeneration:** The research involves manipulating physical and chemical signals to attract nerve cells to the auditory brainstem. This research may yield insight into CAPD if disorganized inputs to the auditory brainstem contribute to the pathology of the disorder.

**Long-term goal:** to regenerate auditory nerves and understand the conditions that promote neural connections with the auditory brainstem, in order to ultimately restore hearing for patients with neural hearing loss and lay a foundation for therapies for individuals with auditory processing disorders.
**Research areas:** otitis media (middle-ear infection), genetics

**Identification of genes that predispose to chronic otitis media in the Ati population of Bolabog, Boracay, Philippines:** The study aims to identify genes predisposing to otitis media via the collection of clinical data and saliva in order to extract DNA within an indigenous population with a high prevalence of chronic otitis media. The study population is ideal for gene mapping due to limited number of founders and marriages only within the Ati population. Collected DNA will be submitted for genotyping and statistical analysis in order to implicate the genomic regions that most likely harbor causal genes for otitis media.

**Long-term goal:** the discovery of genes that are predisposing to otitis media, which will eventually lead to increased knowledge of the disease process behind it and the development of new diagnostic and treatment strategies.

**Zlatka P. Stojanova, Ph.D., House Research Institute**

Stojanova earned a diploma in biotechnology with genetic engineering in 1998 from Sofia University “St. Kliment Ohridski” in Bulgaria and a Ph.D. in human genetics from the University of Utah. Currently she is a postdoctoral fellow at the House Research Institute in Los Angeles.

**Research area:** auditory hair cell regeneration

**Epigenetic regulation of the Atoh1 gene:** Hearing loss is predominantly caused by the death of sensory hair cells in the cochlea. Hair cells cannot regenerate under normal circumstances. The Atoh1 gene is a key player in the development and differentiation of hair cells. The research will test the hypothesis that Atoh1 gene expression in the surrounding supporting cells is actively suppressed by nongenic factors in the nuclei of these cells, preventing sensory regeneration, and if so, whether these can be reversed.

**Long-term goal:** to understand and reverse this failure of hair cell regeneration.

**Jie Tang, Ph.D., Creighton University School of Medicine**

Tang received his B.S. and M.S. from Central China Normal University in 2000 and 2003, respectively. He went on to earn a Ph.D. in biophysics from the Institute of Biophysics, Chinese Academy of Sciences. In 2006 he began his work on auditory cortex plasticity at Washington University in St. Louis as a research associate. Since 2009, he has continued his postdoctoral training as a research fellow in the Hair Cell Biophysics Laboratory at Creighton University School of Medicine, Nebraska.

**Research area:** auditory hair cell regeneration

**Creation of a pendrin with both motor and transport functions:** Prestin is the motor protein of cochlear outer hair cells contributing to the exquisite sound selectivity and sensitivity of our hearing. Pendrin is an anion transporter that is also expressed in inner-ear cells. Tang’s aim is to determine the functional molecular regions of pendrin and prestin by swapping the putative motor and transporter regions between two proteins.

**Long-term goal:** to improve detection and treatment of human deafness caused by prestin and pendrin defects.

---

**Ellen S. Wilch, Ph.D., Michigan State University College of Human Medicine**

Wilch received a B.S. degree in biology and geology from Bucknell University in Pennsylvania and a M.S. degree in hydrology from the New Mexico Institute of Mining and Technology. She went on to receive a Ph.D. in genetics from Michigan State University. She is currently a postdoctoral research associate at Michigan State University.

**Research area:** molecular biology of hearing

**Identification of cis-regulatory GJB2 and GJB6 elements by chromosome conformation capture and investigation of potential cis-regulatory variants in persons with hearing loss and monoallelic mutation of GJB2:** Mutations in the protein-coding sequence of GJB2 cause the amino acid sequence of its protein product, Connexin 26, to be altered or shortened. This leads to loss of function of the Connexin 26 protein with hearing loss as a result. “Chromosome conformation capture” is an experimental approach that will allow us to closer pinpoint those specific DNA sequences that are responsible for regulating GJB2.

**Long-term goal:** to look for mutations of these gene sequences in patients whose hearing loss is not fully explained.

**Hsiao-Huei Wu, Ph.D., Keck School of Medicine, University of Southern California**

Wu received her Ph.D. in biological sciences from Beckman Research Institute at City of Hope in Duarte, Calif. She obtained her postdoctoral training in neural development at the University of California, Irvine, and at Vanderbilt University in Nashville. Currently she holds the position of assistant professor of research at the Keck School of Medicine, University of Southern California.

**Research area:** hearing abnormality in autism

**Regulation of inner-ear development by HGF, the nonsyndromic hearing loss gene, DFNB39:** The objective is to understand the molecular bases of hearing abnormalities in autism spectrum disorders (ASD). The prevalence of ASD diagnoses among deaf children is twice that of the general population among hearing children.
population, and more than 50 percent of ASD patients with clinically normal hearing display an abnormal auditory brainstem response.

**Long-term goal:** to understand at the molecular and cellular level how these mutations in the hepatocyte growth factor gene (HGF) can lead to hearing abnormality.

**SECOND-YEAR DRF GRANT RECIPIENTS**

**Edward L. Bartlett, Ph.D., Purdue University**
Bartlett graduated from Haverford College in Pennsylvania in 1992 with a degree in physics. He received his Ph.D. in neuroscience from the University of Wisconsin-Madison in 1999, and did postdoctoral work at the University of Wisconsin-Madison and at Johns Hopkins University in Baltimore before joining Purdue University in Indiana in 2006. Currently he is an assistant professor in biological sciences and biomedical engineering at Purdue.

**Research area:** CAPD

**Cellular bases of temporal auditory processing:** Auditory thalamic neurons provide sensory input to the auditory cortex, and aberrations here are correlated with dyslexia, Alzheimer’s disease, and impaired cortical development in humans. Despite its importance in normal and abnormal hearing, the cellular and synaptic mechanisms by which auditory thalamic neurons process sound are poorly understood. The synaptic mechanisms of the main inputs to auditory thalamic neurons will be studied using patch-clamp recording techniques in young and developing animals, and these cellular responses will be correlated with noninvasive measures of auditory function.

**Long-term goal:** to predict and redevelop the cellular dynamics that underlie in vivo responses of auditory thalamic and auditory cortical neurons in normal and pathological conditions, including age-related decline, dyslexia, and aberrant auditory experience during development.

**Soyoun Cho, Ph.D., Oregon Health & Science University**
Cho received her Ph.D. in neuroscience from the University of Pittsburgh in 2007. Since beginning her postdoctoral training, she has been working as fellow at the Vollum Institute at Oregon Health & Science University in Portland, Ore.

**Research areas:** cochlea, cochlear implant

**Dynamics of exo- and endocytosis at hair cells:** The sense of hearing starts at the hair cell synapses in the cochlea. Hair
cells convert sound waves into bioelectrical signals. To hear sound correctly, the amplitude, duration, and frequency of sound information should be delivered to the auditory nerve very accurately at this first synapse of the auditory system. It is important for hair cell synapses to manage this task continuously for extended periods of time and throughout the person’s lifetime. However, the mechanisms underlying this unique synaptic transmission are not fully understood.

**Long-term goal:** to investigate the basic characteristics of the hair cell synapse in order to improve the accuracy of auditory nerve stimulations required for cochlear implantation.

Frances Hannan, Ph.D., New York Medical College
Hannan received an honors degree in science education in 1981, and a Ph.D. in genetics in 1990, from the University of Melbourne, Australia. She has extensive postdoctoral research experience obtained at SUNY Buffalo, Cambridge University in the United Kingdom, and Cold Spring Harbor Laboratory in New York. She is currently an assistant professor of cell biology and anatomy and otolaryngology at New York Medical College in Valhalla, N.Y.

**Research area:** sensorineural deafness

The role of diaphanous in the auditory cytoskeleton: Mutations affecting Diaphanous proteins cause deafness in humans and fruit flies. The goal of this research is to understand how Diaphanous proteins function within the stretch sensitive nerve cells of the fruit fly’s auditory organ. Sophisticated live imaging techniques will be used to track the proteins in real time throughout the development of the auditory organ.

**Long-term goal:** to develop new treatments or diagnostics for human hereditary deafness and auditory neuropathy.

Michelle Hastings, Ph.D., Rosalind Franklin University of Medicine and Science
Hastings received a Ph.D. in biology from Marquette University in Wisconsin and was a postdoctoral fellow at Cold Spring Harbor Laboratory in New York. She joined the faculty of the Chicago Medical School at Rosalind Franklin University of Medicine and Science as an assistant professor in 2007.

**Research area:** Usher syndrome

**Therapeutic correction of USH1C splicing in a mouse model of Usher syndrome:** Hastings’ research investigates the molecular basis of neurodegenerative diseases with a focus on Usher syndrome, the leading genetic cause of combined sensorineural deafness and retinal degeneration.

**Long-term goal:** to develop new treatments or diagnostic tools for Usher syndrome.

Here’s a little something to move your patient.

It’s Groove, the microCIC that plays to the rhythms of everyday living. Designed to slip unnoticed into the second bend of the ear canal, Groove has no extraneous hardware to get in the way of personal style. But even out of sight, Groove shines in difficult, noisy situations. Driven by the quality sound Sonic Innovations is known for plus powerful noise reduction, Groove naturally fits the tempo of your patient’s active life.
deafness and blindness. The Hastings Lab is developing approaches to correct the deafness associated with the disease using gene therapy—specifically, antisense oligonucleotides that target the molecular defect caused by a gene mutation in the USH1C gene.

**Long-term goal:** to use gene therapy as a treatment for Usher syndrome and other genetic causes of deafness.

Zhengqing Hu, M.D., Ph.D., Wayne State University School of Medicine

Hu earned his M.D. and Ph.D. degrees from Shanghai Medical University in China in 1999 and a second Ph.D. from Karolinska Institute in Sweden in 2005. He began his postdoctoral training at the University of Virginia in 2005 and is currently an assistant professor in the Department of Otolaryngology—Head and Neck Surgery at Wayne State University School of Medicine in Michigan.

**Research areas:** CAPD, regeneration

**Innervation of in vitro-produced hair cell by neural progenitor-derived glutamatergic neurons:** Hair cells and auditory neurons are usually damaged in patients with hearing loss. This research investigates novel strategies to generate mammalian hair cells in the culture dishes and study whether mammalian stem cell-derived neurons can form neural contacts with newly produced hair cells both in the culture dish and in lab animals.

**Long-term goal:** to develop effective strategies to regenerate auditory pathways using stem cell-based approaches to restore hearing. If successful the research will then explore whether stem cell-derived cells can be used to regenerate auditory cells that are damaged in CAPD.

Judith Kempfle, M.D., Massachusetts Eye and Ear Infirmary

Kempfle received her medical degree from Ulm University Medical School, Germany, in 2008. During medical school, she did epilepsy research but changed her focus to inner-ear regeneration. In 2008, she joined the Eaton-Peabody Laboratories at the Massachusetts Eye and Ear Infirmary in Boston as a postdoctoral research fellow.

**Research areas:** inner-ear stem cells, regeneration

**Influence of bone morphogenetic protein 4 and retinoic acid on differentiation of inner-ear stem cells:** Kempfle’s research goal is to understand and influence the pathways that lead to the formation of hair cells from inner-ear stem cells in vitro and to increase the yield of hair cells with specific compounds.

**Long-term goal:** the better understanding of pathways and drug screening in order to help to develop a drug that can be easily applied to the ear and that can stimulate dormant inner-ear stem cells in vivo for regeneration strategies after hearing loss.

(For more about Kempfle and her research, please see “Meet the Researcher,” page 58.)

Neeliyath A. Ramakrishnan, Ph.D., Wayne State University School of Medicine

Ramakrishnan earned a Ph.D. in molecular biology in 2002 from the University of Calcutta in India. Currently he is studying molecular interactions of ion channels and synaptic proteins in vertebrate hair cells at the Department of Otolaryngology at Wayne State University School of Medicine in Michigan.

**Research area:** auditory research

**Molecular interactions of the hair-cell afferent synapse:** Ramakrishnan’s research will focus on understanding how synaptic neurotransmission from hair cells are regulated. Using tools to detect protein-protein and molecular interactions, the aim is to elucidate the complex regulatory mechanism of hair cell neurotransmission, which is essential for hearing and balance. He will explore calcium sensor proteins in hair cells and focus on otoferlin, a calcium-binding protein essential for hearing.

**Long-term goal:** to provide useful information about the nature and complexity of molecular interaction in hair cell synaptic transmission, which may help develop noninvasive methods of treatment for some forms of deafness.

**DRF CENTURION CLINICAL RESEARCH AWARD RECIPIENT**

James E. Saunders, M.D., Dartmouth Hitchcock Medical Center

Saunders is an associate professor of otology/neurotology at Dartmouth-Hitchcock Medical Center in New Hampshire. He completed his M.D. at the University of Oklahoma, his residency and a research fellowship in otolaryngology at Duke University Medical Center in North Carolina, and a research and clinical fellowship at the House Research Institute in Los Angeles. He currently serves as chairman of the Humanitarian Efforts Committee of the American Academy of Otolaryngology—Head and Neck Surgery (AAO-HNS) Foundation and has been involved in many projects in international medicine related to the etiology, prevention, and treatment of hearing loss in Nicaragua and the developing world.

**Research area:** sensorineural hearing loss

**Genetic hearing loss in remote Nicaraguan families:** Nicaragua is the second poorest country in the western
hemisphere and Jinotega, Nicaragua, is geographically and economically isolated from the rest of the country by mountainous terrain and poverty. Sensorineural hearing loss is common in the children of Jinotega and a common observation in this population is an idiopathic, slowly progressive, flat sensorineural hearing loss. Saunders’ research will investigate three large families with hereditary hearing loss that is characteristic of the Jinotega population, including a detailed search for any distinguishing features of these individuals and an analysis of candidate genes in DNA samples from representative family members. He will aim to determine the specific characteristics of hearing impaired individuals in each of these families through audiometric testing; to collect and isolate DNA samples from one or two members of each family; and to test these DNA samples for known candidate genes appropriate to the characteristics and inheritance pattern of the family.

Long-term goal: to identify genes associated with hearing loss.

This research award is funded by the Centurions of the Deafness Research Foundation. DRF has partnered with CORE Grants Program of the AAO-HNS to offer a one-year DRF Centurion Clinical Research Award for clinical research in hearing and balance science.

DRF C.H.E.A.R. ENDOWMENT GRANT RECIPIENT

Patricia White, Ph.D., University of Rochester School of Medicine and Dentistry

First-Year Grant Recipient

White received her B.S. in biology and a Ph.D. in developmental biology from the California Institute of Technology (Caltech) in Pasadena in 1989. She completed her Ph.D. in developmental biology, also at Caltech, in 2000, where she researched neural stem cells. She began her postdoctoral work in hearing regeneration at the House Research Institute in Los Angeles, and joined the faculty at the University of Rochester School of Medicine and Dentistry in New York in 2010.

Research areas: hearing loss/ototoxicity, sensorineural deafness

The role of FOXO3 in hearing protection: White’s research will investigate whether a stress-induced transcription factor called FOXO3 promotes the cellular death process (apoptosis) in cochlear sensory hair cells. Mice lacking the FOXO3 gene appear resistant to age-related hearing loss (presbycusis). Levels of apoptosis regulators in hair cells of genetically altered and unaltered mice will be measured. Hair cell survival in both kinds of mice will be measured when they are exposed to ototoxic drugs as newborns and to noise as adults.

Long-term goal: to prevent hearing loss from ototoxic drugs and presbycusis.

The C.H.E.A.R. endowment was created to support an annual sensorineural Deafness Research Grant. C.H.E.A.R. (Children Hearing Education and Research) was absorbed into DRF in 1991, and we are very proud to continue its legacy of funding research in sensorineural deafness.

COLLETTE RAMSEY BAKER RESEARCH AWARD RECIPIENT

Kirill Vadimovich Nourski, MD, Ph.D., University of Iowa Hospitals and Clinics

First-Year Grant Recipient

Nourski received an M.D. from Saint Petersburg State University in Russia in 2001. He received a Ph.D. in neuroscience from the University of Iowa in 2006. His Ph.D. thesis project investigated response properties of the auditory nerve under the conditions of combined acoustic and electric stimulation. Currently he is a research assistant professor in the Department of Neurosurgery at the University of Iowa.

Research area: CAPD

Temporal processing in human auditory cortex: Nourski’s research seeks to characterize the capacity of different auditory cortical fields for representation of frequency, intensity, and temporal information (auditory envelope) and to determine where and how utilizing the temporal envelope cues for perception and comprehension by the listener can affect cortical envelope tracking. Understanding auditory cortical mechanisms that underlie temporal envelope processing in normal-hearing listeners is a key prerequisite for advances in diagnosis, treatment, and rehabilitation in individuals with hearing impairments including CAPD.

Long-term goal: to gain better understanding of how the human auditory cortex processes perceptually important temporal features of speech sounds.

THE TODD M. BADER RESEARCH GRANT OF THE BARBARA EPSTEIN FOUNDATION, INC., RECIPIENT

Marcello Peppi, Ph.D., Massachusetts Eye and Ear Infirmary

Second-Year Grant Recipient

Peppi received his Ph.D. in physiology from the University of Cagliari, Italy, in 2003. He began his postdoctoral training at the Center for Molecular Medicine and Genetics at the Wayne State Medical
School in Detroit. He is currently an instructor in the Department of Otology and Laryngology at Harvard Medical School and research associate at the Massachusetts Eye and Ear Infirmary.

Research area: hearing loss treatments

Molecular mechanisms of dexamethasone-mediated protection from acoustic trauma: Corticosteroids are widely used for the treatment of inner-ear disorders. The possible presence of a protective subpathway among the broad variety of pathways initiated by corticosteroids may provide significantly improved therapies for degenerative hearing loss. Corticosteroids have also proven useful in other nervous system traumatic disorders, such as brain tumors and spinal cord injury, though those mechanisms of protection have not been identified. The protein PLZF is present throughout the nervous system. If, as in the cochlea, PLZF is the trigger for protection in those cases, findings on its role in cochlear protection could have broader implications.

Long-term goal: to identify specific proteins involved in hearing protection and in the prevention of cochlear degeneration, leading to new insights for the development of treatments for hearing degeneration and loss.

This research award is funded by The Todd M. Bader Research Grant of The Barbara Epstein Foundation, Inc.