Physiologists Discover Molecular Mechanism for Stabilizing Inner Ear Cells, with Implications for Hearing Loss

University of Kentucky (UK) physiologist Catalina Vélez-Ortega and her collaborators are using an AHRF grant to explore the molecular mechanisms of inner ear sensory cells. An April 17, 2017 UK press release explains:

Mechanosensory hair cells in the inner ear pick up the softest sounds, such as whispers and distant noises.

Unlike other cells in the human body, these sensory cells are fragile and finite. At birth, the human ear contains approximately 15,000 of these cells. They do not regenerate or divide, and therefore are susceptible to permanent damage from exposure to loud sounds. Scientists believe understanding the molecular mechanisms that maintain the structure of these cells throughout the lifespan can provide insight into the fundamental causes of hearing loss and deafness.

UK physiologists Vélez-Ortega, Gregory Frolenkov and their collaborators in the UK College of Medicine have discovered a molecular mechanism necessary for stabilizing stereocilia, the “hairs” of the sensory cells of the inner ear. (Continued on page 5)

Remembering Daniel J. Knight

The American Hearing Research Foundation (AHRF) lost one of its officers and board members last December. Daniel J. Knight passed away from complications related to treatment his was receiving for renal cancer. Dan was 58 years old.

Dan joined AHRF as a board member in 1993, and served as the organization’s Treasurer from 2005 to 2016. He was instrumental in helping to modernize and update the Foundation’s financial processes and investment practices. In his professional life, Dan held a variety of senior level finance positions, including Treasurer of The Chicago Sun-Times.

“Dan was both my brother-in-law and friend, and he will be truly missed by our family,” said fellow AHRF board member Mark Muench. “His dedication and service to the Foundation will be hard to replace.” (Continued on page 3)
Apply for Grants by August 15

The American Hearing Research Foundation is accepting applications for its 2018 grant cycle. Applications are due **August 15, 2017**. Information about grant awards, eligibility, topics, and review process can be found at [http://american-hearing.org/research-grants/](http://american-hearing.org/research-grants/). The application process is described at [http://american-hearing.org/research-grants/grant-application-guidelines/](http://american-hearing.org/research-grants/grant-application-guidelines/).

In addition to its usual grant opportunities, in 2018 AHRF will partner with the American Cochlear Implant Alliance to offer a one-year $25,000 grant for purposes of **planning a future study** of societal costs of severe to profound hearing loss.

In 2017, nine grants totaling $201,000 were awarded to scientists. Several studies are highlighted in this issue. To read more about the 2017 awards, visit [http://american-hearing.org/2017-recipients/](http://american-hearing.org/2017-recipients/).

### Hearing Aid News

- In March 2017, the **Over-the-Counter Hearing Aid Act of 2017** was introduced in both the U.S. Senate and U.S. House. The bill would make certain types of hearing aids – those intended for adults with mild to moderate hearing impairment – available over the counter. The bill would remove the requirement that consumers obtain a medical evaluation in order to get an OTC hearing aid. The bill also would require the FDA to issue regulations containing safety and labeling requirements for this new category of OTC hearing aids. Source: [http://www.hearingreview.com/2016/11/senators-warren-grassley-introduce-otc-hearing-aid-act-2016/](http://www.hearingreview.com/2016/11/senators-warren-grassley-introduce-otc-hearing-aid-act-2016/)

- **Study compares PSAPs and Hearing Aids.** Findings from a small study conducted by researchers at Johns Hopkins School of Medicine in Baltimore, Maryland, suggest over-the-counter products may be an effective alternative to more costly hearing aids for some people. Speech understanding for 42 participants with mild to moderate hearing loss was compared while using five personal sound amplification products (PSAPs) and one hearing aid dispensed in a university audiology clinic. Three of the five PSAPs tested improved hearing nearly as well (within 5 percentage points) of the hearing aid. However, the lowest cost PSAP made hearing worse than using nothing at all. *JAMA*. 2017;318(1):89-90. doi:10.1001/jama.2017.6905

### Who Supports AHRF?

**Kori Hall**

“I recently had an ear infection that made me very dizzy and disoriented for about a week. I have a newborn, and it was really hard to care for her. I went through a whole gamut of emotions including fear, anxiety, and depression (not helped at all by me being two months postpartum!). I decided that after I got well I’d learn more about organizations that work on helping those who deal with this on a regular basis. My hope is that anyone feeling this way can get help when they need it and that research into these debilitating disorders can continue!”
PROGRESS REPORT

ALLISON COFFIN, PH.D.
Assistant Professor of Neuroscience
Washington State University Vancouver

Dr. Coffin’s study “High-throughput drug discovery for prevention of noise-induced hair cell loss” is funded by a 2017 AHRF grant.

Our study examines a carefully selected suite of potential otoprotective compounds for drug candidates that reduce the effect of noise-induced hair cell death. Our lab uses tiny zebrafish larvae to understand how noise damages hearing cells and to develop protective therapies. We have preliminary evidence that MM-201, a new drug candidate developed by M3 Biotechnologies, may reduce hearing loss due to noise over-exposure. We have also shown that antioxidants reduce this hearing loss, which is consistent with previous studies. These studies show that zebrafish hearing cells respond to noise like mammalian hearing cells, meaning we can use zebrafish to test new potential drugs.

Daniel J. Knight (Continued from p.1)

Dan spent a considerable amount of his time working in the service of others. In addition to his duties at AHRF, Dan served as an alderman for the City of Park Ridge, Illinois. Dan was first elected to the city council in 2011 and was set to finish his current term in 2019. His duties as alderman also included being the chair of the finance and budget committee for the city council.

“He was a great alderman, and a better person,” said Park Ridge Mayor Marty Maloney. “He brought a great deal of energy and dedication to what he did for the city.” While serving as alderman, Dan was widely credited for helping to stabilize the City’s precarious financial situation and instrumental in setting the city on course for financial stability.

“Dan was a great colleague and a great friend,” said fellow alderman Marc Mazzuca. “He always wanted the best for the City, and he wasn’t the type who was going to settle for anything less. I’m not sure the general public always knew how much work Dan put in behind the scenes to make good things happen, but everyone on the council did.”

Dan was also a devoted husband and father to two sons. He was married to his wife Sue for 36 years. Together, they raised William and Michael. Today, William works as a project manager for a Chicago-based general contracting firm, and Michael is finishing up his undergraduate studies.

Dan was both a gregarious and outspoken individual. He was never afraid to present his opinion on any number items, and always brought a reasoned, thought-out approach to deal with issues and problems. Dan will be sorely missed by everyone that had the opportunity to know and to work with him.

AHRF would like to ask that you keep Dan, Sue, and their family in your thoughts and prayers.
RESEARCHER PROFILE
NING HU, MD, PH.D.
Associate Research Scientist
Department of Biology, University of Iowa, Iowa City

Dr. Hu’s study “Investigation of gender differences in noise-induced cochlear synaptopathy” is funded by a 2017 AHRF grant.

Dr. Hu explains: Noise, even at sound levels too low to kill hair cells of the inner ear, can damage or destroy the synapses between hair cells and auditory nerve. This is termed “noise-induced cochlear synaptopathy (NICS).” Correspondingly, newly-recognized “hidden hearing loss” has been proposed as well, because such loss is hardly detectable with conventional clinic examinations.

NICS occurs at noise levels that may be commonly encountered in the workplace or during recreational activities. Thus, it is an increasingly significant cause of hearing impairment.

Our studies have shown a sex difference, with female mice significantly less susceptible to NICS than males. We also discovered that there is greater variability in susceptibility among females than among males and the variability in females tends to vary with stage of the estrous cycle. In this funded project we will primarily test if the NICS is correlated to the estrus stages and underlying circulating female hormones. We will define which female hormone(s) will confer protection against NICS. This study will provide information for individuals to better protect themselves against noise trauma and could lead to new pharmacological strategies for prevention and therapy of noise damage.

On a personal note, I was born in Wuhu, a city in southeast China. In China, I studied Clinical Medicine and became an attending physician and Clinical Associate Professor, and then went on to obtain a PhD in otolaryngology and auditory physiology and pathology.

In the mid-1990s, cochlear implants had just been introduced to China. I was deeply attracted to this new technology. In 1997, through the Australia Hearing Service program, I systematically learned the techniques and clinic application of cochlear implantation. I discovered that the University of Iowa is a leading institute for cochlear implant research and I was honored to be offered the position of a Post Doc researcher in the auditory electrophysiology lab in 2000. Iowa City was my first home in the U.S., and the University of Iowa is the institute that helped me launch my research career here.

Outside of work, I like spending time with my family, gardening, hiking and reading historical books.
Phsyiologists Discover Molecular Mechanism

(Continued from p.1)

Stereocilia are nanoscale structures grouped together in staircase-like rows and interconnected by extracellular filaments. Hearing happens when sound vibrations deflect stereocilia, tension the extracellular filaments and open mechanotransduction ion channels allowing calcium and other ions to enter the hair cell.

The UK researchers discovered that blockage of these channels cause the stereocilia to retract and that this retraction depends on the calcium influx through these channels. The scientists uncovered the mechanism maintaining the remarkable staircase-like architecture of the stereocilia. This contribution provides new insight into the molecular mechanisms that facilitate the detection of sound within the inner ear, with opportunities to explore molecular therapies to maintain the stereocilia structure.

Vélez-Ortega notes, “I am extremely excited about this project. I work with a talented group of people in the laboratory of Dr. Gregory Frolenkov at the University of Kentucky. This study started as my postdoctoral work and has become the foundation for my independent line of research.”

Study results partially funded by Vélez-Ortega’s 2017 AHRF grant were published in the March 28, 2017 online journal, eLife digest. https://doi.org/10.7554/eLife.24661.002

Who Supports AHRF?

Nancy Masterson

“I donate because I’m so grateful to (AHRF founders) Drs. Derlacki and Shambaugh.

“I got scarlet fever when I was three, and I came to them when I was 14 [with an ear drainage problem]. I lived about 150 miles from Chicago, and my father drove me once every week or two for three years to see them. After three years, the problem was resolved – after a tonsillectomy and mastoidectomy and Dr. Derlacki straightened my ear canals.

“You wouldn’t think it, but I have very good memories from that time. Dr. Derlacki’s office was very calming. He got along so well with me and especially my father.

“One time I came in and my hearing had dropped. Even though he was an expert in the field, Dr. Derlacki said, “I don’t know why it worked, and I don’t know why it didn’t.” I just appreciated that honesty.”
Partner with Us!

We need your help in funding the vital work of hearing and balance scientists. Please consider donating today to the American Hearing Research Foundation to support their innovative efforts.

General Fund. To support upcoming projects, you can use the enclosed envelope, or donate online at www.American-Hearing.org. If you make an honorary or memorial donation, we’ll be sure to recognize your gift appropriately.

Planning for the Future. Would you like to help the Foundation support hearing research so future generations can have better options and treatments? A simple way to ensure hearing research continues is through a gift to AHRF in your will, known as a charitable bequest.

Charitable bequests can be:
- A specific amount,
- A percentage of your estate, or
- A conditional gift made if your beneficiaries predecease you.

Any amount helps the Foundation with its mission. You can feel confident and develop the best plan for you and your family while also supporting hearing-related research for years to come.

Katie’s run because Initiative

Katie Mertz continues her grassroots effort to raise awareness about Meniere’s disease, a debilitating hearing and balance disorder, as she fulfills her goal to run half marathons in every state plus the District of Columbia. To date, she’s raised awareness in 17 states, and raised $25,000 for Meniere’s disease research. Her 2017 schedule includes Idaho (completed), Washington, D.C., and Arkansas.

You can follow Katie’s efforts by visiting her Facebook page at www.facebook.com/runbecause.

Who Supports AHRF?

David Michael Keeter

“I have severe tinnitus and it’s a tough problem to deal with. So every now and then, I gather up the money in my collection jar and pass it along to AHRF. I want to do what I can. It would be nice to have a cure for this.”
Thank You to Our Donors
This list reflects donations received from December 8, 2016 through June 22, 2017.

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American Hearing Research Foundation
A nonprofit foundation dedicated to scientific research, and the preventive care and cure of deafness.

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General Funds sustain the core programs central to the Foundation’s mission. Donations to the General Fund also can be restricted to use for research into certain areas such as Meniere’s disease. There are several levels from which to choose:

- Chairman’s Circle $5,000 and above
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