

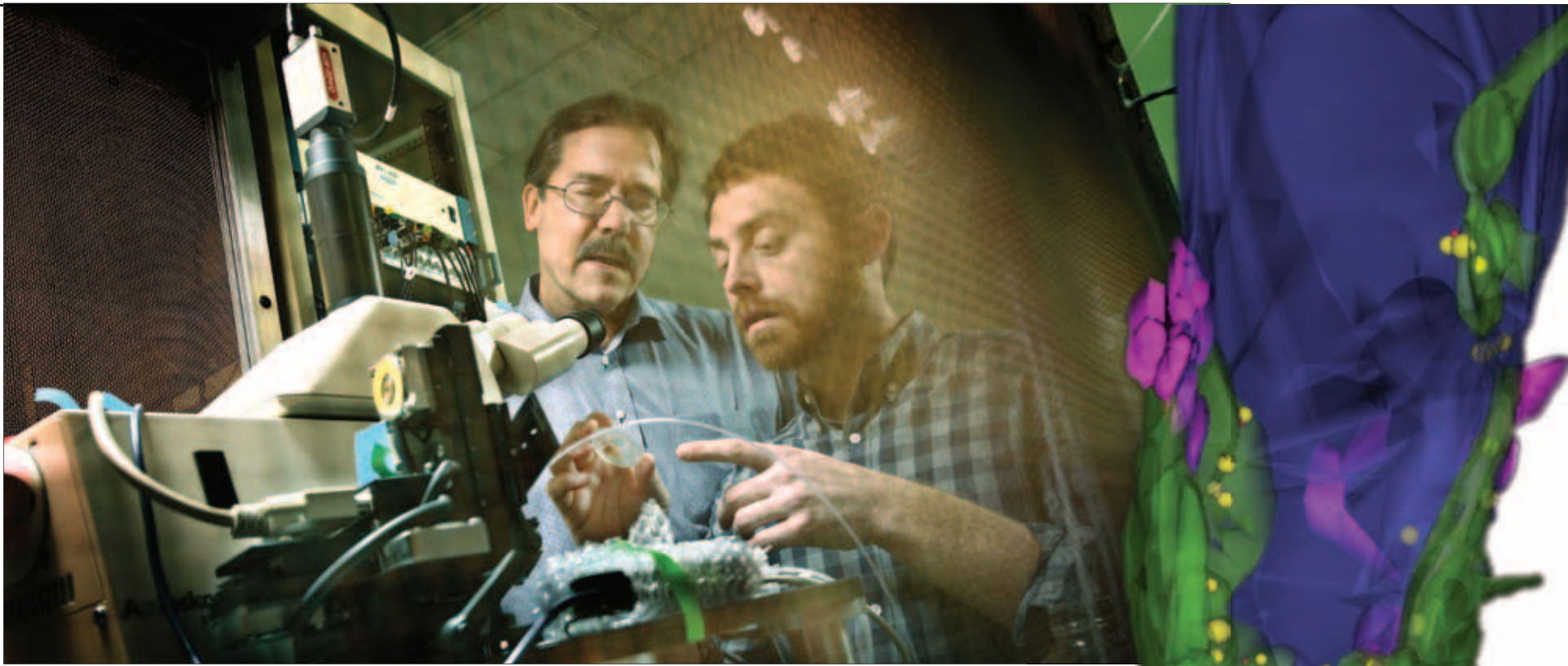
HeadLines



JOHNS HOPKINS
MEDICINE

NEWS FROM JOHNS HOPKINS OTOLARYNGOLOGY-HEAD AND NECK SURGERY

SUMMER 2016



Otolaryngology researcher Paul Fuchs, left, and graduate student Stephen Zachary. Right, inner hair cell (blue) surrounded by afferent contacts (green) and efferent contacts (magenta). Yellow spheres enclose the location of synaptic vesicles in the hair cell.

Better Connections, Worse Hearing

Conventional wisdom has long blamed age-related hearing loss almost entirely on the death of sensory hair cells in the inner ear. However, recent work by otolaryngology researcher **Paul Fuchs** and graduate student **Stephen Zachary** suggests another story. Their studies in mice have verified an increased number of connections between certain sensory cells and nerve cells in the inner ear of aging mice. Because these connections normally tamp down hearing when an animal is exposed to loud sound, the scientists think these new connections could also be contributing to age-related hearing loss in these animals, and possibly in humans.

Researchers have long known that the inner ear contains two sets of

hair cells—sensory cells responsible for converting sound waves into the electrical signals that the brain processes as sound: an inner tier closest to the brain and an outer tier. The outer ones have a secondary function: to amplify the sound waves within the inner ear. Not surprisingly, Fuchs notes, a loss of outer hair cells closely correlates with a loss of hearing. But studies over the last decade have suggested that changes over time also occur in the connections between hair cells and the nerve cells to which they are attached.

Each of those nerve cells is like a one-way street, Fuchs says, taking signals either from the ear to the brain or vice versa. The nerve cells that take signals to the ear are known to turn down the amplification provided by

outer hair cells when an animal is, for example, exposed to a noisy environment for an extended period of time.

Previous research has suggested that with age, inner hair cells in mice and humans experience a decrease in outgoing nerve cell connections, while incoming nerve cell connections increase. To find out if the new connections worked—or worked normally—Zachary painstakingly recorded electrical signals from within the inner hair cells of young and old mice. He found that the incoming nerve cells were indeed active and that their activity levels correlated with the animals' hearing abilities: The harder of hearing an animal was, the higher the activity of its incoming nerve cells.

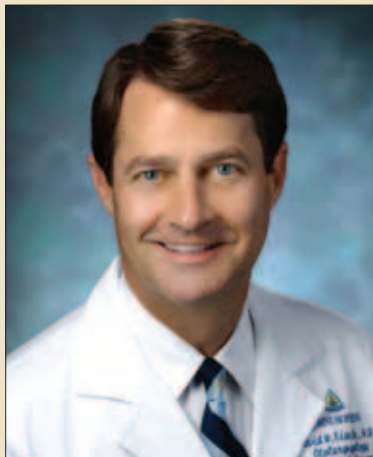
“THESE CELL CONNECTIONS SEEM TO REVERT TO THE WAY THEY WORKED IN EARLY DEVELOPMENT.”

—Paul Fuchs

“These nerve cell connections seem to be reverting back to the way they worked during early development before the animals' sense of hearing was operating,” says Fuchs. “We don't know why the new connections form,

(continued on page 8)

DIRECTOR'S COLUMN



For the Future

Johns Hopkins' reputation for outstanding medical care is well-known throughout the world, and research led by our faculty is regularly featured in the news. But what's not as readily apparent to the public eye is our exceptional programs to train both future and current surgeons—the education component of Johns Hopkins' tripartite mission of teaching, research and patient care. In the early months of this year, our department has already demonstrated our commitment to this mission by offering a variety of training opportunities. For example:

- In February, our team hosted the Inaugural North American Airway Collaborative Symposium on Advanced Surgical Techniques in Adult Airway Reconstruction, a one-day event featuring 10 lectures and panels by 23 faculty members from Johns Hopkins and other medical institutions.
- In April, our department hosted the third International Tracheostomy Symposium, a two-day event that not only offered training for physicians in tracheostomy care, quality and safety, but also featured a patient and family session. Twenty-five national and international speakers participated.
- Last year, Johns Hopkins head and neck surgeon Wayne Koch began leading a new fellowship at Mbingo Baptist Hospital in Cameroon. The fellowship is training doctors in the advanced surgical techniques that are an essential part of our specialty. With only one other similar program in all of Africa, this fellowship is filling a vital need, training surgeons who can effectively provide these services across the continent.

By helping to educate current and upcoming surgeons, we're improving medical care now and in the future.

David W. Eisele, M.D., F.A.C.S.

RESIDENT SPOTLIGHT



Jason Nellis and colleagues are studying the effect of facial paralysis on quality of life and psychosocial health.

Focus on the Face

WHEN JOHNS HOPKINS Department of Otolaryngology–Head and Neck Surgery third-year resident **Jason Nellis** was in the fifth grade, his class took a field trip to a cadaver lab at the University of Utah, sparking an interest in the human body. Later, he became involved in community service, where he discovered a love of forging relationships with new people and helping others through service. By the time he was wrapping up high school, his future career was becoming clear.

“I knew that I wanted to do something that involved biology and working with people. Everyone would always say, ‘You should be a doctor,’” he remembers. “It made sense to me.”

After attending the University of Maryland for his undergraduate degree and the University of Virginia for his medical degree, he landed at Johns Hopkins for his residency. Seeking a lab to join for the research portion of his training, he soon sought out **Lisa Ishii**, a physician-researcher in the department whose work had captivated him as a medical student.

Much of Ishii's work centers on faces, a sight that most people view dozens of times every day without much thought. But minute details of faces, particularly facial defects—such as nasal deformities, an uneven smile or lesions after skin cancer resection—can influence perceptions and emotions, both for the observer in society and the individual whose face is on display.

Nellis currently has several research projects in progress in Ishii's lab centered on this topic. One focuses on patients with acoustic neuromas, benign tumors that affect thousands of patients in the United States each year. Depending on tumor size and other risk factors, surgery to remove it sometimes leads to facial paralysis. To see how the aftermath of surgery in particular affects acoustic neuroma patients, Nellis and his colleagues are following

patients for a year after this treatment to better understand the effect on quality of life and other psychosocial dimensions, such as depression and self-esteem.

Similarly, Nellis and his colleagues are also tracking patients who come to Johns Hopkins with facial paralysis that results from a variety of causes, such as other types of tumors and Bell's palsy. Their early data suggest that this group has a significantly higher likelihood of depression and lower quality of life compared to those who don't have facial paralysis. The researchers plan to follow this group long term to see if these risks change over time or after facial reanimation surgery to treat paralysis.

Both of these studies, he explains, will lend some insight on what life might be like living with various conditions and how this changes after treatment, information that could help doctors and their patients make care decisions.

“A lot of this work is just about understanding what day-to-day life is like for patients when they leave our office, giving us information to provide better patient-centered care and hopefully improve patient outcomes,” Nellis says. “That boils down to why I got into medicine in the first place. Working with patients is the greatest reward.” ■

“I KNEW THAT I WANTED TO DO SOMETHING THAT INVOLVED BIOLOGY AND WORKING WITH PEOPLE. EVERYONE WOULD ALWAYS SAY, ‘YOU SHOULD BE A DOCTOR.’ IT MADE SENSE TO ME.”

—Jason Nellis

Making a ‘Big Bang’ with Medialization Laryngoplasty

COMPLICATION FROM TREATMENTS for Joe Steiger’s muscular dystrophy had left him with bilateral vocal cord paresis since the age of 10. In an effort to return his voice, Steiger, now 28, had multiple surgeries over seven years—the last, a medialization laryngoplasty at age 17. However, when the procedure didn’t have the desired effect due to underlying respiratory muscle weakness, surgeons removed the implants. Steiger soon noticed an unexpected and extremely undesirable consequence: without the implants, he readily aspirated while eating.

He became steadily dependent on a bilevel positive airway pressure (BIPAP) machine and suction to clear his lungs during every meal, leaving him increasingly homebound. After nine years of suffering, a frightening choking incident spurred him to seek help. In 2013, he found himself in laryngeal surgeon **Simon Best**’s office, discussing options to solve this problem.

After a full workup, including videostroboscopy to test Steiger’s vocal cord function, Best suggested possible remedies. Injections of a filling agent might thicken the vocal cords enough to help prevent aspiration, but it wouldn’t be a permanent fix. A total laryngectomy would permanently prevent aspiration from eating, but it would also be a radical and life-changing surgery, altering Steiger’s physical appearance, manner of breathing, and even his sense of smell. A third option was to perform another medialization laryngoplasty, returning the implants that had previously pushed Steiger’s vocal folds together.

“I was confident that the implants would return eating to normal,” remembers Steiger. But it wasn’t an easy decision. After difficult recoveries from several previous surgeries to treat various conditions associated with his muscular dystrophy, he was hesitant to move ahead.

Best was also concerned that another medialization laryngoplasty might not have the results that Steiger was looking for—after all, the procedure is usually used for voice problems, not swallowing issues. “I wasn’t sure of his chances of success,” Best says. “But with few good options, and based on his previous history of better swallowing when these implants were in place, I trusted that this procedure would help him.”

In November 2013, Steiger underwent the two-hour surgery, which is typically performed awake to help surgeons fine-tune the implants’ placement

based on the patient’s use of their voice. Although Steiger can’t phonate very well because of his muscular dystrophy, the movement of his vocal cords was still helpful for positioning the implants.

Within days after surgery, it was clear that the procedure was a success. Steiger was able to eat without aspiration for the first time in nearly a decade, which opened up a myriad of possibilities that wouldn’t have existed otherwise. Now able to confidently eat outside his home, he was able to travel. In January 2015, he achieved his longtime dream of traveling from his home in Sykesville, Maryland to Burbank, California, to attend a filming of his favorite show, *The Big Bang Theory*, and meet cast members.

“That trip was and will be the most memorable thing I ever do,” says Steiger. “It was all possible from successful surgery and never giving up on your dreams.” ■

For information, call 443-287-2124.
Visit hopkinsmedicine.org/otolaryngology.



Following medialization laryngoplasty, patient Joe Steiger was able to eat without aspiration for the first time in nearly 10 years. After the surgery, he traveled to meet the cast of his favorite show, *The Big Bang Theory*.

Honor Roll of Gifts

The entire Department of Otolaryngology–Head and Neck Surgery is deeply grateful to the many donors who have chosen to support our research, patient care and teaching. This generous support allows us to continue bringing the very best care to our patients and their families and inspires us to persist in finding answers to tomorrow's questions. As we move further into the 21st century, your support is a testament to our efforts and provides added motivation to set the standard of excellence in our field.

Thank you for your generosity.

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Survivor Support

Head and Neck Cancer Survivorship and Education Day informs patients about their disease, helping them to restore a measure of control.

PETER RIEKER WAS always thinking of others. Even as his last days as a head and neck cancer patient drew near, one of his final acts was to make life easier for other patients with his diagnosis. Before his death in 2013, he and his wife, along with the couple's two sons and countless family members and friends, started the Johns Hopkins Head and Neck Cancer Survivorship Program with personal gifts and donations requested in Rieker's obituary.

Since then, the thousands of dollars collected for this program have paid for a variety of services for people living with head and neck cancer, a group of diseases that receives little attention compared to other cancers, such as breast, lung and prostate.

"These are still uncommon tumors to the population at large," says head and neck surgeon **Christine Gourin**. "Head and neck cancers only make up 4 percent of all cancers. These can be isolating diseases to get."

That's why a major focus of this program is Head and Neck Cancer Survivorship and Education Day, an event that **Amy Brady** has organized for seven years. As manager of the Johns Hopkins Head and Neck Cancer Multidisciplinary Program, as well as the Survivorship Program, she's seen firsthand the despair patients can experience when they're first diagnosed and the empowerment they can feel once they learn more about their disease.

"It is the fear of the unknown that causes much of the anxiety for our patients," she says. "Educating patients about the disease and its causes as well as the types of treatment is the key to giving the patients power and control over their disease."

Head and Neck Cancer Survivorship and Education Day fills that need by giving patients, caregivers and the general public insight on head and neck cancers in a variety of ways: through talks by physicians, nutritionists, dentists and speech/swallow specialists on current head and neck cancer topics; by providing free educational materials on everything from transportation and



housing during treatment to support groups and what to expect during your treatment; having a panel of head and neck cancer specialists available to answer questions; and many opportunities to socialize with other attendees, where lasting relationships are often formed, Brady says.

Steven Clem, a Johns Hopkins patient who lost his tongue and voicebox to cancer in 2008, has gone to many of these events and has served as a guest speaker. He also regularly meets with patients who need similar surgeries to survive their cancers.

"Over the last four to five years, I have met many people there to talk to," he says. "I tell them that there is a good, if not great, quality of life after surgery. It's my way of paying back and paying it forward."

The Survivorship Program also funds a monthly support group for head and neck cancer patients and their spouses or caregivers, run by Johns Hopkins speech-language pathologist **Kim Webster**. Johns Hopkins patient Sam Digiacommo has become a regular attendee after his laryngectomy for vocal cord cancer in 2011. Meetings are like miniature Education Days, he says, with speakers and a chance for patients to meet and talk.

"It might be months after treatment before you can get out of the house, but I think if you're able, you should come," he says of the support group. "It lets people know that they're not alone. There are other people out there you can relate to."

In the future, Brady says, the Survivorship Program will fund more efforts to educate patients, such as extensive information about these diseases on Johns Hopkins' website, as well as ways to make their lives easier during treatment, such as helping to defray the cost of parking during radiation treatments, which necessitate daily trips to the hospital over the course of weeks.

"After a diagnosis of head and neck cancer, our patients' lives are forever changed," says Gourin. "The Survivorship Program is helping patients to navigate their new normal life on every step of this journey." ■

For information, call 443-287-2124.
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"THE SURVIVORSHIP PROGRAM IS HELPING PATIENTS TO NAVIGATE THEIR NEW NORMAL LIFE ON EVERY STEP OF THIS JOURNEY."

—Christine Gourin

Saving a Career with Cochlear Implantation

ROBERT LONG, 57, of Alexandria, Virginia, had always had excellent hearing—an attribute critical for his job as a litigation attorney who has argued 18 cases before the U.S. Supreme Court. But all that changed in his late 40s, when a condition his doctors struggled to diagnose left him with progressive hearing loss. He wore one hearing aid, then two. Eventually, even with the aids, his hearing declined so much that it threatened his career.



“I love my job, and I really wanted to continue doing it,” Long says. His hearing loss was also beginning to impinge upon family and social contacts. “I told my wife once that it felt like I was on a ship headed out to sea, but everyone else was still on shore. I was losing contact.”

After about five years of steadily declining hearing, he sought help from head and neck surgeon **John Carey**, who told him that he was the perfect candidate for a cochlear implant (CI).

Although news stories often focus on children or young adults as the prototypical CI patients, middle-aged and older adults are two of the fastest growing demographics for receiving these devices, explains **Howard Francis**, who directs the Johns Hopkins Listening Center. CIs are critical for very young patients,

who are in the midst of developing language. But these devices have also proven pivotal in helping adults such as Long maintain their involvement in professional and social pursuits.

Last year, Long underwent surgery to have a CI implanted in his right ear.

About a month after surgery, Long’s audiologist, **Andrea Marlowe**, activated his CI. “I could immediately hear my wife sitting across the room, asking if I could understand her,” he remembers.

Not all patients have the same kind of immediate success that Long experienced, Francis explains. Although at least 95 percent of patients have improvement in their level of hearing and understanding speech, age, length of hearing loss and the amount of hearing deficit can predict how well a patient’s CI will work for them.



Robert Long

A key to achieving the best outcomes is auditory rehabilitation, which helps patients learn to use their CI over time. Although many centers offer CI surgery and auditory rehabilitation is widely offered for children, Johns Hopkins is unusual in making this service available for adults as well, Francis says.

“We find that adults, and particularly older adults, have their own needs that can be addressed with this useful service,” he adds.

Long’s CI has allowed him to continue practicing law, including arguing cases in court. If hearing in his left ear continues to decline, he plans on getting a CI in that ear too.

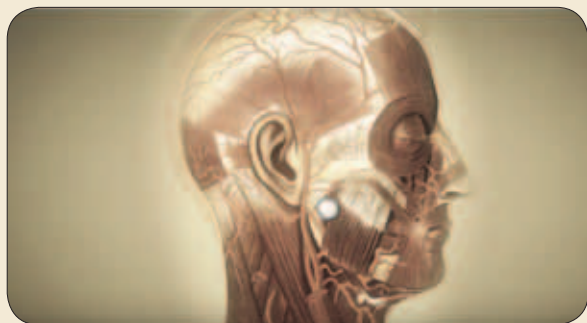
“Hearing loss is a serious loss,” Long says. “After my experience, I’d do a second CI without hesitation.” ■

THESE DEVICES HAVE ALSO PROVEN PIVOTAL IN HELPING ADULTS SUCH AS MR. LONG MAINTAIN THEIR INVOLVEMENT IN PROFESSIONAL AND SOCIAL PURSUITS.

—Howard Francis

For information, call 443-287-2124. Visit hopkinsmedicine.org/otolaryngology.

MOVING PICTURES



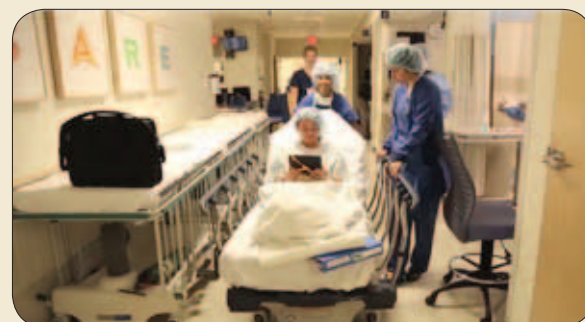
Jubenal’s Story: Parotid Salivary Gland Tumor:

See a video about the powerful journey of Jubenal Gonzales, a Connecticut-based firefighter and paramedic diagnosed with a malignant parotid gland tumor. Gonzales traveled from his home state to Baltimore to seek treatment from head and neck surgeon David Eisele. bit.ly/Jubenals_Story



Sarah’s Story: Pediatric Cochlear Implant

Watch a video about Sarah Aquino, whose hearing loss by the age of 12 was so significant that hearing aids were no longer helpful. Initially scared at the thought of cochlear implant surgery, Sarah and her mother were quickly reassured after meeting with the comprehensive team of experts at the Johns Hopkins Listening Center. bit.ly/Sarah_Aquino

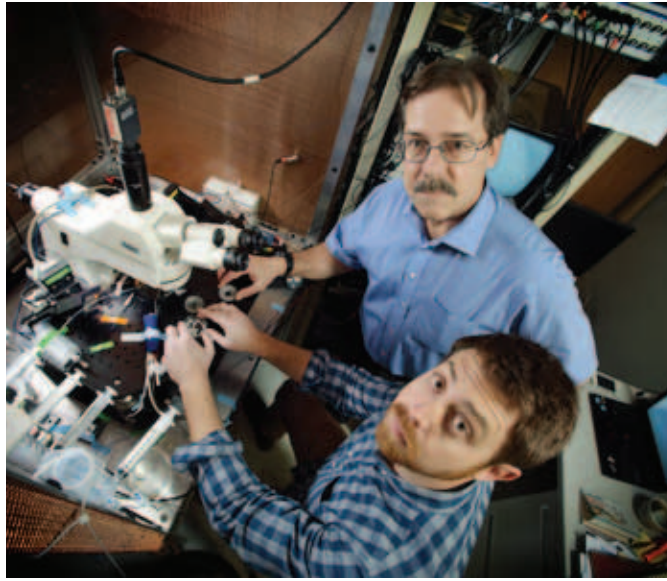


Better Connections, Worse Hearing
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but it might be as simple as a lack of competition for space once the outgoing nerve cells have retracted.”

This basic research could eventually lead to new treatments for age-related hearing loss. If the same phenomenon is occurring in human ears, Fuchs and his team say there may be ways of preventing the incoming nerve cells from forming new connections with inner hair cells, a technique that could help maintain normal hearing through old age. ■

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Otolaryngology researcher Paul Fuchs, above right, and graduate student Stephen Zachary.

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This newsletter is published for the Department of Otolaryngology-Head and Neck Surgery by Johns Hopkins Medicine Marketing and Communications.

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HeadLines

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