

HeadLines



JOHNS HOPKINS
MEDICINE

NEWS FROM JOHNS HOPKINS OTOLARYNGOLOGY-HEAD AND NECK SURGERY

SUMMER 2020

Giving to Stem Hearing Loss

\$15 million gift spread over five years funded professorship and research projects

When businessman, investor and philanthropist David Rubenstein's two daughters were in pre-school decades ago, their teachers told him that his daughters weren't paying attention as well as other children in class. After months of doctors' appointments, tests eventually showed that both girls had congenital hearing loss of about 30%. Although each were fitted for hearing aids, they chose not to use them, Rubenstein says, relying instead on strategies such as reading lips and sitting close.

Now in his 70s, Rubenstein's own hearing has declined due to his age, he says, strengthening his enduring interest in hearing loss. A longtime member of the Johns Hopkins Medicine board of trustees, Rubenstein says he knew exactly where to direct his gift when he donated \$15 million to Johns Hopkins Medicine over five years — he earmarked the funds for hearing loss research and establishment of a professorship bearing his name in the Department of Otolaryngology-Head and Neck Surgery. **Paul Fuchs**, Ph.D., who has spent the last 25 years of his career at Johns Hopkins, is the inaugural David M. Rubenstein Research Professor of Otolaryngology-Head and Neck Surgery.

"My hope is that this gift will eventually make great strides for people with hearing loss, whether it's born or acquired," Rubenstein says.

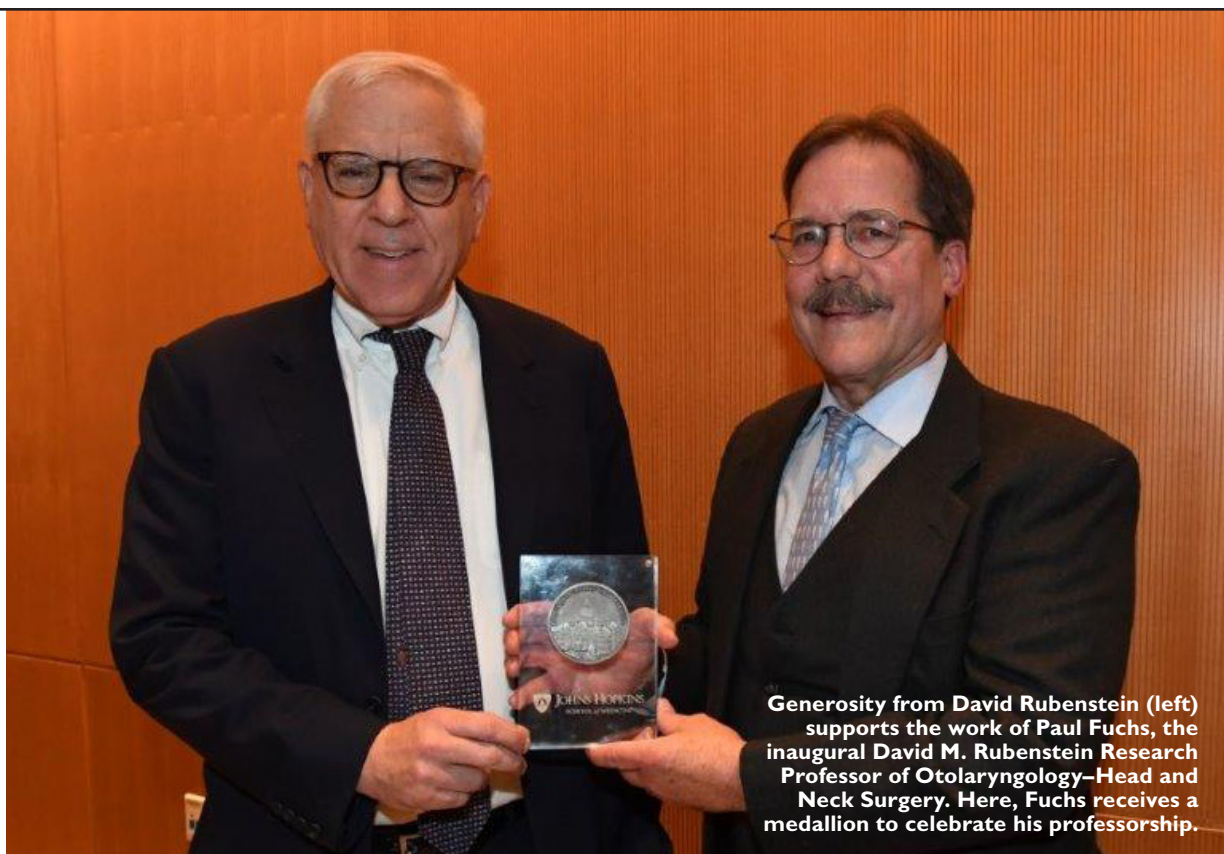
Fuchs explains that his new professorship, along with grants from the National Institutes of Health and other sources, will help fund his salary, allowing him to devote more money to his research projects. "It allows us more flexibility for which

research projects we pursue," he says.

Fuchs has devoted some of this gift to investigations of afferent neurons that send sound information to the brain and efferent neurons that project from the brain back to the ear. His work has shown the existence of a feedback loop that appears to protect the ear from acoustic trauma by inhibiting the effects of loud sounds. He and his team are currently testing the possibility of an applied gene therapy that might protect hearing in noisy environments.

The majority of Rubenstein's gift is in an endowment that funds "accelerator grants" for other investigators at Johns Hopkins — \$100,000 grants that have the potential to speed up ongoing research projects. For example, Fuchs says, some of these grants have funded the work of **Elisabeth Glowatzki**, Ph.D., on ear reinnervation, reconnecting the sensory cells in the ear with the brain when those connections have declined or been lost. Other grants have funded research by **Angelika Doetzlhofer**, Ph.D., on encouraging support cells in the ear to morph into sensory cells — the death of these sensory cells is a major cause of hearing loss.

"We are all using these funds to help push our work forward, collaborate and grow expertise," Fuchs says. "It's a revolutionary gift that will have a big impact for years to come." ■



Generosity from David Rubenstein (left) supports the work of Paul Fuchs, the inaugural David M. Rubenstein Research Professor of Otolaryngology-Head and Neck Surgery. Here, Fuchs receives a medallion to celebrate his professorship.

A New Option for Single-Sided Deafness

Cochlear implants have been approved for decades in the U.S. for bilateral sensorineural hearing loss. However, in July 2019, the Food and Drug Administration green lighted use of these devices for single-sided deafness and asymmetric hearing loss, a significant change from the devices' previous indication.

Single-sided deafness may result from a multitude of causes, says Paul Fuchs, Ph.D., the David M. Rubenstein Research Professor of Otolaryngology-Head and Neck Surgery. These include viral and bacterial infections, trauma to the ear, circulatory disorders, acoustic neuromas and genetic causes. Even when in a single ear, hearing loss can lead to communication, educational, social, behavioral and cognitive problems.

By restoring sound input to the affected ear, children and adults with hearing loss may be able to avoid these issues, Fuchs says, improving their quality of life. Johns Hopkins has long been a pioneer in cochlear implantation, since the formation of its Listening Center nearly three decades ago. ■



To support the department's work, call 410-955-0173

Learning About the Vestibular System with MRI

In 2009, researchers in Italy made an unusual discovery: Under the strong magnetic fields of an MRI machine, healthy human volunteers experienced momentary dizziness and longer-lasting nystagmus. Johns Hopkins vestibular neurologist **David Zee** was a visiting researcher in Italy at the time and witnessed this himself, changing his own research to better understand why this happens and what it could teach us about the vestibular system.

Two years later, when a resident in the Johns Hopkins Department of Otolaryngology–Head and Neck Surgery — **Bryan Ward** — considered his own research project, Zee suggested he take the baton.

“It had been about 80 years or so since any new way of stimulating the vestibular system had been discovered,” says Ward, who joined the faculty in 2018. “This was a fundamentally new interaction with the vestibular system, and it was exciting to be somebody who could help drive this research forward.”

Over the last nine years, Ward has helped lead efforts to define how magnetic fields affect the vestibular system and how researchers can use these findings in broader efforts to determine how the brain adapts to sensory input over time. He and his colleagues published a review of their work in the June 2019 *Journal of Neurophysiology*.

Soon after Ward began his research in this area, he and his colleagues recruited 10 patients with a variety of inner ear disorders to see whether they also exhibited these same effects under magnetic

Johns Hopkins researchers narrowed the effects of magnetic stimulation down to the utricle and superior and horizontal semicircular canals.

“I remember the first time I saw nystagmus in a MRI, I had this realization that it was probably the first time humans had ever seen this before.” —BRYAN WARD

stimulation. Within a strong 7 Tesla MRI, the effect was variable. The dizziness and nystagmus depended on which ear was injured. By ruling out which parts of the inner ear were affected by these volunteers’ disorders, the researchers narrowed the effects of magnetic stimulation down to the utricle and superior and horizontal semicircular canals.

Using mouse models, Ward and his colleagues confirmed that this effect occurs outside of humans. When the team put zebrafish in the MRI machine,

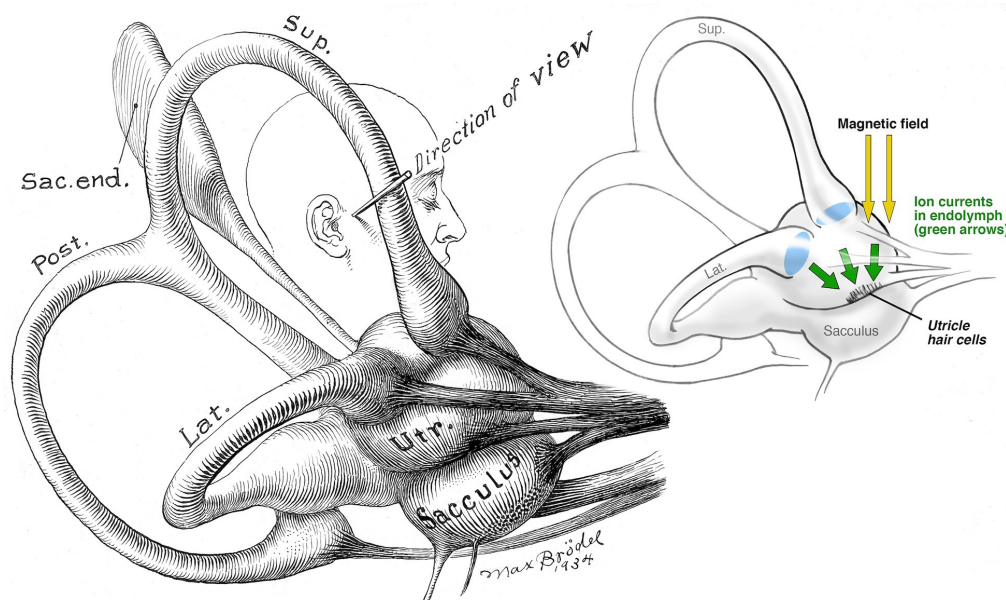
the animals flipped and twisted, suggesting that this phenomenon is highly conserved among animal species.

More recently, Ward and his colleagues have used magnetic stimulation as a model for vestibular system learning. Their research shows that while dizziness from the MRI subsides after a few minutes, abnormal eye movements persist over hours, mimicking what happens when patients lose vestibular function on one side due to trauma or surgery — offering a window into how vestibular sensation adapts to changes over time.

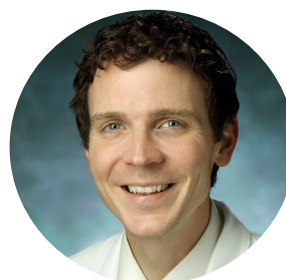
The team’s most recent project is to try to image the movement of fluid in the inner ear, offering the opportunity to identify the first direct evidence that magnetic stimulation is acting on this structure.

“I remember the first time I saw nystagmus in a MRI, I had this realization that it was probably the first time humans had ever seen this before,” Ward says. “I think the work we’re doing will be used by researchers to study the vestibular system for many years to come.” ■

☎ To support this research call 410-955-0173.

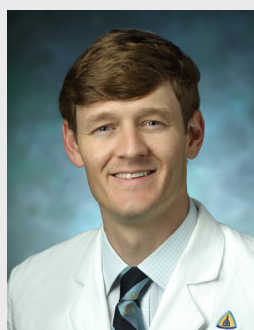


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FACULTY NEWS

3 New Faculty Members Join Department of Otolaryngology–Head and Neck Surgery



Francis “Pete” Creighton, M.D.

Pete Creighton is a fellowship trained neurotologist and lateral skull base surgeon. His clinical practice specializes in surgical and medical treatment of middle ear, inner ear, skull base and facial nerve disorders. These include skull base tumors, vestibular schwannomas (acoustic neuromas), hearing loss, cholesteatoma, cerebrospinal fluid leaks and eardrum perforations. Treatment specializations include cochlear implantation, stapedectomy, mastoidectomy, and middle fossa and translabyrinthine craniotomies.

His research focuses on the integration of robotic and augmented reality platforms to improve surgical safety and efficiency.

Johns Hopkins Among Few in U.S. to Treat Obstructive Sleep Apnea with Hypoglossal Nerve Stimulation

A new device stimulates the hypoglossal nerve in time with each breath, pushing the tongue out to open the airway.

THE GOLD STANDARD for treating the millions of people worldwide who suffer from obstructive sleep apnea is continuous positive airway pressure, or CPAP, a therapy that keeps the airway open by pushing air inside. While CPAP is an effective noninvasive therapy, it is not well tolerated by a significant number of patients, says Johns Hopkins otolaryngologist-head and neck surgeon **Kevin Motz**. Patients must wear a mask that fits over the nose, mouth or both, and this can cause discomfort and disturbed sleep. Until recently, their only alternatives were invasive and often painful surgeries to reconstruct or reorient the pharynx, remove part of the tongue, or advance the jaw.

“It has been difficult to predict who will have a good response from these surgeries and who will go through them and get little benefit,” says Motz.

For patients who qualify, a new device could offer a better option. Placed by surgeons via a minimally invasive procedure, the device stimulates the hypoglossal nerve in time with each breath, pushing the tongue out to open the airway. The treatment is now offered at Johns Hopkins as part of its growing sleep surgery services and is available at only a few institutions across the U.S.

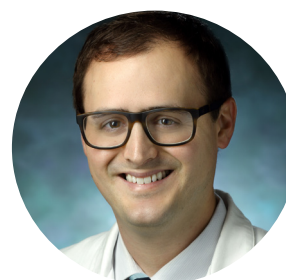
Motz says that patients must have a body mass

“This approach liberates them from being attached to a CPAP machine at night, and it gives them peace of mind that their disease is being adequately treated.”

—KEVIN MOTZ

index of less than 35 before surgery and an apnea-hypopnea index, or AHI, between 15 and 65. Patients who fit these criteria and have failed CPAP therapy must undergo sleep endoscopy — a test to assess their pattern of airway collapse during sleep — to determine whether the approach could provide benefit.

Candidates who progress to surgery undergo a two-hour outpatient procedure to implant the device. Three separate incisions — on the neck below the jaw, just below the collarbone and one on the ribcage — are used to place the pulse-generating electrode, a respiratory sensor, and a processor and power unit. A month after the operation, the device is turned on. Patients turn on the device each night, setting a delay



so that it's only on after they've fallen asleep. Several months later, they participate in another sleep study to objectively test its benefit and fine tune its settings for maximal benefit.

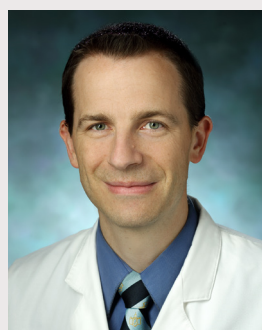
Data from the device maker shows that this hypoglossal nerve stimulator significantly reduces patients' AHIs, with those with the

highest AHIs receiving the most benefit. Daytime sleepiness scores also steadily decreased over time. Together, these numbers suggest that the patients' risk for the most serious consequences of sleep apnea, such as cardiovascular disease and auto accidents, can be significantly lowered with the use of this device, says Motz.

“This approach liberates them from being attached to a CPAP machine at night, and it gives them peace of mind that their disease is being adequately treated,” says Motz. “We're pleased to add this to our toolbox for treating sleep apnea.” ■



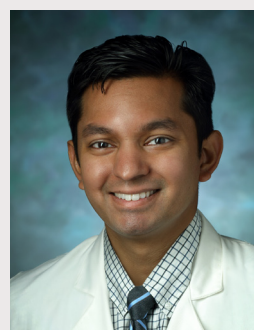
To support the department's work, call 410-955-0173.



Nyall London, M.D., Ph.D.

Nyall London's clinical practice focuses on inflammatory sinonasal disease, skull base disorders, and malignancies of the skull base and sinonasal cavity. This includes chronic rhinosinusitis, skull base defects such as encephalocele or cerebrospinal fluid leak, as well as malignancies such as squamous cell carcinoma, olfactory neuroblastoma (esthesioneuroblastoma) and nasopharyngeal carcinoma. He recently completed fellowship training focused on sinonasal tumors and endoscopic and open approaches to the skull base.

London has published more than 40 peer-reviewed research articles, reviews and book chapters in publications such as *Nature*, *Nature Medicine*, *Science Translational Medicine* and *Nature Cell Biology*. He also directs a laboratory at the National Institutes of Health/National Institute on Deafness and Other Communication Disorders focused on clinical trials and mechanisms of olfactory neuroblastoma (esthesioneuroblastoma) tumorigenesis.



Shumon Dhar, M.D.

Shumon Dhar has expertise in the comprehensive management of voice, upper airway and swallowing disorders. Dhar has unique training in bronchoesophagology, which positions him to treat patients with profound swallowing, reflux and motility problems from a holistic perspective. He uses the latest in diagnostic modalities within a multidisciplinary care model. He also offers minimally invasive endoscopic options for the treatment of GERD and Barrett's esophagus. Advanced interventions performed by Dhar include endoscopic and open treatment of cricopharyngeus muscle dysfunction and

Zenker's diverticulum, complete pharyngoesophageal stenosis, vocal fold paralysis, and severe dysphagia in head and neck cancer survivors and patients with neuromuscular disease-related swallowing dysfunction.

Dhar's research focuses on risk factors, prevention and management of patients with profound swallowing disorders. In addition to dysphagia, he is interested in neurolaryngology, specifically laryngeal dystonia and using implantable medical devices to improve patient outcomes and quality of life.

VIDEO

COVID-19 Conversations: Otolaryngology Alumnus Shares Insights from Hong Kong

Visit bit.ly/hopkinscc to watch the first installment of “COVID-19 Conversations,” a series in which Johns Hopkins experts connect with colleagues around the world to discuss their experiences with COVID-19. In this episode, David Eisele, director of otolaryngology (on left below), and Carole Fakhry, chief of head and neck surgery (on right below), speak with Johns Hopkins alumnus Jason Chan, who is now a faculty member at the Chinese University of Hong Kong.

HeadLines

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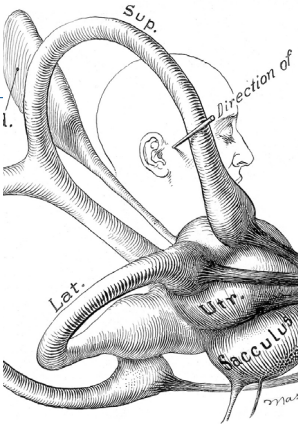


Inside



1

Giving to Stem Hearing Loss



2

Learning About the Vestibular System With MRI



3

Johns Hopkins Among Few in U.S. to Treat Obstructive Sleep Apnea with Hypoglossal Nerve Stimulation