THE Eye & Ear Foundation OF PITTSBURGH

News for Supporters and Friends

2021 SIGHT+SOUND

FALL EDITION

Jonas T. Johnson, MD, FACS: Surgeon, Clinician, Teacher, and Leader

by Lisa A. Goldstein

After 42 years at the University of Pittsburgh, 16 of which were as Chairman of the Department of Otolaryngology, Dr. Jonas T. Johnson is stepping down. But make no mistake, he's not retiring. If anything, he is reinventing himself.

As much as he has loved being in the operating room, where he has helped patients and trained the next generation of physicians, Dr. Johnson felt an obligation to step down.

Once his successor is named, Dr. Johnson plans to focus on helping run the UPMC Head & Neck Cancer Survivorship Clinic, where he hopes to raise awareness and understanding about survivorship among his colleagues and treat patient side effects more effectively.

"Dr. Johnson would not necessarily say this, but his legacy will certainly be his patients, who have expressed directly to us at the Foundation their admiration and appreciation for him," says Eye & Ear Foundation CEO Lawton Snyder. "He connects with them directly, and they appreciate him for those human qualities."

Snyder also cites the many physicians Dr. Johnson has trained over the years, as well as his and the Department's numerous accomplishments as part of his "incredible legacy." He truly exemplifies what a chairman can and needs to do, he adds.

That legacy includes a career spent on the care of people with head and neck tumors, a rank of Distinguished Service Professor at the University of Pittsburgh School of Medicine, and joint appointments in the Department of Radiation Oncology, Department of Oral and Maxillofacial Surgery at the University of Pittsburgh School of Dental Medicine and Department of Communication Science and Disorders in the School of Health and Rehabilitative Sciences. Dr. Johnson is also a Past President of the American Academy of Otolaryngology-Head and Neck Surgery (2003), the American Head and Neck Society (2004), and the Triologic Society (2014).

"What I truly enjoyed was being able to practice medicine in this environment of research and education," says Dr. Johnson.

Near and dear to his heart is the Survivorship Clinic, initiated when he was approached in 2015 by Dr. Marci Nielsen, a postdoctoral candidate at the time. She asked if she could join him in the Clinic to recruit patients for a research project. Through this, Dr. Johnson learned that patient care was inadequate, and a more comprehensive standard of care was needed with long-term follow-up care.

In the years since the Clinic has grown, it now includes surveillance (checking people for recurrence), prevention, assessment, and help in navigating the health system.

"The first thing I've done is try to help people understand what was missed and how there is this huge opportunity to do better in the future," says Dr. Johnson.

Lawton Snyder, CEO, Eye & Ear Foundation and Jonas T. Johnson, MD, FACS, Distinguished Service Professor and Chairman of Otolaryngology, Dr. Eugene N. Myers Endowed Chair

Dr. Johnson is also passionate about health literacy or helping patients understand what is happening to them. He is also concerned about the burden of the caregiver and financial toxicity. With three million data points, Dr. Johnson has the perfect ammunition to educate other doctors, who he says, love data.

And now, after participating in several chairman searches in other departments, Dr. Johnson is helping with his own.

"It is my enduring belief that the key aspect of outstanding organizations is successful succession," Dr. Johnson says. "I believe that my successor will be better than me, and I fully want that to happen. They'll have all these new things to work on that will make the future brighter than the past."

To donate to the Head and Neck Cancer Survivorship program, visit eyeandear.org or return the attached envelope.

An Amazing Breakthrough for Retinitis Pigmentosa and Big First Step

by Lisa A. Goldstein

After 40 years of total blindness, a 58-year-old French man saw stripes in a crosswalk. He was also able to reach out and touch a large notebook, a staple box, and hydroalcoholic gel on a table and discern whether two or three glass tumblers were before him.

This newfound ability didn't just happen overnight. A new kind of gene therapy called optogenetics finally paid off after 13 years of research.

The breakthrough study published in the May 2021 issue of Nature Medicine detailed how a team of researchers led by Dr. José-Alain Sahel, Chair and Distinguished Professor of the Department of Ophthalmology at the University of Pittsburgh School of Medicine, delivered a protein called ChrimsonR directly to the retina to make it more receptive to light.

This was accomplished by genetically manipulating a common non-pathogenic virus so that it carries the protein. After about four months, the ganglion cells in the retina could transform and absorb light. Shapes and shadows on the amber color spectrum became visible.

In other words, optogenetics was used to make the top layer of ganglion cells at the front of the retina photosensitive, bypassing the non-functioning degenerated layer of photoreceptors.

There are caveats, of course. So far, the treatment had only proven successful in one patient (the only one trained before the project was interrupted due to COVID), and in only one eye, with the help of special goggles to deliver sufficient and safe levels of light. The goggles also scan the field of view, register light changes in each pixel, and send pulses of amber light to the eye.

The 58-year-old man's retina was given five months to accept the treatment, followed by seven months of training with the goggles. Prior to the injection, he could not visually detect any objects with or without the goggles, nor could he after injection without the goggles. According to the study abstract, "This is the first reported case of partial functional recovery in a neurodegenerative disease after optogenetic therapy."

The significance of this work will be recognized at the Falling Walls Conference in November, an event that commemorates the fall of the Berlin wall that the BBC calls a convening of "the brightest minds on the planet." Dr. Sahel is one of seven winners from 1,000 nominations in 115 countries for Science Breakthroughs of the Year. His award in the Life Sciences category is "Breaking the Wall to Restoring Vision for Retinal Degeneration."

Meanwhile, more research is needed to determine dosing levels, improve the goggles and training to use them, and consider the best treatment timing. The trial is now expanding, with new patients being treated and trained in Paris and Pittsburgh. Right now, the trial only patients with very advanced disease. Additionally, researchers cautioned, the patient's vision is not expected to get to the point of reading or recognizing faces. Yet this is undeniably a significant achievement that is expected to have long-lasting, maybe even permanent, results for patients with retinitis pigmentosa. S+S

José-Alain Sahel, MD in his Insitut de la vision Lab, Paris, France. (CNRS Photothéque/Céiline Anaya Gautier)

Sad News to Share

by Lisa A. Goldstein

Peggy Smyrnes-Williams

The Eye & Ear Foundation (EEF) lost a beloved friend and supporter in Peggy Smyrnes-Williams who passed away on October 9, 2021. A vice president at the Ladies Hospital Aid Society (LHAS) and an EEF Ambassador, Smyrnes-Williams, leaves a legacy of stalwart support.

"Peggy loved both Otolaryngology and Ophthalmology fiercely," said Heather Chronis Danek, EEF Director of Development – Individual Giving and Corporate Relationships.

Smyrnes-Williams co-chaired the LHAS Gala in 2017 that honored ophthalmology, helped spotlight Morgan Fedorchak as one of the young researchers at the 2018 Gala, and championed the idea of Survivorship for the next LHAS Gala. "Up until her final days, she was thinking of creative ways to highlight the innovative work of the Survivorship team," said Chronis Danek.

Smyrnes-Williams got involved with EEF for several reasons. Her late mother had diminished eyesight, so Smyrnes-Williams directed her mother's memorial donations to the Eye & Ear Foundation. As VP of LHAS, she championed Dr. José-Alain Sahel's arrival in Pittsburgh. "She got to know all of us and fell in love," Chronis Danek said. "Peggy would constantly state that EEF had the best fundraising team at the University of Pittsburgh. Period. With her raising funds for the Foundation, she became one of us."

In addition to being a renowned classical pianist and trial attorney, Smyrnes-Williams was a devoted wife to Dr. Karl Williams and mother to their son Michael.

"Peggy will be sorely missed by all of us at the Eye & Ear Foundation," said EEF CEO Lawton Snyder. "In addition to being one of our Ambassadors, she was always looking for ways to apply her many talents to help our cause. She was a wonderful person and a dear friend." S+S

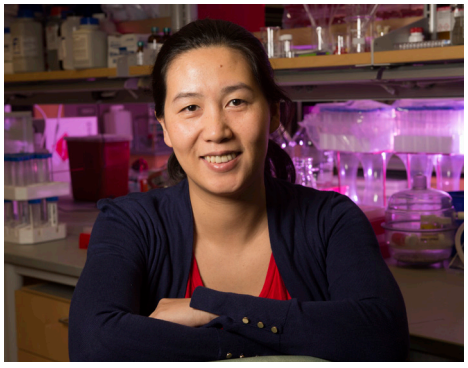
Focusing on our Community

by Lisa A. Goldstein

This year, for the first time through the Hillman Vision Scholars program, four local high school students from low-income and underrepresented communities were exposed to the field of ophthalmology research. This summer, the eight-week program was part of the Hillman Summer Scholars program made possible through the Henry L. Hillman grant to the Eye & Ear Foundation (EEF).

Due to the pandemic, the program was hybrid. All four students attended daily in-person training. Some were exposed to clinical ophthalmologists co-mentored with vision scientists to observe a medical doctor's life and basic research. They also participated in hands-on experiments with side-by-side training. The program concluded with each student successfully presenting their research project virtually on Zoom.

"Despite some hurdles with the hybrid format, all mentors and students made a great effort to complete the program successfully," says Dr. Yuanyuan Chen, Assistant Professor in the Department of Ophthalmology at the University of Pittsburgh and the faculty coordinator. Faculty in the Department showed high enthusiasm. In fact, Dr. Chen states, they have more faculty – including clinical and basic research faculty members -- willing to take on students than the number of students available.



Dr. Yuanyuan Chen

The program is a win-win for faculty and students. The students have limited information about college, and becoming a scientist may not have been in their realm of possibility. "This program is also important to the department and mentors to have an opportunity to reach out to the community and contribute," says Dr. Chen. S+S

What is the Pittsburgh Hearing Research Center?

by Lisa A. Goldstein

The Pittsburgh Hearing Research Center (PHRC) is a unique place that attracts talented and dedicated scientists, physicians, and staff.

Founded in 2017, PHRC's goal is to perform basic science and translational research to develop solutions, cures, and products to treat and prevent hearing loss and hearing loss-related disorders, as well as to develop more general solutions for neuroscience-based disorders, according to PHRC Director Dr. Thanos Tzounopoulos. "We have one of the fastest-growing hearing research communities in the country," he says.

PHRC Research Focus

Research at PHRC covers a lot of ground. Projects include:

- Cellular, molecular, and circuit auditory neuroscience
- Systems and computational auditory neuroscience
- Genetics of hearing loss
- Cell biology and anatomy of the cochlea
- Drug discovery and development for hearing loss and tinnitus
- Gene therapy for hearing loss
- Neuroprosthetics and brain rehabilitation for hearing loss
- Auditory nerve regeneration
- Age-related hearing loss and dementia

"We are well poised and positioned to drive research and innovation in the hearing and neuroscience space and become the top hearing research team in the country,"

- Dr. Thanos Tzounopoulos

New Faculty Members

Two new faculty members have joined the team in the past year: Michele Insanally, PhD and Christopher Cunningham, PhD.

Dr. Michele Insanally

Michele Insanally, PhD, Assistant Professor, Department of Otolaryngology, started in June 2020. She moved to Pittsburgh from NYC. "I am thoroughly enjoying getting to know the city, especially all of the wonderful cultural institutions and my super friendly colleagues and neighbors," she says.

As a graduate student, Dr. Insanally became interested in studying the auditory system when she learned that passive exposure to environmental sounds could significantly influence the developing brain. She's been fascinated by the brain's ability to change and adapt ever since.

PHRC attracted Dr. Insanally because of the "unique combination of amazing basic and translational science." Translational science turns observations from the lab, clinic, and community into ways of improving patients' health.

In her lab, Dr. Insanally studies neural circuits that support flexible behavior. "As we've all experienced, sounds encountered in daily life are rarely neutral," she explains. "You may not notice the sound of a honking horn while stuck in traffic, but one heard while crossing the street might startle you. How do we learn to interpret what we hear and act on it?"

Specifically, the lab investigates how the brain adapts, how this affects learning, and how these behaviors are disrupted in hearing disorders. By incorporating a wide range of techniques – behavioral studies, monitoring of brain signals, computational tools, neural circuit manipulations, and neuroprosthetics – they seek to identify the neural basis of perceptual learning and improve cochlear implant-based perception.

"These are challenging, ambitious projects," Dr. Insanally says. "I've been extremely fortunate to work with four very talented laboratory technicians, a rotation graduate student, and several undergraduates!"

Dr. Christopher Cunningham

Christopher Cunningham, PhD, Assistant Professor, Department of Otolaryngology, started at PHRC in July 2021, when he moved to Pittsburgh from Johns Hopkins University in Baltimore, Maryland. Dr. Cunningham loves Pittsburgh. "It has so many great opportunities and charming neighborhoods," he says. One of his favorite parts of the city is the people, whom he calls extremely friendly and down to earth with a reputation for working hard.

When looking for research areas for his postdoctoral fellowship in neuroscience, Dr. Cunningham was intrigued by the possibility of studying a fascinating biological system with direct impacts on human health. He calls the auditory system "extremely elegant, with many unique specializations contributing to sound processing." He loves going to work every day, knowing he gets to study it and that his work can have a significant impact.

PHRC's "very exciting" vision and structure appealed to Dr. Cunningham. Now that his lab is established, Dr. Cunningham is hiring and ready to begin experiments. He is interested in understanding how genetics and cutting-edge experimental technologies can be utilized to better understand how the auditory system functions and to develop novel therapies for hearing loss. One of the main projects right now is developing gene therapies for a form of deafness linked to a gene called LRTOMT/Tomt.

Drs. Cunningham and Insanally join an impressive team. PHRC is actively recruiting two more experts, one focusing on regeneration and stem cell research and an additional physician/scientist.

"We are well poised and positioned to drive research and innovation in the hearing and neuroscience space and become the top hearing research team in the country," says Dr. Thanos Tzounopoulos. S+S

Thanos Tzounopoulos, PhD and the Pittsburgh Hearing Research Team

The 2021 Wiegand Entrepreneurial Research Awards

by Craig J. Smith

The Departments of Ophthalmology and Otolaryngology are proud to announce that this year's winners of the Wiegand Entrepreneurial Research Award are Christopher Cunningham, PhD, in the Department of Otolaryngology and Debasish Sinha, PhD, in the Department of Ophthalmology.

Dr. Cunningham received his Bachelor of Science degree in Biology from Brigham Young University-Idaho, where he realized his interest in neuroscience. He obtained his PhD in Neuroscience at the University of California-Davis, studying cerebral cortex development. After a postdoctoral fellowship at The Scripps Research Institute and Johns Hopkins University, Dr. Cunningham joined the Department of Otolaryngology and the Pittsburgh Hearing Research Center in 2021. Dr. Cunningham's winning research project, titled *Development of Gene Therapies for LRTOMT-Associated Deafness*, involves studying the gene LRTOMT/Tomt and its association with hereditary deafness. The goal is to understand this association better and develop gene therapy strategies for patients with LRTOMT/Tomt-associated deafness.

Dr. Sinha is the Jennifer Salvitti Davis, MD Chair in Ophthalmology Research and Professor of Ophthalmology, Cell Biology, and Developmental Biology. He is also adjunct faculty, Ophthalmology, The Johns Hopkins University School of Medicine and the Department of Environmental Health & Engineering, The Johns Hopkins Bloomberg School of Public Health. His primary research focuses on understanding the mechanisms that regulate the lysosome/autophagy degenerative process in retinal pigmented-epithelial cells that contribute to the early stages of age-related macular degeneration. Dr. Sinha's winning research project, titled *Targeting Lipocalin-2 (LCN-2) as a Therapy for Age-related Macular Degeneration (AMD)*, is a unique pharmacological approach targeting inflammation in the early stages of AMD.

Christopher Cunningham, PhD

Debasish SInha, PhD

These awards were created by Eye & Ear Foundation Board Member Bruce Wiegand and his wife Barbara in 2018 to provide support in the form of grants for pre-clinical research, which has the opportunity for commercialization, at the University of Pittsburgh Departments of Ophthalmology and Otolaryngology. Additionally, the Wiegands have generously agreed to match any contributions made to the Entrepreneurial Research Award fund up to $100,000. This unique agreement ensures that if a product funded by the award reaches commercialization and goes to market, a portion of profits will be returned to the Eye & Ear Foundation for future research support. S+S

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Pioneers Establishing a Legacy for Skull Base Surgery

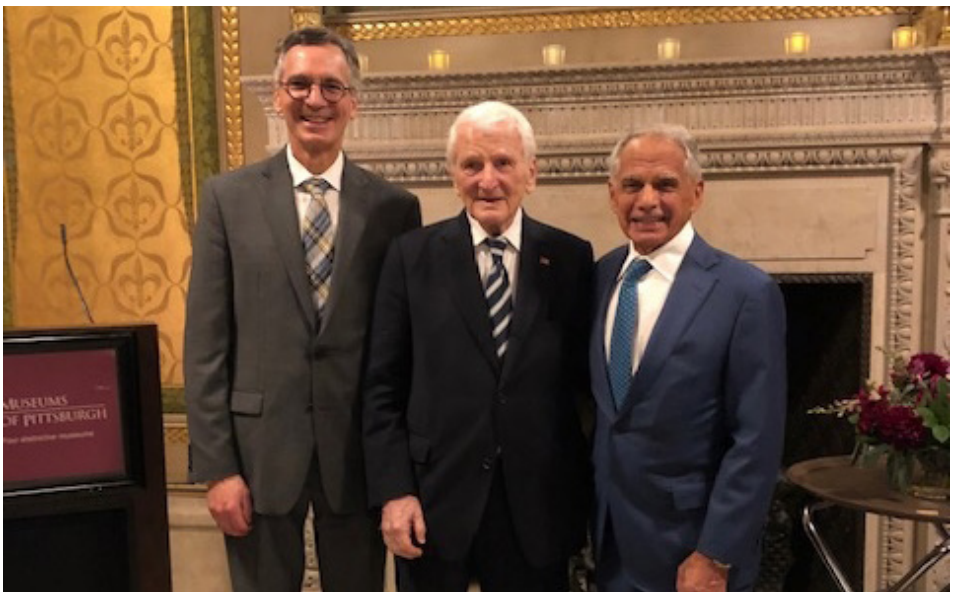
by Lisa A. Goldstein

In 1975, Dr. Eugene Myers, now Distinguished Professor Emeritus of Otolaryngology and Emeritus Chair, Department of Otolaryngology, saw a patient who had been referred by a radiation oncologist. She had cancer originating in her hard palate and had endured 17 operations and two courses of radiation therapy. She complained of severe, unrelenting pain that had her contemplating suicide if he couldn't help her.

Unfortunately, Dr. Myers' exam revealed residual cancer in several areas. But he had recently read an article by Dr. Alfred Ketchum, a well-known surgical oncologist at the National Institutes of Health, in which he described a new technique called the Cranial Facial Approach. Dr. Myers felt he could do the transfacial portion of the surgery on this patient, but he needed to collaborate with a neurosurgeon who could do the cranial aspect.

"I walked over to the office of Dr. Joseph Maroon, a newly minted neurosurgeon who I had known during his training and whose character, surgical skills, and judgment I admired," Dr. Myers recalled. Dr. Maroon was a bit surprised when Dr. Myers said he had a patient scheduled for this daring surgery, but he said he was in. The surgery went very well with no complications. The patient lived free of pain and cancer for five years before dying of a second unrelated cancer. Subsequently, the physicians collaborated in operating upon many similarly afflicted patients with excellent results.

Based on their early success, Dr. Myers worked with neurosurgery to establish a Center for Cranial Base Surgery, the first of its kind in North America. With the recruitment of additional faculty, Pittsburgh rapidly became the destination for such patients from the U.S. and abroad. Over the years, the leadership of the Center for Cranial Base Surgery has changed. Currently, the Co-Directors are Drs. Carl Snyderman, head and neck surgeon, and neurosurgeon Paul Gardner. They have carried out thousands of operations using a minimally invasive technique, many of which they have pioneered, called endoscopic endonasal surgery. They remove tumors from the brain and the base of the skull, often times through the nose, without needing external incisions.



Carl H. Snyderman, MD, Eugene N. Myers, MD, FACS, FRCS Edin (Hon), and Joseph C. Maroon, MD, attending the Pioneers in Skull Base Surgery Dinner

The need to continually refine existing techniques, conduct further research in the biologic behavior of these tumors (some quite rare), and train the next generation of surgeons requires considerable funding, which could be provided from the endowment of a Chair. "This would provide sustainability for this important endeavor," says Dr. Myers.

To that end, Drs. Maroon and Myers are each making significant contributions and will help raise additional support to establish a Chair named in their honor, *"Pioneers in Skull Base Surgery."* To endow a Chair at the University of Pittsburgh is $2 million. Once $1 million has been raised, UPMC has agreed to match the funds up to $1 million. Fundraising is already underway to reach the total amount so that Dr. Carl Snyderman, who is Professor of Otolaryngology, Neurological Surgery, Bioengineering, and Vice-Chair for Quality & Patient Safety, can officially hold the Chair. There is a new rule that an endowed chair cannot be named for an employee until six months after the employee's retirement. Once Dr. Maroon retires, the Chair will be known as the Eugene N. Myers and Joseph C. Maroon Chair in Skull Base Surgery.

"Dr. Maroon and I are thrilled to lend our names to the establishment of the Eugene N. Myers and Joseph C. Maroon Chair in Skull Base Surgery," Dr. Myers said. "It means a great deal to us that our pioneering efforts in establishing the University of Pittsburgh as the world's center for this unique surgery are being recognized in this way."

Dr. Maroon said he's doing this to honor Dr. Carl Snyderman, who he called the consummate caregiver, physician, and surgeon, as well as one of the most creative minds in his specialty.

As for the recipient of this honor, Dr. Snyderman recognized Dr. Myers' leadership and called him a mentor and role model. "With this Chair, I hope to continue his legacy of surgical innovation," he said. S+S

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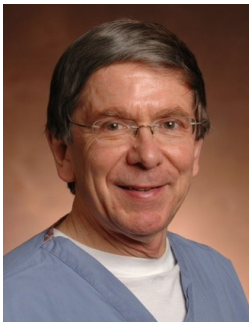
Congratulations to the 2021 Albert C. Muse Prize recipient for Excellence in Research and Advancement of Care in Otolaryngology



Robin T. Cotton, MD, FACS, FRCS(C)

Professor Emeritus

Department of Otolaryngology and Head and Neck Surgery

University of Cincinnati

Congratulations to the 2021 Albert C. Muse Award recipient for Excellence in Innovation and Service in Otolaryngology

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Professor Emerita of Otolaryngology

University of Pittsburgh School of Medicine

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Dinner and Ceremony Monday, November 15, 2021

Please visit eyeandear.org or email Katherine@eyeandear.org for more details.