

HeadLines



JOHNS HOPKINS
MEDICINE

NEWS FROM JOHNS HOPKINS OTOLARYNGOLOGY-HEAD AND NECK SURGERY

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Advising the White House on Hearing Loss

Otolaryngologist, **Frank Lin**'s research connecting hearing loss with conditions such as dementia and brain atrophy led to his presidential advisory post.

While he was growing up, Lin was well aware of the negative impact that hearing loss had on his grandmother's life, an observation that steered his path into the specialty in which he now practices. Helping individual patients he sees in the clinic to hear better has never been enough. Lin says that to really have an impact on this problem, he would need to show that the consequences of hearing loss are so blatant, and that treatment is so beneficial, that policymakers who could make a difference on a broader magnitude couldn't help but notice.

He's accomplishing this goal on a grand scale. In the past two years, Lin was invited to co-chair a workshop on hearing loss and aging at the National Academies of Sciences, Engineering, and Medicine that led to a formal consensus study on the affordability and accessibility of hearing care for adults. Then, last year, Lin was asked to advise the President's Council of Advisors on Science and Technology (PCAST) at the White House, a group that gives advice directly to the president, on hearing loss matters. His work influenced PCAST's recent recommendation to propose new regulations to make hearing aids more accessible to the general public.

Lin's path to advising the White House began with basic epidemiology research that he started in 2010 when he became a faculty member at Johns Hopkins. Through a series of studies, he connected hearing loss with a diverse set of consequences, including dementia and brain atrophy.

"For many people, this line of research would just continue to be pursued," he says. "For me, though,

"I'VE FOUND THAT IT'S ONLY WHEN YOU CAN WORK AT THE INTERFACE OF DIFFERENT AREAS THAT YOU CAN REALLY MAKE A DIFFERENCE."

—Frank Lin

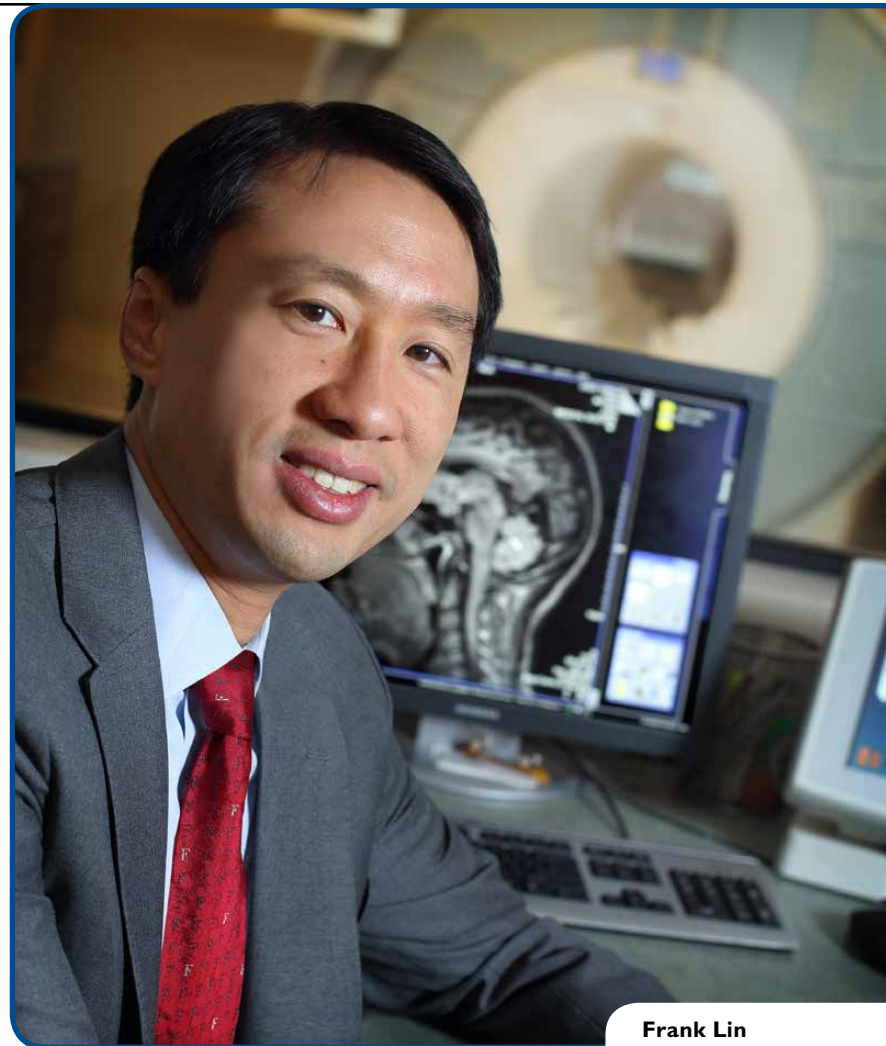
this wouldn't really be too satisfying since what we really care about is whether treating hearing loss can make a difference."

Over the last three years, Lin has worked on launching the ACHIEVE (Aging, Cognitive, and Hearing Evaluation in Elders) study, an offshoot of the Atherosclerosis Risk in Communities - Neurocognitive Study, which will investigate if treating hearing loss reduces the risk of cognitive decline and dementia in older adults.

With promising early results, Lin says that he's not interested in waiting around until the study's conclusion to make hearing help more accessible to older adults. In 2015, he co-founded AccessHEARS with otolaryngology resident **Carrie Nieman**, a nonprofit that's distributing personal sound amplification products at no cost to older adults in Maryland using trained community health workers to provide accessible and affordable hearing care.

To ultimately effect change, however, Lin needed to go to the top—hence his governmental work with the Institute of Medicine and PCAST.

Lin notes that working at these levels has been



Frank Lin

fun but challenging. In any given day, he says, he's constantly switching tracks to speak "different languages"—conversations with fellow scientists, White House staff members, and AccessHEARS colleagues flow back to back. But it's ultimately worth it to achieve his goal of improving the lives of older adults by improving their hearing.

"I've found," Lin says, "that it's only when you can work at the interface of different areas that you can really make a difference." ■

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To Find Promise for Patients with **Laryngotracheal Stenosis**

Kevin Motz, a fourth-year resident in the Department of Otolaryngology–Head and Neck Surgery, admits that he wasn't a particularly strong student in high school. However, when he arrived at Stevenson University for his undergraduate education, he says, members of the science faculty saw something that he didn't see in himself.

"KEVIN'S PRODUCTIVITY IN MY LAB HAS BEEN STRONG, BUT REALLY, IT'S HIS GROWTH AS A CLINICIAN-SCIENTIST THAT HAS MADE ME PROUD."

—Alexander Hillel

"They invested in me," he remembers. "They helped me unleash my true potential and realize what I was capable of."

After completing his undergraduate degree in biology, he attended Georgetown University School of Medicine. Now at Johns Hopkins, Motz is showcasing that potential in

a big way. As a recipient of one of the coveted National Institutes of Health T32 training grants as well as a CORE grant from the American Academy of Otolaryngology–Head and Neck Surgery, he is spending two years in Johns Hopkins physician-researcher **Alexander Hillel's** laboratory studying the mechanisms behind laryngotracheal stenosis (LTS), a step that could lead to new, more effective treatments for this debilitating disease.

LTS is characterized by scarring and subsequent narrowing of the airway, eventually restricting breathing and hindering some patients' ability to speak. The most common trigger is prolonged tracheal intubation, a procedure in which a tube is inserted into the airway, to help patients breathe. Other cases arise from autoimmune disorders or from no known cause.

Current treatment for this disease involves periodically dilating or stretching the airway or releasing scar tissue with a laser; however, these methods provide only temporary relief. The only permanent cure is surgery to remove the scarred portion of the airway and reconnect the healthy ends. Neither is an ideal option, Motz says.

Developing new treatments requires learning what causes patients to develop airway scarring in the first place—the

"THEY INVESTED IN ME. THEY HELPED ME UNLEASH MY TRUE POTENTIAL AND REALIZE WHAT I WAS CAPABLE OF."

—Kevin Motz

focus of Hillel's and Motz's research. Motz is currently investigating the role of groups of immune cells known as CD4 T cells. Using mice with induced airway scarring as a model, Motz has identified increased numbers of these cells in tracheal scar tissue. Currently, he and his colleagues are working on blocking a subset of these cells known as T helper 2 cells. Preliminary evidence demonstrates that mutant mice carrying an induced genetic defect that blocks the development of these cells form significantly less airway scarring and stenosis.

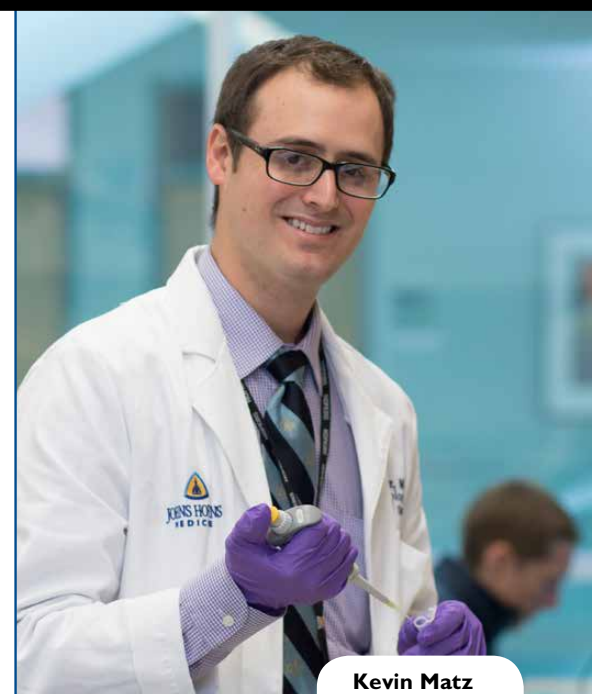
Motz says he hopes that gaining a basic understanding of this disease will lead to drugs that can block the development of LTS, perhaps by blocking immune system involvement. In addition to advancing the field, these studies are also giving him a strong

background to follow in the footsteps of his mentor, Hillel, who also did a T32 fellowship during his own training at Johns Hopkins.

"Kevin's productivity in my lab has been strong," Hillel says, "but really, it's his growth as a clinician-scientist that has made me proud."

After completion of his residency, Motz intends on pursuing advanced fellowship training in otolaryngology and says he looks forward to continuing his research in LTS. ■

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Kevin Motz

RESEARCH

Cracking the Code of Adenoid Cystic Carcinoma

Of the estimated 566,000 new cancer diagnoses made each year in the United States, adenoid cystic carcinoma (ACC), a cancer mainly found in the salivary glands, represents only 0.2 percent.

"But, if you're the one with the disease, percentages don't matter," says **Mariana Brait**, assistant director of the Head and Neck Cancer Research Division of the Johns Hopkins Department of Otolaryngology–Head and Neck Surgery. That's why she's made ACC a central focus of her research career.

Unlike other more common cancers, such as breast, prostate or lung, there are no effective

treatments for ACC. Researchers still know virtually nothing about the causes of this cancer and how it develops and spreads, says Brait, a roadblock that hampers discovering new therapies.

The key to finding new tools to attack this disease, she explains, is gathering clues about how ACC cells are different from those in healthy tissues. In the past several years, researchers working on this disease at Johns Hopkins and other institutions have discovered that these cancerous cells often possess an unusual trait: a rare type of genetic alteration in which part of one gene has broken off and fused to another. Though these fusions affect the same two genes, they're not



Mariana Brait

A Patient's Journey: Recurrent Respiratory Papillomatosis

The first time Otolaryngologist head and neck surgeon **Simon Best** met Morgan Toll was relatively unremarkable. The 25-year-old was visiting Best's clinic for simple follow-up after surgery from another provider for complications of recurrent respiratory papillomatosis (RRP), a disease that causes benign tumors called papillomas to grow inside the airway and the condition that Best has focused on for his entire career.

However, the second time that the two met would prove to be one of the most dramatic patient encounters of Best's career. Toll returned with severe breathing difficulties and scarring of her vocal cords, with an airway measuring only 3 millimeters. It was the narrowest he had ever seen in a patient who didn't already have a tracheostomy, or a breathing hole, in the neck.

"IT'S A LONG ENDEAVOR, BUT IT'S EXTREMELY GRATIFYING THAT WE CAN MAKE PROGRESS STEP BY STEP, AND AT THE END, WE CAN GIVE HER A NEW LEASE ON LIFE."

—Simon Best

"She came to my clinic saying that her breathing had gotten worse, and it was unbelievable. There was hardly any space in her airway at all," Best remembers. "It was so severe that the only thing to do that day was an emergency tracheotomy."

Rushed to surgery, the procedure would start Toll on a two-year surgical journey with Best that would

give her back her ability to breathe and her voice.

Toll was diagnosed with RRP at only 8 months old. By the time she was 11, the trained theater actress had endured more than 200 surgeries. These procedures were necessary not only to remove the papillomas that constantly grew, but also to remove scar tissue that became a byproduct of so many surgeries.

In remission for more than a decade, Toll's disease began to resurface when she was 22. Eventually, her voice barely rose above a whisper, and simple physical tasks—such as climbing a small flight of stairs or even walking across a room—became extreme challenges.

After her emergency tracheotomy, Toll and Best formulated a plan to improve her health and her voice. He's now performed a series of seven surgeries, each one removing a little more scar tissue using special techniques to reduce regrowth. Part of the novelty of her case is that her vocal cords had to be removed due to overabundance of scar tissue there, but Best has been able to return her voice by injecting a filler into other parts of her larynx, thickening the tissue there to act as a new set of vocal cords. Eventually her airway was widened to the point that the tracheotomy was able to be removed—the culmination of a long journey.

Being able to breathe—a gift that she'll never take for granted, Toll says—and speak has allowed her to start pursuing acting work, a vocation that runs in her family. (Toll's sister is Olivia Holt, an actress who has starred in several Disney Channel shows and movies.)

"I'm now looking to start my life again," she says.

Although Toll's treatments are far from over—the nature of RRP means that new papillomas will continue to arise—Best says he's incredibly

encouraged by her success so far.

"It's a long endeavor," he says, "but it's extremely gratifying that we can make progress step by step, and at the end, we can give her a new lease on life." ■

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When patient Morgan Toll came to Johns Hopkins with severe breathing difficulties and scarring of her vocal chords, Simon Best first performed an emergency tracheotomy and devised a long-term surgical treatment plan that has restored her voice and ability to breathe.

identical from patient to patient—the genes are broken and fused at different points.

Whether these alterations have differing effects on the disease is unknown. However, it could make a big difference for patients, says Brait. If researchers had an idea of how each mutation type affected individual patients, they could tailor conventional therapy to be more effective or, eventually, develop targeted medicines aimed at disrupting the products of these fused genes.

To make those possibilities a reality will require research to identify mutation types in hundreds of tissue samples from patients and try to correlate them with the disease characteristics that doctors see in the clinic. This includes identifying whether

"IF YOU'RE THE ONE WITH THE DISEASE, PERCENTAGES DON'T MATTER."

—Mariana Brait

patients' disease recurs after treatment, whether recurrence was local or in distant metastatic sites, or how long patients ultimately survived after diagnosis, for example. Performing this work will take research funding that's in short supply.

Determined to make this research concept a reality, Brait applied for a grant from the Adenoma

Cystic Carcinoma Research Foundation (ACCRF), a nonprofit that supports research to improve therapies and accelerate a cure for this disease.

In February, she learned ACCRF had fully funded her grant proposal for two years at \$100,000 each year.

"It was a huge deal to me because this is really my first independent grant, ideas that I initiated and that I can bring to fruition. It also means a lot in the head and neck community," says Brait. "With these funds, my colleagues and I will have a chance to make a real difference for patients with ACC." ■

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Amber's Story:
Reconstruction After
Mohs Surgery

See a video chronicling the medical journey of patient Amber, who was left with a large wound on her face after undergoing Mohs surgery for the removal of squamous cell carcinoma. Willing to travel anywhere for the best facial plastic and reconstruction surgeon, she found surgeon Lisa Ishii at Johns Hopkins, and she is delighted with the outcome.



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