



NIDCR Dr. Janice Lee leads underbite operation

Procedure has never been conducted in the CC before

In August 2018, Trina Davis walked into her first college course at Bowie State University with her mouth shut, elastic bands constricting her ability to communicate. Testimony to her strength and resiliency, Davis was overjoyed to be in class and recovering from a life changing surgery.

Ironically, 17-year-old Davis, a senior at Bowie High School, started taking English 101, Public Speaking and Oral Communications just four weeks after orthognathic surgery. The five-hour operation to fix her underbite, caused by underdevelopment of her upper jaw and overdevelopment of her lower jaw, was the first time the procedure was performed in the Clinical Center.

While her appearance has changed drastically since surgery – the operation was not for cosmetic purposes. Patients with skeletal underbites (or severe overbites where there is underdevelopment of the lower jaw) deal with headaches, trouble breathing, speech impediments and problems eating and chewing, which can lead to gastrointestinal upset.

“Before surgery, I had trouble chewing certain meats and I had a speech impediment,” Davis said. “I had to go to speech therapy to figure out how to form my words correctly. Now, I’m able to better pronounce my words the way I want to. I’m still learning how to chew certain things but I’m get-

ting better. It’s getting easier because my bite changed. I feel good that I’m able to contribute to research [and that this] research will help somebody else. I’m appreciative for that.”

Davis and her family are part of a natural history protocol led by Dr. Janice Lee, the clinical director at the NIH National Institute of Dental and Craniofacial Research (NIDCR). The study focuses on craniofacial anomalies or developmental growth abnormalities of the head, face and jaw. Lee is on a mission to see if part of the abnormal growth that Davis and others experience has a genetic connection and can be predicted.

“Our genes dictate how we look and those genes run in families. For example, I often hear how a patient recalls having an uncle or grandparent who has the same facial trait, whether it is a large lower jaw or a small nose, etc.,” Lee said.

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Trina Davis in March 2019, nine-months post surgery.

Cochlear surgery at CC provides hearing to pediatric patient

In the NIH Clinical Center operating room, a surgery was performed in Fall 2018 that marked two milestones for Building 10 – the first time *two cochlear (bilateral) implants* were placed and the first time the surgery was performed for a *pediatric* patient, allowing them to hear once again.

Dr. Michael Hoa, an otolaryngology surgeon-scientist with the NIH National Institute on Deafness and Other Communication Disorders (NIDCD), successfully performed the surgery, which lasted 4.5 hours. Others have previously performed cochlear implantation at the Clinical Center, but in *one ear of adult* patients.

“It had been over 10 years since a [single] cochlear implant surgery was performed at the Clinical Center,” noted Hoa, who is a specialist in neuro-otology (the study of the anatomy and diseases of the ear). Hoa established the cochlear implant programs at both MedStar Georgetown University Hospital, where he serves as co-director of the cochlear implant program, and the Washington D.C. Veterans Affairs Medical Center.

COCHLEAR page 4

From the House of Hope to the homeless: operating room staff bring warmth to the local community

With a needle, thread and desire to give back, members of the NIH Clinical Center operating room have repurposed surgical cloth into blankets for those experiencing homelessness in Montgomery County.

Seven surgical technologists and nurses donate their personal time to sew a special double ply, water resistant cloth that

wraps sterilized surgical instruments prior to any operation. The cloth, which is never inside the operating room during a procedure, cannot be recycled. It’s been estimated that the blue sterile wraps account for roughly 19 percent of an operating rooms waste (<https://go.usa.gov/xm9pK>).

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Back Row: Maureen George, Dr. Jonathan Hernandez, Ellie Custis and Melissa Wheatley. Front Row: Carrie Wellen, Phuong Nguyen and Audrey Murphy.

"We wanted to reduce the operating rooms' footprint while finding a meaningful way to support our community and give to those in need. The blue wraps are water resistant, durable and warm," said Carrie Wellen, the surgical technologist who began the sewing initiative.

On Feb. 27, Wellen delivered the first batch of 30 blankets to Bethesda Cares, a non-profit outreach organization for the homeless. Bethesda Cares works closely with people experiencing long-term homelessness, helping them find paths to homes (<http://bethesdacares.org/>).

There are nearly 850 homeless individuals in the Montgomery County area, according to a 2018 survey by the Metropolitan Washington Council of Governments (<https://tinyurl.com/yyaz2rh6>). Many will not go to shelters even when the temperatures drop to below freezing. The blankets provide a comforting, warm solution for the homeless.

"The blankets are a wonderful item for our street outreach staff to distribute during this extremely cold period," said John Mendez, Bethesda Cares executive director. "We're truly grateful for the support of the NIH Clinical Center throughout this challenging winter period."

Using their suture skills, the volunteers can typically produce one blanket an hour. Dr. Jonathan Hernandez, surgical oncologist with the National Cancer Institute, donated a sewing machine for volunteers to use before or after work. To become a sewing volunteer, email carrie.minard@gmail.com

Wristbands let patients check-in clinics, day hospitals

The Clinical Center has added a new component to the electronic medical record system to allow patients to check into an outpatient clinic or day hospital appointment by simply scanning their wristband identification bracelet. Immediately, a patients' location is updated in their record, allowing for staff to improve the time in which patients are seen by clinicians and to better coordinate patient care and enhance patient safety.

Previously, a clipboard, pen and paper were used for the check-in/out process and clinic staff were dependent on someone notifying them of patient arrival. The paper process also did not update the location in the medical record. With this enhancement, departments no longer have to call the clinic or the nurse to determine the patient's location. Using an electronic process allows clinic staff to remotely identify when a patient has arrived.

When patients scan their wristband, their name will appear on a patient list in the medical record via an application called Display Board. The checked-in location updates

in their record with a suffix of – DB (Display Board). As patients check out, the patient list is updated to remove patients from the location and return them to their home clinic location.

After each successful scan in/out, patients will hear, "Thank you for checking in," or "Thank you for checking out."

"By decreasing the number of phone calls and interruptions and improving identification of patient location, we have taken a step in improving patient safety and patient care coordination," said Dr. Tina Patel, with the Clinical Center Department of Clinical Research Informatics.

The electronic check-in/out technology may be brought to hospital procedure areas in the future.

The implementation of this new feature was led by Dr. Jon McKeeby, and supported by Keith Adams, Marcus Anderson, Seth Carlson, Larry Harris, Dr. Rachel Khoo, Yenshei Liu, Frank Mickey, Mary Myers, Mindy Nghiem, Dr. Tina Patel, Jeanne Preuss, Richard Walker and Betsy Wendell.

Saul Rosen, former Acting Director of CC, passes at 90

In February, former Acting Director of the Clinical Center Dr. Saul Rosen died at the age of 90. For more than three decades, Rosen served the NIH. Under former CC Director John Decker, Rosen was named Deputy Director (1984) and then Acting Director from (1990-1994).

In 1984, the NIH Record (page 10 <https://go.usa.gov/xEzFQ>) quoted Rosen saying, "My job will be to make sure the Clinical Center continues to function as a high-quality hospital, which is easy because it is already very good indeed." The CC News covered his retirement in 1994 (<http://tinyurl.com/y65hn2uu>). Rosen said, "The CC stands for more than the Clinical Center to me. It also represents competency and collegiality. And I hope that's what we have been."

In his interview, Rosen astutely identified scientific processes and technological advancements that would continue well beyond his time: MRI and PET scanning, a strong protection of human subjects in investigative research and revolutions in molecular and cell biology.

In addition, Rosen said, "The thing that knocks my socks off from here to Prince

George's County is the new work in gene therapy, work that was pioneered here at the Clinical Center [in 1990]. Dr. Francis Collins...will be heavily involved in pushing this technology forward."

Rosen began his path forward in NIH research when he arrived as a clinical associate in 1958 at the National Institute of Arthritis and Musculoskeletal and Skin Diseases. Later, he conducted research as a senior investigator at the National Institute of Diabetes and Digestive and Kidney Diseases (NIDDK).

A Bostonian, Rosen graduated Harvard College at the age of 18, cum laude, then pursued a Ph.D. in chemistry at Northwestern University. He then returned to Harvard Medical School and graduated in 1956. View NIH Oral History, a testimony with Rosen: <https://bit.ly/2O4ENUi>



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FEVS

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"A well-known example is the 'Habsburg' jaw, a famous European family whose genealogy is linked with the distinct family trait of an underbite," Lee said. "We still don't know exactly which genes cause these features. One of our goals is to see if we can predict who's going to have a certain type of development – specifically abnormal development that causes functional problems."

"When a patient develops abnormally or, for example, the lower jaw grows too far out for too long, it's no longer just an aesthetic concern or dental concern," Lee added. "It's to the point where nothing can be done about it except surgery. Our hope is to identify kids and families at risk for these extreme growth patterns and potentially intervene to avoid surgery. Gene analysis and new imaging technologies have opened up the possibility of predicting these growth patterns."

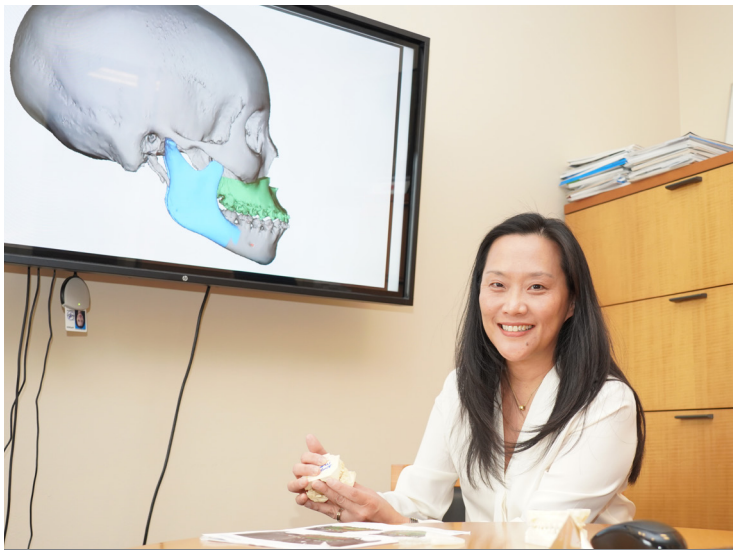
Just Keep Rowing

Before arriving at the NIH in 2013, Lee had performed hundreds of surgeries to fix a skeletal underbite or overbite and craniofacial birth defects. But Davis's operation would be the first time she conducted it in the Clinical Center. Patient safety was paramount. But to Lee, patient safety comes naturally. In addition to her NIDCR duties, she serves as the chair of the Patient Safety, Clinical Practice & Quality Committee at the NIH.

With a few months to prepare, she gathered a multidisciplinary team and got started.

"It took a lot of coordination," Lee said. "I started off by giving a presentation of what I was going to do and examples of the surgery to several groups, including a team of representatives from the Perioperative Medicine Department, Intensive Care Unit, Office of Patient Safety and Clinical Quality, and the unit where Davis would recover. What surgeons have learned is that the more you communicate with everybody who's working with you, the less we will have errors but also just the appreciation of everybody's perspective. Then came the heavy lifting and it could not have been done without Pam Orzechowski, my research nurse, and our craniofacial team. She orchestrated the nursing and support staff."

Lee continued training with the operating room team, specialized surgery equipment was gathered from nearby Washington



Dr. Janice Lee, the clinical director at the NIH National Institute of Dental and Craniofacial Research, holds an old stone model of the mouth while showcasing the new 3-D model of the mouth on screen, used to better prepare for surgery.

Hospital Center, and the National Naval Medical Center, and surgical checklists were created. As the clinical director, Lee had implemented a proctoring policy for new clinicians, a typical practice in academic medical centers, and chose to apply the policy to herself. Thus, she also brought in Drs. Kal Shastri and George Obeid from Washington Hospital Center to "proctor" her. Dr. James Gilman, the CEO of the Clinical Center, stayed closely attune to the preparation and activities. Virtual surgical planning was initiated about four weeks prior to surgery. With new technologies like 3D cone-beam computed tomography (CT), Lee was able to simulate the surgery on the face, have a surgical guide fabricated and prepare for surgery. These plans were shared with Davis and her parents.

Finally, the day of surgery came.

"I love operating. This hospital is unlike any other," Lee said. "This hospital has been extremely supportive of doing this well and doing this right. The huge trust to do things safely and in a team approach, I think, really resonates with me. When you're in the operating room, you have to work well as a team to be effective. That has really been what I've experienced here. Everybody wants to do what's best for their patients and everybody wants to work together to make it safe."

Lee and her team set forth to fix Davis's jaw. Twenty screws, four plates and several large bone screws were placed.

"You don't think about breaks," Lee said. "When you get going, it's like a rhythm. It's like a rowing team. You just start moving down the river and lo and behold you cross that finish line. As a senior surgeon, one of the things I really work to refine is the economy of motion – meaning every move, every effort has to matter and has to be ef-

ficient. We trim off minutes here, minutes there which becomes hours and then the patient no longer requires six hours of anesthesia, its five hours or less."

"I've built surgical teams [before coming to NIH and] it took years to get everybody to work smoothly," Lee added. "Here, I don't know what it was. Maybe the laser focus everybody brought. Maybe the fact that everybody was super engaged. It [was] remarkably smooth."

More Than a Guess

Lee's goal is to enroll 1,500 patient partners and healthy volunteers over the course of 20 years to start looking at the genetic etiology and to see if she can predict these conditions.

"People often ask me when they bring in their first child, 'Is this going to happen to my second, or third, or fourth child?' and I've never been able to give them a good answer," Lee said. "The best answer is 'Let me look at mom and dad. Let me get an idea.' But it's a guess."

Through the course of the research, "if it turns out to be very difficult [to identify] mutations or variants, we will work on predictive tools using computational methods and high-resolution/low-radiation cone-beam CT scan," Lee said. "We need a multi-pronged approach because in medicine it is rarely a one answer for every patient."

Two-dimensional x-rays are a thing of the past. Lee is excited to see how three dimensional cone-beam CT scans can bring high resolution, low radiation, better surgical simulations and prediction capabilities in the years to come.

With this technology, "We get to see if there are certain things about the shape of the skull base and the rest of the face that would give us some ideas that somebody will grow differently. When they're born, it's not obvious. When they hit puberty, for some reason, the lower jaw may continue to grow."

Pamela Orzechowski, Lee's research nurse coordinator, said "Perhaps we can eventually provide some early intervention or stop the overgrowth of the jaw so a patient will not have to undergo surgery."

In January 2019, Davis returned for a check-up. With a smile on her face, and all A's on first semester of college courses, she said "once you start seeing actual results in your face as the swelling goes down, you know it was worth it. You see how things have changed for the better."

Hoa's expertise in establishing cochlear implant programs and his experience with building the multidisciplinary medical teams necessary for pediatric cochlear implant programs were essential for success.

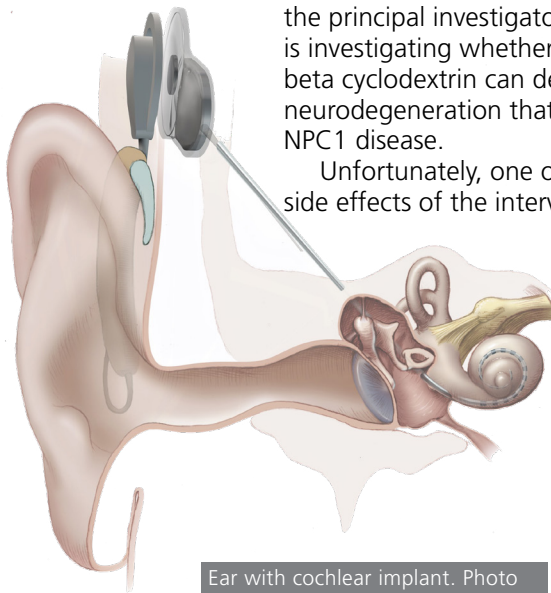
"The clinical care team at the NIH Audiology Unit, coordinated by Dr. Kelly King, was critical to mapping out this patient's surgery and recovery program," added Hoa. He also engaged community based partners, such as the Treatment and Learning Center in Rockville, Md., and Gallaudet University in Washington D.C.

A cochlear implant is a small, electronic medical device that can help a person who is deaf or severely hard-of-hearing experience sounds. The implant consists of two parts: an external portion that sits behind the ear and a second element that is surgically placed under the skin and inserted into the cochlea in the inner ear. A cochlear implant bypasses damaged portions of the ear and directly stimulates the auditory (hearing) nerve. Signals generated by the implant are sent to the brain through the auditory nerve, which the brain recognizes as sound.

The cochlear implant surgery was conducted on a patient participating in a research protocol to treat Niemann-Pick Type C1 (NPC1) disease at the Clinical Center. NPC1 disease is a rare, progressive genetic disorder in which cholesterol and other fatty substances (lipids) accumulate inside of cells and tissues of the body damaging the brain, peripheral nervous system, liver, and other organs and tissues. The disease is often fatal - many patients with NPC1 develop debilitating problems such as trouble swallowing and walking.

"Niemann-Pick robs people of their experience of the world," said Hoa.

Hoa is the consulting otologist on the protocol Hydroxypropyl Beta Cyclodextrin for Niemann-Pick Type C1 Disease (<https://go.usa.gov/xmxGe>) which is sponsored by a Mallinckrodt Pharmaceuticals, and is being conducted by the NIH Eunice Kennedy Shriver National Institute of Child Health and Human Development and the NIH National Center for Advancing Translational Sciences with Dr. Forbes D. Porter serving as



Ear with cochlear implant. Photo courtesy of NIDCD.

the principal investigator. The protocol is investigating whether hydroxypropyl beta cyclodextrin can delay or stabilize neurodegeneration that happens in NPC1 disease.

Unfortunately, one of the potential side effects of the intervention is hearing loss, an outcome that patients and their families were notified of and viewed as an acceptable trade-off for the potential benefit for people with NPC1.

One participant in the study experienced a profound hearing loss, as a result of intervention with the study drug. When they first developed hearing loss the study participant was seen by the Treatment and Learning Center, which donated hearing aids to the patient. However, the patient continued to lose more hearing and eventually had so much hearing loss that hearing aids were no longer effective. The patient underwent a cochlear implant candidacy evaluation with an external pediatric audiology team led by Dr. Claire Buxton, based at Gallaudet University.

The team concluded that the patient did not have appropriate access to sounds, including speech, and that they were a candidate for the technology. The intricate surgical procedure, entailing just over two hours for each ear and a life-long implant of medical technology, was successful.

The patient's journey didn't end once the surgery was done, however. It can take a year for a patient to adapt to the device. Successful rehabilitation after implant surgery includes speech and auditory rehabilitation therapy as well as sessions with cochlear implant audiologists who adjust and reprogram the implant as the brain learns how to process the new sounds it receives.

The unique nature of the NIH and the integrated care teams and researchers provided a positive model for this surgical procedure that was new to NIH in pediatric patients. The ongoing care and commitment to collaboration, not only among colleagues, sections and departments - but also with study volunteers at the NIH - is an enduring model.

Medical musician brings soothing sounds to patients



The NIH Clinical Center recognizes its volunteers, such as Michael Bard pictured above, during National Volunteer Week, April 7-13, and throughout the year. In 2018, more than 170 people volunteered 11,000 hours in the hospital.

For volunteer Michael Bard, a guitar is his instrument of healing. Bard uses the power of his music to address the psychological, social and spiritual healing of individuals.

Bard, a medical musician, who volunteers in the Pain and Palliative Care Department, comes every Monday, Wednesday and Friday for patients and families, and the clinical staff in the intensive care, pain and palliative care and oncology units.

Although Bard specializes in Spanish, Latin and classical guitar music, he can easily transition from Bach to the Beatles and from show tunes to the Rolling Stones. He has performed at the John F. Kennedy Center for the Performing Arts and Strathmore, a renowned visual and performance arts center in North Bethesda.

Bard refined his skills at the Medical Musician Workshop: Medicine for Musicians in July 2017 and 2018. The workshops, held at the Berkshire Medical Center in Massachusetts, included lectures by physicians from various medical departments, and hands-on training in the hospital's critical care unit.

Volunteer at the CC: <https://go.usa.gov/xEMb4>