SUMMER 2019

DELIVERING ON A PROMISE

A Voice for Cooper

Le Bonheur surgeon first to successfully create airway, voice box for patient with total laryngeal agenesis

THE UNIVERSITY OF TENNESSEE HEALTH SCIENCE CENTER

Le Bonheur Children's Hospital

Le Bonheur surgeons successfully complete first-ever pediatric transoral robotic surgery

Surgeons at Le Bonheur Children's recently performed the first known pediatric transoral robotic approach to the cervical spine. The surgery team used the newest generation robot to remove a bony tumor from the C1 vertebrae of a 14-year-old female patient.

"This is a unique approach for pediatric spine surgery," said Le Bonheur Surgeon in Chief Trey Eubanks, MD, a pediatric surgeon. "While this method is currently used to remove lesions in adults, as far as we know this approach has never been used in a child."

The surgical team included Otolaryngologist and Robotic Surgeon John P. Gleysteen, MD; Neurosurgeon L. Madison Michael, MD; and Otolaryngologist and Rhinologist Sanjeet Rangarajan, MD, M. Eng.

Use of the robot and transoral approach had multiple benefits for the surgeons and the patient. Without use of the robot, surgeons would need to split the palate to create the space to remove the lesion. Avoiding opening these critical tissues significantly reduces recovery time, risk of wound problems post-surgery and need for narcotic pain medicine. The patient is left with no visible incision or scarring as a result of the transoral approach.

The robot also gave the surgeons specific advantages: detailed, three-dimensional view of tissues as well as the ability to perform fine movements in a miniscule workspace.

"This gives us another tool with which to approach difficult cases when the surgical plan would be more difficult or require a longer hospital stay," said Eubanks. "Our technology is only as good as our faculty who are skilled at these types of approaches."

Otolaryngologist and Robotic Surgeon John P. Gleysteen, MD, sets the robot in place for surgery. Robotic surgery reduces recovery time, risk of post-surgery wound problems and need for narcotic pain medicine.

The surgery began with a robot set up by Gleysteen and Rangarajan, who prepared the patient for surgery and made the initial incisions. They dissected the tissue until reaching the spine. Once the instruments reached the spine, Michael worked to remove the patient's vertebrae lesion. Rangarajan closed the tissues and finished the surgery.

Le Bonheur uses robot technology for surgical treatment of other conditions, including performing robotic sleeve gastrectomy as part of Le Bonheur's Adolescent Bariatric Surgery program. The pediatric urologists also utilize robotic surgery to treat congenital abnormalities of the genitourinary tract, including ureteropelvic junction obstruction and vesicoureteral reflux in addition to performing nephrectomies.

Use of robotic surgery across pediatric specialties allows for better outcomes, a faster recovery time and more precise procedures for surgeons.

Le Bonheur Children's Hospital

Le Bonheur Children's Hospital in Memphis, Tenn., treats more than 250,000 children each year in regional clinics and a 255-bed hospital that features state-of-the-art technology and family-friendly resources. Our medical staff of more than 240 physicians provide care in 45 subspecialties.

LE BONHEUR LEADERSHIP

Michael Wiggins, MBA, FACHE – *President*
Jon McCullers, MD – *Pediatrician-in-Chief*
Barry Gilmore, MD, MBA – *Chief Medical Officer*
James "Trey" Eubanks, MD – *Surgeon-in-Chief*
Harris Cohen, MD – *Radiologist-in-Chief*
Kathleen Seerup, BSN, MSHA, RN, NE-BC – *Chief Nursing Officer*

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A Voice for Cooper

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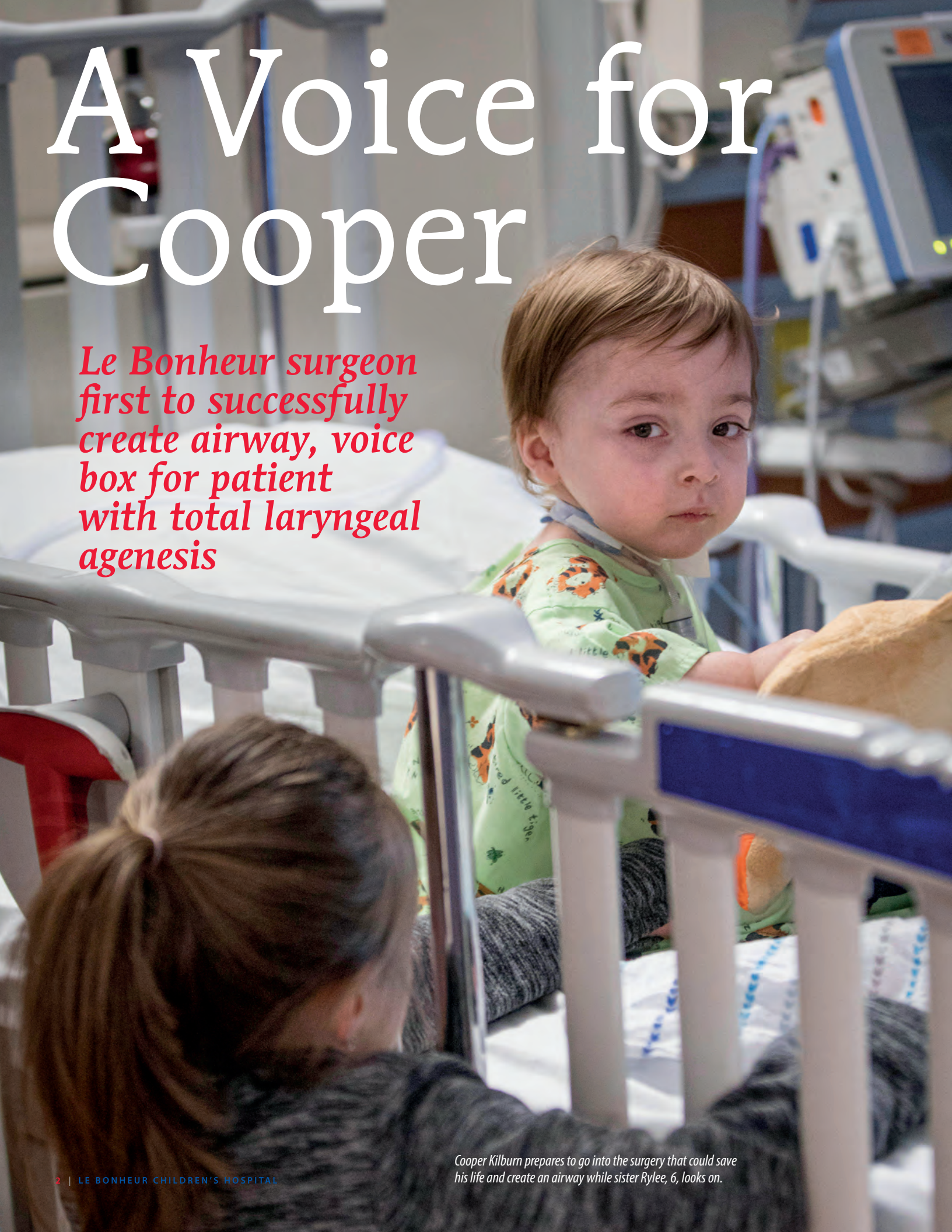


Cooper Kilburn prepares to go into the surgery that could save his life and create an airway while sister Rylee, 6, looks on.

First a car wreck would save Cooper Kilburn's life. Two years later, an innovative surgery would change it.

Cooper was born with no airway, no larynx and, therefore, no voice. He relied on a trach to breathe. He slept on a ventilator to survive the nights. And at 2 years old, he had never made a sound. Not a cry. Not even a whimper.

And that's how this story begins. Lucky for Cooper, his medical team made sure that wasn't the end.

On February 27, 2019, Cooper became the world's first recorded child born with no airway or larynx to successfully undergo voice box reconstruction. The surgery, led by Le Bonheur Otolaryngologist Jerome Thompson, MD, used a rib graft to create the voice box and establish a viable airway.

Ten weeks later, he made sounds for the first time – laughing with his mom in the comfort of his home.

"I thought I must be dreaming," said Cooper's mom, Brooke. "I called Dr. Thompson to listen over the phone and confirm — we were finally hearing Cooper make his first noises."

The story of Cooper's voice, Brooke will tell you, is an unlikely one. Because if it wasn't for a car wreck at the start of a family vacation, the Kilburns would have lost their son before he was born.

ALL IT TAKES IS FIVE PERCENT

Brooke Kilburn of Adamsville, Tenn., was 16 weeks pregnant with Cooper when she was in a car accident. Shaken but unhurt, she went to her obstetrician for an ultrasound to make sure all was well with her child.

"The obstetrician saw something wrong but didn't know what it was," said Brooke. "A specialist in Jackson, Tenn., told us he had never seen anything like this before. He prepared us for the worst case scenario."

An ultrasound revealed fluid was building in Cooper's chest because of a rare condition – total laryngeal agenesis due to congenital high airway obstruction syndrome (CHAOS).

Cooper had no airway and no way to breathe outside the womb.

"The fluid had started to compress Cooper's heart," said Brooke. "If we hadn't had that wreck it would have been too late for him."

As it was, Cooper had a five percent chance of being born alive.

BEATING THE ODDS

Brooke and Brad Kilburn quickly began searching for ways to save Cooper's life.

Their first stop was St. Louis Children's Hospital where a surgeon was able to take enough fluid out of Cooper's chest in utero to relieve the pressure on his heart. But that was all he was able to do. His recommendation – take Cooper to Memphis and Le Bonheur Children's.

"Both our specialist in Jackson and surgeon in St. Louis mentioned Le Bonheur's expertise for Cooper," said Brooke.

After Otolaryngologist Jerome Thompson, MD, performed the EXIT procedure that allowed Cooper to survive birth, he spent 324 days in Le Bonheur's Neonatal Intensive Care Unit (NICU).

"We knew that Le Bonheur specialized in children and that with the help of Dr. Thompson, Le Bonheur was the only place that Cooper could be born."

In conjunction with Le Bonheur Children's Hospital's Fetal Center, Thompson put together a plan for Cooper to survive delivery. Cooper would be delivered at a neighboring birthing hospital and then transported to Le Bonheur's Neonatal Intensive Care Unit (NICU).

Brooke and Brad Kilburn pray over Cooper before his life-saving surgery to create an airway and voice box.

At birth, Thompson immediately performed an ex utero intrapartum treatment (EXIT) procedure, a partial caesarean section that allowed Cooper to continue to receive oxygen via the placenta.

"Cooper's lack of airway required that we leave him attached to his mom via the umbilical cord as long as possible," said Thompson. "The EXIT procedure allowed us to remove the fluid from his lungs and perform a tracheostomy prior to his full delivery."

After remaining in Le Bonheur's NICU for 324 days,

Cooper Kilburn:

Laryngeal agenesis

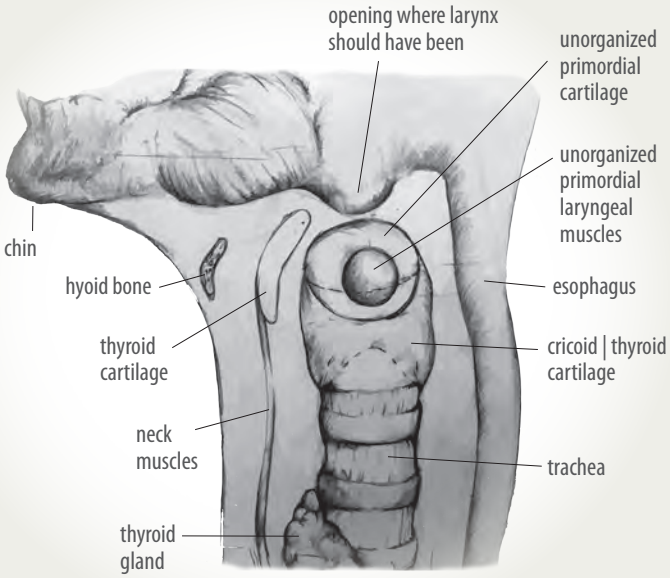
Laryngeal agenesis occurs when larynx development is halted in utero leading to no larynx or airway. The reasons for this cessation of growth are currently unknown. Children can also be born with partial laryngeal structures with the condition known as laryngeal atresia. The types of laryngeal atresia correspond with where in embryonic development the larynx growth was halted.

The facts

- Larynx development begins in the fourth week of fetal growth. The windpipe starts as a solid tube and hollows out from the top and bottom to create an airway.
- About 50 documented cases of total laryngeal agenesis in history exist.
- Most fetuses do not survive to term due to lack of diagnosis.
- Diagnosis depends on the discovery of indirect results of laryngeal agenesis – such as Cooper's fluid-filled lungs. The lack of airway is rarely identified in fetal screenings.
- Survival depends on immediate diagnosis in utero and a rapid tracheostomy at birth.

Valcamonico, A. V., Goncalves, L. F., & Jeanty, P. (1992, August 17). Larynx, atresia. Retrieved July 16, 2019, from The Fetus.net website: https://sonoworld.com/TheFetus/page.aspx?id=401

Anterior Neck View



Cooper's anatomy before surgery

Cooper had beaten the odds. But it was just the beginning of the road for the Kilburn family. Even at home Cooper remained dependent on a trach at all times and a ventilator at night. If his trach clogged, he would not survive. The Kilburns knew that Cooper's chances for mortality increased every year that he remained on the trach.

Cooper needed an airway.

A REVOLUTIONARY SURGERY

Two years and eight surgeries later, the Kilburns were back at Le Bonheur. This time they saw light at the end of the tunnel.

"Over the first two years of his life, Cooper had done remarkably well and continued to follow normal development milestones," said Thompson. "Our goal for surgery was to create an anatomical airway, get him off the trach and, possibly, give him a voice."

To date, there have only been a handful of documented attempts to create an anatomical airway from scratch – all of which have been unsuccessful. If Cooper's surgery was a success, he would be the first recorded child born with total laryngeal agenesis to have reconstruction of an airway and a larynx. During Cooper's previous procedures, Thompson had seen embryological vocal cords and knew Cooper still had a

"We planned for this surgery from day one of Cooper's life. Everyone from the nurses to surgery technicians to the neonatal team rose to the challenge and worked together for this surgery to be a success."

Otolaryngologist Jerome Thompson, MD

Otolaryngologist Jerome Thompson, MD, celebrates after removing Cooper Kilburn's airway stent and confirming successful airway construction.

Creating an airway

Rib removal

• Pediatric Surgeon Ying Zhuge, MD, harvests two ribs – one from each side of Cooper's body. These ribs will be used to construct the voice box.

Modeling of the ribs

• While Zhuge closes the incision in Cooper's abdomen, Otolaryngologist Jerome Thompson, MD, begins shaping the ribs to be the appropriate size and shape for the front and back of the voice box.

First incision

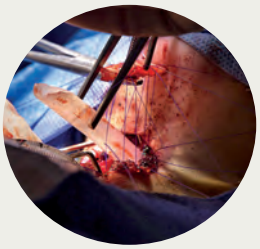
• Thompson takes over the surgery and makes the first incisions to access the cartilage structures in Cooper's throat. He discovers a rock-hard triangular-shaped cartilage structure where the airway should be.

Marking needles inserted

• Because of the solid nature of the cartilage in Cooper's throat, Thompson passes a needle through the thyroid lamina, with the help of Otolaryngologist Jennifer McLevy, MD, and a video camera, to find the midline of the cartilage between Cooper's vocal cords. He marks the space for the new airway.

Posterior graft placement

• After making an incision down to the esophageal muscles, Thompson inserts the first rib to form the back wall of the voice box. The first rib is secured with 20 stitches smaller than a human hair.



Stent placement

• A stent is placed in Cooper's throat to prevent cartilage movement during the healing process. The hope is that the stent allows a mucous membrane to heal over cartilaginous walls to create a mucosalized airway. The stent remains in place for six weeks.

Anterior graft placement

• The final rib is put into place to form the anterior wall of the voice box. This is secured with stitches.

chance to make sounds.

Cooper's surgery and care was a collaborative effort between various divisions of Le Bonheur, including ENT, neonatology, pulmonology, pediatric surgery and the ICU.

"We planned for this surgery from day one of Cooper's life," said Thompson. "Everyone from the nurses to surgery technicians to the neonatal team rose to the challenge and worked together for this surgery to be a success."

Thompson and his team had confidence that they could create an airway for Cooper because they had done it before – just not to this extent. Previous airway reconstruction patients at Le Bonheur had some sort of laryngeal structures, but Cooper had none. Cooper's health improvement had plateaued. The time for the surgery was now.

The four-hour surgery was conducted in tandem by Pediatric Surgeon Ying Zhuge, MD, and Thompson. Zhuge removed two of Cooper's cartilaginous ribs to use to construct the voice box.

Subsequently, Thompson made his first incisions to uncover the abnormalities in Cooper's laryngeal anatomy — a "conglomerate of embryological mistakes," according to Thompson. Where there should have been hollowed out spaces, there were round marbles of cartilage. Underdeveloped muscles and cartilage were in all the wrong places.

While his thyroid cartilage was normal, his

Six weeks after airway reconstruction, Thompson tests to see if Cooper is able to breathe through his new airway by simply holding a string in front of his nose and watching for movement.

cricoid, a signet-ring shaped piece of cartilage below the thyroid cartilage, was unlike anything Thompson had ever seen. A rock-hard triangular-shaped structure sat where his windpipe should have been. Through this rock-hard cartilage, Thompson and his associate, Otolaryngologist Jennifer McLevy, MD, used needles and a camera to establish a midline for the airway between Cooper's vocal cords.

"He saved our son's life and even gave him a voice we didn't think he would ever have."

Brooke Kilburn

Thompson then used the ribs to create the front and back walls of a voice box, thus establishing a new airway. The surgery ended with a stent inserted to hold everything in place while mucous membranes grew around the newly inserted ribs.

The next step was the hardest — waiting six weeks for the stent to be removed to see if Cooper would have an airway.

COOPER'S FUTURE

Six weeks later, Thompson removed the stent to find what he had been hoping for — Cooper now had a new anatomical airway. During the six weeks that the stent was in place, a fibrous envelope had grown around the grafted ribs, allowing the airway to retain its structure when the stent was removed.

At a recent follow-up appointment, a small piece of string determined whether or not Cooper's airway was viable. The smallest movement of the string as Thompson held it to Cooper's nose showed that, yes, the surgery had been a success and Cooper was breathing through his new airway.

Cooper's journey is ongoing. He undergoes a laser treatment every two weeks to remove scar tissue from the new airway and still has surgeries planned for the future to reconstruct his epiglottis so he can eat safely.

But the benefits of the surgery are already evident. While only the size of a pinky finger, this new airway gives Cooper a better chance at life. Brooke and Brad no longer have to worry about Cooper's trach plugging. He has a new airway.

And he has even started making sounds for the first time in his life.

"We can't thank Dr. Thompson and Le Bonheur enough for what they have done for Cooper," said Brooke. "He saved our son's life and even gave him a voice we didn't think he would ever have."

"Le Bonheur's greatest strength is its culture – it's inspiring to watch this team commit themselves to improving the health of children and their families."

Michael Wiggins, MBA, FACHE
President,
Le Bonheur Children's Hospital

PROFILE: MICHAEL WIGGINS, MBA, FACHE

New Le Bonheur president looks to continue momentum, expand relationships

Michael Wiggins had watched Le Bonheur Children's from afar – he'd seen it grow and earn a reputation as a great children's hospital. For more than a decade, the hospital built premier programs, recruited top-notch specialists and continued that momentum.

With that foundation laid, Wiggins jumped at the chance to help lead Le Bonheur when then President and CEO Meri Armour announced her retirement late last year. Wiggins was named the hospital's new president in April.

"I already knew that I was joining a successful team," Wiggins said. "What I learned when I arrived is that Le Bonheur's greatest strength is its culture — it's inspiring to watch this team commit themselves to improving the health of children and their families."

Wiggins comes to Le Bonheur from Children's Health in Dallas, where he served as senior vice president of Clinical Operations and the Northern Market and administrator for Children's Medical Center in Plano, Texas. Before that, he served as vice president of operations at Children's of Alabama in Birmingham.

Wiggins says he's focused on continuing the foundational momentum that's already been laid and is focused on what he calls the four pillars of an elite children's hospital: maximizing the health status of all children, without regard to their economic status or demographics; providing the highest quality and safest care; training the next generation of providers and scientists; and investing in research to advance care.

He's also interested in focusing on creative opportunities to engage children within the community and expand relationships with children and families — whether that be through community partnerships, telemedicine or networking with other hospitals.

> ***"We want to be engaged in every aspect of this community — always finding better ways to improve children's health. The greatest risk we have is being satisfied with our success."***
>
> Michael Wiggins, MBA, FACHE

"I want us to be in relationships with children and families before they know that they need us," Wiggins said. "We want to be engaged in every aspect of this community — always finding better ways to improve children's health. The greatest risk we have is being satisfied with our success."

As he settles into Memphis, Wiggins is intent on meeting as many people as he can and learning more about the Memphis region and its complex needs surrounding children. He's also working to settle his family — his wife, Robin, and three school-aged children — into their new city.

"I'll spend the summer learning how I can help represent Le Bonheur and engage people to invest their time and talents in improving the lives of children," he said.

Michael Wiggins, MBA, FACHE

Education and Training
University of Alabama at Birmingham – Bachelor of Science
University of Alabama at Birmingham – Master of Business Administration

Honors
American College of Healthcare Executives – Fellow
Beta Gamma Sigma

Work Experience
Sr. Vice President of System Clinical Operations, Children's Health, Dallas, Texas
Administrator, Children's Medical Center Plano and Northern Market Executive, Children's Health, Dallas, Texas
Vice President, Operations, Children's of Alabama, Birmingham, Ala.

Hitting the Right Note

Music therapist brings healing, encouragement to patients and families

Two-year-old Zecari Taylor spent more than 200 days waiting for a heart transplant at Le Bonheur. Those long days were filled with needle sticks, procedures and an endless stream of doctors. And through it all, only one person could make Zecari perk up just by entering the room: Music Therapist, Taylor Brown, MT-BC, NICU MT.

"Zecari loves music and loves to sing," said Zecari's dad, Christopher. "Taylor helped her a lot – she helped her recover faster."

As Le Bonheur's music therapist, Brown is part of the 31-member Child Life team who helps children cope with the difficulties of being in the hospital. Through the use of musical therapy, Le Bonheur is able to provide distractions, encourage development and reduce costs across the hospital.

BENEFITS FOR BABIES

Music therapy is the clinical- and evidence-based use of music by a credentialed music therapist to help patients reach individualized goals during their hospital stay.

Neonatologist Mark Weems, MD, is one of several physicians who regularly employs music therapy techniques with his patients. He has seen firsthand the measurable medical benefits that music therapy provides babies in the Neonatal Intensive Care Unit (NICU) – the area of the hospital in which Brown spends the majority of her time.

"Music therapy during stressful procedures in the NICU helps our babies maintain lower heart rates and higher oxygen saturation," said Weems. "The babies are less stressed and more able to tolerate the painful procedures they must frequently undergo."

In addition to assisting with procedures, Brown works with parents to teach increased communication and interaction with their child, whether singing their favorite songs or educating parents on noise levels and infant hearing.

"Parents are able to be involved as much as they want," said Brown. "They get to be a secondary 'patient' and participate in music therapy or just sit back and watch their child be a kid again."

"Music therapy during stressful procedures in the NICU helps our babies maintain lower heart rates and higher oxygen saturation. The babies are less stressed and more able to tolerate the painful procedures they must frequently undergo."

Neonatologist Mark Weems, MD

One technique Brown teaches parents is multimodal neurologic enhancement – progressive muscle relaxation used for auditory, tactile and vestibular stimulation. This process uses singing, humming, touching and rocking for babies to acclimate to the stimulation of the world and create a bonding experience for parents.

Research from the *Journal of Pediatric Nursing** shows music therapy is also a cost-saving measure. Providing a calm and soothing environment reduces the amount of patients who have to be sedated to undergo procedures such as CT scans, ECHOs, IV starts and EEGs. Hospitals can save time on

*DeLoach Walworth, Darcy. Procedural-Support Music Therapy in the Healthcare Setting: A Cost–Effectiveness Analysis. *Journal of Pediatric Nursing*: Nursing Care of Children and Families, Volume 20, Issue 4, 276 – 284.

How music therapy is used

Decreasing pain perception

Music therapists are part of the hospital pain team that works on long-term pain management for patients. It can reduce pain perception by providing distraction, focusing on relaxation and lifting spirits.

Promoting development

Patients can work on a range of developmental skills. Shakers help develop motor skills. Drums encourage exploring new textures. Singing develops communication and verbalization skills and helps with cognitive development.

Distraction during procedures

For NICU babies, distraction is an essential component of a successful procedure. Music can provide a soothing atmosphere and a reduction in pain perception.

Normalizing life in the NICU

Music helps children acclimate to the world around them especially if they have been in the NICU since birth. Babies are introduced to pleasant sounds instead of the beeping and machine noise they have been accustomed to hearing.

sedation medication and cost of staff presence.

Weems commonly uses music therapy during routine infant retinal eye exams. This process can be stressful, leading to increased oxygen requirements, periods of exaggerated mood change and frequent apneic episodes after the exam.

But since Brown has begun implementing music therapy during these exams, Weems has seen measurable improvements among the tiniest babies in the unit.

"Music therapy during the eye exam seems to help these fragile infants tolerate the exam and remain stable afterward," said Weems.

A UNIQUE SKILL SET

Music therapy encompasses a unique combination of disciplines: psychology, counseling, medicine and music. Musical ability and a love of music is key, but a pediatric medical music therapist must also be an expert in childhood development, hospital procedures and the medical field, according to the professional requirements delineated by the American Music Therapy Association.

For Brown, the intersection of these aspects was the perfect fit. She received her undergraduate degree in music and psychology and subsequently completed an equivalency program in music therapy. She finds that the results of music therapy are a thrill to watch.

"Music therapy so beautifully combines all of my passions rolled into a profession – psychology, helping people, counseling and music," said Brown. "Seeing a baby with a rough experience from day one give the faintest smile in response to music makes what I do worth it every time."

FUTURE IMPACT

Brown currently focuses her time in the NICU and consults with patients elsewhere in the hospital who could benefit from music therapy. But she has hopes to further expand the music therapy program at Le Bonheur as part of a growing Child Life team led by Director of Child Life, Jessica Kellough Liles, CCLS.

"Taylor is able to connect the power of music with

research and science to support even the tiniest patients in our care," said Liles. "We hope to continue to grow this program so more patients and families can receive the services provided by a board-certified music therapist."

In the NICU, Brown's work with the babies in Weems' care lays the foundation for a positive future impact on a child beyond the temporary reduction of stress and anxiety.

"Music therapy has the potential to improve long-term neurodevelopmental outcomes," said Weems. "While stressful and painful procedures are usually necessary for babies in the NICU, frequent pain and chronic stress has a negative impact on long-term neurodevelopment. By reducing stress in the neonatal period with music therapy, we can positively impact the child for years to come."

Music therapy helped Zecari recover from a heart transplant

Zecari Taylor hardly knew life outside of a hospital for her first two years. Born with hypoplastic left heart syndrome, she was brought to Le Bonheur immediately after her birth.

After several failed rounds of medical therapy, Zecari was added to the heart transplant list.

"Before her transplant, she was very sick," said Zecari's dad, Christopher. "She just wanted to lay down and rest."

But thanks to Le Bonheur Music Therapist Taylor Brown, Zecari's hospital stay was eased, and she was able to heal faster.

"Music played an essential role in calming her nerves and anxiety," Brown said. "Through various music therapy techniques, we were able to make sure that she didn't fall too far behind developmentally."

Brown worked with Zecari on verbalization and communication using songs to teach concepts like animals and colors, as well as transitioning between activities and following directions.

Zecari waited 200 days at Le Bonheur, finally receiving her new heart at 18 months old. Music therapy continued to play a role in her hospital stay post-transplant as Brown was able to sing the songs that Zecari learned before her transplant. This familiar music provided a sense of normalcy and continuity when Zecari was in pain in a new unit surrounded by new people.

Zecari started changing immediately after surgery. She used to be a sick kid who needed constant rest. Now she is very playful, eating well and even learning to get along with her siblings.

Now 2 years old, Zecari still delights in seeing Brown when she comes in for her regular checkups. For Zecari and her family, music was the language that helped Zecari heal and become a kid again.

A NEW STANDARD

Pediatric surgeon develops first comprehensive guideline for management of pilonidal disease

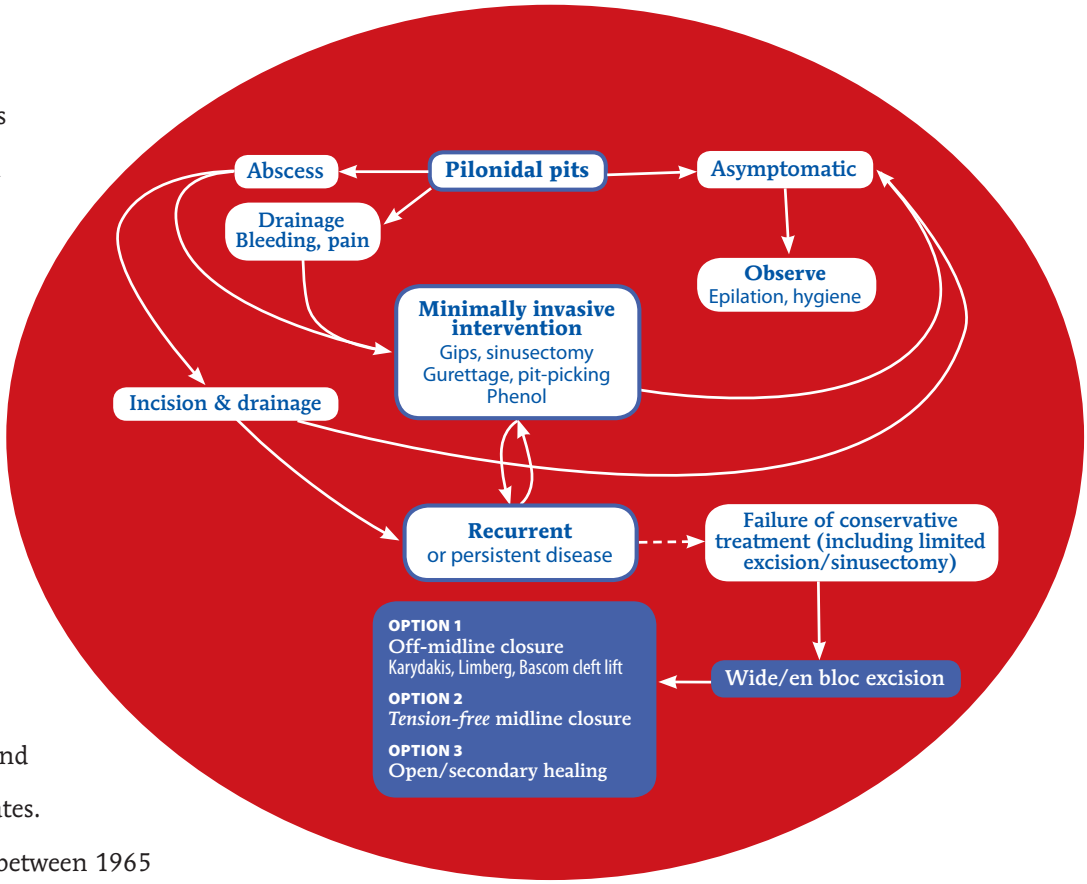
Le Bonheur Pediatric Surgeon Tim Jancelewicz, MD, recently conducted a systematic review of papers addressing the management of pilonidal disease, showing that less invasive management should be first-line treatment, and wide excision should no longer be the management of choice except possibly in the case of recurrent or chronic disease.

The research was published in *Journal of Pediatric Surgery* on behalf of the American Pediatric Surgical Association Outcomes Committee. The aim of the study was to develop clinical practice guidelines for surgeons regarding the management of the disease based on the best available evidence.

"To date there have been no comprehensive guidelines for the management of pilonidal disease," said Jancelewicz. "With these guidelines, we can give the best care up front in order to minimize the risk of recurrence, avoid maiming operations and minimize the patient's time away from school or work."

Pilonidal disease (PD) is a common and often debilitating infectious and inflammatory condition of the gluteal cleft and sacrococcygeal region characterized by sinus

and abscess formation. Recurrence is common, and the peak ages affected by the disease are between 15 and 22 years. For decades the standard definitive care has been excision with either secondary healing or primary closure of the wound, but recurrence is very common. PD is painful, debilitating and embarrassing for adolescents and young adults. The ideal approach to treatment would minimize pain and suffering and have low recurrence rates.



Pediatric Surgeon Tim Jancelewicz, MD, developed the clinical practice guidelines above for the best management of pilonidal disease.

The study reviewed 193 articles between 1965 and June 2017 that addressed the operative or non-operative management of PD. The review excluded cases before 1964, low quality and irrelevant articles. The following questions were asked during the systematic review:

1. **Are non-operative and minimally invasive management strategies effective treatment for pilonidal disease?**
2. **What are the indications, contraindications and associated complications with various operative techniques for pilonidal disease?**
3. **What operative and non-operative approaches for the treatment of pilonidal disease are associated with the highest quality-of-life and patient satisfaction?**

Various methods of intervention for PD were reviewed. Non-operative procedures include standard hair removal and hygiene, laser hair removal, phenolic acid and fibrin glue. Minimal surgery techniques included incision and drainage, minimally invasive pilonidal excision (MIPE), sinusectomy and endoscopic approaches. Finally, the study reviewed the use of excision or removal of affected tissue with en bloc resection. This has been the most common definitive surgical treatment of PD for decades.

The review identified that there has been a clear trend towards less invasive management because of unacceptably high failure rates seen after traditional wide excision and patient preference of less invasive procedures. The study created a management algorithm for PD that recommends minimally invasive intervention unless PD is recurrent or chronic.

"It is crucial to have guidelines for the pediatric surgeon because we often see pilonidal patients at their first presentation," said Jancelewicz. "Minimally invasive pilonidal excision should be used as a first approach – it is safe, effective and much easier for both the patient and surgeon."

Calming Measures

Le Bonheur develops new protocol for children with sensory sensitivities

Bright lights and beeping monitors. Unfamiliar faces clothed in caps and masks and gloves, holding syringes and sutures and strong-smelling wipes.

Hospitalization can cause sensory overload for anyone, but for a child with autism, the clinical setting can make treatment seemingly impossible.

That's why Le Bonheur Children's Hospital is working to create a more therapeutic environment for sensory sensitive patients, a trend on the rise in hospitals across the country. Last year, Le Bonheur launched a quality improvement project to improve experiences for patients on the autism spectrum. Hospitalist Medical Director Cynthia Cross, MD, a physician champion for the project, said the initiative is essential for patient safety.

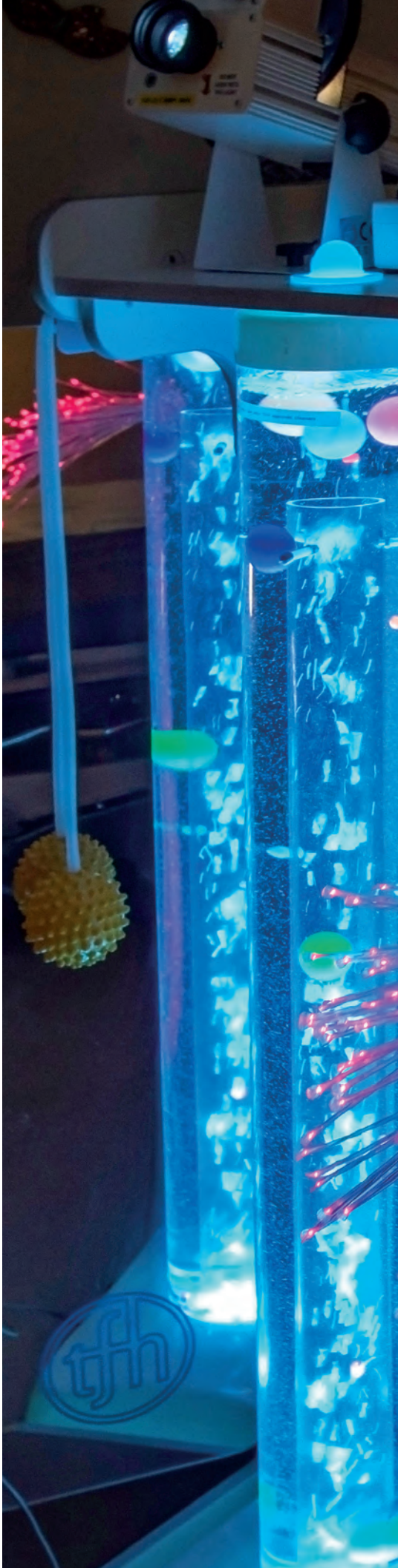
"You can't always tell by looking at a patient that they have special needs," said Cross. "While they may appear on the surface not to have special needs, there are underlying features for individuals on the autism spectrum that affect care greatly. The consequences of not paying attention to those details can lead to major reactions."

In fact, a review of incident reports was a significant catalyst for this change, said project manager Marley Lemons.

"We noticed that many of the occurrences we were having involved patients with documented triggers, but they were buried in the medical record and not addressed until we were in service recovery mode," said Lemons. "We talked with other hospitals about best practices and put a team together to identify how we can intervene to prevent those situations."

Sensory Friendly Spaces

Toward the end of May, Le Bonheur EEG Tech Ashly Smith entered 14-year-old Kaleb Bishop's room to attach electrodes to his head in preparation for an electroencephalogram. In addition to her usual supplies, she wheeled in a new machine designed to create calm. The mobile station is one of three available in the hospital and part of a sensory "toolkit" located on the floor. The Vecta mobile sensory station is armed with aromatherapy, calming music, an illuminated bubble column, mirrors, a wall projector and weighted light strands.



Child Life Specialist Elissa Gargivlo prepares the Vecta mobile sensory station for a patient.

As Ashly worked, Kaleb focused on the sensory station, changing the colors of the bubble column with the simple push of a button. He watched as ping pong balls shot up the tube and fell gently back down. Then his eyes closed.

"Makes me sleepy," he said. "It's good."

Nursing leaders in Le Bonheur's Neuroscience Institute presented their implementation of the sensory toolkit at this year's American Association of Neurological Surgeons (AANS) scientific meeting.

Key to their success, said Iva Scroggins, MSN, CNRN, RN-BC, was buy-in from staff to embrace the tools as helpful options rather than added responsibility.

"From a staff perspective, I think our nurses feel more empowered and supported," she said. "We have a challenging population. Many children with neurological conditions have problems with touch, noises and change in environment, and usually the first things we do when a patient comes to the epilepsy monitoring unit are blood tests, an MRI and hookup for an EEG."

To help combat the whirlwind of confusion admission to the EMU can create, the unit collaborated with the hospital's Child Life Department to develop a script to screen for sensory-sensitive patients prior to arrival.

This type of screening has now been piloted for inpatient gastroenterology admissions. Also piloted were care plan discussions each morning during team huddles for patients identified with autism spectrum disorder (ASD). After the huddle, nurses were asked to complete a form containing interventional information for the patient's chart. They also placed a sticker on the patient's door to increase staff awareness.

These measures are now under review by the team tasked with scaling up to the hospital level with a goal for implementation in 2020. Forms are being customized according to departmental needs and in the process of becoming electronic for easier access.

A questionnaire for parents is currently in front of the hospital's Family Partners Council for suggestions to help personalize responses for patients with specific sensitivities.

Families as Partners

For Le Bonheur mom Brittany Schwaigert, communication prior to arrival signifies an enormous improvement in patient experience. Brittany's son, Greyson, has tuberous sclerosis complex (TSC) with a secondary diagnosis of autism. The family comes to Le Bonheur for MRI scans routinely, and until recently, Brittany struggled with consistency in clinicians'

adherence to Greyson's special needs.

"For parents of autistic children, studies have shown that the stress level associated with coming to the hospital can be comparable to that of a front-line soldier," she said. "Preparation from the hospital can alleviate a lot of that stress for both parents and kids prior to their appointment."

Today, about one in 59 children has been identified with ASD according to estimates from the Centers for Disease Control and Prevention's Autism and Developmental Disabilities Monitoring (ADDM) Network. ASD commonly co-occurs with other developmental, psychiatric, neurologic, chromosomal and genetic diagnoses. The co-occurrence of one or more non-ASD developmental diagnoses is 83%. The co-occurrence of one or more psychiatric diagnoses is 10%.

"The question of, 'Is autism more common now or are we better at diagnosing it?' I'm not sure the answer has been fully elucidated," said Cross. "But the fact is that we have more patients coming into the hospital identified as having autism or being somewhere on the spectrum."

Many children with ASD have trouble with sensory regulation; the way they experience sights, sounds and touch can be different from others in similar situations. A child with ASD may respond negatively to bright lights or loud noises. Inversely, they may not respond readily to sound, touch or pain.

Knowing a child's potential challenges can make a world of difference during treatment. For example, Brittany's son Greyson has issues with intravenous sedation, specifically the uncomfortable sensation of having a needle in his arm for sustained periods of time. Greyson's instinct is to jerk the needle out of his arm, and he has had the unfortunate experience of an hours-long, failed IV sedation attempt.

Now, Greyson's chart specifies that he is unique in needing mask sedation as his first option before an MRI.

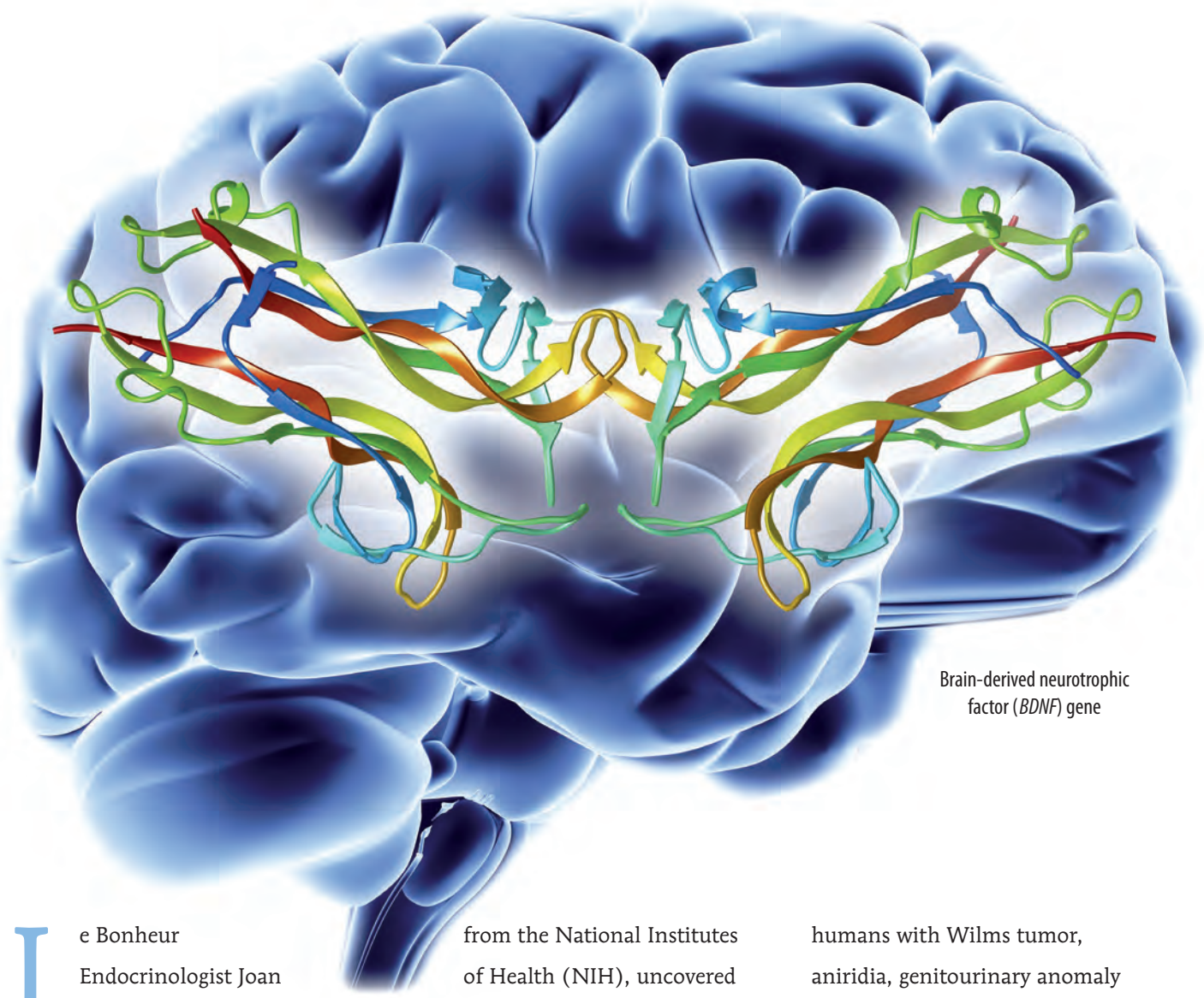
"We've realized that individualized care plans are best," said Le Bonheur Nurse Jennifer Cummings, RN, a project team leader involved in the pilot project. "Before, there was so much guesswork in how we tried to take care of these kids, but now our families can let us know what works."

The process, said Cross, will continue to evolve.

"To formally create this proactive process is the outstanding part of all this," said Cross. "Families can see that we have prepared for their loved ones and have confidence in the care that we give. It's just the right thing to do."

UNLOCKING A CRUCIAL PAIN GENE

Le Bonheur researcher uncovers the effects of BDNF *gene deletion on pain sensitivity*



Brain-derived neurotrophic factor (*BDNF*) gene

Le Bonheur Endocrinologist Joan Han, MD, recently published an article in *Pain Journal* exploring how haploinsufficiency of the brain-derived neurotrophic factor (BDNF) gene effects pain sensitivity. Her research of the BDNF gene, funded by a grant from the National Institutes of Health (NIH), uncovered insights into pain circuit function and future strategies for pain control.

Han and her team conducted several tests to explore the effects of BDNF gene loss. First, they investigated pain sensitivity in humans with Wilms tumor, aniridia, genitourinary anomaly and range of intellectual disabilities (WAGR) syndrome. These children have variably sized heterozygous deletion of the 11p13 region.

Out of the 12 patients with WAGR who completed sensory testing, six had heterozygous

deletion (+/-) of BDNF. They first analyzed parental reports about their child's pain sensitivity. Parents of BDNF +/- subjects were significantly more likely to provide descriptions of their child having pain insensitivity.

"With the BDNF deletion there is an impairment in nociception or an increased pain tolerance," said Han. "However, there is not a complete lack of pain sensation as observed in other gene mutations."

Subjects also underwent quantitative sensory testing over a range of hot and cold stimuli. Again, BDNF +/- WAGR subjects rated these stimuli as significantly less painful confirming the hypothesis that BDNF hemizygosity may lead to impairment in nociceptive processing. In motor and sensory nerve conduction studies, results were similar between subjects with and without BDNF deletion, indicating intact peripheral signaling and suggesting that central nervous system processing of pain signals may be what is disrupted with BDNF loss.

Secondly, Han conducted nociceptive testing on rats in order to further assess the role of Bdnf haploinsufficiency. The rats were exposed to various hot and cold stimuli including noxious heat, cold plate and infrared diode laser and observed for behavioral assessment. The Bdnf +/- rats exhibited a significantly longer latency to withdraw from each of the stimuli.

"Our data suggests that BDNF may be a part of a larger gene network that modulates the complex trait of pain sensitivity in the general population," said Han.

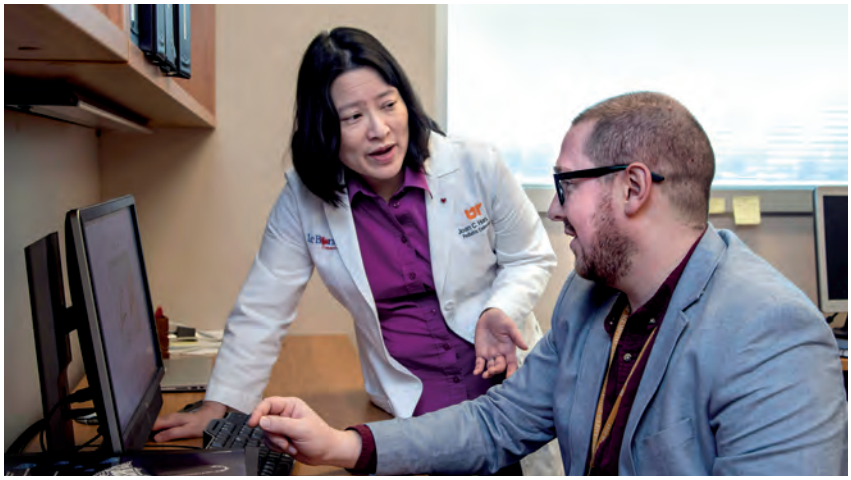
"BDNF as a pain modulatory gene is significant because pain drives individuals to seek medical intervention. Diagnosis and treatment of painful medical emergencies can be delayed in patients with pain insensitivity."

Le Bonheur Researcher Joan Han, MD

The results of the study show that BDNF haploinsufficiency produces an elevated pain threshold, therefore suggesting a system where transmission in peripheral afferent nociceptors is still intact. However, the synaptic transmission of these signals to spinal second order neurons or higher CNS regions is potentially disrupted requiring stronger stimuli to elicit responses.

"BDNF as a pain modulatory gene is significant because pain drives individuals to seek medical intervention," said Han. "Diagnosis and treatment of painful medical emergencies can be delayed in patients with pain insensitivity."

Han and her team posit that reduced BDNF signaling causes a deficit in processing incoming painful sensations. Further research hopes to uncover the larger gene network in the trait of pain sensitivity as well as apply these findings to potential analgesic approaches.



Joan Han, MD, (left) and her team have uncovered insights into pain circuits that can lead to future pain control stategies.

THE WORD GAP

Le Bonheur partners with Seeding Success, University of Memphis and Urban Child Institute to increase parent-child talk time

Early talk with babies is the strongest predictor of cognitive development and school readiness, and the most important period of time for a baby's brain development is before the 3rd birthday. But in some lower socioeconomic families, children may hear as many as 30 million fewer words than children from more advantaged backgrounds, eventually resulting in an academic achievement gap.

In an effort to close the word gap in children between 0 and 30 months, Le Bonheur has partnered with local nonprofit Seeding Success, the University of Memphis and the Urban Child Institute to implement programs from the LENA Foundation. One program, LENA Start, is a small group program for parents offered through Le Bonheur's Family Resilience Initiative that aims to educate parents on the importance of early, quality talk to promote brain development.

Le Bonheur's Family Resilience Initiative began its first LENA Start group in Memphis in Fall 2018. The LENA Start program combines three elements to help parents build brain development in babies. First, the LENA system, a "talk pedometer," measures how much parents are talking and provides ways for parents to increase words and conversational turns. Second, weekly parent group meetings are held for 13 weeks, and parents receive reports of their progress. Finally, each family receives a shared reading book and materials to take home and practice simple talking tips and techniques learned during each session.

This LENA Start group had six families graduate from the program with an 80% increase in conversational turns.

"The LENA Start program creates a supportive

Parents in LENA Start receive children's books to encourage interaction and talk time with their children.

place where parents feel connected," said Lisa Rogers, manager of the Family Resilience Initiative. "This exceeded our expectations in terms of the level of commitment from parents as well as the increase in relationships and rapport."

Participants in the LENA Start program will continue to have follow-up and ongoing observation for several years. For six months after the program, parents receive text messages with talking tips, resources and other ways of increasing talk time. Seeding Success will be able to view and track how these children fare in kindergarten through a data sharing agreement with the local school districts.

In addition, parents from the original group have expressed interest in continuing support groups. "We are always excited to see organizations look for sustainable and innovative ways to support parent engagement outside of programs like LENA Start," said LaDora Watkins, network specialist with Seeding Success. "This group wanted to continue meeting, so the Family Resilience Initiative leadership took the initiative to find other ways to continue helping these parents work with their children."

Preliminary results from this group demonstrate the positive changes enacted by the program. Eighty percent of those in the group increased conversational turns and interactive talk between parents and children, and 75% of families showed gains in total scores including reading, inclusion in routines and lowered parental stress. Nationwide, the LENA Start program has shown that children whose parents participated in the program are gaining nearly two months of developmental skill every month.

LENA Start is one of three programs the LENA Foundation is using to close the word gap and thereby improve school readiness. LENA Home is a one-on-one program where family support workers visit families on a weekly basis to focus on early language development. Le Bonheur implemented LENA Home into its existing Maternal Child home visitation programs in February 2019. The final piece is LENA Grow which focuses on early childhood educators and day care centers increasing talk interaction with the children in their care.

Briefs

McCullers adds new UTHSC responsibility

Jon McCullers, MD, chair of Pediatrics and Pediatrician-in-Chief at Le Bonheur recently took on the role of Interim Senior Executive Associate Dean of Clinical Affairs and Chief Operating Officer with University of Tennessee Health Science Center (UTHSC). In this new role, McCullers is charged with supporting the alignment of the clinical, education and research missions within the College of Medicine and with hospital partners. He will provide oversight of academic practice plans and associated affiliation agreements. He will also promote the growth of service lines and academic clinical practice. McCullers retains his positions as chair of Pediatrics and Pediatrician-in-Chief at Le Bonheur.

Jon McCullers, MD

Heart Institute announces two-story, $37.6-million expansion

Le Bonheur will expand its Heart Institute by adding 19 beds and creating a 31-bed dedicated cardiovascular unit. The expansion will add 10 additional Cardiovascular Intensive Care Unit beds and create room for an 11-bed step-down cardiac unit. A new MRI-guided hybrid catheterization lab will be added for a total of three catheterization labs.

Operating room expansion opens to patients

Le Bonheur's recent expansion includes the addition of four new operating rooms with space to add three more and is part of a $16-million expansion taking place on the east side of the hospital. The addition addresses the growing need for pediatric surgical services at Le Bonheur. The expansion includes renovated central sterile storage, now adjacent to the new operating rooms, allowing for better workflow and efficiency.

Le Bonheur's Fetal Center receives ultrasound accreditation

The Fetal Center recently was recognized by the AIUM Ultrasound Practice Accreditation Council as an Accredited Ultrasound Practice in the areas of Obstetric – First Trimester, Obstetric – Second Trimester and Obstetric – Third Trimester.

Neuroscience Institute hosts 13th annual Pediatric Neurology Symposium

This spring, more than 100 neurologists and neurology providers from around the country attended this year's Pediatric Neurology Symposium learning about the state-of-the-art practices and trends in treating pediatric neurology patients. Kim Meador, MD, was the recipient of this year's Kayden R. Vinson Distinguished Scholar Award and Lecture. Meador is clinical director of Stanford Comprehensive Epilepsy Center in Palo Alto, Calif.

International PDA Symposium provides live case for cardiologists, cardiac surgeons

Le Bonheur's Heart Institute recently hosted the multispecialty International PDA Symposium. Special focus was on transcatheter PDA closure in extremely low birth weight, premature infants. The program featured panel discussions, review of literature in multiple subspecialties, hands-on echocardiogram and catheter workshops and a PDA closure case streamed live to the symposium.

Professor emeritus receives gold medal from NASCI

UTHSC Professor Emeritus of Radiology and Pediatrics Ina Tonkin, MD, recently received the gold medal from the North American Society for Cardiovascular Imaging (NASCI) for her work in congenital heart disease. Prior to her retirement, Tonkin worked at Le Bonheur for more than 30 years with primary interests in heart diseases and interventional radiology.

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Le Bonheur named a 'Best Children's Hospital' by *U.S. News & World Report* for ninth consecutive year

Le Bonheur is listed as a "Best Children's Hospital" by *U.S. News & World Report* for the ninth consecutive year, according to the list released last month. The hospital ranked in eight specialties, earning a Top 10 honor in Cardiology and Heart Surgery. "Our physicians, nurses and associates are consistently dedicated to providing the best care and outcomes for every child who enters our doors," said Le Bonheur President Michael Wiggins. "This honor recognizes our commitment to children and their families, and we are incredibly proud to be named a 'Best Children's Hospital' for the ninth year in a row."

Le Bonheur is ranked in cardiology and heart surgery, gastroenterology and GI surgery, neonatology, neurology and neurosurgery, nephrology, orthopedics, pulmonology and urology.