Experience and expertise

A family finds the compassionate care they needed at the Pediatric Heart Center—and a legacy of innovation behind it

Marissa Wooten’s maternal instincts kicked in minutes after her son was born in 2012. Unlike her two very vocal daughters before him, baby Nolan cried for just a second and then lay lethargically on her chest. Something didn’t seem right.

The delivery nurses told Wooten that some infants transition after birth differently and that everything was OK. But she noticed that Nolan had a blue tint.

“Usually babies are mad when they come out and they cry and cry. He just lay there,” Wooten says. “It was scary.”

Wooten continued to express her concerns, prompting nurses to do a pulse oximetry reading. Nolan’s oxygen level was 60 percent, far below a normal 95 to 100 percent. His care quickly became more intense. Staff intubated him and brought him by ambulance to University of Minnesota Masonic Children’s Hospital.

Nolan was admitted to the hospital’s neonatal intensive care unit and started

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undergoing tests. Doctors suspected a rare congenital heart defect called transposition of the great arteries (TGA).

Often fatal if uncorrected, TGA is a condition in which the two main arteries leaving a baby’s heart are reversed. So rather than having oxygen-rich blood flowing from the lungs to the body, it flows right back to the lungs. This leaves babies struggling to get enough oxygen, explains Rebecca Ameduri, M.D., a pediatric cardiologist at University of Minnesota Masonic Children’s Hospital who cares for Nolan.

Finding expertise

When he was 6 days old, Nolan had an open-heart operation in which surgeons switched his arteries back to normal. The medical staff kept assuring Wooten and her husband, Shawn, that TGA is the best of the serious heart defects to have, but the family still was extremely nervous. After all, a newborn’s heart is the size of a walnut, and the coronary arteries are like strands of hair, adding to the intricacy of the surgery.

Though TGA occurs only in 1 in 3,300 live births, the University’s pediatric cardiac surgeons have ample experience correcting it. The hospital treats about five to eight infants who have TGA annually, Ameduri says.

After six weeks in the hospital, the Wootens were able to take Nolan home to Plymouth, Minn.

Despite the stress of having a sick baby, two other children to care for, and a husband who works on the road as a minor league baseball coach, Wooten felt well cared for at U of M Masonic Children’s Hospital. The medical care was excellent, she says, and every time her girls came to visit Nolan, they were welcomed into family playrooms and entertained by volunteers. (In fact, the experience made such an impression on Nolan’s sister Kylee that she raised money to give 500 books to patients.)

“Obviously, nobody wants to be in the hospital in that situation. But because we had to be, it was the best experience I could imagine,” Wooten says. “Everyone was so nice and caring and answered my 7 million questions.” The nurses and physicians always were available, she adds, even checking on their patients on days off.

Nolan sees Ameduri now for annual checkups, where she monitors his arteries and overall heart health. “Children with TGA tend to do very well long term,” she notes. “Everything is connected as it should be.”

The Wootens are thrilled that Nolan can live his life as a normal, active, 3-year-old boy without medication or restrictions on his activities. It’s all thanks to his successful treatment at Masonic Children’s Hospital.

“He’s doing really well, and we’re really fortunate,” Wooten says.
Building on a legacy

Treating children like Nolan who have complex heart conditions is part of the University’s long and storied history of pediatric cardiac care. It’s been a center of innovation since the 1950s, when U surgeons performed the world’s first successful open-heart surgery on a child.

U pioneers also developed the first heart-lung machine and the first wearable, battery-operated pacemaker, and they completed the first repair of a hole in one of the heart’s chambers.

This legacy of leadership continues today. U physicians are spearheading a $13 million National Institutes of Health grant focused on identifying childhood predictors of adult cardiovascular disease—a first-of-its-kind study. In addition, researchers here are investigating insulin resistance and metabolic syndrome and their effects on children’s heart health.

This deep commitment to innovation and excellence in care are two of the reasons Anthony Azakie, M.D., C.M., wanted to join the University’s team. In February Azakie became codirector of the Pediatric Heart Center and chief of the Division of Pediatric Cardiac Surgery. He considers the University of Minnesota the birthplace of pediatric heart surgery, and he aims to continue breaking ground in improving treatments and techniques.

Some of the areas on which Azakie plans to focus are developing primary complete one-stage repairs of complex heart defects in newborns, minimally invasive surgeries, and heart valves that grow with children to decrease the need for follow-up surgeries. And with more children today thriving after heart surgeries, the University will continue to help adolescents under pediatric care transition to adult cardiac care.

“We are creating a new era of care for children who have heart defects. We are developing the premier pediatric cardiac program, where we achieve superb outcomes while advancing the field,” says Azakie. “That’s consistent with the history of the University of Minnesota.”
When Jenna Zona was born in 2013, it became immediately apparent that something was seriously wrong. She had numerous blisters, and her skin was extremely fragile.

Doctors diagnosed her with epidermolysis bullosa (EB), a devastating disorder in which a missing protein keeps skin layers from adhering to one another. The slightest friction can cause painful blisters and open wounds. They occur all over the body and internally, too, making it difficult to eat and digest food.

These injuries run the constant risk of infection and, over time, predispose the child to skin cancer. Often called “butterfly children” because of their fragile skin, those with EB live in agony.

Now 2 years old, Jenna wears extensive protective bandages, which her parents, Dwayne and Adriana Zona, must change every other day. It’s extremely painful for Jenna, and the bandages still don’t fully prevent blisters.

But Jenna is fortunate in many ways, says her grandfather, Richard Zona. Her case is less severe than some children’s, and she lives in Minnesota, one of two places in the United States where researchers are tackling EB.

Jenna is under the care of University of Minnesota Masonic Children’s Hospital’s Jakub Tolar, M.D., Ph.D., a pediatric blood and marrow transplant physician and director of the U’s Stem Cell Institute, and pediatric dermatologist Kristen Hook, M.D. After caring for numerous children who have EB, Tolar was inspired to find more effective treatments.

“I make a living treating children with leukemia and other cancers, and it’s one of the most horrific disorders I’ve ever seen,” he says.

Since 2007 Tolar and his colleagues have studied the effectiveness of bone marrow transplants in children who have the most severe types of EB—becoming the first and only hospital in the world to use this experimental strategy. For most of the kids in the clinical trial, healthy cells from donated marrow provide the missing protein and reduce internal and external blistering.

This trial and the care Jenna receives at the University motivated Richard Zona and his wife, René, to make a six-figure donation to support Tolar’s research. Zona’s commitment also inspired a six-figure donation from the TCF Foundation.

“If we can get greater awareness about EB, that’s a big plus in coming to a cure,” Zona says. “You need the research dollars to get to the therapies that will result in a cure.”

With gifts like the Zonas’, Tolar is undertaking additional research to better understand this complex disorder. He aims to find the best combination of therapies (and ultimately a cure) and to prevent cancer in EB patients, a common outcome. This research also can lend knowledge to treatment strategies for other ailments such as skin burns.

“If we didn’t have this support, it would be almost impossible to keep pace with the depth and quality of the research we need to keep bringing these new therapies to the clinic,” Tolar adds.

Such research gives families like Zona’s hope for a better prognosis for his granddaughter and other children who have EB.
A multidisciplinary team of specialists helps a girl with a challenging autoimmune condition stay in the game

For 13-year-old Josie Adams, battling the inflammatory disease dermatomyositis has meant a childhood dominated by intense medical care and hospital stays, taking long lists of medications, enduring numerous lifesaving surgeries, and contending with painful skin rashes all over her body. Diagnosed at age 2, Josie has not yet reached that sought-after remission of her dermatomyositis.

This rare autoimmune illness causes inflammation of the body’s small blood vessels, especially in the skin and muscles. It primarily presents itself as red and purple rashes on the face, elbows, hands, and knees, along with muscle weakness. More severe inflammation causes calcinosis—which can appear as small pebbles of calcium up to large, tumor-like rocks in the skin and muscle—and other life-threatening complications.

Unfortunately for Josie, her dermatomyositis is the most aggressive and treatment-resistant kind, affecting skin, muscle, and other vital organs, says her physician, Richard K. Vehe, M.D., director of pediatric rheumatology at University of Minnesota Masonic Children’s Hospital.

In addition to the rashes and rampant calcinosis it causes, Josie’s dermatomyositis almost turned fatal in 2006 when it caused a tear in her small intestine. She spent nearly seven months in the hospital, where she endured four surgeries and many other treatments.

“I think she saw every kind of specialist then,” recalls her mother, Deb Adams. “She almost died during that time.”

But Josie’s medical team saved her life, and they continue to tamp down her disease with anti-inflammatory and immune-modulating medications. That’s made it possible for Josie to do many things she loves: play hockey and lacrosse, go ice skating, and spend time with her friends.

“At the beginning, we didn’t know if she’d survive,” Vehe says. “Josie is back in the game—she goes to school, plays sports—but in no way would I paint this as being fixed. It’s a challenging condition.”

Josie, a seventh grader in Edina, feels pretty good physically, but dermatomyositis has its trials. She gets upset that she can’t wear tank tops or shorts because of her condition. Her care team at the University “helps a lot, and they understand my frustration,” she says. “They try new medicine as soon as possible if one of the other medicines isn’t working.”

Her parents, Deb and Jay Adams, can’t say enough about their daughter’s treatment. Caregivers from multiple disciplines—including endocrinology, gastroenterology, physical therapy, and pediatric rheumatology—constantly team up to care for Josie.

“The team is so supportive of our family and [Josie] living as normal a life as she can,” Deb Adams says. “Everything they’ve done means the world to us.”

The expertise of Josie Adams’ care team allows her to keep doing many of the things she loves, like playing hockey.
Kids and sleep

Why is sleep so important?
In general, we sleep for physical recovery as well as mental recovery, to be able to improve our memory, learning, attention, and mood. When you don’t have adequate sleep, you’re at risk of having problems in any of those areas.

When children are sleep-deprived, they tend to have more mood, hyperactivity, and attention problems, which can look similar to symptoms of behavioral problems like ADHD.

In addition, scientists suspect that there’s brain remodeling happening every night when we sleep. Without this remodeling, new connections in the brain would not be made as efficiently. This explains why sleep is so important for learning.

Sleep also has been found to have a detoxifying function in our brains. It appears that sleep is the only time this circulatory system gets rid of toxins.

What’s normal sleep behavior for children?
It really varies. The younger the child, the wider the range of normal for hours of sleep. But generally, as a child gets older, the need for sleep decreases.

Babies may need 12 to 16 hours of sleep in a 24-hour period. That shifts to about 10.5 hours for younger kids and then 9 hours for a teenager. Naps are also normal for children in the first years of life, with kids stopping naps sometime between 2 and 4 years of age.

Teenagers commonly have other sleep time problems, with a propensity for staying up late. The reason for this has long been thought to be behavioral, but we know now that there is a biological predisposition to have a later bedtime, which can be exacerbated by the alerting signal of light in electronic devices. This problem is common but can lead to chronic sleep deprivation with multiple health consequences, such as obesity and excessive daytime sleepiness, which can result in lower levels of school achievement, mood problems, and more car accidents.

What kinds of issues does a pediatric sleep specialist help families address?
We see families when children are having insomnia, which can be seen as having a hard time getting to bed or having night awakenings. We see children who are having dangerous sleep behaviors like sleepwalking or who are having night terrors too frequently. We take care of children who aren’t getting good quality sleep, which happens with obstructive sleep apnea or when natural sleep timing does not match school schedules, possibly resulting in insufficient sleep.

We also have expertise working with children who have other medical problems that can make sleep disorders more likely, such as neuromuscular disorders, Down syndrome, autism, anxiety, and depression.

How do you identify the cause of sleep problems?
We ask families to give us a 24-hour sleep history for the child. This usually helps us identify many sleep disorders. There is a subset of patients that may need further testing in the sleep lab, as is the case with suspected obstructive sleep apnea, nighttime hypoventilation, and hypersomnia.

To help prevent sleep difficulties, a good place to start is keeping a regular sleep schedule every day of the week—even on weekends—as well as avoiding caffeine and electronics use at night.

To make an appointment with a pediatric sleep specialist, call 612-365-6777.

A Q&A with Helena Molero-Ramirez, M.D., a University of Minnesota Masonic Children’s Hospital pediatric pulmonologist and sleep medicine specialist

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Join us for WineFest No. 21 – A Toast to Children’s Health

Friday, May 6, and Saturday, May 7, Renaissance Minneapolis Hotel, The Depot

Twenty-one. That’s how many years WineFest has been counting its lucky stars. This event supports the work of U of M Masonic Children’s Hospital’s stellar physician-researchers—dedicated not only to caring for patients but to finding new cures and treatments for their injuries and illnesses, from the common to the complex.

That’s why we’re calling this celebration Lucky No. 21. Don’t miss this chance to enjoy exceptional wine, gourmet fare, incredible auctions, and fabulous entertainment. Learn more or purchase your tickets at thewinefest.org.

(Left) WineFest No. 21’s featured art is “Objects of Desire” by Layne Kleinart.

Champions for Children Celebrity Golf Classic

Monday, June 6 • Windsong Farm Golf Club • Independence, Minn.

Join Minnesota Viking John Sullivan as he hosts the eighth annual Champions for Children Celebrity Golf Classic. Since its inception, this tournament has raised more than $750,000 for University of Minnesota Masonic Children’s Hospital’s Adopt A Room program. This year we’re driving toward $1 million raised. Visit give.umn.edu/events/champions to learn more.

A message of hope

What do you say to someone who saved your life? Cancer survivor Travis Moore (at right in photo) admits he was speechless when he reconnected with University of Minnesota Masonic Children’s Hospital’s John Wagner, M.D., in February, 20 years after he received an experimental bone marrow transplant under Wagner’s care.

Moore, who is now 30 years old and a globe-traveling fisheries scientist, made a visit to Minnesota from his Florida home to thank Wagner and other members of his care team—and to give hope to kids who are battling leukemia today.

Wagner, a renowned pioneer in blood and marrow transplantation, says the therapy that Moore received 20 years ago provided a framework for the leukemia therapies patients receive today.

Spotlight on pediatric blood and marrow transplantation

Join us Monday, April 18, at University of Minnesota Masonic Children’s Hospital for an opportunity to meet John Wagner, M.D., and learn about the world-renowned and transformative practices of the U’s pediatric blood and marrow transplant team. Through a series of medical cases, Wagner will reveal the successful formula that has led to groundbreaking advancements in treatment that will ultimately save the lives of more children.

Space is limited. RSVP online at www.rsvp.umn.edu/BMT.
To advance care for abused and neglected children in the region, Otto Bremer Trust has donated $2.5 million to the Center for Safe and Healthy Children at University of Minnesota Masonic Children’s Hospital.

It’s one of the largest donations ever made in the United States for a public health crisis that now causes more childhood deaths than cancer annually.

The Center for Safe and Healthy Children became the first-of-its-kind regional program in 2014 when it started collaborating with Hennepin County Medical Center and Children’s Hospitals and Clinics of Minnesota to provide comprehensive care to maltreated children. The new gift will ensure that kids in Minnesota and the region will get the care they need, help to train more child abuse specialists, and enhance the center’s telehealth capabilities.

A relatively new subspecialty, child abuse pediatrics focuses on diagnosing and treating those who are suspected victims of any form of child maltreatment, including physical abuse, sexual abuse, factitious illness (medical child abuse), neglect, and psychological and emotional abuse. These physicians are also trained to recognize medical conditions that are often mistaken for abuse and work closely with law enforcement, the judicial system, and community agencies advocating for child safety.

In recognition of the gift, the center has been renamed the Otto Bremer Trust Center for Safe and Healthy Children at University of Minnesota Masonic Children’s Hospital.