PEDIATRIC ENT SAVES BABY WITH A RARE SACCULAR CYST

At 5:30 in the morning, Amal Isaiah, M.D., Ph.D., answered his phone. Just that week he had been credentialed at the University of Maryland Medical Center (UMMC). A resident was on the line, in need of help. An infant, just a few hours old, had been transferred because he was breathing noisily and thrashing about in respiratory distress. Dr. Isaiah rushed to the hospital. Soon he saw an extremely rare cyst blocking Noah Schultz's airway, leading him to perform multiple surgeries seldom done in a baby.

Dr. Isaiah, an assistant professor of otorhinolaryngology–head and neck surgery at the University of Maryland School of Medicine (UMSOM), first saw Noah on the operating table. "The baby was desaturating and in pretty significant distress," he recalls. He needed oxygen, fast, but three previous attempts to place a breathing tube had failed.

Dr. Isaiah had just completed a clinical fellowship in pediatric otorhinolaryngology at the University of Texas Southwestern Medical Center and the affiliated Children's Health Dallas. Before that, he had trained as an ear, nose, and throat resident at the University of Maryland. Now he was back, inspired by two professors, chair Scott Strome, M.D., and Kevin Pereira, M.D, both of UMSOM's Department of Otorhinolaryngology–Head and Neck Surgery.

A CONGENITAL SACCULAR CYST

In the operating room, Dr. Isaiah could not identify Noah's airway. Rather, he saw something he had never seen before, a saccular cyst. "Even in tertiary centers, it's once in a lifetime," he says. Congenital saccular cysts, sometimes diagnosed after the baby's death, occur in perhaps 1 in 100,000 live births.

In Noah's case, the fluid-filled cyst, about 2 centimeters wide, started in the voice box. "Basically, Noah was breathing through a pinhole," recalls Dr. Isaiah. He weighed his options: He could try his luck with a breathing tube or perform a tracheotomy. The latter would carry a high risk of complications.

Using an endoscope, the doctor moved the cyst aside, uncovering the larynx. He quickly decompressed the cyst, which was lodged deep inside the voice box. He tried to insert a breathing tube, and it settled into place. Immediately, Noah started breathing.

"Really, somebody looked out for me, and somebody obviously looked out for the baby," Dr. Isaiah says. Two days after the surgery, he was able to remove the breathing tube. Finally, Noah was breathing on his own.

A LONG-TERM PLAN

"In medicine, we always say, 'It's subject to change," notes Dr. Isaiah. Sure enough, a few days later, the cyst reappeared. Again, he removed as much of it as possible.

As Noah recovered in the Neonatal Intensive Care Unit, Dr. Isaiah thought about the coming winter and flu season. If the cyst returned, Noah could suffer respiratory distress at home. The doctor could see only one definitive way to prevent that: a tracheostomy, a procedure uncommonly done in babies, especially during the first month of life. Hence, three weeks after removal of the second cyst, Noah underwent his third surgery, one Dr. Isaiah hoped to later undo.

As a Rhodes scholar, Dr. Isaiah had studied developmental plasticity at Oxford University in England. He knew that Noah's chances of speaking normally depended on how soon he could learn speech and language skills; however, a combination of problems with his airway kept the boy from developing those skills normally. Reversing it soon would let Noah start speech and language therapy when his brain was still developing, giving him a chance to catch up with his peers.

Yet, before the tube could come out, Dr. Isaiah had to make sure the cyst was gone for good. He waited nine months, then checked Noah's windpipe again in the OR. He explains, "The airway is a magic box; you can't assess it well until you get to the OR." There he saw only a tiny cyst, no longer obstructive, and unlikely to grow back.

A SETBACK

With the cyst out of the way, another problem became apparent: subglottic stenosis. Noah would never be able to breathe on his own through such a narrow airway. Widening it could give Noah a better life--without the tracheostomy.

KEY POINTS

- According to most reports, congenital saccular cysts occur in about 1 in 100,000 live births.
- Often, they go undiagnosed until after the infant's death.
- Noisy breathing at birth, though not always pathologic, may indicate a blocked windpipe and requires evaluation.

Just after his first birthday, Noah underwent a single-stage laryngotracheal reconstruction. The operation took several hours. Fortunately, Dr. Isaiah's training included performing many of these procedures. He knew how to make a small split into the trachea, insert a small cartilage spacer, and stitch it in.

At the same time, Dr. Isaiah removed the tracheotomy tube and closed the opening, rather than wait and do that later in a double-stage reconstruction. "I had enough confidence in his recovery that I was able to do both in one go," he says. Noah spent 10 days in the hospital, half of them in intensive care. Meanwhile, Dr. Isaiah watched for graft prolapse, infections, or other complications that could spoil Noah's recovery.

Furthermore, Noah had to learn to breathe normally, through his mouth and nose, a process that involved sedating him to prevent agitation. "That's a big change for an infant who's had no time to learn that since birth," Dr. Isaiah says.

ONE DAY AT A TIME

After all the ups and downs, Noah has gone home. He was doing well at his first doctor's visit after the reconstruction. Dr. Isaiah says the treatment succeeded due to the collaborative work of "very capable" groups, including the pediatric anesthesia team and the teams in the neonatal and the pediatric intensive care units.

Still, Dr. Isaiah thinks about Noah frequently. He calls the parents often to make sure his breathing stays quiet. "Every day that goes by, we've won 1 percent, 2 percent of the battle," he says.

Noah is starting to speak better, but speech and language therapy should further improve his articulation and voice quality. He might need even more surgery later for his voice.

Dr. Isaiah says, "Noisy breathing at birth doesn't mean danger all the time, but a relatively high degree of suspicion is required when the noisy breathing occurs very early in life." It should spur investigation of mechanical airway blockage.

Because Dr. Isaiah found what hindered Noah's breathing and carried out four operations to fix it, Noah should live a normal life. "It was a very good thing I picked up the phone right away," says Dr. Isaiah.

To reach Dr. Isaiah, please call **410-328-5837.**

Rounding Out Care & Research

UM RESEARCHERS IDENTIFY POTENTIAL PROTECTIVE AGENT AGAINST FLU

A new study by researchers at the University of Maryland School of Medicine, published in the Journal of Leukocyte Biology, has identified an innovative strategy for treating influenza, and perhaps other infectious diseases. Scientists showed that a small protein called retrocyclin-101 (RC-101) could potentially improve the symptoms and mortality associated with the flu and possibly other types of infectious illness. The protein is unique in that it not only targets the flu virus itself, but also the harmful inflammation the virus triggers in the host.

While the effect of RC-101 has been studied as a flu treatment in cells before, it has never been studied in animals. When looking at human immune cells, the researchers found that RC-101 had two positive effects. First, it blocked the flu virus from infecting the cells; second it blocked the runaway inflammation that is behind most symptoms of influenza infection, such as fever, pain, lethargy, and trouble breathing.

"We think that this protein could lead to medicines that could be a powerful tool in the battle against the flu, and against inflammation in general," says the study's lead author, Daniel J. Prantner, PhD, a research associate in the Department of Microbiology and Immunology at the University of Maryland School of Medicine.

NEW CHARLES COUNTY LOCATION

University of Maryland Specialty Care is now in Waldorf at Pembrooke Square, providing surgical consults, pre- and post-op visits, minor in-office procedures and specialty care services in one convenient location.

The office is staffed by board-certified physicians, nurses and support staff from the University of Maryland Department of Surgery. Services offered on-site include: Adult Congenital Heart, Bariatric Surgery, Colorectal Surgery, Pediatric Cardiac Care, Thoracic Surgery, Transplant Surgery (liver & kidney), and Vascular Surgery.

Most patients can be seen within two weeks of calling for an appointment, and sooner for urgent cases.

For more information or to refer a patient, please call **240-607-2010** *or visit* **umfpi.org/SC.**