Parents' fast actions lead to world-first heart procedure (Print)

Parental instincts and quick reaction time led to two central Illinois parents saving their child's life.

It started at home when the parents noticed something was extremely wrong with their 2-year-old son. They performed CPR and resuscitated him. He was transported to OSF HealthCare Children's Hospital of Illinois in Peoria where it was determined he had suffered sudden cardiac arrest and had a malignant heart condition known as Brugada syndrome.

What is Brugada syndrome?

Brugada syndrome is a rare genetic disorder that causes heart arrhythmias and sudden cardiac death.

"Most kids that go through that, unfortunately, don't survive," says Harma Turbendian, MD, a pediatric cardiac surgeon with OSF Children's Hospital of Illinois. "So, this was fast-acting parents who knew what they were doing, then EMS, followed by our team here in the ICU."

Meet the team

The medical team at OSF Children's Hospital who treated the toddler was led by Sunita Ferns, MD, the director of Pediatric and Adult Congenital Electrophysiology, Mark Plunkett, MD, the chief of Pediatric and Congenital Heart Surgery, and Dr. Turbendian, who was the primary surgeon in the procedure.

OSF HealthCare surgical teams have been using a device known as the EV-ICD, or extravascular implantable cardioverter-defibrillator, made by Medtronic, since spring 2024. But the new technology had only ever been used on adults or much older teenagers. The purpose of the device is that if or when a malignant heart arrhythmia occurs in the person, it will shock the heart back into rhythm.

By analyzing the 2-year-old's response to a trial of antiarrhythmic medications in the ICU and incorporating insights from his preliminary genetic results, Dr. Ferns made the diagnosis of Brugada syndrome and brought together a team of specialists.

"We needed buy-in to trial a new device for the first time ever in a patient his size from the surgeons and good technical support. Medtronic was excellent. We called our field engineers and had a whole team here," Dr. Ferns says.

Time to start the procedure

Sudden cardiac arrest is quite rare in kids, only happening in approximately one out of 100,000 children each year. However, the survival rate after one of these events is very low as well, with only 5% of pediatric patients surviving an out-of-hospital cardiac arrest.

Dr. Turbendian explains how the device is inserted.

"They're fairly small incisions that are required for implantation of the device. You actually don't have to access the heart in order to place this thing," Dr. Turbendian says. "You can make a small incision under the breastbone and one part of the device just slides through that small incision to sit under the sternum."

The possibility of implanting an EV-ICD takes away the need for open heart surgery to select patients.

Traditionally, a transvenous pacemaker has been used for older kids and adults. It was determined by Dr. Ferns and her team that the transvenous device was too large for the 2-year-old patient, and the EV-ICD was the better option. The EV-ICD incisions are much smaller, making this the more minimally invasive option.

"The lead was inserted, and the device was implanted outside of his rib cage," Dr. Ferns says. "Given that a 2-year-old is going to grow leaps and bounds over the next decade or two, it's an excellent choice for him. When we tried this, it was the first time in the world that someone trialed it on someone this small."

Dr. Plunkett calls the device a game changer. One that will send shock waves across the globe in the world of medicine.

"Children in the past had to undergo much larger procedures, oftentimes a sternotomy or thoracotomy. The devices were often bigger, and we were sewing patches on the heart or implanting coils," Dr. Plunkett says. "And some of those devices were not as effective as this new technology."

History-making procedure

"It's the first of its kind. This implantation is the smallest and youngest child ever to have this device," Dr. Plunkett says. "So that has expanded its application and market in a huge way. Its design initially was for adults or adult-sized patients. The fact we've proven it to be effective and safely implantable in a child this size really expands its application immensely."

Dr. Ferns agrees, hoping the team's work will get published soon, which helps medical centers across the world learn when and how to perform this procedure.

"It tells the world that we can now use (the EV-ICD) in a smaller child. That's how a lot of pediatric implants or newer technologies happen. It's one or two centers that do it and they publish their results," Dr. Ferns says. "We expect that to be published shortly. There are kids all around the world who sustain cardiac arrests, and they now have this option."

EV-ICDs are just a "safety net"

"We don't want it to keep shocking a child, because that's no way to grow up. To keep worrying about when your next shock is going to happen," Dr. Ferns says. "So, we monitor these devices constantly. If we see any arrhythmias in the background, despite the medication he's on, we are able to offer him other technologies. Ablative technologies can help modify the substrate, which is the tissue that's responsible for the bad rhythm."

Dr. Ferns is in charge of the child's post-surgery care moving forward. She says Quinidine, a drug that was initially made to treat malaria and now treats irregular heart arrhythmias, was the perfect match for the patient's condition.

"He has responded exceptionally well to this treatment," Dr. Ferns adds.

This decision was made after the child's EV-ICD did its job and went off while at home. His parents had to bring him back to the team at OSF HealthCare, who then had to ensure everything was meshing correctly.

While Dr. Ferns says the patient will forever be "married" to cardiology, she's confident in the techniques and technology the Electrophysiology (EP) Lab at OSF HealthCare Saint Francis Medical Center has to treat these patients. The EP Lab manages arrhythmias from unborn fetuses all the way to patients in their 70s with complex congenital heart disease. Technology like contact force sensing and high-density mapping are available, along with many other resources.

If the patient's symptoms were to worsen as time goes on, Dr. Ferns and her team in the EP Lab have the ability to perform ablations which target any abnormal heart tissue causing the arrhythmia, suppressing the abnormal electrical focus.

The generator of the device is in the axillary region, or the armpit. So as the child gets older and bigger, the strain sometimes found on conventional epicardial leads as a child gets taller is eliminated. The battery of the ICD should last 10-12 years.

Saving the patient's life started at home

All three doctors say they can't overemphasize the importance of the parents' actions at home.

"When we look at the magnitude of this, first we have to go back to the fact this child survived an arrest at home, was successfully resuscitated, was neurologically intact and presented to our hospital," Dr. Plunkett says.

This leads Dr. Ferns to say learning CPR is one of the most important things someone can do. Especially as kids enter middle and high school age and are involved with high intensity sports.

Another benefit of an EV-ICD is that if/when a patient experiences an arrhythmia, it will notify the congenital heart team in the EP Device Clinic.

"Most arrhythmia syndromes tend to happen at higher levels of activity. Many abnormalities can be detected by a simple noninvasive test such as an electrocardiogram (ECG)," Dr. Ferns says. "Universal screening is not yet common in the United States, but it is a part of the health care system in many European countries."

Strong family history is a sign to get checked out

Strong family history of sudden cardiac arrest or heart disease is a sign to get seen by a cardiologist. Dr. Ferns recommends starting off with an ECG which she advises doing before kids participate in high intensity athletics.

To learn more about the Electrophysiology Lab at OSF Saint Francis Medical Center, visit the OSF HealthCare website.