

Forging the Path – TAVR for Congenital Heart

Two teams of OSF HealthCare heart specialists worked together to give a 30 year old congenital heart patient a new lease on life, without the need for open heart surgery. The route they took put Kayla Pepmeyer on the map – she is now one of the only adult congenital heart patients in the world to receive a new heart valve via transcatheter aortic valve replacement, or the TAVR approach.

TAVR is typically a procedure for patients with severe aortic stenosis (narrowing of the aortic valve opening). It uses a balloon-expandable aortic heart valve – placing it into the body via a catheter-based delivery system. The valve is designed to replace a patient’s diseased aortic valve while the heart continues to beat – avoiding the need for open heart surgery which requires stopping the patient’s heart.

In Pepmeyer’s case, a heart valve that was surgically replaced several years ago was beginning to fail, and it now needed to be replaced as soon as possible.

“One of her valves, the tricuspid valve, did not form. So half of her heart didn't form, including the right ventricle, which pumps blood to her lungs. She was left with one single pumping chamber which only pumped blood to her body, so she required several surgeries to redirect the blood flow to her lungs,” said Dr. Marc Knepp, pediatric cardiologist and director of the Adult Congenital Heart Program at OSF HealthCare Children’s Hospital of Illinois.

Usually TAVR is performed on high-risk and intermediate-risk patients with symptomatic severe aortic stenosis. While randomized TAVR trials for low-risk patients are underway, using the approach for adult congenital patients is a new procedural frontier. Pepmeyer was willing to forge the path.

“I've always been the one where they say, ‘we’ve never done this before, but we're going to do it, and if it works, awesome. If not, we'll figure something else out.’ So I've always been that guinea pig, per se. It's a little more terrifying now that I'm older, but it's cool to say, ‘okay, this can be done,’” Pepmeyer said.

Dr. Knepp has been working as part of Pepmeyer’s care team, and felt strongly that TAVR was the right move for her case. The numerous surgeries Pepmeyer has endured in her lifetime puts her at greater risk during large, invasive procedures.

“By having multiple surgeries, that increases the amount of scar tissue in the chest wall, the difficulty of getting into the heart to operate on it, and we know that multiple surgeries is not good for someone's kidneys, their brain, their liver, or even their heart function long-term. As patients with a single ventricle age, doing repetitive surgeries becomes more and more dangerous with more complication risk, and more risk for even death,” explained Dr. Knepp. “So trying to do things minimally invasive, or less invasive, are the ways to look ahead and to develop our field.”

The adult congenital heart team at OSF Children’s Hospital of Illinois worked closely with interventional cardiologists, surgeons, valve coordinators and other care specialists at OSF HealthCare Cardiovascular Institute to create Pepmeyer’s surgery plan.

“Her case had a lot of complexities,” explained Dr. Marco Barzallo, an interventional cardiologist with OSF HealthCare Cardiovascular Institute. “Her aorta is extremely tortuous. We normally try to go through the femoral artery, which is the easiest procedure, but in her case she had some other complications, so that made the easier way not very feasible. So we came up with a second, better option which is going through her carotid artery, which is the second-most, less-invasive procedure. In her case, this worked very well for us. So coordinating with them, getting all the studies read, getting to understand the physiology and what is at stake, and play all the potential options was actually a great experience for all of us.”

“Having that teamwork - that's so necessary for adults with congenital heart disease. Having providers who talk to each other, who want to help take care of patients like this is very important,” added Dr. Knepp.

The TAVR surgery was successfully performed in October of 2021, and Pepmeyer was on her way home to St. Louis after just two days in the hospital – a marked difference from her last valve replacement, which required open heart surgery and a lengthy recovery.

“I was up the same day and surprised – very surprised – about the amount of pain I was not in, honestly. I'm so used to having to hold my chest to cough and everything, and it was not that. It was easy. Up the same day and they were telling me I was leaving the next day,” reflected Pepmeyer.

Now the 30 year old mother is fully recovered, happily living life with her fiancé and six-year-old daughter. Both Drs. Knepp and Barzallo say outcomes like Pepmeyer's are the success stories that keep them going.

“It's just amazing. It encourages us as we watch babies sometimes struggle a little bit after the their cardiac surgery, to know that later on they're going to be doing well. I think pioneers like Kayla help us actually prepare for the future and to make sure we're doing all the good things that we need to do when the babies are small.”

“It's one of the feelings that you live for,” Dr. Barzallo agreed. “It's not that you just do a procedure, but when you see someone come back to your office, and not just for Kayla, but every patient that has TAVR. It's very rewarding when you have someone, even the same day after the procedure saying, ‘I feel good.’ Normally after an open heart surgery, you'll have many tools and a wound to heal that takes a longer time to heal. But someone who has instant results and feels the difference right away is actually very rewarding.”

Learn more about the Congenital Heart Center at OSF HealthCare Children's Hospital of Illinois, including conditions treated and services available [here](#). For more information about TAVR, or to see if you are a TAVR candidate, visit OSF HealthCare's TAVR information page [here](#).