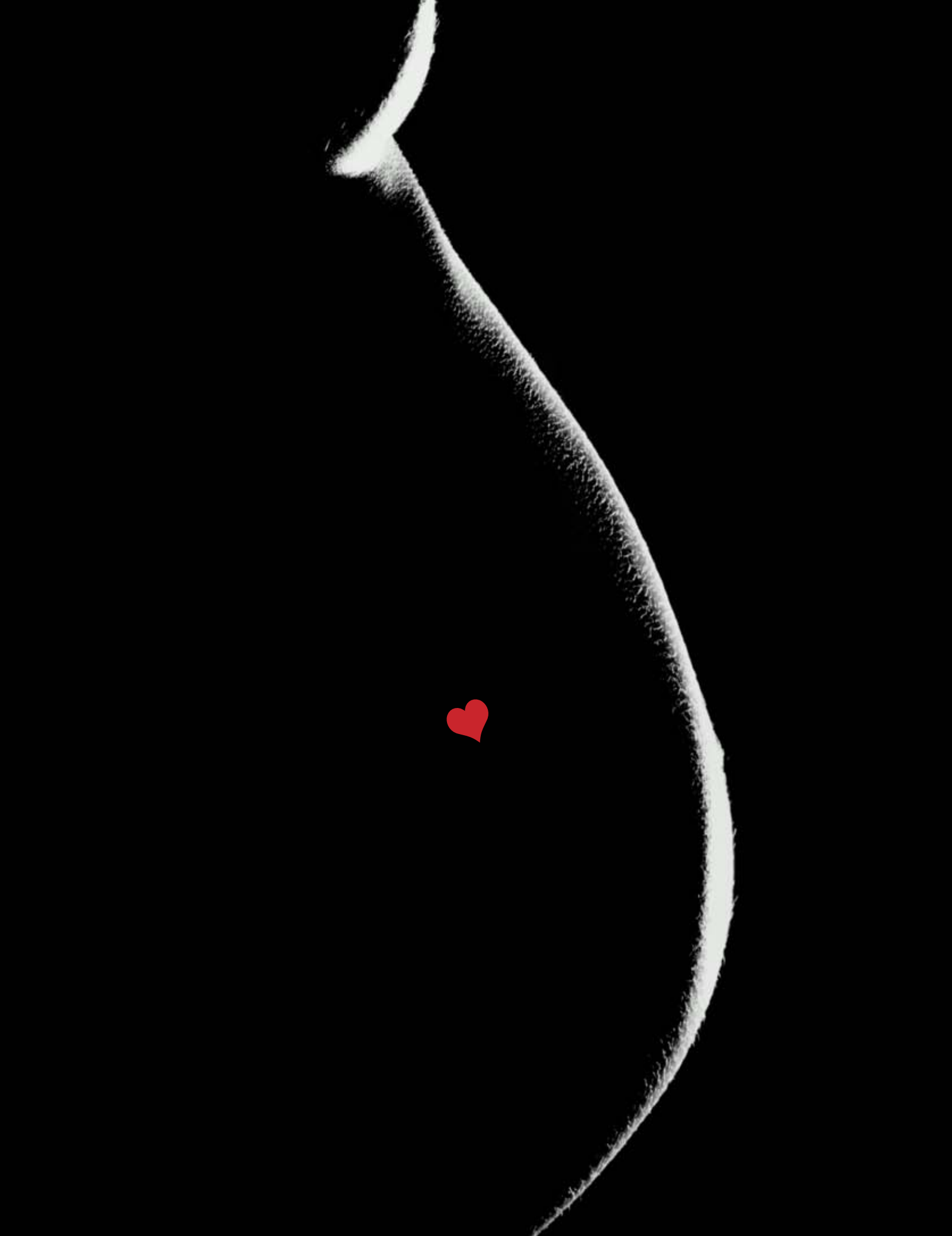
g (the greatest gift)

Early in her pregnancy, at her first ultrasound appointment, Katie Larrison, 27, of Algonac, Michigan, knew something wasn't right. "The technician took the disk out and said, 'I have to show this to the doctor.' Then the doctor came in and said, 'I'm not going to beat around the bush. There's something wrong with her heart, but I don't know exactly what it is.' "

by Jeff Mortimer

LAURENCE MONNERET/GETTY IMAGES



As Katie and husband Jeremy, 29, hugged in the parking lot outside the doctor's office, she said to him, "Whatever it is, we'll get through it. It'll be OK in the end."

Still, Katie was in tears when she called her mother, a registered nurse who works in adult critical care, on the way home. "She asked a lot of good questions and kind of set the tone," Katie says. "She was strong about it, so I knew I could be."

Their baby girl, whom the Larrisons named Mira, was diagnosed with hypoplastic left heart syndrome (HLHS), a condition in which the left side of the heart fails to develop; it's often described as having half a heart.

Jeremy and Katie had been married almost eight years and already were the parents of an energetic and outgoing 3-year-old daughter, Rayne. They grew up and fell in love in the little upstate Michigan town of Mio, but had been living since 2004 in Sitka, Alaska, where Jeremy, an aviation maintenance technician for the U.S. Coast Guard, was stationed. They had just learned that Katie was expecting again when Jeremy was transferred back to Michigan, which meant they would be near their families during her pregnancy.

The Larrisons decided to have Mira treated at the University of Michigan, largely because they had done their research and knew about its Congenital Heart Center's Prenatal Heart Program, but their decision had some serendipitous support.

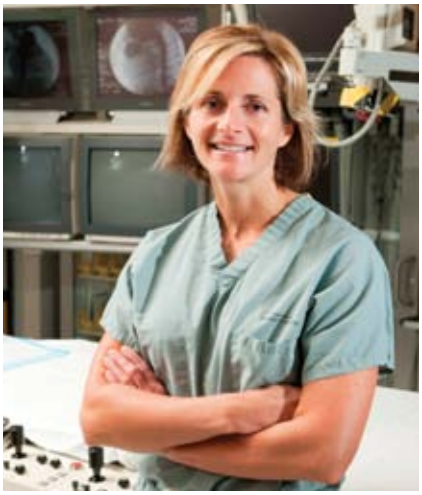
"I was talking to one of the guys at work, and he was telling me about his son who has a similar condition," says Jeremy. "His boy is three or four and just had his last surgery. They were going to U-M and were pleased. Two weeks later, we found out about Mira."

Not long thereafter, Katie was having her hair done. She was starting to show, and struggling with whether or not "I should tell people she has a heart defect, and I might not have her in three months. That might be a hard conversation."

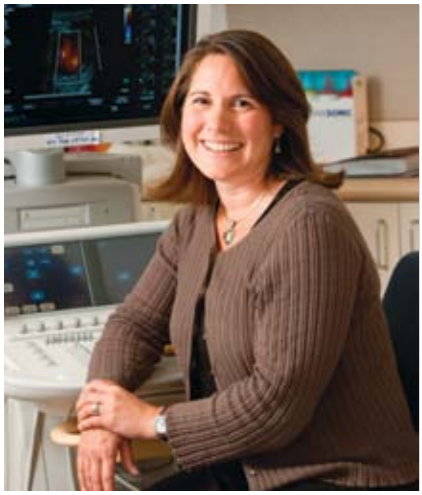
The moment arrived when her hairdresser popped the question. "I decided to just tell her the baby had a problem and we were at U-M," Katie says. "She stopped washing my hair and asked what was wrong. I said she had a heart defect, HLHS. She said her son had had the same thing 15 years ago and they were at U-M too."

The boy survived only five-and-a-half months, but his mother spoke highly of the care he received. "It's weird that I ended up in her chair," says Katie. "She was the first parent I met whose baby had the same condition that Mira was going to have. It was kind of surreal."

The Larrisons soon learned that Mira's HLHS, so serious in and of itself, was only one of her problems. University of Michigan doctors discovered that she also lacked an adequate hole in the atrial septum, the wall between the upper chambers of her heart, which resulted in pressure build-up in the undeveloped left heart and higher pressure in the developing lungs. Mira also had Turner syndrome, a rare genetic defect that compromised her ability to successfully withstand the procedures needed to fix the other problems.



Aimee Armstrong, pediatric cardiac interventionalist



Carlen Fifer, pediatric cardiologist

Anne Wuerth, pediatric nurse practitioner

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"Whenever we went, it seemed like it was never good news," says Katie.

Katie and Jeremy were informed of their options: aborting the baby; going through with delivery and keeping her comfortable until she died; or trying to save her with a series of surgeries, including a rare, risky fetal intervention to create a larger hole in her atrial septum.

"We made our decision, I think from the very beginning, that we were going to give her a chance," says Katie.

The prospect of the fetal intervention, daunting as it was, didn't change that. "I thought, of all the surgeries she's going to have, this is the only one I could physically help her through," she says. "I'm pumping the blood for her, helping her in there. If I can support her that way, that's the greatest gift I can give to her."

It was due only to a bureaucratic snafu that the Larrisons were living anywhere near a pediatric cardiologist, much less a program like Michigan's, one of only two or three medical centers in the country that perform fetal cardiac interventions. "They messed up my orders," says Jeremy. "I was supposed to be in Alaska for another year."

Then there was that savvy ultrasound technician, and the chance meetings with the parents of similarly afflicted children that helped nudge them toward the University of Michigan, where the way they were treated made it easier to persevere.

"We knew what was going on was very serious," says Katie, "and they presented it that way, but they always seemed to have a plan. A nurse practitioner named Anne Wuerth would stay with us through all our appointments so we knew how it all came together."

According to Aimee Armstrong, M.D. (Fellowship 2003), a pediatric cardiac interventionalist and clinical assistant professor in the Medical School who is an integral part of the team of specialists that operated on Mira, "About 6 percent of babies with HLHS have an inadequate atrial septal defect. When those babies get a hole created after birth, only about 50 percent of them live to get out of the hospital. And patients who have Turner syndrome and HLHS, even if they have a good-sized hole, have a poor prognosis. Only a few survive. I don't know of anyone else, even in the literature, surviving with HLHS, intact septum and Turner syndrome."

Children's Hospital in Boston was the first in the country to perform fetal cardiac interventions, and groups of U-M doctors went there twice to observe the procedure.

"One of the things we learned from Boston is that you really need to have the whole team involved and invested," says Carlen Fifer, M.D., a clinical associate professor and director of the Prenatal Heart Program and Fetal Cardiac Intervention Program. "This is something that requires multiple team members across departments."

Katie and Jeremy were informed of their options: aborting the baby; going through with delivery and keeping her comfortable until she died; or trying to save her with a series of surgeries, including a rare, risky fetal intervention to create a larger hole in her atrial septum.

She ticks off the list: pediatric cardiologists, interventionalists, fetal echocardiographers, maternal fetal medicine specialists whose focus is obstetrics and dealing with babies who have high-risk problems, and maternal and fetal pediatric anesthesiologists, not to mention social workers and nurses.

"You have eight people who need to work as one unit, basically," says Armstrong. "We all have different specialties and there isn't any one person who could do this entire procedure on his or her own."

On December 2, 2008, the team was ready, and so was Katie. Recognizing so many of the faces she saw in the operating room helped soothe her nerves. "There were over 20 people in there during the actual

intervention," she says, "and you could see people in the next room over. It was just crazy. But they were all people that we seemed to know."

Katie was put to sleep and monitored by Jill Mhyre (M.D. 1999), an obstetric anesthesiologist. Pediatric anesthesiologist S. Devi Chiravuri, M.D., sedated and monitored Mira. Marjorie Treadwell (M.D. 1984), an obstetrician/gynecologist and maternal fetal specialist, guided the ultrasound operated by Fifer and colleague Sarah Gelehrter, M.D. (Residency 2002, Fellowship 2005), both fetal echocardiographers, and the image was projected on several television monitors in the room.

Cosmas van de Ven, M.D., also an obstetrician/gynecologist and maternal fetal specialist, inserted a needle

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through Katie's abdomen, then through Mira's chest, into her beating, grape-sized heart, and through the atrial septum. Steered by the ultrasound image, Armstrong threaded a deflated balloon through the needle and across the septum, and Albert Rocchini, M.D., a pediatric interventional cardiologist, inflated it several times, opening a hole in the septum and relieving the stress on Mira's heart and lungs.

Constant vigilance was essential. "We have several pairs of eyes making sure we're in the right place and doing the right thing," says Treadwell. "You're talking about very small spaces, and sometimes it can be difficult to see on the ultrasound. It's very helpful to be able to double-check with each other."

The fetal intervention kept Mira in the game, enabling her at least to survive until birth. But on the day she was delivered by caesarean section, her care team prepared Katie and Jeremy for a baby who would be blue and listless, weighing about four pounds, with a blood oxygen saturation level, or "sats," in the 30s.

Instead, "She came out pink and screaming," says Katie. "I heard someone yell, 'Her sats are in the 90s!' "

Mira weighed five pounds, 11 ounces, "and every little bit of that helped her," says Jeremy. She was far from out of the woods, but she was here. "There were a bunch of surprises that day," adds Jeremy. "They had a plan for her, but she came out having a different plan."

There was, of course, considerable planning involved in getting her that far. The team that treated her met regularly to evaluate how she was doing, coordinate what they were going to do, assess what they had done so far, and prepare for every conceivable eventuality.

It was van de Ven who delivered Mira on January 27, 2009. The original date had been changed so teams could be on standby at several locations, depending on what kind of procedures her condition would dictate.

Her destination turned out to be a cardiac catheterization laboratory. Although she was in far better shape at birth than anyone predicted, Mira wasn't strong enough to undergo the Norwood procedure, the standard surgery for newborns with HLHS.

"We found with other children with Turner syndrome in addition to HLHS, that if you put them on a cardio-pulmonary bypass machine for a major heart surgery right after birth, they often are very sick afterward and may not survive hospitalization," says Armstrong. "So we took a newer, innovative approach, a hybrid approach in the catheterization laboratory, so she didn't have to go on bypass."

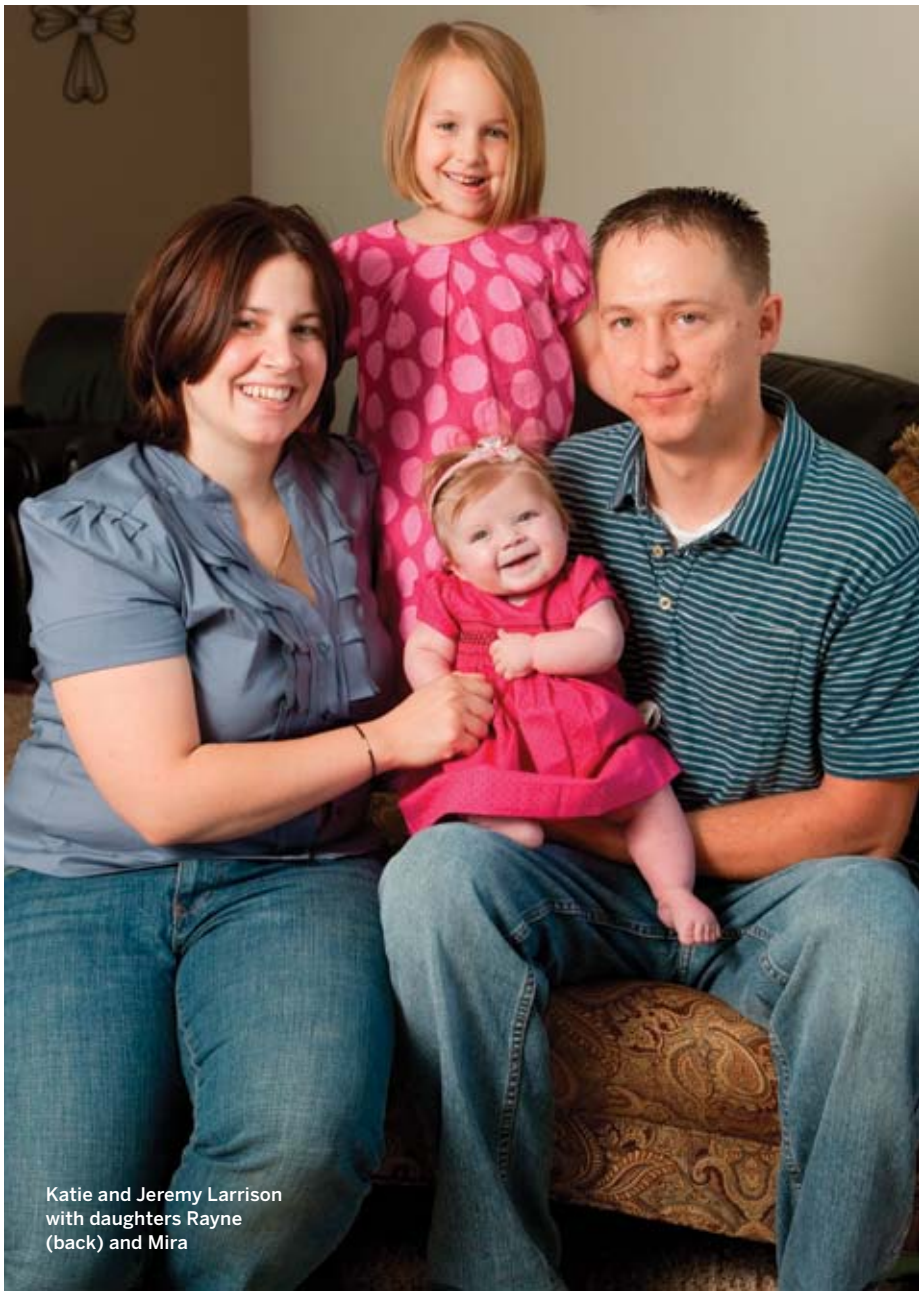
A few hours after Mira was born, Armstrong and Jennifer Hirsch, M.D. (Fellowship 2008), a pediatric heart surgeon, opened Mira's chest and placed bands on her pulmonary arteries to decrease the flow of blood to her lungs, and placed a stent across her atrial septum to keep the hole open.

Two days later, she received another stent in her heart that improved the blood flow to her body by holding open the duct between the aorta and the pulmonary artery. This passage, called the *patent ductus arteriosus*, normally closes in the first few days after birth, but Mira's needed to stay open so the right side of her heart could effectively do the work of both sides.

She eventually had the Norwood procedure when she was four months old, and she tolerated it well because of her age. In the meantime, she had made history. Just two weeks after her birth, Mira Larrison went home.

Not only was Mira's just the fourth fetal cardiac intervention performed at the U-M since the Fetal Cardiac Intervention Program was created in 2008, but "she was the highest risk fetal intervention patient we've had so far," says Armstrong, "and she's done the best out of all of them."

"Mira is the only one right now who has been able to get through those surgeries and go home," says Fifer, "and she had three strikes against her."



Katie and Jeremy Larrison with daughters Rayne (back) and Mira

The Larrisons' home is in a small, pleasant, older subdivision, nestled between a woods and the northeast shore of Lake St. Clair, a mile or two from the Canadian border. Children fill the otherwise quiet street in the summer, laughing and calling to each other as they ride their bikes and pull their wagons.

Her parents' fondest hope at the moment is that, someday, Mira will be one of them. "I want her to be able to walk and play and run," says Katie, then adds, her eyes twinkling, "and I saved some of her really high insurance statements, just to show her when she's older."

Right now, she's able to be an almost regular baby, the latest family miracle. She bounces gently in her swing, gurgling and chuckling as she eyes the world around her. Except for her feeding tube, she's as extraordinary and ordinary as any infant. And except for the dry-erase board affixed to the outside of a cupboard door, which Katie and Jeremy use to keep track of her 11 medications, it could be any home where an infant lives.

"We tell people that it just becomes our new normal," says Katie. "I don't think we could picture being without her."

Through all the medical dramas, the Larrisons also had Rayne to care for. And, for a while, she had no trouble picturing being without Mira.

"She's a great big sister now," says Katie. "She's always the first one to get Mira a toy when she's cranky. But in the beginning, she didn't want anything to do with her. One time at the hospital, she asked if we could leave her now."

But their mother knew the sisters had turned the corner when, on the way home from the U-M one day, she turned to look at her children in their adjacent car seats. What she saw brought tears to her eyes.

Rayne, unbidden, had taken her sister's hand. [M]

Meet Mira's team **MORE ON THE WEB**

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