

November 20, 2006

A tiny girl, a big triumph

Remi Parker Howard was born with a closed heart valve. To save the twin, her cardiologist would have to pull off a risky procedure.

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The identical twins were born eight weeks early.

Kennedy Jain Howard arrived first, at 3 pounds, 5 ounces. Her sister, Remi Parker Howard, came 30 seconds later.

Remi was a little larger, 3 pounds, 7 ounces, but she was the one in danger. Monitors showed her blood wasn't getting enough oxygen.

At first, doctors told the twins' parents, Ingrid and Kevin Howard of Charlotte, that Remi's heart defect might require surgery. But the challenge of operating on a heart the size of a walnut didn't sink in immediately. Everything had happened so fast.

Ingrid Howard's plan had been to have the twins by Caesarian section in late September, two weeks short of full-term pregnancy. But on Sunday night, Aug. 13, her water broke.

The babies arrived the next day – eight weeks early – and went straight to neonatal intensive care, where the preemies were monitored closely.

Ingrid was exhausted. She had lost a lot of blood during the C-section. Neither she nor Kevin really understood when doctors referred to Remi's condition as "pulmonary atresia."

Later, they looked it up and questioned friends and relatives who have medical backgrounds.

Ingrid recalled her fear: "We could never get anyone to say, 'It will be OK.'"

Remi's heart didn't develop properly in the womb.

The valve that forms the opening to her pulmonary artery, one of the two great vessels of the heart, they stayed closed.

Instead of swinging flaps, which let the blood through and then keep it from flowing back, the valve was blocked, like a solid wall. Without that opening to her lungs, Remi's blood wouldn't get oxygen to deliver to her brain and the rest of her body.

To keep her alive, doctors immediately prescribed a drug, called prostaglandins. It would help for a while by keeping open a special fetal blood vessel, called PDA, for patent ductus arteriosus.

Every fetus has a PDA, which connects the pulmonary artery directly to the aorta, the heart's other great vessel. In the uterus, a fetus's lungs are filled with fluid. So the PDA sends blood to the mother's placenta to get oxygen, bypassing the lungs.

Once a baby is born, the PDA closes in a day or two, and the pulmonary artery takes over, carrying oxygen-depleted blood from the right ventricle to the lungs.

Thanks to the drug, Remi's PDA stayed open, and her oxygen levels improved. But that wouldn't last long.

Non-surgical approach

Most babies born with pulmonary atresia require surgery as the primary treatment or as follow-up. In Remi's case, doctors agreed that Dr. Herbert Stern, who specializes in therapeutic heart catheterization at the Sanger Clinic, should start with a non-surgical approach similar to heart catheterization. Guided by continuous X-rays, he would maneuver a thin catheter from Remi's groin to her tiny heart. Then, he would snake a hair-thin wire through the catheter, delivering radiofrequency energy to burn a tiny hole in the pulmonary valve and into her pulmonary artery.

To widen the opening, he would insert another wire with a small balloon attached. Inflating the balloon would dilate the valve. Successive balloons, each one a little larger, would gradually open the space.

Stern, 48, who graduated from the Medical University of South Carolina and did a fellowship in pediatric cardiology at Duke University, had done the procedure before, but never on a child so small. Even in bigger babies, the risk of complications is high because that section of the heart is paper thin. A hole could accidentally get burned in the wrong place.

In case something went wrong, Stern asked a surgeon to stand by.

Twins together again

The morning of the procedure, when the twins were 4 days old, Ingrid and Kevin Howard were surprised when they arrived in intensive care.

Nurses had moved their healthy twin, Kennedy, into Remi's incubator, so the twins could spoon, just as they had for 32 weeks in utero.

While their parents watched, Kennedy's arms flailed, finally resting on her sister's belly, as if to give her a hug.

In pictures taken by the nurses, Kennedy looks pink and healthy. Remi appears listless, with yellow, jaundiced skin. They are entwined in tubes.

Then nurses took Remi, and Ingrid and Kevin Howard waited with their families.

An hour passed, then two – longer than they expected. The room grew quiet. At four hours, Ingrid couldn't stand it. She got up to take a walk.

Opening the door, she was met by a smiling Stern. Remi's going to be OK, he said.

The family cheered and hugged. Ingrid held out her hand to shake Stern's.

Aren't you going to give me a hug, she remembers the doctor asking. She wrapped him in her arms.

'Nice and pink'

Remi stayed on prostaglandins for 12 more days, to give her right ventricle, the right pumping chamber of the heart, time to grow and heal. When doctors stopped the drug, they watched her reaction. Remi stayed, in Stern's words, "nice and pink."

That meant she wouldn't need surgery after all. "Her life expectancy should be normal," Stern told the parents.

On Sept. 22, five weeks after the procedure and two weeks after her sister, Remi came home.

Today, at 14 weeks old, Remi weighs 8 pounds.

Kennedy is only a little bigger, at 8 pounds, 9 ounces.

They look like dolls, dressed in pink print dresses with white socks trimmed in pink over their tiny feet.

Ingrid, a petite person herself, holds Remi in one arm, the baby's head cradled in her palm. Kevin holds the other twin, after first scrubbing his hands with antibacterial cleanser.

The only reminder of Remi's heart condition is the band around her chest that is connected to a laptop-sized heart monitor. It has gone off only a few times. Not one was cause for alarm.

Stern has searched medical literature and contacted other heart specialists around the country. As far as he can tell, Remi is the smallest child to have survived the procedure without needing surgery.

Stern has written the case of "Baby RH" for a medical journal, so other doctors know it's possible. When the paper gets published, he plans to give a copy to the Howards for Remi's baby book.

He wants her to know, someday, that she is "Baby RH."

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