Liza Arguelles-Brady and her son, James, who was born with his aorta and pulmonary artery transposed.

SOMETIMES BABIES ARE BORN WITH ARTERIES THAT ARISE FROM THE WRONG PLACES.

A CHANGE OF HEART

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AS THE ULTRASOUND TECHNICIAN SLID THE TRANSDUCER OVER HER SWOLLEN BELLY, LIZA ARGUELLES-BRADY WAITED WITH NERVOUS EXCITEMENT TO SEE THE FIRST IMAGE OF HER UNBORN CHILD.

Instead of hearing those magic words, "Congratulations, you have a healthy baby!" she was greeted with silence. For some reason, the technician couldn't get a good image of the fetal heart. A second test, days later, yielded the same result. Liza was advised to get a more sophisticated sonogram at NYU Langone Medical Center, just to be safe. If something was amiss, no one was letting on.

This time, the images were crystal clear, and the news wasn't good. Liza's baby, a boy, had a serious heart defect called transposition of the great arteries (TGA), in which the aorta and the pulmonary artery, the major vessels carrying blood away from the heart, arise from the wrong ventricle. The vessels are transposed, so that the aorta, which normally arises from the left ventricle and carries oxygen-rich blood to the body, arises instead from the right ventricle, which is filled with oxygen-poor blood. Meanwhile, the pulmonary artery, which normally emerges from the right ventricle carrying oxygen-poor blood to the lungs where it will be loaded with oxygen, now emerges from the left ventricle. Because of this anatomical mix-up, oxygenated blood flows in a closed loop, cycling endlessly between the lungs and the left side of the heart, never reaching the body.

As Liza learned from Achiau Ludomirsky, MD, professor of pediatrics and director of the Division of Pediatric Cardiology at NYU Langone, her baby would need open-heart surgery—an arterial switch procedure—just days after birth. "I couldn't believe what I was hearing," Liza recalls. "My head was spinning. I had never heard of anything like this."

Actually, this scenario is more common than you might imagine. About one in 125 children in the U.S. is born with a congenital heart defect, says Dr. Ludomirsky. Of these, 5 to 7 percent—approximately 2,000 babies—have TGA. Until the 1970s, when the arterial switch procedure and a heart-lung bypass machine small enough for neonates were developed, TGA was lethal or severely debilitating. Today the survival rate is greater than 90 percent. However, the arterial switch remains a complex procedure, and the Brady case would turn out to be more complex than most.

After hearing the diagnosis, Liza and her husband, Peter, could do little but count down the days until the delivery date, still four months away. "The first weeks were tough," says Liza. "I did the worst thing possible, which was to search *YouTube* for videos about TGA. All you see is babies in intensive care units attached to all kinds of tubes, with music playing in the background." Peter worked hard to focus on the best possible outcome. "Although it was a setback, my mindset is you deal with what you've got," he says. "What was helpful was hearing from Dr. Ludomirsky that this procedure has a high success rate, and that our child could essentially have a normal life. Also, he promised us his team would be ready, no matter when the baby was delivered."

During those tense months, the Bradys visited with Ralph Mosca, MD, professor of cardiac surgery and director of the Division of Pediatric and Adult Congenital Cardiac Surgery at NYU Langone, who would perform the operation, and toured the neonatal and pediatric intensive care units, better known as the NICU and the PICU, which would become familiar territory. "That was tremendously helpful down the road," says Peter. "We knew what to expect."

When Liza's water broke on March 31, the Bradys headed to Tisch Hospital at NYU Langone, a short drive from their home in Jersey City. Hours later, James Brady, 7 lb., 10 oz., was delivered by C-section (a clinical decision unrelated to the baby's heart condition). He was whisked off to the NICU before Liza or Peter had even a moment to cradle him in their arms. "I was able to give James a kiss," Liza remembers.

In the beginning, James looked like a healthy newborn. Most babies with TGA develop normally in the womb, where the fetus's circulation bypasses the lungs and its blood is oxygenated by the mother, via the umbilical cord. At the moment of birth, however, circulation begins to change in various ways. Most crucially, a healthy baby's blood is immediately shunted to the lungs, allowing the baby to oxygenate its blood by breathing, a milestone announced by the first cry. For James, however, this would be one of his last gasps—unless his mismatched arteries were realigned quickly.

He wasn't taken to surgery immediately. "It's better to wait a few days," says Dr. Ludomirsky, explaining that babies with TGA sometimes have additional birth defects. "You want to bring the baby into the OR in the most stable state that you can." Waiting is possible because newborns with TGA are usually able to pump some oxygenated blood to the body through temporary remnants of the fetal circulation, namely, the ductus arteriosus (a small blood vessel that connects the aorta with the pulmonary artery) and the foramen ovale (a hole between the left and right atria). Both connections allow limited mixing of "red" (oxygenated) and "blue" (unoxygenated) blood, until they seal shut in the months after birth.

MOST BABIES WITH TGA DEVELOP NORMALLY IN THE WOMB. AT THE MOMENT OF BIRTH, HOWEVER, CIRCULATION BEGINS TO CHANGE IN VARIOUS WAYS.

LEFT: Dr. Ralph Mosca, who operated on James. CENTER: Liza with her son. RIGHT: Dr. Achiau Ludomirsky, director of the Division of Pediatric Cardiology.

By day two, however, James began to turn a sickly blue. Despite supplemental oxygen and a drug called prostaglandin, which slows shrinkage of the ductus arteriosus, his body was starved for oxygen. Dr. Ludomirsky called Michael Argilla, MD, director of the Pediatric Cardiac Catheterization Laboratory, to perform a balloon septostomy: A balloon-tipped catheter is inserted into a leg vein and snaked up into the heart, where it is passed through the foramen ovale into the left atrium. Then the balloon is inflated and withdrawn, widening the foramen ovale, which increases mixing of red and blue blood. The procedure worked, buying James precious time.

Several days later, James was wheeled into the OR for the arterial switch procedure. Dr. Mosca opened his patient's chest and examined his heart, barely the size of a walnut. As part of the repair, the coronary arteries, which supply the heart muscle with blood, need to be rerouted. (In a normal heart, the coronary arteries arise from the aorta on the left side, branching immediately off the ascending aorta just beyond the aortic valve. In an infant with TGA, the aorta emerges from the heart's right side. When the vessels are switched, the coronary arteries must be freed from the right side of the heart and moved to the left side so that they receive oxygen-rich blood.) Complicating matters, James's coronary arteries were abnormal—in size, shape, and position. With a jazz musician's flair, Dr. Mosca began to improvise, using parts of James's aorta and pericardium (the membrane that surrounds the heart) to reshape the top of his heart, making for a better fit with the coronary arteries.

"The challenge with this operation is all mechanical," says the surgeon, who performs about 10 TGA repairs a year. "You have to line up all the arteries in such a way that they are not twisted, in which case you won't get proper blood flow. If you get the plumbing right, these hearts can do very well."

Back in Liza's hospital room, the family waited for news. After four hours, the normal length of a TGA repair, Dr. Mosca sent word that there were complications. It would be a while, but when the operation was finally over, James was fine. "That was the beginning of a tough three or four days, sitting with him in the PICU. And then having to leave him at the end of the day..." says Liza, her voice trailing off.

The couple began to breathe a little easier when doctors told them James's replumbed heart was strong, but he wasn't flourishing quite yet. "At first, he didn't want to feed," Dr. Ludomirsky explains. "It's not uncommon with any patient after major surgery, and here we had a little baby who had never fed on his own." James's natural impulses eventually took hold, and he began to suckle. "The nurses encouraged us, and they showed us all the little tricks to get him to feed," says Liza. "We could see progress every day." When James left the hospital, his parents were finally able to strap him into his car seat for the drive home.

THERE IS ANOTHER CHAPTER TO JAMES'S STORY. A follow-up exam revealed that his heart had developed some scar tissue—which sometimes occurs as a consequence of cardiac surgery—at the suture line in the new aorta. In mid-September, James underwent another procedure and was sent home five days later. Soon, he was giggling and cooing like any other six-month old. His prognosis, according to Drs. Mosca and Ludomirsky, is good.

A week later, Liza brought James to watch his dad run in a race preceding the famous Fifth Avenue Mile. "Of course, James didn't know what was going on," Liza laughs. "But the weather was beautiful and James was in a good mood. All in all, it was a great day."

"We have him signed up for baby swim class, play groups, and music class," says Liza, who is looking forward to the normal joys of parenthood. "We can't wait to use the Baby Jogger with him." ●