

# To mend young girl's heart, Texas doctors tried something new

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The first hint came in a grainy black and white ultrasound in a doctor's office in West Texas.

Elizabeth Chacon, 35 weeks pregnant, reclined on the exam table and stared at the screen.

The doctor pointed to the blotches and began to explain what he saw, but Elizabeth thought he was most likely worried for nothing. This was her fourth baby, and nothing about the pregnancy had seemed alarming or even unusual.

On the screen, she remembers thinking, the baby looked fine. Still, the doctor insisted she drive to Fort Worth to see a specialist.

Less than a week later, Elizabeth and her husband, Efren, each packed a change of clothes into a small overnight bag, hugged their children goodbye and headed east from Odessa.

Ivy Celeste Chacon was born in the afternoon on June 9, 2015, a Tuesday.

So many doctors and nurses and specialists crowded into the delivery room at Texas Health Harris Methodist Hospital Fort Worth that Elizabeth realized then something was indeed wrong.

"I was in denial until she was born," Elizabeth says. "But there were so

many people in that room. It was packed, and I knew."

'GOT VERY QUIET'

Elizabeth can no longer remember whether Ivy cried immediately or even how much she weighed. She remembers only this: a tiny baby, surrounded by people in scrubs and gloves and masks.

Someone, maybe a nurse, told Elizabeth that her daughter was OK, just before they whisked the baby next door to Cook Children's Medical Center, connected to the hospital by a special corridor. In an instant, the noise in the delivery room stopped.

"Quiet," Elizabeth says. "I remember it suddenly got very quiet."

Ivy was born with congenitally corrected transposition of the great arteries, an exceedingly rare [heart](#) defect in which the heart's lower two chambers are reversed.

In a normal heart, the right side pumps blood to the lungs to pick up oxygen, while the left side then pumps that oxygen-rich blood to the rest of the body.

In Ivy's heart, however, her weaker ventricle formed on the left side, pumping blood to her body, while her stronger ventricle pumped blood to the lungs.

Cook Children's has surgically corrected about a dozen congenitally corrected transposition cases, says Dr. Lisa Roten, a pediatric cardiologist at Cook's who treated Ivy.

"This is a very rare, random occurrence," Roten says.

About 1 percent of all babies are born with a [congenital heart defect](#). Of those babies, less than 1 percent has the same condition as Ivy. Doctors estimate only 5,000 to 10,000 people in the entire United States have congenitally corrected transposition of the great arteries, or CCTGA.

Frequently, babies with CCTGA have additional heart defects. Physicians at Cook Children's soon learned the full extent of Ivy's heart complications.

She also had dextrocardia, a rare condition in which the heart points toward the right side of the chest rather than left. Her heart had an atrial septal defect, a hole in the wall that separates the top two chambers of the heart, and a ventricular septal defect, a hole in the wall that separates the bottom two chambers. She had pulmonary valve stenosis, a deformity on the pulmonary valve that limits the blood flow.

"I would say Ivy had the most complicated heart we have ever seen," Roten says. "We have never seen quite this combination of issues."

Babies with CCGTA are often born with a blue skin tone because their blood is not fully oxygenated. Until the 1980s, Roten says, little could be done to help so-called blue babies.

"Where we are in 2016 is far from where we used to be," she says.

### 'SHE LOOKED PERFECT'

After Ivy was born, a nurse wheeled Elizabeth to Cook Children's to see her.

Ivy, tiny with a smattering of dark hair, lay in an incubator, a tangle of tubes monitoring her heart and delivering medication to increase her blood flow.

"I thought she looked perfect," Elizabeth says.

Days then weeks passed in a blur of doctor's appointments and video chats with Elizabeth's children at home in Odessa.

At 23 days old, Ivy underwent her first surgery to insert a shunt, or a small tube made of Gore-Tex, into her heart to increase the flow of blood to the lungs. It was a temporary fix, but it would allow Ivy and her family to return to Odessa.

On Aug. 1, Elizabeth and Efren loaded their suitcases into the SUV and tucked Ivy into a car seat. For most of the drive home, Ivy wailed.

"I was scared," Elizabeth says. "I didn't know if I would know how to take care of her."

Doctors at Cook Children's were considering how to repair Ivy's heart.

Because her heart was so complex, most hospitals would have reduced the organ to a single ventricle. Doing so would represent the lowest risks, but patients often suffer early heart failure, requiring a heart transplant to survive.

Dr. Vincent Tam, a cardiothoracic surgeon at Cook's, had an idea. What if physicians could combine two difficult procedures that would correct the anatomical structure of her heart?

Tam would perform a complex switch operation, known as the Senning procedure, along with an aortic translocation, called the Nikaidoh procedure.

First, the Senning procedure would create a baffle, or conduit, at the top of Ivy's heart that would reroute the flow of blood. Second, the Nikaidoh

procedure would correct the rare combination of transposition of the great arteries, pulmonary stenosis and a ventricular septal defect, moving the aortic root to her left ventricle.

First performed in 1983, the Nikaidoh procedure is named for Dr. Hisashi Nikaidoh, a cardiothoracic surgeon at Cook's who pioneered the operation. Together, Drs. Tam and Nikaidoh have performed more of these procedures at Cook's than any hospital in the country.

Surgery was scheduled for Oct. 3, 2016, a Monday. Physicians at Cook Children's did not believe this exact procedure had ever been performed.

To prepare, Tam turned to one of the newest frontiers of medicine - 3-D printing.

#### 'IT WAS LIKE A PIECE OF HER'

Dr. Steve Muyskens held a 3-D plastic model of Ivy's heart in his hand and pointed to the transposed arteries, demonstrating how the blood flowed.

Muyskens, a cardiologist at Cook Children's, developed the hospital's cardiac MRI program, which offers advanced imaging techniques. It is among the few pediatric laboratories of its kind in the country.

But MRIs have limits. They create two-dimensional images, so doctors study the scans and then try to imagine what the heart would actually look like.

Enter 3-D printing. Using MRI scans, Muyskens created a digital image of Ivy's heart and uploaded it to a 3-D printer. A few hours later, Cook's had an enlarged three-dimensional model of Ivy's heart.

"I could hold her heart in my hand, rotate it, look inside and figure out how to repair it," Tam says. "This advancement absolutely facilitates better discussion and planning."

This marked the first time Cook Children's used 3-D printing to plan for a surgery. The hospital will open a new 3-D laboratory in January to support pre-surgical planning and education for patients and their families.

Before the operation, doctors showed Elizabeth and Efren their daughter's heart and walked them through the procedures. Elizabeth remembers holding the piece of plastic in her hand.

"It was amazing," she says. "It was like a piece of her."

In Odessa, Ivy was turning into a toddler, with the help of heart medication and A surgical shunt.

Small for her age, she still managed to climb on top of the dining room table to swipe fruit sitting in a bowl. She adored Mickey Mouse, loved to scribble with markers and followed her siblings - Iris, Sandy, Ely and Efren Jr. - everywhere they went.

Seeing Ivy growing stronger, Elizabeth and Efren began to wonder if the surgery was necessary.

"After we brought her home from the hospital, we never thought of her as sick," Elizabeth says. "She never got colds. She slept through the night. She was a normal kid."

Doctors told them the medicine and shunt were only temporary. Ivy, they said, needed surgery to survive.

But Elizabeth and Efren worried about the consequences. What if their daughter never fully recovered from the operation? Or worse, what if she did not make it?

The day before the surgery, the three drove from Odessa to Fort Worth. The couple wept most of the drive. They imagined turning around, returning home in time for dinner and tucking Ivy into her own bed.

But they kept going.

On surgery day, a Monday, the Chacones arrived at Cook's at 6:30 in the morning to prepare.

Three hours later, they kissed Ivy goodbye and collapsed in seats in the waiting room.

At 1:30 the next morning, Elizabeth and Efren met Ivy in recovery. Sedated, she was covered beneath a blanket of tubes and an oxygen mask, her tiny chest still open. She wore a mint-colored bow in her hair. Elizabeth cried.

"We're still here," she told her daughter. "We love you so much."

For a week, Ivy's fever soared, then plummeted, again and again. She remained sedated. Elizabeth thought she looked tired and uncomfortable.

They prayed. At night, one parent slept with Ivy in the recovery room, while the other stayed at the nearby Ronald McDonald House.

"I was worried she would never wake up or be the same," Elizabeth says. "I don't really know how we got through it. We just did what we had to."

Day by day, Ivy regained strength, and the surgery appeared successful.

On Nov. 4, a Friday, Ivy and her parents returned home.

## 'HER HEART IS WONDERFUL'

Ivy is now 18 months old with a sly, mischievous smile and a shock of wavy, dark brown hair.

A bright red scar starts at the top of her chest and snakes down toward her stomach. Ivy's sister, Iris, calls it her zipper.

At a recent doctor appointment in Fort Worth, Roten told Elizabeth that Ivy's heart is working perfectly.

"Her chest X-ray looks great. Her heart is wonderful. Every valve is functioning. It looks fabulous."

"It's like y'all put a new battery in her," Elizabeth joked. "She is nonstop."

At home, Ivy often pushes a stool to the front door, trying to escape to the front yard to play on the swing set. She recently discovered cookies are tucked into the pantry, and she sneaks in when she thinks no one is looking. She loves to eat pizza, pasta and enchiladas with green sauce.

"When she's being bad," Elizabeth says, "I just remember her lying in the hospital, and I feel happy she gets to be bad."

Sometimes, people call Ivy a miracle baby. Relatives call her la nina guerrera, the girl warrior.

During a recent checkup, Ivy lay on an exam table, while Kim Hill, an echocardiographer at Cook Children's, hooked her up to the ultrasound machine.



An echocardiogram, Hill explained, uses sound waves to produce images of the heart.

Fussy at first, Ivy settled in to watch Mickey Mouse on her mom's cellphone.

Blotches of blue and red appeared on the monitor, showing a clear image of Ivy's heart and [blood flow](#). In a moment, the sound of her heartbeat filled the quiet room.

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