

Operation on Fetus's Heart Valve Called a 'Science Fiction' Success

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Denise Grady

BOSTON, Feb. 21 — Soon after he was born last November at Brigham and Women's Hospital, a baby named Jack was wheeled through a passageway to Children's Hospital, where cardiologists stood by, ready to perform a procedure to open a narrowed valve in his heart.

To the doctors' surprise and delight, Jack did not need their help. His aortic valve, though a bit narrow, was wide enough to do its job, and the chambers of his heart looked healthy. Even though he had been delivered six weeks early, he was robust and breathing on his own.

The medical team had already widened the valve once, in a daring procedure months before, while Jack was in his mother's womb. But the doctors were not sure the valve would stay open. Now, examining the baby, they realized that their repair job had worked even better than expected. They had, for the first time in the United States, corrected a deadly heart defect in a fetus.

"We couldn't believe it," said Dr. Stanton Perry, one of the cardiologists who had worked on Jack. The team was so elated, he said, that it did echocardiograms over and over, just to watch Jack's beating heart.

Jack's doctors believe that by opening the pinched valve during the 23rd week of pregnancy, they prevented a devastating disorder, hypoplastic left heart syndrome, in which the left ventricle, the heart's main pumping chamber, stops growing and becomes scarred and useless. Such babies are, in essence, born with half a heart.

Untreated, the condition is fatal soon after birth. Treatment requires extreme measures: three heart operations, with a 30 percent death rate and a total cost of at least half a million dollars. Even when the surgery works, the heart is still far from normal, and survivors may ultimately need transplants. The syndrome is what drove doctors at Loma Linda University to perform a desperate operation in 1984, in which they transplanted a baboon heart into an infant who died 20 days later.

An estimated 600 to 1,400 children a year are born in the United States with hypoplastic left heart syndrome, and doctors say that when it is diagnosed in a fetus, many couples end the pregnancy. Many cases, but not all, are brought on by a narrowed aortic valve.

Jack's heart repair, performed through a needle inserted in his mother's abdomen, may offer hope of preventing at least some cases of the syndrome. The operation may also encourage doctors to consider whether other heart problems might be repaired in fetuses.

Even though doctors have been performing fetal surgery since the 1980's, most fetal heart surgery is considered too risky and difficult.

The success here, Dr. Perry said, "has the potential to open up the whole field."

Dr. Audrey Marshall, another of Jack's cardiologists, said, "We think that with a lot of congenital heart disease, some insult or injury occurs early in development, and the sooner you correct it, the more normal these kids can become."

Doctors caution, though, that fetal procedures must be done only for conditions that would be fatal or severely disabling, because operating exposes both mother and fetus to all the usual hazards of surgery and anesthesia, plus the risk of early labor and the physical and developmental problems of prematurity.

Jack's story began last summer, when ultrasound scans at about 20 weeks of pregnancy showed that his aortic valve was severely narrowed and his left ventricle barely working. Doctors felt sure he would have hypoplastic left heart syndrome.

To his parents, Jennifer and Henry G., who spoke on condition that their last name not be used, the options seemed bleak. They could end the pregnancy. They could have the baby, decline surgery and let him die. Or they could put him through three heart operations.

The couple, college-educated professionals in their 30's, sought help at Children's Hospital.

There, Dr. Wayne Tworetzky, a cardiologist, described a fourth option, what he called "the science fiction procedure." He said the ventricle could not pump blood past the pinched valve. If the valve could be opened, the ventricle might be able to grow and function normally. But the procedure would have to be done soon, before the damage was irreversible.

The approach was science fiction because no one in the United States had ever made it work. From 1989 to 1997, only 2 of 12 babies survived attempts at the procedure, according to an article in *The American Journal of Cardiology* in May 2000. The operations were done at six medical centers in England, Germany, Brazil and the United States. In the case of one survivor, the technique had failed completely, and the baby required surgery after birth. In the other survivor, the valve closed up again and had to be reopened several times after the child was born. Nor was it clear whether hypoplastic left heart syndrome would have developed without the procedure.

The team at Children's had also tried the technique in 2000, but it was unable to get the needle into the fetus's heart.

This time the fetus was younger, and Dr. Tworetzky and his colleagues hoped he might have a ventricle that was less scarred and easier to pierce.

Mr. and Mrs. G. decided in favor of science fiction. If it failed, they would accept the three operations.

Opening a narrowed heart valve may seem like a simple idea, but the task itself becomes incredibly difficult when the patient is a 23-week fetus whose heart, a moving target, is the size of a grape. There are no standard instruments for a procedure on such a tiny heart.

On the morning of Sept. 13, a dozen doctors, nurses and technicians gathered in an operating room at Brigham and Women's Hospital. Mother and fetus were anesthetized, and Dr. Louise Wilkins-Haug, an obstetrician, carefully kneaded Mrs. G.'s abdomen and rolled the fetus over to give the doctors better access to his heart. When the fetus was in the right position, Dr. Rusty Jennings, a fetal surgeon, held him firmly in place. The positioning, guided by ultrasound, was a crucial step, and it took several hours.

Since obstetricians, who routinely perform amniocentesis, have the most experience inserting needles into the uterus, it would be Dr. Wilkins-Haug's job to insert a long needle into the fetus's left ventricle. She would be guided by ultrasound images on a monitor.

The needle had to be perfectly angled toward the aortic valve — a spot about an eighth of an inch in diameter — and it had to avoid piercing coronary arteries or parts of the heart that might touch off disruptions in heart rhythm. Dr. Wilkins-Haug had only one shot. It would not be safe to jab the heart repeatedly.

With her first thrust of the needle, she passed through the mother's abdomen and the uterine wall, stopping just outside the fetus's skin. Studying the monitor, she realized her angle was slightly off, and corrected it. She pushed the needle into the ventricle. Its position was perfect, and she held it tightly in place.

Now it was time for Dr. Perry and Dr. Marshall to step in. Watching the monitor, they passed a threadlike wire through the center of the needle held by Dr. Wilkins-Haug, into the ventricle and through the tiny opening of the aortic valve. With the wire in place, they slid a catheter over it, carrying the same kind of balloon used to dilate blocked arteries in adults. They inflated the balloon to about an eighth of an inch, and passed it back and forth several times. Then they pulled out.

Blood flow through the valve began to improve almost immediately. The work had taken 20 minutes.

"I remember coming through the anesthesia and being aware of this general happy chatter around me," Mrs. G. said. "Someone, I think Dr. Marshall, put a hand on my shoulder and said: 'We did it. We dilated the valve.'"

Eleven weeks later, doctors decided to induce labor so that they could widen the valve again.

Even though he was born six weeks early, Jack emerged "kicking and screaming," his mother said, weighing 5 pounds 8 ounces and breathing on his own. His valve did not need more widening.

Jack's outlook is good, although the valve remains narrower than average. As he grows, Jack may need procedures to widen it, and someday it may need to be replaced. But what counts more is that Jack has two ventricles, not one.

"Basically, he'll be a normal little boy with absolutely no restrictions," his mother said. "He can do anything, play any sports he wants to."

Jack's doctors are well aware that medical victories cannot be declared on the basis of one patient.

"The next 10 cases could be unsuccessful, of course," Dr. Tworetzky said. Nonetheless, he said, "We showed it can be done."

Experts at the University of California at San Francisco, a leading fetal surgery center, received the news about Jack with caution and enthusiasm. Dr. Michael Harrison, director of the fetal treatment program and widely regarded as the father of fetal surgery, said: "I think it's fabulous. It's the right thinking, and the right way to get at it."

But, he added, "Single cases can never prove a concept."

At home last Wednesday night, peering out elfishly from beneath the pointy hood of his blue sweater, Jack sucked down a bottle of formula and dozed off, snuggled up on his mother's chest. Jack's 2-year-old sister, up too late, burst into tears over a missing book and was swept off to bed. Visitors fussed over Jack. His mother looked down at him and said, "We were handed a miracle." Jack, oblivious, smiled in his sleep.

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