

CHAPTER FIVE Intervention

"We had to do something."

Dr. Guleserian knew of Rylynn's case almost from the start. Her surgical partner, Dr. Joe Forbess, performed Rylynn's first two surgeries, and the surgeons often briefed each other on cases. In her capacity as surgical director of transplantation, Dr. G was part of the team that evaluated Rylynn and approved her listing for transplant the previous December. Rylynn and her failing heart worked their way on to Dr. G's caseload for good when the two-year-old was admitted to Children's on July 18, 2011.

When I was first called to the CVICU it was obvious that Rylynn was in severe heart and lung failure. She was on the verge of intubation. In December, she was in heart failure but, as is often the case, the cardiac transplant team could manage her as an outpatient on oral heart failure therapy. By May she needed IV milrinone (to increase the heart's pumping function and decrease vascular resistance) and that's when her UNOS status was bumped up to 1A. Her heart was failing but at least her other organs—her kidneys, liver, and intestines—were still OK. Now, in July, things were much worse—she looked terrible. The IV milrinone was already maxed out. I didn't think she would survive.

It wasn't just her heart. Her other organs were failing because of low cardiac output, so the next logical step was mechanical circulatory support. But Rylynn was already a high-risk transplant candidate. Not only was she a single-ventricle patient, she was what we call 'highly sensitized.' She had markedly elevated panel-reactive-antibodies (PRA—the higher the PRA number, the more difficult it would be to find a matching heart) from her prior surgeries and from exposure to blood products. We knew that if we implanted a ventricular assist device, in her case one called a Berlin Heart, she would be exposed to more blood products, which theoretically could increase her already high antibody level, making it even more difficult to find a suitable donor heart. Unfortunately, you never know when a donor heart will be available. There is no crystal ball. Knowing that only one-third of single-ventricle patients who are on VAD support even survive to transplant, the odds were against her. But we had a child whose heart was failing right in front of our eyes. We had to do something.

From SURGEON'S STORY by Mark Oristano with Kristine Guleserian, MD

Beginning in the early 20th century different artificial hearts and ventricular-assist devices were developed by any number of people not always trained in medicine. In the 1930s, Alexis Carrel and Charles Lindbergh created one of the earliest perfusion devices, designed to supply blood to a body organ to keep it alive. Each man was already world famous. French physician Carrel won the Nobel Prize in medicine in 1912 for his vascular suturing techniques. Lindbergh was the first man to fly solo across the Atlantic, in 1927. The perfusion device they invented was never used with humans, but it was their theory that led to the development of the heart-lung bypass machine in use today. In the 1950s Paul Winchell, a popular children's television ventriloquist, designed a rudimentary artificial heart, among many other inventions. Winchell met Dr. Henry Heimlich, the man credited with the maneuver to prevent people from choking on food, and Heimlich encouraged Winchell to work to improve his artificial heart design. Winchell patented the heart in the early 1960s. It was never used in human patients.

The first patient to receive an artificial heart was Barney Clark, a dentist, in 1982. He lived for over three months tethered to a machine weighing 400 pounds, powering the device in his chest. Dr. Michael DeBakey and Dr. Denton Cooley, two legendary heart surgeons from Houston, contributed significantly to the development of artificial valves, and the total artificial heart. The technology dreamed of in those earlier days would play a major role in Rylynn's treatment. But before they could take advantage of technology, the team treating Rylynn had to deal with bureaucracy.

The Berlin Heart is a German invention, as its name would suggest. It's a ventricular-assist device designed to augment or totally replace ventricular function in patients with end-stage heart failure. The Berlin Heart wasn't even approved by the Food & Drug Administration for pediatric use in the US during the period when Rylynn's condition was worsening, except on a case-by-case "compassionate use" basis.

While we were taking care of Rylynn, trying to do the right thing for her, I couldn't reach anybody from the FDA or the insurance companies to get the special approval required to implant a Berlin Heart in a pediatric patient. I was at the mercy of the system, watching Rylynn get sicker because it wasn't normal business hours. It always seemed like every holiday weekend a child like Rylynn would be admitted to the CVICU in the middle of the night in de-compensated heart failure, maxed out on medical therapy, and in urgent need of mechanical support. But the

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government and the insurance companies only work Monday through Friday, nine to five—no holidays.

Rylynn was born with hypoplastic left heart syndrome. Her left ventricle never worked. And now her right ventricle, her only functioning ventricle, was failing. Rylynn initially did very well, but she developed severe right ventricular dysfunction and severe tricuspid regurgitation. Her right ventricle, her only pump, was failing, and the valve between her right atrium and right ventricle was leaking badly. She would never be a candidate for the Fontan procedure, the third in the series of surgeries for kids with HLHS. The only surgical option would be a heart transplant.

Not everybody who needs a heart is automatically an eligible candidate for transplantation. There's a rigorous order of medical, logistical, and financial considerations for people seeking a place on the list. U.S. Department of Health and Human Services statistics show that an average of 79 people get organ transplants every day in the United States, while 22 people who are waiting for transplants die. Recent statistics show that on a given day, some 400 children are on the waiting list for new hearts, and around 50 die each year while on the list and waiting. Supply and demand is the first barrier.



Andrea and Rylynn in the Children's Medical Center CVICU.

Rylynn was officially on the transplant list, but nobody knew when a donor heart would become available. Berlin Heart implantation meant another round of open-heart surgery, more

time on the pump, more blood thinners, and a new list of potential complications. Rylynn and her family would have to adjust to living with external tubing and a pump moving blood into and out of her body, possible infection, the need for more blood products adding to her PRA problem, and the added risk of potentially fatal bleeding and clotting. Rylynn was now committed to living in the hospital until a donor heart became available. Rylynn's family was waiting for somebody else to die.

On July 21, 2011, Rylynn was again kissed by her parents and taken to the OR, this time for the implantation of a Berlin Heart-EXCOR, standing for "extra-corporeal," meaning the mechanical pump is outside the chest. Dr. G wrote in her operative report that Rylynn's "liver was palpable (able to be felt) down to her right iliac crest and well across the midline. Her transaminases (an enzyme to catalyze amino acid reaction—an important indicator of liver damage) and INR (a blood-clotting indicator) were markedly elevated, she was severely hypoalbuminemic (low levels of protein in the blood), her creatinine was elevated (indicative of kidney failure) and she was oliguric (not producing sufficient urine), which is indicative of end stage organ dysfunction." In other words, Rylynn's heart had become so weak and inefficient that it was now killing off her own liver and kidneys.

Dr. G ran a scalpel across the already prominent sternotomy scar on Rylynn's chest from her previous surgeries. After sawing open and separating her sternum, Dr. G got her first real look at Rylynn's heart, which she later described as "massively dilated." She spent some time freeing up the adhesions surrounding and obscuring the aorta, vena cavae, and ventricle. As Dr. G freed up the scar tissue, Rylynn's heart rhythm dramatically changed. She went into ventricular fibrillation, a dangerously erratic heartbeat. Her blood wasn't going anywhere.

Dr. G immediately began CPR with open cardiac massage, and then, using defibrillator paddles, shocked the heart back into normal rhythm as if she'd done it a thousand times, which she probably had. It's not nearly as dramatic in the OR as it is on TV. Except that this is a real person and there are no re-takes. Rylynn was given heparin to prevent blood clotting before being placed on the heart-lung bypass machine. Connecting Rylynn to the Berlin Heart meant suturing a cannula in the right ventricle, then sewing another cannula onto the aorta, and tunneling both through the chest wall and into the Berlin Heart pump, all in an effort to supplement the work of her failing right ventricle. The VAD was connected and the Berlin Heart was now the driving force behind Rylynn's circulation. Rylynn was weaned from the heart-lung

bypass machine successfully. The transesophageal echo confirmed that everything was in position and the Berlin Heart pump was working properly. The swelling in Rylynn's liver was already better or, in the words of the operative report, "... it was quite remarkable how her hepatomegaly (liver enlargement) had nearly completely resolved."

Rylynn came out of the OR attached to a blue, file-cabinet-size device. That cabinet was the driving unit of the Berlin Heart pump, controlled by a laptop computer that sat atop it. A small pump, resembling a paperweight with arms, beat outside of Rylynn's chest. Tubes filled with blood ran to and from the pump, to and from her chest. This technology would have to keep her alive until a suitable donor heart was found.

So basically, blood from her right ventricle passes through the inflow cannula, into a pneumatically driven pump, and is redirected back to her aorta via the outflow cannula. The perfectly functioning Berlin Heart pump has essentially replaced her diseased, failing, native heart pump.

Adjusting to life attached to a large machine required both physical and occupational therapy. Rylynn's medications were adjusted—and readjusted—as needed. Only four days out of surgery, the two-year-old could sit up on the edge of her bed, which may not seem like much, but with a series of tubes and gadgets pulling on your chest, your center of gravity becomes displaced, and you have to learn all over again how sitting up actually works.

Andrea and Gilly continued their vigil. Rylynn was able to leave the ICU one week after this latest surgery to visit the hospital lobby for a "Christmas in July" event the Child Life staff organized for patients. When Rylynn's transplant coordinator, Laurie Hinman, heard the tough little girl was eating again she brought Rylynn some Sprinkles cupcakes, the high-end variety. Rylynn ate half a cupcake, her biggest meal since the implantation of her VAD.

It was difficult for Andrea to watch Rylynn go through physical therapy. Her little girl, who should have been running, playing, and carefree, would struggle to stand up, or bend over to pick a ball up off the floor. It was a major effort.

"We were so proud of the way she worked. I know it was hard for her, but she tried her best every time. She can stand up now if she's holding on to something, or holding somebody's hands," Andrea wrote in her blog. "The weekends are the best time. There's only one therapy session a day, and a whole lot less people popping in all the time."

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Lack of sleep was an issue, and Rylynn was given sedation medication. There are numerous reasons for sleep deprivation after major surgery. The bottom line is that lack of sleep slows healing. Sleep deprivation was one thing to be avoided at all costs.

Rylynn and Andrea were interviewed by the Children's Medical Center public relations department for an upcoming TV special called "Children's Med: Dallas." Rylynn's celebrity was beginning to build. That's not surprising, since everybody who walked into her room and saw those huge, gorgeous eyes fell in love with her at first glance. In addition to becoming a celebrity, she was also something of a medical first.

Literally as we were implanting the Berlin Heart in Rylynn, the FDA began the formal process of approving its use in pediatric cases.