



Complex Child E-Magazine

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The Heart of a Fighter: Lorelei's Journey with Hypoplastic Left Heart Syndrome by Nicole Hoagland

My daughter Lorelei was born with Hypoplastic Left Heart Syndrome. This is a story of a fighter beating the odds and surviving.



Lorelei's Story

Lorelei was born on June 2, 2005, at a whopping 8 pounds 12 ounces. She was a perfect, beautiful healthy little girl. We spent two wonderful days in the hospital enjoying her and trying to gear up for parenthood and midnight feedings. Two days later we were discharged and took Lorelei home. As new parents, we were very nervous to start this new chapter in our lives. We loved on her, fed her, changed her diapers and worried about any little thing that was different.

For four days we had the perfect healthy child. And then it happened.

On our second day home I remember feeling a little uneasy during one of Lorelei's feedings. She just didn't seem hungry. Several minutes later she started to breathe unusually, like an asthmatic or an anaphylactic reaction. My husband and I immediately called the pediatrician's office, and in that short amount of time Lorelei's breathing became labored. When the nurse answered, she said, "Hang up the phone and call 911."

June 6, 2005 is forever seared in our heads and hearts. This is the day our picture perfect world was ripped out from under us. When the paramedics arrived they couldn't get Lorelei out of the house quick enough. I remember one of them saying, "Leave the stretcher...she's a baby." As quick as they were in, they were out. Lorelei was in the back of the ambulance with paramedics frantically working on her.

By the time we arrived at the nearest hospital, the paramedics had already called for an airlift team to take Lorelei to the nearest children's hospital. They had no idea what was wrong with her. They couldn't even get an IV line in because her body was going into failure.

At first, I sat in the front of the ambulance trying to pull it together. It felt so surreal. All I could think of was to call my mom and dad because I didn't know if I would ever hold Lorelei alive again. I truly believed that she was not going to make it. I felt like I was having an out of body experience while I watched a dozen doctors and nurses work on her. I could see more tubes and lines than I could see of Lorelei. Watching the helicopter leave was the hardest moment yet as I knew it would be an hour before I could be in the same building as Lorelei. I just wanted to hold her and see her every second.

An hour later, we arrived at Cardinal Glennon Children's Hospital in St. Louis, Missouri to join Lorelei. They still did not know what was going on. We anxiously awaited any news that might give us hope. Finally, after five hours of feeling like our lives were over, we received some news. The hospital's cardiologist came out to talk to us. He said, "You have a very sick little girl, but we can fix her if we can get her well enough for surgery."

I held onto those words like they were pure gold.

Hypoplastic Left Heart Syndrome

Lorelei was diagnosed with Hypoplastic Left Heart Syndrome. In short, this means that Lorelei's two left chambers of her heart are severely underdeveloped and they do not function. Normally, the left side supplies oxygenated blood to your body and the right side to your lungs. But Lorelei only has the right half of her heart to live on.

The next logical issue was determining how this could be repaired. Surgeons typically perform a series of three open-heart surgeries over approximately three years. They are

called the Norwood (typically done at birth), the Glenn (done around four to six months old) and the Fontan, which is completed when the child reaches approximately 35 pounds. During these three open-heart procedures, surgeons redirect the blood so that the right two chambers can pump blood to both the lungs and body. See this Flash Movie on Hypoplastic Left Heart Syndrome for more detailed information.

[<http://www.cincinnatichildrens.org/health/heart-encyclopedia/anomalies/graphicsummaries/hlhsswf.htm>]

So essentially, Lorelei is living with half a heart.

Delayed Diagnosis

In the days, weeks and months ahead I asked myself the same question many times. **How do you go home from the hospital with half a heart?**

Unfortunately, there are no tests in place to detect Congenital Heart Defects (CHDs) prior to discharge, and many CHDs are not apparent until four to six days after birth. During pregnancy, the mom is providing the oxygen to the baby's body, which effectively masks any problems that might result from a heart defect. For the first four to six days of life, a temporary duct remains open in the baby's heart that allows oxygenated blood to flow through. Until this closes, it can be very difficult to detect CHDs. Depending on the severity of the defect, the baby can show distress signs, go into heart failure or even die.

There have been recent studies on why and how to prevent heart failure and death in babies with CHDs. One recent study suggests using Pulse Oximetry, a measure of oxygen in the blood, on newborns after their first 24 hours of life. This test is inexpensive, painless and has a 70% success rate in detecting CHDs. This test is particularly good at detecting the severely affected babies who would be at risk of death if sent home undetected.

Also, there has been talk of performing ultrasounds around 26 to 28 weeks when the heart is more developed. This would greatly reduce the risk of babies being sent home in a critical state, since some CHDs aren't even detectable until 26 weeks gestation.

Norwood Surgical Repair

Lorelei struggled day and night to stay alive for two long weeks in the NICU before she could have her first open heart surgery, called the Norwood. It had been a long two weeks, and I believe I was only able to hold her three times. It was quite a chore just getting her into my arms, but once there, I was in heaven and didn't move an inch for fear of alarming something or clamping off a line.

Lorelei had her Norwood surgery at two weeks of age and did as well as could be expected. That was the second longest and hardest day of our lives. Watching a sick little

baby being wheeled away for open-heart surgery is one of the hardest things in the world. You don't know when or if you'll see her again. And at this point I didn't know if I would ever hold my little angel tube free again.



Of course she had her ups and downs after her open-heart surgery, but overall, recovery went well. I was able to hold her nine days after surgery. She continued to recover and we were able to go home from the hospital on July 4th. Obviously, this is Independence Day, but Independence Day holds a whole new meaning for the Hoagland family. It was our true freedom day from the hospital.

Bringing home Lorelei this time was nothing like bringing her home when she was two days old. We went from worrying about how to change a diaper and how to mix her formula to learning how to hook up the heart monitor, checking her pulse oximeter, drawing up her medicine, giving her medicine, mixing high calorie formula and many other little things. I felt like our world was upside down and that we would never sleep again for fear that she might stop breathing.

During the next four months we took her to numerous doctor appointments and had many sleepless nights and days. As hard as it was to face the future, we had to. We knew in four short months that we would have to walk our baby into the operating room for yet another open-heart surgery. How do you live for today when you are so scared of tomorrow? We did our best...we loved her, hugged her and spoiled her all that we could.

Glenn and Fontan Surgical Repairs and Complications

When she was five months old, we took Lorelei in for her second open-heart surgery called the Glenn. Her surgery went extremely well, and the surgeon even said that it was

“picture perfect.” Within 24 hours of her surgery she was extubated and drinking formula in her bed. I couldn’t believe how good she looked. On the second day, she had juice and was playing.

But then something didn’t seem right again. She didn’t want to drink her juice and I had that really uneasy feeling once again. The doctors mentioned having her transferred out of the PICU that evening, but I was having no part of that, so we stayed.



Later that evening the surgeon’s nurse called to check on us and I said that she was doing fine. As soon as those words came out of my mouth, her heart rate and oxygen levels decreased. Within seconds her numbers were in the single digits. The nurse hung up and Lorelei’s room was immediately flooded with doctors trying to figure out what was going on.

After stabilizing and reintubating her, we were off for a CT scan of her brain. We were informed hours later that she suffered a stroke and was having seizures. The stroke affected the entire left side of her body and they did not know what caused it. You are at risk of having strokes when you have a CHD, but typically it will happen during the open heart surgery and not two days after. We figured there wasn’t much we could do about the stroke. We just wanted to get her better again.

Little did we know what getting her better would involve. After another week in the PICU, we figured out that Lorelei had lost substantial use of her left arm and leg, and she also was showing signs of vision problems. Her left eye would not cross midline. They informed us that she has a form of focal seizures. She stares off to the left, stops breathing, and her oxygen levels and heart rates drop to the single digits, which is not a good thing with only half a heart.

We were discharged ten days later with new medications for her seizures, more oxygen and a whole new set of problems. Once we settled in at home, Lorelei was thrown into all the therapies that we could get her. Lorelei had numerous severe seizures and 911 calls over the next few years. She worked three and a half hard years to walk, she is just starting to talk, her left arm is delayed, her vision is unknown, she wears glasses and has depth perception problems, and she still has a hard time self-feeding and chewing foods. Everyday is a challenge and frustration for her.

We believe that her mind can comprehend so much more than she can express. She struggles to communicate the simplest of things like, "I want a drink," "I'm hungry," or "Help me," and most importantly, she cannot tell us if she hurts or doesn't feel well. This is very stressful when you have a child with a disability. But there is one thing that I always find truly amazing: she never complains and she lights up the world. Lorelei is truly one of the most loving children who you could ever meet.



Now it was time to wait for the next open heart surgery. Lorelei's third open-heart surgery, called the Fontan, happened when she was three. This surgery is supposed to be one of the most challenging ones, but for Lorelei it was probably one of the best. She was in and out of the hospital in one week. Although she did go home on oxygen, she was off of that within a week of discharge. Lorelei's recovery went well and we continued to work hard on her therapies.

Life with Half a Heart

The future for Lorelei is unknown. Someday she may need a transplant, her heart could just fail, or she could have another stroke. The list is endless. All we do know is that we have a FIGHTER on our hands. She fights day in and day out to live.

We are honored and blessed to have Lorelei as our daughter. When the hospital first told us we had a “perfect” daughter, they were right. Lorelei is perfect. She’s perfect just the way she is and I wouldn’t change a second of our lives together.

Lorelei has shown us all what living is really about. It’s not about how much your house costs or who has a brand new car. Life is about living life for TODAY. It’s about showing and telling everyone around you how much you love them.

It’s about a little girl with half a heart who taught us all how to love with a whole heart.



Visit Lorelei’s CarePage at www.carepages.com/carepages/Loreleih