

Samaria



I'm a strong believer that things happen for a reason. My husband and I were living the good life we planned: we had purchased our first home, had a child named Miranda, and were now expecting our second. The day after Christmas 2006, was the day I was scheduled to find out the sex of our baby. My mother attended this appointment with me, to hear the news and help with our then 18-month old. Nothing could have prepared us for what unfolded.

The ultrasound technician discovered an abnormality and our life as we knew it instantly changed. I called my husband and he left work to attend the first of many appointments with specialists. We received sound advice the following week with regards to the Children's Hospital of Philadelphia (CHOP) as a doctor told us, "Take ANY appointment they can give you." This was the best advice we received and it is why we are so lucky to have our little girl alive and well today. She will need additional surgery, but our journey to this point has been a dream come true.

The Fetal Heart Program at CHOP

Our first visit to the Children's Hospital was overwhelming. We met with the director of the Fetal Heart Program, Dr. Jack Rychik, as well as some of the members of his team, which included Denise Donaghue, the nurse coordinator, Dr. Sumekala Nadaraj, an attending pediatric cardiologist, and Joy Haldeman-Englert, a social worker. After a physician performed a fetal echocardiogram, they met with my husband and me to discuss what they knew. Samaria was diagnosed with Ebstein's anomaly of the tricuspid valve with pulmonary atresia. Dr. Rychik told us on a scale of one to ten with severity, Samaria was an eight. Frightening!

The Fetal Heart Team really prepared us for what was to come.



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They explained the whole process from start to finish and answered our questions thoroughly. Each time we visited, many more appointments were arranged. We met with the surgeon, Dr. Thomas Spray, during one of our visits for a fetal echocardiogram. He arrived fresh from surgery and was still in scrubs. Dr. Spray was very thorough, as he explained the three stages of the surgery and the timeline. The first surgery would be performed shortly after birth, the second around the age of six months, and the third at eighteen months. Dr. Spray was very personable and exceptionally knowledgeable.

We toured the Cardiac Intensive Care Unit (CICU) during a following visit for another fetal echocardiogram. I am very glad we took this tour because we saw other little babies with their chests cut open and lots of tubes, wires, and devices attached. This initial experience was both frightening and surreal. However, the tour gave me peace of mind that when our time came, our baby would be surrounded by a calm and healing place. The staff was very warm and friendly.

Another time we met with the lactation consultant, Rachelle Lessen. She was the best! She prepared us for what to expect post delivery. Rachelle explained the importance of pumping every 2-3 hours initially to stimulate the milk production. While I already knew the importance of breast milk for the antibodies, my new challenge became learning how to accommodate my baby with a congenital heart defect. Little did I know the crucial role Rachelle was going to play in my baby graduating from a feeding tube.

Samaria's Birth

The birth of my second daughter could not have been any easier. I endured 36 hours of painful labor with Miranda, but Samaria was different. Prior to delivery, I lived at my mother's house with

Miranda to be closer to CHOP and the Hospital of the University of Pennsylvania (HUP). The plan was that I would deliver at HUP and Samaria would be stabilized before taken through a connecting tunnel to CHOP.

My mom lives in the Northeast, around 45 minutes away from both CHOP and HUP. My biggest fear was not being where I needed to be when I needed to be there. Unfortunately, the Penn Relays were occurring which meant additional traffic in Philadelphia. Fortunately, luck was on our side and the day was ours! We made it to the hospital and I will never forget what my mom told the valet parking attendant: “She’s in labor, REALLY!” I have to love my mom!

My husband drove in from Lancaster while I was being checked out and he made it while I was still early in labor. They moved us to the labor and delivery section, where we were in room #4. It is important to note that 4 is my husband’s lucky number and there was a picture of a little Amish boy in our room, which was an unusual coincidence because we are from Lancaster County. I was given an epidural and the rest of the day was spent watching one of my favorite movies: *Happy Gilmore*. It seemed that fate was on our side!

At 8:00 p.m. Eastern Standard Time with all the doctors in place, it was time to push and Samaria entered the world! I could not even hold her, as she was taken immediately to CHOP, but we were able to get a picture of her. The toughest part of her birth was simply not seeing her as she was moved into another room and stabilized. I was still hooked up to the epidural and could not leave the labor and delivery room. My mother, step-father and husband were able to watch my little baby exit the hospital and I was stuck in that room, crying.

However, we saw her the next day and she was beautiful! The



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plan was set in motion: the doctors were going to observe her and determine what their next move would be based upon her status. If she fell under the 50 % mortality rate, she would be put on the heart transplant list. If she was thriving, she would undergo the three surgeries. We received the news that her first surgery was scheduled for the following Monday.

Time in the Hospital

We were the lucky ones. Everyone we spoke with told us just how talented Dr. Spray was, simply stating; “He is the best.” They were absolutely right and our baby’s precious life was in his hands.

In hindsight, the day of her first surgery and time immediately thereafter all blend together. Our time seemed spent driving to and from the hospital, being there for my baby in the hospital, and spending quality time with my now two-year-old while not in the hospital. My husband worked to provide for us Monday through Friday and then drove to my mother’s house to spend the weekends with us. The cycle continued: little sleep and lots of stress. We spent each day trying to take it all in and make sense of what was going on with our child. In addition to the travel, I pumped my breast milk for when my baby could actually eat. I brought in a cooler full of breast milk to the hospital from what I had pumped at my mother’s house.

I remember every chirp and bell from the alarms. The CICU is full of chirps from the ventilators and bells from medicines finishing. I remember the hours ticking away while sitting bedside of our little baby in a tiny, plastic bed. She shared her unit with three other babies and their families. Some babies would eventually “graduate” and be taken to the step-down unit. The nurses in the CICU are so dedicated to their position. They truly cared for their “kids,” as they would call them.

At one point, I came into the hospital and a night nurse had put a little bow on my baby's head. I was so happy to see her looking so pretty with that tiny bow. When you cannot even hold your child, it is the little things that make the bigger things seem bearable. The day I was able to finally hold my baby, was the day her A-line was removed and she, too, graduated to the step-down unit.

My husband and I were required to take a course on nasogastric tube (NG Tube) feeding and infant CPR before leaving the hospital. The day we expected to have a "car seat test" with Samaria, was the day she decided she did not want to come home just yet. She either regurgitated or had an infection, which resulted in a code blue and CPR. The medical team reacted quickly and gave her CPR for 15 minutes. They contacted the surgeon as well, to discuss whether they should put her on the heart lung machine. Fortunately, Samaria was a fighter and the "episode" or "hiccup" was a just a little bump in the road. We were so blessed that it happened while she was still in the hospital.

When she was five weeks old, we were finally discharged with our little bundle of joy. Samaria had an NG tube for food and medicine. An NG tube is a tiny little tube that is taped to her cheek which enters her body through her nose and goes down into her stomach. She ate with the feeding tube every 3 hours and was allowed one feeding from the breast. We kept little gloves on her hands to prevent her from pulling the NG tube out.

Maintaining Murphy's Law, twice on the first day home she pulled the tube out of her nose. It was nerve-racking to say the least, as she took all of her medicines via the NG tube – and there were lots of medicines! It took about twenty minutes each night to prepare the medicines for the next day. We quickly learned the importance of keeping things organized.

We had a visiting nurse come to check on her, in addition to



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regularly taking her to her pediatrician's office. We kept very busy.

Our cardiologist Dr. R. Lee Vogel was an attending physician from CHOP, who also worked in the hospital's satellite offices. We met with him when we first came home, and he suggested contacting CHOP's Feeding Team for information about no longer needing the feeding tube. The big challenge with cardiac babies is weight gain.

I was instructed to contact the lactation consultant for CHOP, Rachelle Lessen. She guided me over the next few weeks on how to calculate the amount Samaria was taking in from nursing. We rented a baby scale because we had to weigh Samaria prior to and immediately after feeding. We recorded Samaria's weight difference to calculate how much sustenance she had actually consumed. In the course of a day, our goal was 600 CCs of food. All day, every day, and each night, we had to record how much she had been fed by her weight.

Our Little Surprise

Samaria appeared to be flourishing in our eyes. Things were going well and we were, for the first time, excited to go to the cardiologist appointment, in hopes of a good report. We were wrong. At our June cardiologist checkup, Samaria's blood oxygen level was not good, in the 60s. Her "normal" level should be between 77 and 83 due to her surgery. We thought Dr. Vogel was going to be happy with her, little did we know he really saved her by sending us back to CHOP for observation. We left for the hospital with the clothes on our backs and I am sure I was in shock. She looked good; she did not look blue, pale or ill.

As instructed by Dr. Vogel, we went through the emergency room and were admitted. The doctors decided to have a heart catheterization performed to see what was going on inside her

heart. The physician who performed the heart catheterization said he opened the shunt a little bit where it was becoming too narrow. How frightening it was because she did not look sick! We would never have known anything was wrong because Samaria appeared to be flourishing in our eyes.

We left the hospital with Samaria on an oxygen tank. The oxygen was just one more thing to add to her medications and NG tube. I was frightened. It was scary to care for an infant that was so medically fragile. Little things like taking your infant on a walk around the neighborhood suddenly became a big feat because of her tubes and oxygen tank. We were forced to stay indoors the majority of the summer until her surgery because it was so challenging. Germs and the sickness they cause were another big fear and Purell hand sanitizer became our best friend. Before we let anyone touch her, we required them to “purell” their hands first.

Samaria’s feeding tube was taken out in July 2007. I was excited but cautious because this meant that she would take all of her medicines via her mouth in addition to eating. We received additional directions from Rachelle, who gave me lots of encouragement with the feeding tube transition. We were still required to weigh her before and after feedings, and the big challenge now was making sure she was gaining weight. Plus, we had an added challenge of Samaria potentially spitting out the medicines because would have to physically ingest them herself.

Bi-Directional Glenn Surgery

Samaria’s second stage surgery was scheduled for August 23rd, 2007. The plan included our admission to the hospital on August 22nd, for a heart catheterization before the surgery the following day. I am still amazed with the talent and care that exists at the



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Children's Hospital of Philadelphia! Her surgery went smoothly and she recovered peacefully. We were in the hospital for only seven days this time! Samaria left the hospital this time on less medicine and with uncertain need for her to be on oxygen.

Being discharged after her second stage surgery was a different experience. Now we knew what to expect and could anticipate the next steps. We knew we needed to set up an appointment with her pediatrician and her cardiologist again. Life was looking up!!!

Fall and Winter 2007

The fall season was wonderful! In October, Samaria, Miranda and I took part in the American Heart Association's Start! Heart Walk in Lancaster. The heart walk was a great opportunity to meet other families of children with menacing heart conditions.

Samaria began receiving a medication called Synagis that was to help prevent her from getting RSV. She was required to take it every 28 days at the pediatrician's office. Our biggest fear now was the cold and flu season and we were committed to keeping Samaria healthy. It was very frightening when Samaria got her first cold. Both of our girls were sick. Her cough sounded terrible and I slept with my arm around her to prop her up. Fortunately, Samaria was a very happy sick baby!

The winter months were also wonderful and Samaria looked great! Looking at her one could never realize just what she had been through! Our baby's first Thanksgiving, Christmas and New Year's Eve were extremely wonderful. She had her second cold shortly after the holidays and the new concern was Samaria looking blue. She had the appearance of a blue strip down her face at times and back to the cardiologist we went! This is normal apparently for her make-up because her blood will mix at times.

At this point, Samaria's pediatrician was pleased with her physical

progress, but now wanted to focus on developmental progress and referred us to Early Intervention. Early Intervention sent a coordinator and two occupational therapists to our home to evaluate Samaria. Surprisingly, our challenge now became Miranda. The occupational therapist had put dice and a cup in front of Samaria, and waited for Samaria to react, but Miranda kept taking the dice away. When the occupational therapist was ringing a bell, Miranda wanted to ring the bell. Miranda wanted to be involved! She wanted the occupational therapist to be playing with her, and not Samaria!

After the evaluation, they concluded that Samaria could receive physical therapy and special instruction. We were open to any suggestions!

The Strange Tick

As if someone had tickled Samaria's neck, her little head would go to one side for no reason. She would do this frequently and there was no rhyme or reason to it. I really became scared when we were eating dinner one night and she had eight ticks in a row – something had to be going on. While getting ready to meet with an eye surgeon to discuss correcting a blocked tear duct, I came across literature in her binder on one of the medications she had been taking. It read, "Stop taking this medication immediately and contact your doctor if you experience head, neck or jaw spasms." I immediately contacted her pediatrician's office and was advised to discontinue the one medication for the next 48 hours and to report back to them. I was frightened. Was I giving her a medication which was causing her to have this tick? As time passed, she still had ticks after going off the medication. Our next step became having her evaluated to see if the ticks were seizure related.

Back to the hospital we went for a twenty minute electrencephalogram (EEG). In an EEG little electrodes are placed on her



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head and they evaluate her brain waves. I found this test to be the most emotionally challenging one because Samaria was swaddled in a blanket and then further restrained with Velcro straps. Samaria cried in terror, but left the twenty minutes never having had the tick. So our next step was to return to the hospital for a 24-hour EEG in hope that the tick would appear.

Once again, Samaria cried hysterically when they applied the electrodes to her head. For the 24-hour EEG, they had to glue the electrodes to her head verse using sticky wax for the 20-minute test. Once the electrodes were applied, her little head was wrapped with gauze and they put her wires in a little back-pack that was placed on her back. Samaria was a trooper and once we were back in her room, the child life specialist got Samaria a mat for the floor and some toys. Samaria was still upset, and then the best thing happened, a volunteer came in with a beautiful dog for pet therapy. Samaria was instantly calm and so delighted to pet the soft dog. The older dog just laid there, allowing Samaria to touch and pet him. I had brought a few books and a stuffed animal from home, and we now resigned ourselves to make the best of the hospital room, in hopes that Samaria would have the tick. The hospital room was video-taped and when she would have an episode, I was to push button to alert the nurses on duty.

Entertaining a 13-month old is challenging when you are out of your environment. Fortunately, my mother and step-father brought my three-year-old Miranda to the hospital to visit us. Their visit was really wonderful because Miranda has an uncanny ability to put Samaria at ease. Samaria adores her big sister and I am certain that seeing Miranda made her hospital visit more bearable. After they left, we went to the playroom for a little bit to break up the evening.

Fortunately, at 6:30 a.m., Samaria finally had the tick! I pressed

the button to alert the staff and the physician reviewed the videotape along with the brain waves. Her head movements were not epilepsy, just a tick! Thank God!

Early Intervention

A few months passed with Samaria's physical therapy and special instruction every other week. Samaria was beginning to really emerge developmentally! Week after week, we could see her progress. Samaria was given little physical exercises to do. For example, she would go from sitting on a pot to standing. When she started with physical therapy, Samaria would only crawl three feet and get winded. By the time she graduated from physical therapy, Samaria was walking and climbing!

As for special instruction, we practiced speech and incorporated sign language, in hopes of advancing Samaria's communication skills. Samaria now understood more of the world around her, but remained mostly nonverbal. Samaria mastered the signs for "more," "again," and "eat".

Summer 2008

This summer was a gift. Miranda took swimming and dance lessons and Samaria was able to attend her big sister's events. Samaria loves going to the pool and playing in the water. Sometimes she would appear blue in her fingers and lips while in the pool. After checking with her cardiologist, Samaria was okay. She went on her very first vacation this year as well. We went to Sea Isle City, New Jersey. She got to experience the sand between her toes and take in the smell of salt air. Samaria was able to do all the fun things "normal" children do and she was quite happy. Samaria was able to enjoy boardwalk soft serve and go on some rides.

The merry-go-round at Ocean City was her favorite. We went



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to the local playground a lot while on vacation. I'm certain an onlooker would not have been able to guess my little girl was ever sick or ever needed so many surgeries. She looked like a cute little one-year old, trying to keep up with her busy three-year old sister!

The Here and Now

Fall is here and now we are preparing for the third stage of her open heart surgery, the Fontan. Her heart catheterization will go in through her groin and also through her neck due to her heart "plumbing". After her bi-directional Glenn surgery, the blood from her head and arms runs directly into her lungs. The Fontan surgery will connect the bottom half of her body's blood flow to the top half of her body's blood flow and send it directly into her lungs. Somewhat confusing, but the procedure has been around for decades. I find comfort in the saying over the Cardiac Reception Center's wall. It reads, "If it can be done, it can be done here".