Going To Battle With My Granddaughter

Four years ago I became the proud grandmother of the sweetest little soldier you have ever seen. She was a full term baby weighing over 9 pounds and seemed normal at birth. It wasn't until her 8-week checkup that problems were found. The key was her tiny thumb without a joint (knuckle). I know it sounds strange and I could not imagine this being a flag - but during her 8-week checkup her pediatrician indicated that this was a warning that something was wrong. They did tests and discovered several problems including Occult Spinal Dysraphism (OSD), Cross Fused Ectopic Kidney (double kidney), and Capillary Hemangioma (active skin cell benign tumor). I can remember at the time the pediatrician's comments on how unusual her spinal cord looked in the scans. The main concern at the time was the Hemangioma on her shoulder and eye lid. The physicians prescribed tiny glasses and on her way she went.

By the time our little soldier was one she was walking, all the physicians could do was watch in wonder. She seemed normal with the exception of the rather thick glasses and the large hemangioma on her shoulder which required several laser surgeries to stop the benign tumor growth. The year went by quickly and her wonderful personality began to come through. We knew she was in pain but could only comfort her and wait (physician recommendation) as her team of physicians watched to see if she outgrew these problems.

By age two there were more tests and more specialists. Traveling to different specialist became the norm. She became a very active two year old but rarely spoke - other than indicating what she wanted. With her wonderful personality she played and was able to compromise for her speech problem. During this time she had several sinus problems and sore throats so her pediatrician decided to remove her tonsils and adenoids. She seemed to be less congested after this operation.

At age three she was a very active child, had become a big talker (only I found it very difficult to understand what she was saying in her gruff voice). When she said Wal-Mart it was Bac-Dok. She began attending speech class three times a week. She was also diagnosed with Asthma and began using an inhaler. A few months later specialists were called in to examine a small knot on her lower spine and some other problems and the decision was made to continue to watch the problems.

Then came age four (2004) and the mega blow - she went in for her regular checkup and another Magnetic Resonance Imaging (MRI) was done that resulted in a diagnosis of a Urochal Cyst on her bladder, Chiari I Malformation with Cervical Syrinx (on her upper spine) and Coccygeal Aplasia (on her lower spine). I was in shock; I thought the original diagnosis back in 2000 was simply wrong, because she had managed to overcome so many obstacles. We all began an intense search for definitions, resources, other patients and luckily stumbled on the Chiari & Syringomyelia News Website. This sight gave a great deal of information.

I read that Chiari I Malformation is usually considered a congenital malformation characterized by a small or misshapen posterior fossa (the compartment in the back of the skull), a reduction in cerebrospinal fluid pathways and a protrusion of the cerebellar tonsils through the bottom of the skull (foramen magnum) into the spinal canal. This all seemed so hard to grasp and when I thought of what was ahead for my Granddaughter all I could do was pray. My son described our little soldier's MRI as looking exactly like the diagram at the hospital. Her tonsils were elongated and protruded down the spinal canal. The surgery was scheduled almost immediately for the following month. The Neurosurgeon told us he performed several of these operations each month and that if this surgery was not successful they would do a second surgery to remove the cerebellar tonsils. I was so worried about the possible complications from the surgery itself and infection afterwards that I cannot express my emotions.

I come from a family that believes knowledge is power and we would all go into battle with our little soldier. She did not exhibit the usual symptoms such as headache, dizziness, double vision, numbness, hearing loss, etc. so surely they had misdiagnosed her condition. She never seemed tired and kept going like the Energizer Bunny. A month before her diagnosis while walking down the airport concourse she told us she could not walk anymore and that she needed to sit down. For an extremely active child this was very unusual and somehow it all started to come together.

After the diagnosis we made calls and attempted to reach other families that had experience with Chiari but it seemed to us that the Health Insurance Portability and Accountability Act of 1996 (HIPAA) kept us from getting the first hand patient information on Chiari surgeries we sought. I simply wanted to talk to another parent or grandparent that had been through this with a young child so we would know what to expect after the surgery.

On July 16, 2004 our little soldier went into battle against Chiari armed with all her Care Bears. The surgical procedure involved enlarging the posterior fossa (back of her skull) and patching it with skull tissue from above the opening. The procedure took about three hours and during this time the staff prepared us for what to expect. We knew she would be sedated and hooked up to all kinds of tubes and monitors. When they took her to recovery we were allowed to see her in the Pediatric Intensive Care Unit (PICU) where she was heavily sedated on morphine and sleeping calmly. When the pain killer wore off during the night she would wake slightly until they gave her another dose. This went on all through the night and into the next morning. We were encouraged to give her liquid to drink but each time she threw up so it was a long night.

About 24 hours after the surgery the nurses began limiting the pain killer and allowing her to wakeup. We brought her favorite talking turtle and she gleamed with a big smile. That afternoon she was moved to a private room and was allowed to play for a little while. The next day around noon she was released to go home, we were shocked, because she was still in a great deal of pain. She stayed near the hospital that night and on Monday the family drove home. We were told that children bounce back quickly but we all thought that a two day hospital stay was short for that type of surgery.

About 20 days after the surgery she woke up screaming in pain from an excruciating headache so she was taken back to her surgeon and a Computed Tomography (CT) imaging scan was done that did not reveal any swelling or fluid buildup. I was relieved there was no infection and apparently this is normal while the skull is healing. We were told we won't know the outcome of the surgery for several months but we all pray that this will be her last surgery. It's been over a month and a few terrible headaches since the surgery but our little soldier is back to her active self with the exception of no more head-butting (bumping heads with her Daddy).

The research I did on Chiari for my Granddaughter reveled that this problem was discovered over 100 years ago and today there continues to be limited medical research on Chiari. The diagnosis is difficult and the treatments are extremely painful (and not always successful). From what I understand early diagnosis is imperative and can save these children from years of pain and possible paralysis. If children have speech problems, headaches, or any of the other symptoms parents should demand an MRI. We need to arm our little soldiers with knowledge before we send them into battle because with research and early testing Chiari can be defeated.

Would you like to share your perspective? Submit to: director@conquerchiari.org Type "Perspectives" in the subject line.