My Spiritual Journey with ECD

By William Dean, Christmas 2020

What's Going On with Dad?

3 weeks into my hospital stay, my daughter who is working a summer internship 5 states away calls her brother to ask, "What's going on with Dad?" My two college age kids could not figure out why my doctors had not identified my problem. The reality is after three weeks, doctors did not know what we were dealing with but my medical team was working hard to rule out possibilities and come to a diagnosis. Now I was facing my next big step, a pericardial window in the sac that surrounds my heart to allow drainage of the fluid.

My symptoms started at the end of May with an echocardiogram that identified a significant amount of fluid around my heart. I was direct admitted into the hospital….didn't even get to go home to hug and kiss my wife before being admitted. The first catherization immediately pulled 2 liters of fluid off my heart and eventually 4 liters before being discharged for a few days. Doctors were sure it was probably from a virus which typically causes pericardial fluid. But after 5 days at home and a routine follow up echocardiogram, the fluid had returned and I was direct admitted again - no hug, no kiss.

A second catherization pulled another liter of fluid and I was held in my hospital room in hopes that fluid discharge levels from my heart would get down to an acceptable level. The hospital was lonely. COVID-19 was in a spike and the hospital was locked down. With no visitors, the nurses, doctors and even our custodian became my family. So, I just waited and waited for my fluid discharge to drop to an acceptable level – it never did. I was boosted with prednisone to tame the "inflammation" and hopefully aid in the fluid reduction.

I communicated with my Endocrinologist at John's Hopkins who had treated me for 12 years for Diabetes Insipidus. I wanted to keep him in the loop. He responded that it could be Sarcoidosis or Erdheim Chester disease. He said the second is very rare and he had never seen a case before. My doctors were still scratching their heads but ready to release me when my Cardiologist said before I was discharged, let's get a chest CT just to rule some things out. The chest CT came back with an inflamed aorta. My diagnosis was moved to an anureised aorta. So, in my hospital bed, with no family to lean on, I began reaching out to a friend of mine who was the only person I knew who had been through an aorta replacement.

After a couple of days of consultation between doctors, my cardiologist returned to tell me the interior diameter of my aorta was fine and the exterior diameter was due to some form of tissue build up. No anureised aorta. But what was causing it? This is when things started changing just about every day. Lymphoma ruled out, Multiple Myeloma ruled out, Leukemia ruled out. Doctors now focused on my immune system, so a Rheumatologist was called-in to lead my case.

The Rheumatologist explained that to cure the fluid discharge, we had to address the underlying cause and he insisted this could only be determined by taking a biopsy off my aorta that was inflamed. The biopsy was completed. In the meantime, my fluid drainage was not slowing and the Thoracic Surgeon was called in to discuss a pericardial window.

While waiting for the biopsy results, the hospital loosened the COVID restrictions to allow one family member to visit. After three weeks, I was finally able to see my wife and so thankful that she could now be a part of a big decision on my cardiac procedure. So, when my daughter called asking, "What's going on with Dad?", we really did not know. But we did know some very serious stuff was going on with my health.

When the Rheumatologist returned with the biopsy results, it was a good news/bad news message. Good news, it's not your immune system. Bad news is you may have a rare blood disorder - Erdheim Chester Disease, the very disease my Hopkins Endocrinologist had mentioned earlier. Erdheim Chester Disease, or ECD, is a rare blood disorder affecting less than 1000 people worldwide. A diagnosis could only be confirmed with further testing of the sample. Hematologists who had taken themselves out of my case the prior week, were now back in a lead role. I now had a preliminary diagnosis of ECD and I was the first case doctors had ever seen in my small community.

The pericardial window was completed and now doctors set a new drainage target to allow me to be discharged. We monitored the drainage for two more weeks continuing to up the prednisone while waiting for biopsy staining to be completed to confirm the diagnosis which would take another 3 weeks.

At the fifth week of my hospital stay, a decision point arrived to again boost my prednisone when the Thoracic Surgeon decided not to risk infection and pull my drainage tube. We would count on the pericardial window to keep the symptoms at bay while the Hematologists worked on the ECD diagnosis. Five weeks after it began, my heart drain was pulled and I was sent home to figure out where my next step with my health would lead.

My Spiritual Journey

Just a few years ago, the prognosis for ECD was generally a life expectancy of less than 3 years and when diagnosed with a rare disease with this outcome, your life flashes before your eyes. What am I going to do? How long do I have to live? Are there any experts who can help? Where are the clinical trials? Are there treatments? Do I have any options? How will I provide for my family? What quality of life do I have ahead of me? ….or is it time to reconcile the disease may lead to the end of my life?

My challenge was navigating through all of my questions logically so I could make sound decisions, select a team of doctors and a treatment, and with their guidance make informed decisions that will provide the most effective care possible. While in the hospital, I launched into a literature survey to create a short list of physicians who did research on the disease and who could work with my local team of physicians to provide the best knowledge and care for my condition.

At the same time, my spirituality and my faith in God had to be at the center of my healing. Doctors and medicines are the means while my Creator provides the grace for the miracle of healing. I have always tried to be a faithful, practicing Christian for my entire life. When I taught youth Sunday School, if often told students that I didn't have a "rags to riches story" but I am always impressed with people who do and share their stories. That would change with my ECD diagnosis.

Life Goes On … Life is Good

Within a few weeks after my discharge, I had committed to my ECD specialist. Doctors performed a baseline heart and brain MRI to assess the extent of my disease before beginning treatment with Zelboraf. Zelboraf is a new immunotherapy just approved by the FDA within the last 5 years to treat ECD. It has greatly changed treatment options and the prognosis for the disease is now measured in years to possibly a decade. My local doctors worked closely with my expert doctor guiding my plan and treatment. After just two weeks on my new treatment, I noticed a significant improvement in blood flow in my legs and my overall well-being. Over the last few years, just the simple act of standing would weaken my back, making me have to sit down. I could see the problem was dissipating and I could now work in my yard much longer without sitting down. In a few more weeks, blood flow in my legs had improved to where I did not have numbness after standing up. For years, I thought these were just symptoms resulting from getting old since I just hit the 50 mark a couple of years prior. Little did I know, they were symptoms associated with a much bigger problem and now I had a means to treat it.

In December, just 7 months after ECD sent me to the hospital, I completed a new PET scan and a new Cardiac MRI that showed a significant improvement in the disease in all parts of my body. For me, life has improved and I am able to do many things as I did several years ago. I'm on a pathway to healing and I am fortunate that the rigor of my doctors' efforts led to a diagnosis likened to finding a needle in the haystack. My diagnosis may have been made early enough to have minimal scaring on my body at this point in my life. There is no cure for ECD, but I now have a plan to move forward managing the disease and living life to its fullest.

God sends angels to me every day. They come in the form of talented doctors and nurses; they come in the form of those leading and giving their time to the ECD Global Alliance; they come from other patients I have met in the ECD community; they come in the form of friends who have delt with their own health struggles and give me spiritual counsel; and they come in the form of my brave and strong family who encourage me and give their love to me each day.

The answer to my daughter's question is revealed. Today we do know what's going on with Dad. We know what is in front of us and we know our plan to handle it. At this point, my story has moved from “rags” to “riches” because I can safely say that God is in control as I am blessed every day as one of his miracles.

For this Christmas and New Year’s season, I’ve been blessed with a cup that runneth over. “May the Lord bless you and protect you. May the Lord smile upon you and be gracious to you. May the Lord show you his favor and give you his peace.” Merry Christmas.