

Kelly Paprota - ADOLESCENT FMD

I was 17 years old when I was first diagnosed with FMD. I was very active—played on my school’s soccer team, was Class Treasurer for Student Council, sang in my high school show choir, and played violin in the orchestra. As far as I knew I was healthy...I didn’t really think much about my health to be honest. My mom is an RN, and she was good about keeping us on track with routine checkups and things. I figured I was covered!



The only complaint I had was severe headaches that occurred about once a week. My doctor said they were migraines, then he said they were tension headaches...none of the medicine they gave me for them did any good so I just learned to live with them. In the spring of my junior year of high school, Student Council hosted a blood drive in the school’s gymnasium for any student that wanted to donate blood. I decided to donate, even though I felt a little queasy at the thought of it. Those of you who have donated blood will remember that you start off in two little booths, where they do routine checks on your vital signs and your blood iron levels to make sure everything is ok for donating. One of the nurses put the blood pressure cuff on my arm, waited a minute, and said “Hmmm, that can’t be right”.

She pumped up the cuff a second time, and again shook her head. She called for another nurse to come over and give it a try. The second nurse pumped up the cuff again, waited, then exclaimed “What?? This little thing?? Hmmm, I just don’t know...” The nurse then told me that for some reason, my blood pressure was too high today for me to be able to safely donate blood. She said it was probably some sort of fluke, and that I could come by their office later this week if I still wanted to donate. I shrugged it off, having never been told my blood pressure was high before. I was more upset that I didn’t have an out for soccer practice that evening than anything else! My mom was out of town until late that same night, and she came in and woke me to say hi when she got home. Sleepily, I hugged her and told her welcome home. Then, I said, “Oh, and by the way...the nurses at the blood drive today said something about having high blood pressure...I’m not sure what that’s about.” She immediately got her own cuff out of the car to see for herself. (At the time, she was a Home Health nurse so the family car was pretty much a mobile hospital.)

She checked it and calmly said, “Change your clothes; We’re going to the hospital”. Of course I protested...I mean it was midnight, I was in my pajamas, and I felt totally fine! She insisted. She tells me now that my blood pressure was upwards of 200/140 at that time.

We got to the ER, and were seen by the triage nurse. My mom shared her concerns, and the ER nurse checked my BP with some sort of machine there in the nurses’ station. I remember that it started beeping loudly both times she checked it (a warning or error beep?). They put me in an ER room, hooked me up to some machines, and monitored me for a while. They admitted me after a few hours, when my BP dropped dramatically upon standing and I almost passed out. Over the next few days in the hospital, I had lots and lots of tests done to help the doctors figure out what was making my BP so high. I was very frustrated all this time, because I felt totally fine and couldn’t understand why I needed to

be missing school and other activities. The doctors were frustrated too, because all their tests kept coming back normal.

I had MRI's, CT scans, CAT scans, blood tests, you name it. On the fourth day, my doctor came in to tell me that I was being released since they couldn't find anything wrong. "It must be just essential hypertension", he said. My mom was very upset at this decision, and knew this couldn't be right. I just wanted to go home. Then, about 20 minutes later, my doctor came back in the room and said "Never mind, we just got the results of your renal scan back and we need to run some more tests". They did more renal and arterial scans, and gave me the diagnosis of FMD the next day. Of course I hadn't heard of it, and my mom hadn't either. The doctors went on to tell us that my right renal artery was 90% occluded (blocked), and my left was about 20% occluded. This was what was causing my high blood pressure, they said. They also told me this was the likely cause of my severe headaches, and that I was lucky I hadn't had a stroke. These were sobering words, for sure. The treatment the recommended was to do a "routine angioplasty" on the right artery to open it back up for blood to flow through. The left was not severe enough to warrant treatment. I agreed to the procedure, and they prepped me for it. I remember the doctor quickly giving me his spiel on what they were going to do during this and how it would all work right before going in for the procedure. He told me I would be conscious, because I had to be able to hold my breath and move slightly if needed for them to get the wire where it needs to go. I told him I understood, and we were ready to go.

Right at the end, the doctor added, "This NEVER happens, but I have to tell you just in case...in some cases during an angioplasty, the artery can tear...and we do have a backup surgical team on call to go in and repair that tear if that were to occur. But it never happens, don't worry." And then I was wheeled in. The angioplasty itself was quite a strange experience. It's an interesting sensation to have this little wire poking around inside your body...and a big screen above you where you can see it moving around in your arteries...and doctors talking about you to each other as if you aren't awake yet you are...its very surreal. Finally, they got the wire into position and inflated the tiny balloon. I heard them say they weren't entirely satisfied, so they were going to inflate it a second time. All of a sudden, an indescribable gut-wrenching pain ripped through me, and I heard all sorts of confusion and shouting of orders from the doctors.

There was chaos that to me seemed like hours...I remember screaming in pain and not understanding what was going on. A nurse told me that my artery had ripped during the procedure, and that I was going into emergency surgery because I was bleeding into my body cavity. I thought I was dying. I might have been. I cried for the anesthesiologist that they told me was coming....I remember asking over and over when he was coming in because I wanted the pain to stop. Later I was told this all happened in less than 10 minutes, but it felt like a lifetime. The next thing I knew, I woke up in ICU struggling to breathe. I had a respirator tube in my throat and I didn't realize it was breathing for me. I think my whole family was in the room, too. I was confused at first, but they told me what had happened.

My artery had "shredded" during the procedure ("That NEVER happens, don't worry" ...) and I was bleeding internally. They had rushed me into emergency surgery, put me under and made an incision from the bottom of my sternum all the way down my abdomen,

stopping 3 inches below my belly button (they went around it!). Then, they made an incision on the inside of my right thigh. They grafted a piece of artery from my leg onto my right renal artery to replace the portion that had torn. They stapled me back up again, and I woke up a while later in ICU. The recovery from this emergency surgery was painful, both physically and emotionally. I was in the hospital for another 8 days following the emergency procedure. I was fed through a feeding tube for the first few days, and I lost 13 pounds (when already thin) and looked completely emaciated by the time I went home. As a 17 year old girl, you can imagine the horror and shock I felt when I saw my abdomen stapled all the way down for the first time. Also, it was painful to stand up straight and walk since they had cut through my abdominal muscles.

All of my many visitors from school were given the task of making me walk around the hospital wing with them, holding onto my IV tree, to help the healing process. Even though it was a near-death experience (the doctors admitted afterwards that they weren't sure what the outcome would be), the solution that had ultimately occurred ended up being the best treatment possible for my FMD. Now that I have a graft leading into my right kidney, the FMD can't affect that area again. As for the other side, I am keeping an eye on it. I do have to take medication every day for mild hypertension, however.

My blood pressure returned to normal for a few years, but then started staying at about 130/90 consistently. It is well controlled with a low-dose ACE inhibitor and regular exercise. I no longer have severe headaches. Even though what happened to me was pretty traumatic and also painful, I can still say I am glad things turned out the way they did. It gave me a better appreciation for my life and my body, and I don't take those things for granted anymore. I wouldn't even want to get my scars removed if I could, because I see them as fond reminders of a hard part of my life that made me stronger. Few people with FMD will experience what I did, but I hope that reading my story will prepare them for the worst... the part that doctors like to say "NEVER happens".