Tracey & Alex Toto - (Mother and Daughter)

When I was 13, I needed a physical to go to camp over the summer. At that routine physical it was noted that my blood pressure was 180/110. The doctor told my mother that I was probably just anxious and not to worry about it. After a few repeat readings and a few weeks spent in the hospital over the next summer for tests, it was determined that the likely culprit was renal artery stenosis due to fmd. Over the next summer, by now I am 15, I had 2 unsuccessful angioplasties and one unsuccessful renal artery bypass. Lucky for me it was only affecting my right renal artery. My parents at that point got cold feet, and declined any additional surgeries.

A wonderful hypertensive-nephrology doctor who was just starting his own practice 'found' me and I have been medically managed since then. I have been on vasotec, an ACE inhibitor, since I have been 16 (with brief periods on different drugs during my pregnancies and when I was trying to get pregnant). I carried 2 babies to full term, with just a brief scare of preclampsia at the end of my second pregnancy. My right kidney, after the unsuccessful bypass, started to get smaller, while my left kidney compensated by getting larger and larger. My most recent urine and blood test showed normal kidney function, even with a 3cm right kidney. I have been very lucky.

Unfortunately, I had no inkling this could be genetically passed down to my children.

A year and a half ago, I was out playing tennis and a sitter was home with my then 3 year old daughter. She was to turn 4 in one week. When I arrived home they were playing in the driveway. I immediately noticed that the left side of my daughter?s face was totally drooping and her eye totally popped open. I feared she had had a stroke and took her straight to the pediatrician. My doctor calmly told me that she had a bell's palsey and that it would go away over time. No need to panic.

Well, I panicked. The next morning I brought my daughter to her ear, nose and throat doctor. She had never seen a bell's palsey in such a young girl and prescribed an MRI to make certain nothing else was wrong. I had to literally strong arm my pediatrician into agreeing with the MRI. She didn't want to subject Alex to unnecessary anesthesia. Ha ! Who could know how many times my little girl would be put out in the next 4 months?

At the MRI, the pediatric anesthesiologist came out after the MRI was complete. He did not want to panic us, but unless we agreed to go directly to the nearest ER he was going to have to call an ambulance. He explained to us that even under anesthesia my daughter's blood pressure was 200/100.

We went directly to our local ER. There a fantastic cardiologist took my history and explained that she probably had fmd, and that if he were us we should get ourselves transferred to Columbia Presbyterian, in case she had to have surgery or something. What a surreal ambulance drive. Once we were there we had to do the same ER thing and wait until 2am for a room. They did a nuclear scan there and decided that she had unilateral renal artery stenosis. (Which turned out to be wrong). They insisted that the bell's palsey had nothing to do with the high blood pressure and the fmd and they released us with no medication, just an appt to come back in one week to see the nephrology doctor. Alex turned 4 during that hospital stay.

I could not get out of my mind one line I found on this web site: "and sometimes the severe high blood pressure causes facial palsey in children." I emailed Pam Mace for the name of a pediatric doctor familiar with fmd in the northeast. She immediately called me back with Dr. Meyer's number at CHoP. I called him, he took my call immediately and we went down there that very day. That day marked the beginning of a 20 day stay at CHoP. I think I cried one million tears during those 20

days.

Alex underwent 2 CTA scans, 3 angiograms with attempted angioplasties. Thirteen of her 20 nights were spent in the ICU. It was determined that her right renal artery was 75% closed and her left renal artery 90% closed. Alex and I both have the smooth kind of fmd, not the typical string of beads. The angioplasties didn't really do the job and the best they could do was get her blood pressure under control with medicines and send us home with the name of a surgeon in Michigan who might be able to help her.

Alex was sent home on a Sat in mid November and we went to Michigan at the very beginning of December. Dr. Stanley performed her bilateral renal artery bypass (one artery was bypassed, while on the other the affected part of the artery was cut off and the good end re-implanted in her aorta). After a few scary moments in the PICU, Alex went home 12 days later, right before Christmas. We were overjoyed.

That was when the real trouble began. Almost immediately she had pain after ever meal she ate. I spoke with Dr. Stanley repeatedly. She kept getting dehydrated and ending up in the local ER once a week for fluids because she wasn?t drinking. She didn?t sleep at night because she was in so much pain. I took her to a gastroenterologist who performed numerous tests and eventually told me "it was all in her head." She lost 25% of her pre-surgery weight. I paced with her in my arms for almost 6 weeks.

Eventually I took her to my son's gastroenterologist who called Dr. Stanley's office. He was on vacation but he spoke with the other surgeon in Alex's case. It was known that her SMA was affected by the fmd, but she had never had pain after eating before. Apparently the significant drop in her blood pressure was enough that the blood was no longer getting through her SMA and she had intestinal aschemia. She would have to have another bypass surgery, this time of her SMA.

I couldn?t believe it, but was almost joyful to have a reason for her 8 weeks of pain. However, I worried about her physical state. I asked the gastroenterologist if there was any way to 'bulk her up' before her surgery. His suggestion was to get a PIC line put in and feed her with TPN. We did this 2 days later at a children's hospital in NJ, yet another 3 nights in the hospital.

February 5th we arrived in Ann Arbor, Alex had a quick angiogram to see if her SMA was indeed affected. It was. She had her next bypass surgery the next morning. This surgery ended up being a re-implantation of her SMA into her aorta. The surgery was a success. She ended up with a blood clot after the surgery which actually mimicked her pain before the surgery. That took about 2 ½ weeks to clear up. We went home to NJ after she ate her first McDonald's hamburger (yes I cried in Mickey D's).

In May Alex had an MRA of her carotid arteries to see if they were affected by the fmd. It did appear that her brain arteries were affected, not the lower carotids, but up higher in the brain. We elected to go to Boston, to Dr. Scott, for an evaluation. Her brain angiogram in Boston showed normal arteries, she had had a false positive on that MRA. We felt like we dodged a bullet.

It is now one year since Alex got home from Michigan and we are a very thankful family. She has had access to the most amazing doctors I think I have ever come across. Both her nephrologist Dr. Kevin Meyers and her surgeon Dr. James Stanley have been wonderful. Not only are they both fantastic at what they do, but also patient with me as the parent (and I don?t let much go without questioning it!) Alex takes a very small dose of vasotec twice a day to keep her blood pressure under control (same medicine as her Mom). She is monitored with blood draws every 3 months and I take her blood pressure every Monday morning. She is now 5 ½ years old and will start kindergarten in the fall. Her SMA is monitored by whether or not she has pain after she eats.

Eventually she will have to have another angiogram. We have decided to go back to Michigan for any additional tests, but until Dr. Stanley says we have to, clinical follow-ups are enough for me. Knock on wood my disease hasn?t progressed since I am a teenager (and I am now 39 years old) and hopefully Alex will follow the same path.

My next concern is to do everything I can to help the scientists out there that are trying to find the gene responsible for this disease so that when my little girl goes to have children in 20 years she will be able to screen her embryos for this. It is done for other diseases, so I can dream, right? We can't be sure but my grandmother had unexplained high blood pressure her whole life and my great-grandmother died of renal failure in her late 40s. It appears that fmd strikes the women in my family.

Lastly, we spent over 70 nights in hospitals and hotels away from our home trying to heal our little girl. Flying 5 states across the country. We recognize that it is a huge financial burden on parents and since then have worked with the University of Michigan to set up a fund for travel and living expenses for parents who would otherwise not be able to bring their children to Michigan to seek out surgery by Dr. Stanley. We have hit up friends, colleagues and family to contribute to this. Please contact me if you would like to contribute to this fund or if you need financial help. A social worker attached to the University of Michigan Medical Center is charged with determining the use of the money.