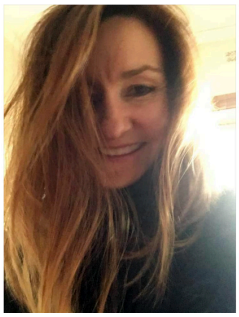
A Teenager's Immortality



I remember, as a child growing up in the country, helping my Dad to gut and prepare chickens for our family to eat. My family were big believers in self-sufficiency as much as possible and using all resources fully to reduce our footprint on this world. I guess we were environmentalists before that was a thing. I always wanted to know how things in the world worked so I would ask him a thousand questions about the organs we removed:

"What is it? What does it do for the body? How does it work? How is blood made? Are all animals the same?"

My Dad was a very patient man and very knowledgeable but there would be things he couldn't answer and then he would tell me that I should go to University to study and find the answers for myself. So, I did. At 17 I went to University to do my first year of Medical Science and loved every minute of it. In one of my holiday breaks I organised to do work experience at a small country hospital laboratory near home. During my stint there I was called into the boss's office. I was really nervous. Had I broken the million-dollar blood analyser I had used that morning? What he had to say was something that had never crossed my mind.

Earlier that day when I was running the routine CBC's (complete blood pictures) for the lab we had taken my blood to use as the normal control. Unbeknownst to me the staff had swapped it out as soon as they noticed my blood was far from normal and had spent the day doing blood films and other tests on it to check the results. It turns out that I had a platelet count of over 1.5million. This of course was well over the normal range. The boss told me they had discovered this and that it could be something transient and benign but could also be a type of leukemia so they had made an appointment for me to see a specialist haematologist that they worked with who would see me the next day in the city. They called my parents and talked to them about it all too. They were incredible people who did as much to soften the blow as they could. The thing I remember most was my Mum being really upset and I couldn't understand why. Maybe it was my teenage feelings of immortality, but I remember feeling calm and had absolute confidence that whatever it was I would be alright, and I would handle it.

And so, the initial ride began. My haematologist was amazing. There were blood tests and clotting assays that he had his staff do for me with no appointment when I would pop in to the lab in between lectures, and a bone marrow biopsy that he scheduled around my exams. It turned out that it wasn't leukemia but rather uncontrolled high platelets due to overactive megakaryocytes in my bone marrow. A myeloproliferative disorder (MPD as they were known then) called Essential Thrombocytosis. Or ET like the 'phone home'

character in the movie as my brother liked to say. As I was otherwise healthy and there was no known cause I was left untreated and just had regular monitoring. As far as I know I was the only patient with this disorder in the state at the time and no genetic markers had been identified world-wide. There was not much known about the condition, so I basically forgot about it and got on with life. My strange bone marrow became a part of me that I owned, and it would never own me. Looking back, I now know that I was having symptoms but didn't recognise them as such. I had nothing to compare them to as I have had ET for my whole adult life. I suffered migraines with huge visual disturbances, I had massive night sweats, I was tired (but what teenager isn't) and my periods were just all clots but (in case any males are wondering) this isn't a thing females usually share with each other so I didn't know this wasn't normal. There were a couple of patches where my legs felt a little numb, but it always returned to normal again, so life was fine. I was a runner and was fit and as healthy as I could be, and I ate well. My platelet numbers reduced slowly with time, untreated and settled at about 750K for many years with the usual fluctuations.

Years later in my 30's after finishing study, working and travelling abroad, and once again back in Australia I had an abdominal ultrasound for an unrelated reason. They found that I had a major clot in my portal vein that was old. I still don't know when it occurred, but my body had set up collateral circulation and got on with its job. The clot was slowly breaking down on its own, but my liver function tests were a little off and I was exhausted all the time, so I went to visit a liver specialist. My haematologist had retired and the next person I saw was a career doctor whose interest lay with becoming the head of department rather than with low priority patients like me, but I did learn that they now knew I carried an acquired genetic mutation, Jak2 that explained my ET and that it was now classed as a chronic, rare form of cancer. Luckily, my new gastroenterologist / hepatolgist was totally on the ball and did a full search for gastric and oesophageal varices and a liver biopsy to check for ischaemic damage. He ended up treating me for the ET as well as the gastric complications. He started me on Clopidogrel as I already had some varices in my gut (so aspirin was not a great idea) but I had no liver damage apart from the altered blood flow through it. I settled into a new balance and was healthy and happy and life continued. I started running properly again and over time my varices disappeared. At 40 I had a baby boy after a carefully monitored but uncomplicated pregnancy. During the pregnancy I experienced what my GP called a state of "super health". My LFT's were normal, my platelets sat in the 500-600K range, my hair, nails and skin were the best they had ever been and I had no night sweats. All was smooth sailing again for another 7 or so years.

Just recently at 48 I had another clotting episode even though my platelets were at an all time low for me. Although we couldn't find the clot, I suffered extraordinary abdominal pain and had a significant rectal bleed where the lining of the bowel stripped out. They think I had an ischaemic bowel after a running race, albeit temporary and with no lasting bowel damage but with high risk varices appearing in my stomach. With my platelet numbers as low as they have ever been for me there was no call to lower the numbers further with cytotoxins so I was put onto Apixaban (a NOAC) as well as Clopidogrel and blood pressure medications.

The last two years have been a massive trial of changing and trialling many different regimes of NOACS and blood pressure meds to try to find some that don't cause me horrendous side effects eg. mental confusion to the point of not knowing the alphabet or being lost in my own workplace, huge sweats, pins and needles in my limbs, heart palpitations, chest pain, closed throat and laboured breathing, heart rates as low as 40 with huge dizziness and two trips to emergency. I can't say it has been fun but after trying all that 3 specialists from different fields and a GP have recommended I have done some of my own experimentation on myself

(with my GP's blessing) and have determined which drugs were causing me grief and at what levels I could tolerate some of them. I think I have finally found a regime that seems to allow me to live a normal life again and protects me as best I can. But with my two biggest risks being bleeding out due to ruptured gastric varices while on dual anti-platelet and anti-coagulation therapy and clotting again it's a complicated balancing act. On the plus side my body seems pretty robust and has coped fairly well with the two major clotting events I have had. I am extremely fortunate to have known about my condition from a young age and have learnt to read my body well and have always known what to look out for. I don't think of my ET as a separate thing from myself. It's a mutation that occurred in one of my cells long ago that my body didn't recognise as foreign and so it has become part of me.