

**So, you got the news** and it wasn't good. Five years tops and the only way out was to have another person die. A haunting thought that would drift to me at night. Imagining someone out there, marked, fated to die unexpectedly and pass life to me. It's a precipice that you are dared to peer over. The edge falls off into a spiral of deep philosophical and religious thoughts that are beyond comprehension. You lie in the dark and just stare into it. My mind would not let me form a picture of that person. At the moment of contemplation, they are living life and loving: leaving you to wonder and trust. As it turned out I would never know the person that I am now forever connected beyond a few details of his medical history. They were disclosed to me in a rush during the small hours of the morning. It was an intense moment. I had to decide if I should take the offer of life he was extending. That event is much later in my story but now when those thoughts drift to me it is not his face I look for but the picture of his family, whose choice it was to make the offer at the time of their deepest grief. What charity is that?

I looked fine and felt fine but a fuse was glowing somewhere inside my body. It's just that there was no smoke. Little was obvious except the occasional puffiness around my left ankle and even that dissipated by the time I awoke. This was the first sign of what would become a growing water retention problem. It is surprising to know that the body is a pretty leaky system. If fluid builds up, it falls, dropped by gravity to search out the lowest spaces. The ankles are an obvious and when severe, the stomach cavity fills: edema and then ascites. A less obvious low spot, for men at least, is the scrotum. There is a small passage between the low stomach and the "equipment" through which the testes had dropped into place during puberty. The pressure of the ascites can force its way through and engorge. It feels like a lead weight pulling down taught on a bunch of wire like tendons in your groin. I once overheard a patient say to a nurse that he thought he was going to have to get a wheelbarrow to carry his cantaloupe sized balls around. Humor is always a way to help manage to the new normal. That particular challenge was well ahead. At that early time, my liver was just beginning to show signs of stress. It was not producing enough Albumin: the protein that keeps water locked up inside cells. Just this small signal was enough to remind me and cast a shadow of concern. As soon as there was an unfocused moment it would muscle its way into my mind, shove aside everything else and root itself. Even in those early days I was tossed by a tempest of uncertainties, every problem I faced had another dimension lurking just behind it. This would be a constant for more than five years and its height would ebb and flow with a tide of symptoms and testing.

An initial anxiety is to decide who, what and when to tell. Of course, my wife had been part of the voyage of discovery, navigating almost eighteen long months of tests and changing theories of why my blood was abnormal. The twists and turns rippled out in diminishing circles like waves through my family. Those closest felt the bumps. Increasingly though, I would have to put things in context for everyone else. Silicon Valley is a place where people define themselves by their work and to run its race you need to clock a 5K in under 20 and have a plan for 15. The symptom that tests your ability to run at that pace is fatigue. As things get worse, no matter how long you sleep you wake up in the morning feeling like you just got off the SFO to JFK red eye. Late night phone calls and video conferences to Asia became a supreme burden to get through. You sense your edge becoming duller but do others see it as well? You convince yourself that there is a silent awareness that something is wrong with your performance. You must reach out to someone to register. It was my fortune that my boss was exceptional and until I was formally listed for a transplant I told no one else. It was enough for me to regain my focus and off load the psychology.

If someone were to come up to you and tell you they had a liver problem I am willing to bet that you would move quickly to the thought that it was alcohol related. When there was a need to

explain what was going on with me it felt like I had to ring a bell and shout, “Unclean! ... Unclean!”. I sensed a rush to judgement that the condition must have been self-inflicted. Indeed, chronic alcoholism is the leading cause of liver disease but it's not the only one: not by far. As a side note I remember filling out a questionnaire on various health topics, a necessary part of medical profiling. When it came to detailing my alcohol consumption I tried to answer honestly and noted that, while I rarely drank at home I would often drink on my frequent business trips to Asia. Somehow that morphed into being labeled a “binge drinker” and that I would need to attend AA meetings in conjunction with any treatment I received. The newness of everything and the power of the bureaucracy had me on the verge of agreeing, until I imagined my introduction. “Hi, I’m Calum and I’m not an alcoholic, not even a heavy drinker. My Doctor asked me to come and check a box.” It was a ridiculous request, a disrespect to those trying to manage their real addiction and the first inclination that the system doesn’t always get things right.



Explaining that I had a genetic condition seemed never enough to satisfy. It was a stretch to get from there to a liver transplant. The problem is, it's quite a story. Around two thousand years ago a genetic mutation expressed itself in some remote portion of the Viking community. The mutation caused a condition known today as alpha-1-antitrypsin deficiency. It provided a benefit to those who had it and so by selection it became prolific in the Viking gene pool. Alpha 1 is a protein designed by the body to throttle back the

inflammatory response in the lung. Deficiency in this “buffer” allows the white blood cells to become so aggressive they will pretty much eat anything: infected tissue, pathogens as well as the good stuff. When lung diseases like bronchitis would savage, having a souped-up immune response in the lungs was a real plus. So, the gene was spread and became entrenched. Driven by overpopulation and enabled by their shipbuilding technology the Vikings ravaged and traded through Europe from Ireland down through the rivers of Russia. Dublin, a trading hub, became their largest city. Alpha 1 came along with them and their progeny inked the blood of many on the Emerald Isle. But, what was a real benefit in the Early Middle Ages turned sour as lifespans grew. The over aggressive immune system in the lungs would finally carve out voids presenting emphysema. But the liver?: the reason for the deficiency in Alpha 1 was because the rest of it had mutated and was trapped inside it. Building up since birth, until the chronic insult gradually damaged it beyond repair: the condition known as cirrhosis. Much of this whole mystery was unraveled in the late 60's by research in Stockholm: except why the lung and liver conditions don't occur together. They could even put a clock on it. Start of the fifth decade of life. And mine was running.

Next up: A voyage of discovery: The winding path to my diagnosis.