



## View from inside

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I do not recall the exact moment I was told that my kidneys were failing. Perhaps because I was slowly drip-fed the news over the course of 5 years. As soon as each outpatient appointment at the Charles Dent unit in London was over, I'd begin reviewing and researching the latest my consultant shared—GFR, creatinine clearance, ultrasound findings and anything else I was quick enough to mentally take note of.

The tests would reveal poor kidney function—my body had finally succumbed to my mismanagement of glycogen storage disease type 1A. My body was overwhelmed and failing in basic self-sustenance.

I work in a clinical environment—I understand standard lab panel data, the Krebs cycle, G6P gene mutation, Kaplan-Meier curves, but when it came to seeing myself clinically, I could not fit myself into any of the models or articles that the internet would throw at me.

I hoped I could draw upon my science background but the GSD paired with renal failure just did not quite make sense.

Meticulously reading journal papers, scouring the internet and social media, I kept trying to find the one piece of information that would tell me that renal failure in those with a rare metabolic disease could be reversed. I'd make detailed notes, action plans and conscientious decisions to implement x, y and z changes—and by the end of the day was too exhausted to do any of it. Physically, mentally and emotionally—just drained.

I realised quickly that facing my present had changed both nothing and everything. Before kidney failure, I spent a lot of time in and out of hospital. After kidney failure, I would spend a lot of time in and out of hospital. So the problem wasn't really that “my life has changed”.

So I wondered why, in the waiting room waiting for my first dialysis session, was I so grief-stricken? Time began to

feel less like the ticking clock and more like a state of being. Stillness settled in but my heart beat frantically.

I knew all the motivational concepts—“everything happens for a reason”, “this too shall pass”, “born survivor”, “hard wired for struggle”. But in that waiting room, I disregarded them as baseless delusions. I eventually settled for pondering upon how I got to this point in life in the first place. This point, where I'd finally succumbed to my lifelong passenger, one that I had named Defeat.

The certainty of the past was easier than this capricious present and this simple act of reflecting was almost comforting.

Like everyone, throughout my 30 years of life I'd followed the question of what makes human life meaningful, but in this moment, Defeat had won and here I was even further away from answering this question.

My childhood with GSD was very much controlled and dictated by a number—the time on the clock, centimetres of an NG tube, scoops of Caloreen and later starch.

Alongside this, I was also wrapped up tight and securely by those around me so no external factors could harm me. My mother's classic line was “my daughter can't take part in school assembly because she has a liver problem”. Soon that word “assembly” started being replaced with other significant milestones in any child's life—nursery, swimming, playing in the sun, camping, dancing, jumping in puddles, and going on holidays—anything you did, I did not. I felt left out and wanting to break free.

Then there was my father who generally never used the “liver problem” card. Until one day, he came to know about the benefits of a disabled parking badge. I was angry. If there was a reason I was handicapped it was because my wings were tied, not because I did not have any.

On reflection, both my parents had seen me at my worst—childhood seizures, epistaxis and emergency trips to the hospital for vomiting events so severe and constant that not even a molecule of Caloreen would stay inside me.

So to keep me as healthy as possible, they cocooned me, secured me and protected me by implementing these added

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strategies so that even during danger, I could be helped as quickly as possible.

Hindsight gives 20/20 vision, but at the time I resented this. And then came the introduction of corn starch when I was around the age of 8 years. I was told “you might feel butterflies in your stomach after the first few times you drink this”—how exciting! This magic elixir would mean no more restrictive and face deforming NG tube—now mum can no longer point to it during the “my child has a liver problem” conversations. And the added benefit of course is that I’ll generate butterflies!

Reality check: it was disgusting, the texture made me gag...and for an already large child, I gained a lot of weight—and lost it and then gained it—my body is still covered with stretch marks from the years of fluctuating weight. Nonetheless, overall, I remember being on the bigger side.

I wish I could say I was an elated human being and this was no hindrance to me—but it was. I struggled with it all, the

resentment, the weight fluctuations, the stretch marks, the lack of confidence, the jibes and comments that came with it all.

To sum it up, I was a large child, drinking starch, fluctuating weight, did not know much in the way of exercise because “my daughter has a liver problem”. Even if I knew how to exercise, GSD made it really difficult. I was always last in the cross country, the one no-one picked for their teams. I was consistently told that exercise is not any harder for me than in non-GSD children. I begged to differ—it was a mammoth task for me.

I started developing a negativity towards the starch and GSD in general. We will always find ways to circumvent what we perceive as inconveniences, leading us to take our own measures. I cannot recall how old I was when I took “control of my life” and decided that starch and I were no longer



compatible—but I was old enough to know when I was hypoglycaemic and old enough to know a bottle of a commercial energy drink would (what I thought then) fix it.

I vividly remember one day when I was 14 years old. I begged my older cousins to take me to a Michael Jackson concert. I took my starch container with me (my parents still thought I was taking it). After several hours of waiting for the superstar to appear on the stage, I started feeling hypoglycaemic. I knew the only answer was to drink the starch, but I avoided it like the plague. How would I explain I am drinking what looks like milk? This is the age everyone else has started exploring alcoholic drinks, and there's me drinking "milk" at a Michael Jackson concert!

The hypoglycaemia progressively worsened and fairly quickly too. I started feeling dizzy, sweaty, spaced out and claustrophobic. I needed to get away, so I walked away from the crowds. My heavy body found the comfort of the portable toilets and sheer exhaustion took over as my body just buckled. I gave into the starch and gulped the gritty, dry, thick liquid and physiologically calmed down after 10 minutes or so.

Internally though—I broke. Through the cracks of my broken spirit, I entered a feeling that I would soon become closely acquainted with—my life long passenger, Defeat.

This complicated relationship between Defeat, starch and I went on for years. Times changed. Corn starch turned into Glycosade; I met someone, went to University, found a job, but my intimate three-way relationship never left me. But with all the added expectations of life, I was drained by the end of the day. I was lethargic and fatigued every single moment from morning until evening. I would keep feeding the monster that was hypoglycaemia with the rush of energy drinks and then crash from exhaustion, sleeping anytime and anywhere. A wooden floor was as comforting to me as a soft bed. There was no ounce of energy left in me to do anything more than consume energy drinks, sleep and subsequently get bigger.

Work suffered as did my relationship with my husband. How could either of them even try to understand when I'd hidden the extent and impact of the GSD from them in the first place? So much so that I'd forgotten the severity of it myself.

Four years after my wedding, I sat in this waiting room. My life was about to start depending on a machine. Alive, yet not really living. Spiritless.

Kidney failure as a result of mismanagement of the underlying GSD. Kidney failure, because I was consumed by Defeat.

Defeat is toxic and yielding to it deforms our spirit. It makes us less than we are, less than we were born to be—it prevents us from achieving our potential in life. Here I was, a 30-something-year-old and Defeat had thrown me into the pit

once and for all. All these years, I looked everywhere for the enemy and failed to see it was me.

But tragedy is also where you find light. Pandora found Hope in her box—what was in mine?

Fast forwarding to post transplant.

You must have heard this story...woman learns she has a severe illness. By some miracle, she survives, and within days, she does everything she ever wanted. She lives with a new purpose, a new bounce in her step. Everyone thinks she's crazy but she is happy, carefree and full of life and spirit.

The afterword? Woman is no longer Defeated. That was me. New liver, new kidney but somewhere along the journey my same old mind had recalibrated overnight.

Is that what it took? Did I have to stare at the bottom of the pit to make me stand up and confront Defeat? Did Defeat have to cripple me to mere existence before I woke up to the magnitude of life?

In my Pandora's Box, I found Gratitude.

Post-transplant, time for me is double-edged. The most obvious reaction to feeling life in my veins is frantic activity. So now I learn new languages, I swim, I master complex Yoga poses, I travel all over the world from the heights of mountains to the depths of valleys. I lift weights heavier than my own body weight, I run with the wind, I sleep under stars, I sing tunelessly and am notoriously eccentric in my very own stretch-marked skin. I do everything and anything, because I can.

But I also think, speak and act with intent—I plod, I ponder and spend a little more time looking over my shoulder at the postcards from my past. Hindsight affords me to see with perspective.

Firstly, that my parents have loved me beyond belief (irrespective of what I understood it to be).

Secondly, that despite my poor management of GSD, despite the pain my GSD doctors must have felt to see me deteriorate and not take heed of advice, despite not being worthy of their efforts—they did something that no other doctor in a white coat ever did for me.

My first best friends, Professor James Leonard and Dr. Philip Lee, took off their white coats and reached out to a struggling human being—giving me value as a human first, GSD second. From the beginning, they were involved in creating the gift that is my life—like many readers of this publication, they studied, toiled and worked hard to give me life and teach me life.

Here was the integral answer to my quest for understanding what makes life more meaningful.

Believing in the sanctity of a human life. Upon that realisation, Defeat exited.

Life has been messy, wrinkled, torn in pieces but it is magnificently woven together with love, integrity, wonder and my new life passenger, Gratitude.