

*Some stories need to be told. They are the ones of undying spirit – of hearts that won't let go. They are the stories that gently remind us of where we came from and sharply hone our perspectives for where we're going.*

*This is our story.*

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To be young is to be invincible ... or so we thought.

Four years ago, my husband Michael and I acted as if we “had our whole lives ahead of us”. Little did we know how quickly that would change.

It was the summer of 2000 and our two major concerns in life were raising our four-year-old son and (to a much less noble cause) figuring out how to afford a boat.

We loved spending our free time on the water. Actually, Michael (then 29) enjoyed most things athletic – from soccer to sky diving. But, both of us revered water skiing as absolute therapy for our souls.

We never missed a chance to be on the lake and so it was no wonder then that, after a weekend trip in June, we attributed the onset of itching to us having the bad luck of collecting a rude batch of chiggers. Turns out, only I had.

Michael's itching never stopped. In fact, it progressed to a ridiculous point and I'd wake up in the middle of the night to see him asleep, but still clawing at his legs. There was no visible irritation on his skin, save the scratch marks he inflicted himself. Asleep or awake, the itching was unrelenting, maddening and our very first clue that youth didn't necessarily equate immortality.

Dr. John Ramsay did what every family physician would at first – treat for an allergic reaction. Michael changed soaps, avoided certain foods, took oatmeal baths and applied topical medicines. All failed.

It was after a visit to local dermatologist Dr. Michael Braden that we realized the problem was more than skin deep.

Blood tests were ordered and I can still hear Dr. Ramsay's voice on the phone gently informing us that the results indicated a “problem with Michael's liver” and further testing was necessary.

I think we sensed that something might be seriously wrong. So, while we still could, we decided to slip away on a long weekend to New Orleans to celebrate our 10th wedding anniversary before coming back to face the test.

To call it “testing” is kind. The diagnostic procedure they were to perform on Michael was an ERCP (Endoscopic Retrograde Cholangiopancreatography) and a more accurate descriptive would be “modern-day torture”.

An ERCP requires the patient to lie on his stomach with his head turned sideways so that a flexible, lighted scope on a tube can be inserted through the mouth and guided deep into the body. Then, doctors inject contrast dye so that they can take X-rays to view the liver and the ducts inside it that carry bile to the intestine.

Michael's ERCP was performed in Kerrville and lasted for four hours, during which he was not sedated adequately and became “combative” against the attempts to intrude his body. It took several attendants holding him down to get the job done.

When the doctor finally came out to talk to me, his confidence in his findings was unnerving. It was the first time I heard the word “transplant” in the same sentence as “Michael will need”.

The diagnosis was “PSC” (Primary Sclerosing Cholangitis), a rare, progressive disorder that is characterized by ducts inside the liver scarring and narrowing. When the ducts narrow, it interrupts the flow of bile and causes a buildup in the liver, which eventually leads to total organ failure.

In the process, the backup of bile causes yellowing of the skin and eyes (jaundice) and itching from excessive bile acids.

The doctor added that there is no known cause of PSC and, more importantly, there is no cure.

“No cure.” Those words hung in the air and time really did stand still. I can remember that very moment as vividly as people remember where they were and what they were doing when monumental events take place – like a president being shot, the space shuttle exploding or the attacks on 9-11.

This time, though, the tragedy was hitting home and the doctor said Michael could need a liver transplant in 15-20 years.

The news was not good, but at least we now had a name to our enemy and we would have two decades to adjust and prepare ourselves for his fight for life.

I didn't even show the same level of concern as did my mother and my mother-in-law who were also there hearing the news. I realize now that I foolishly thought there was nothing that Michael and I couldn't overcome together.

After that day, treatments began to aid Michael's liver by freeing up the constricted bile flow. The only thing doctors could do was to perform another ERCP and place plastic, straw-sized stents in his common bile duct, the major draining point for the liver.

For that procedure, we would head to San Antonio to Methodist Transplant and Specialty Services. Thankfully, the doctor there had learned of Michael's high threshold for anesthesia and used the "big darts" to put him out, allowing the stent placement to take place in a much shorter and easier procedure. Michael's lab numbers improved and we returned home to deal with our news.

It was only a few short days later when Michael developed a high fever with chills. It seems that the stent had become clogged with bile ("sludge", for lack of a better word) and an infection was brewing.

We went to Hill Country Memorial Hospital where Dr. Leonard Bentsch replaced the stent with a clean, fresh one in a third ERCP that went much more smoothly than the first two.

That would be the cycle for the next few months: replacing the stents every six to eight weeks, clogging, infections, replacing.

More frighteningly, the x-rays taken each time he had an ERCP revealed that Michael's condition was deteriorating rapidly, rather than slowly as had first been predicted.

Our time frame of 15-20 years before liver failure shrank and in just a couple of months we found ourselves at Baylor University Medical Center in Dallas for transplant evaluation.

After submitting to a week of rigorous testing, which included work-ups on everything from psychological stability to kidney function, Michael was accepted as a viable candidate.

We were relieved when he was officially put on the waiting list but then grew impatient when the actual wait began. Michael had come to terms with transplant being the only way he would survive and so he was ready to get on with it before his physical condition weakened.

Still, as the months trailed on and Michael underwent ERCP after ERCP, infection after infection, his body began to show the signs of disease and his stamina dwindled. It was like having a never-ending flu.

Yet his attitude held steadfast. Always the optimist of the two of us, he kept his upbeat disposition – even when he felt extremely bad. He poured himself into work and it seemed his goal not to let anyone see the damage taking hold.

But the day came when his despair was too much to hide. Our son, Austin, then five, was diagnosed with Type I diabetes. As we left the doctor's office after hearing the news, I remember Michael's eyes tearing up in one of the very few times I've ever seen him cry. He was so tired – and not just physically. Life seemed to keep throwing wrong turns our way and though he wanted to bear the burden for Austin, he knew he could not and that made his heart ache.

I, too, was profoundly sad. But, by then I was committed to handle anything thrown our way. The realization had sunk in that we were, in fact, not invincible just because we were young. We were human and inherently vulnerable.

Our lives had become a continual onslaught of health challenges and the stress was nothing if not gnawing. My only choice was to dig in. I was bound and determined our family would survive.

The very next day I faced up to my fear of needles and learned to give my child insulin shots without letting him see the trembling in my hands. Children smell fear and I wasn't about to let him have a whiff of mine. Michael, on the other hand, was more of a "natural" at giving the shots and helped ease the process for me.

Austin's diabetes came as a blow, but it was at least something we could control. We educated ourselves on preventing long-term complications and we worked together to manage his condition.

Then, the pendulum of misfortune swung back to Michael's side. Somewhere along the way, he began developing painful bumps on his elbows and palms. It wasn't characteristic of his liver disease and so we really didn't know what to think of it, except that it meant he could no longer open a jar or grip a steering wheel without gloves.

To the doctors' amazement, further blood tests revealed that Michael's cholesterol was 1,985 mg/dL – virtually off the charts since 240 mg/dL is considered to be very high and in the "at risk" level. His cholesterol was so elevated that it literally seeped out of his blood and collected as deposits (the bumps) in his joints and just underneath his skin.

At that point, we stopped asking ourselves, "What next?"

A specialist in Dallas diagnosed Michael's problem as "Lipoprotein X Deficiency", an extremely rare, genetic disorder that results in elevated cholesterol. It was so rare worldwide that my attempts to research it on the Internet failed completely and we found that disturbing since information, to us, aids coping.

It was decided that the best treatment would be to filter Michael's blood somewhat like kidney patients do in dialysis. It was imperative to get the cholesterol under control before it clogged his main arteries and rendered his heart unable to withstand a transplant. So he began weekly "pheresis" treatments in San Antonio at the Cancer Care Institute.

There, he would be stationed in a hospital recliner for four to eight hours straight with both arms outstretched and hooked up with large needles to lines leading to and from a machine. His blood, visibly yellow from the high cholesterol content as it left his body, would be run through two different filtering cylinders but would sometimes be so thick it clogged the machine.

Each time his blood was filtered, his cholesterol would drop to between 1,100-1,200 mg/dL, but would jump by several hundred points before the next week's visit.

It was obvious that pheresis was only buying time – time that seemed to tick by in slow motion as Christmas and the New Year 2002 came and went.

As the Fredericksburg Publishing Company neared the finish of its new office building at 712 West Main Street, Michael, who had been in the computer department there before leaving on disability, still was determined to make his life useful. He worked to cable the new building and set it up so that the computer system/server could transfer sites.

It was a major project he felt determined to finish and it served as his driving force.

Time for the move closed in and by Feb. 13, 2002 – three days prior to the scheduled relocation – Michael was still at the construction site having pulled several late nights in a row. As the evening wore on, I convinced him to come home and rest.

As we got in the car, he let out a heavy sigh and said he felt relieved that the computer system was at a point that it could move forward if for some reason he were not there.

It was a chilling prediction.

We were no sooner home and had Austin to bed when it happened.

I was on the cell phone talking with my mother on one line when I heard the house phone ring and Michael answer in the back room. From the bits I could hear, it was obvious that it was the transplant coordinator calling and with shaking hands, Michael began frantically writing down instructions. We had only a few hours to get to Dallas, they said.

We were in shock. Truly the call had not been expected until March or April because of where Michael was on the waiting list. We had not even packed a bag and so we did that then at a frenzied pace.

While waiting for my mother to get to our house from Ingram so that she could care for Austin, we tried to wake him back up to say our good-byes and give him our Valentine cards. He was so sleepy that I'm sure he doesn't remember it, but we do and I think the gesture was probably more for our sake than his.

Shortly after that, we flew out the door on a journey we would never forget.

*Next week, the second in this series will focus on Michael's first liver transplant, his episode of rejection and then his road to recovery...which would only turn out to be a loop.*