

Dextrocardia

By Gregory Fagan

Robert had scheduled his vasectomy for a Tuesday morning. Within a couple of weeks, he thought, he would be back to his normal self – working, commuting, kids, ball games, and the occasional date night with his lovely wife of seven years. It was to be a routine visit to the urologist, uncomfortable, but routine. Many of his friends went through the same procedure, and none had regretted it – except for his French friend who called it a form of mutilation, but other than that – he was ready.

It was late June, a Tuesday morning. The mid-morning heat began settling in as Robert and his wife, Sara, parked at the urologist's office for his 11 o'clock appointment. This was going to be the consult, but the plan was to then take immediate action. He had the rest of the day off, and the kids were at Grammys. Sara, who would be the designated driver, giggled and made nervous jokes as they made their way into the front office. 'Are we sure this is it?' The answer: 'Yep, that's it.'

Robert was brought into the exam room where he met the doctor. Risks were explained, the positives, the negatives, details about the procedure and what to expect moving on. He then undressed, and they prepped. At the start, the urologist began searching for the vas deferens, and found one, but not the other. He joked, but he just could not find it. Two nurses came in to help with the process, and Robert, now extremely sensitive to the fingers poking, pushing and pulling, nervously exclaimed after about five minutes, 'Forget it – I think we are all done here.' But his outburst wasn't what caused the urologist's concern.

During the consult, Robert explained his previous medical history. At age nine, he was diagnosed with severe bradycardia during a bout of pneumonia. A pacemaker was implanted, and he was told he would need it for the rest of his life but would still be able to live a normal healthy life. When he was 13, the leads became dislodged during a summer vacation, and he had to have surgery again. Then again, at age 21, as he was working a summer job at a beach restaurant in Florida, he cut his finger washing dishes. He hadn't even noticed it because there was very little blood flow, but then his friend and backup dishwasher mentioned he looked very pale. And he was. The pacemaker had failed, and he was sent to Philadelphia for a new one, where the first one was implanted. Now, not only does he have to contend with the pacemaker, but he also makes the clinician's role a bit more interesting with his dextrocardia situs inversus totalis, which for the most part, is the cause of his cardiac problems.

'No one knows of my reversed organs. It's messed up,' he says – and he also doesn't tell many people about his pacemaker. Robert is a funny guy, educated, serious at times, and loving, but when it comes to this issue – it is way too personal. Perhaps it has something to do with his childhood: living with large chest scars at only eight years old. I can imagine kids teasing him.. But this is the type of thing that sits deep in a child and then is carried with him

into adulthood. He never even told his wife, even after eight years of dating, until they were married. She knew, for the most part, because mothers can sense secrets. But her guess was the pacemaker had failed, and that's why he was sent to his cardiologist.

'Daddy has another boo-boo,' his wife had explained to their lovely girls, who were four and six. They reacted as kids do, wanting to see the cut and "Band-Aid". But he didn't give many more details – as he felt the kids did not need to know anything else. Robert and his wife had both agreed: keep it simple. There would be plenty of more time for explanations.

Robert was scheduled for another "routine" procedure. He was supposed to be in and out within a day. Just like before, arrangements were made at work, and he had taken the Tuesday off. He arrived early that day, hungry, but in good spirits. His outward philosophy was to just hold his breath and let the clinical world take him, fix him, and then release him, but, after his earlier procedure, I found he also carried a deeper and more profound emotional response. There is a sense of shame associated with conditions we have no control of – especially when we internalize these struggles and do not share them with others. Though Robert is emotionally intelligent, to him, there is an associated weakness that accompanies his psyche when dealing with this issue. It is then buried, for the most part, but comes out during doctor visits, insurance questions, airport security checkpoints, and during times like these – unexpected.

Wheeled into the OR, he is asked by the surgeon what type of music he would like to listen to.

'Phish, if you have it.'

'How about Grateful Dead – live in '84 from Indianapolis.'

'Sounds great.'

Robert was given local anesthetics; after the initial operation, he said he never wanted to be conscious during a procedure again. Around 15 minutes into it, the whole story began to unfold, as the surgeon noted there were three leads dislodged from previous failures. They were deep. The leads were covered in tissue, and one was very long – long enough to concern the surgeon that any attempt to pull the lead out may result in further complications of the abandoned lead falling into the right ventricle. Previous replacements neglected to correctly dispose of the initial leads.

The call to stop the surgery was made.

Afterwards, there was a call to Robert's mother, who is a lung cancer survivor, and recently lost her husband to sudden heart failure. He was reluctant to call, primarily based on the guilt his mother has regarding his condition. His condition is a rare genetic disorder, affecting only 1 in 22,000 of the general population, and there was no way of knowing of his condition, as no one in the family was ever diagnosed. It was only after he was hospitalized at age nine that confused doctors became aware of his condition. Still, the call was made, and she began her 1100-mile drive from Florida to Boston.

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Family health in the U.S. is a strange occurrence. It seems everyone knows, to some extent, their family histories – they share with friends, they base diets and lifestyle changes on it, they brag, they deny, they do a lot of communicating; however, it is only when an emergency occurs that families seem driven to act. Only then does it become apparent that all the communication, all the knowing, all that continuous emotional chatter wasn't beneficial at all. Health crises seem to be treated as discrete events, rather than a continual pursuit of lifetime care. Robert's case is a prime example. There were seldom visits to the cardiologist; there was a lack of communication to loved ones, and there was no long-term health plan to ensure he got the treatment he needed. This may also be the reason for his mother's guilt as well.

There is another perspective here: We tend to ignore and push aside pressing health issues until they occur. Why? We treat disease and illness as separate events, until they become serious enough to warrant long-term attention. For Robert's case, he never knew how to acknowledge his heart condition and never allowed it to become part of his life or his story. It was a source of shame and a reminder of mortality, which he wanted to forget.

Robert was transported to Boston for surgery. His cardiac surgeon, an enthusiastic and passionate innovator in cardiac medicine, met with Robert and his wife. The procedure was explained. An effort to communicate within the gap of patient and clinician understanding, however, made Robert uncomfortable. The cardiac surgeon described the procedure as going into the heart and 'ripping' the old leads out. While Robert appreciated the surgeon's foresight to dumb down the description of the central venous puncture technique by using a 'street' approach, it did more harm than good. The visualizations disturbed him – and ultimately increased his family's anxiety, as it prevented him from properly describing and understanding the procedure.

The procedure took a little over four hours. The pacemaker was replaced, new leads were placed, and there was an attempt to remove the older, dislodged ones. Two were removed, but there was an issue with the longest and oldest lead, which was not addressed during this surgery; instead the lead was just cut short. Post surgery, a decision to go back in to remove the lead was debated, but considering the risk, the decision to leave the lead in was made.

Recovery was hard for Robert. He spent almost a week in the cardiac unit, and this meant an extended duration of not seeing his children. He choose no painkillers – no oxycodone, as this made him anxious and sick – so he was relaxed with a modest dose of Acetaminophen. The initial procedure was planned as a day surgery, so he could go back to work within a few days, but the reality was far from the ideal and frustrated Robert, who now had to contend with his work schedule, used-up vacation and sick time. He had to think about the need to explore short-term disability, resulting in a reduction in pay, which he could not afford. To top this off, communication in a busy urban, academic hospital is slow, and some of the answers he needed to make decisions took time to get.

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Healthcare is dynamic and multidimensional, especially when we factor in the family and personal elements. Robert is 35 years old. He has two children, a beautiful and wonderful wife, a job, a home—which has been unstable as of late because of mortgage, taxes, and all the stuff in between. It is easy in our society to push illness aside, especially when we have invested so much into our lives. There is personal responsibility to tackle, and there is the air of stigma to contend with as well, but Robert is young and stubborn. He strives for success, but only waits until a serious health issue to deal with his genetic disorder. These are common problems. How do we acknowledge illness? How do we communicate with our families, and extended families? How do we tell our story? How do we ensure we communicate with the clinician and the inverse happens as well, especially when charts detail only data points?

There is a communication gap between the patient and clinician. We react to the clinician's handling of illness as a discrete event, by turning our illness into a discrete event. Recently I was able to listen to a patient who spent a lifetime managing her disease – a rare genetic condition called Mucopolysaccharidosis. She had been treated with enzyme replacement for many years and worked hard to advocate for herself and others like her. She had decided to end her treatment, thus, for the most part, ending her life. Her reasons were personal and profound. She discussed the issue of the gap between patient/clinician relationships. And though she had made great relationships with her physicians over the years, and they are indeed passionate researchers, the main reason for discontinuing treatment, she said, was the clinician was treating her disease, not her. Every day in the clinic was day one – there was no connection between the patient, the disease, life and the clinical world. She could not bear being an advocate on one side, and then a trial on the other.

Back to Robert, however. He was released, and sent home to recover. His children, scrambled again, to see the “Band-Aid” but Robert, sore, weak and tired, was only able to muster a kiss. His arms were in pain, and he was ordered to avoid strenuous movements because of the high risk of dislodging the newly implanted leads. A follow-up was scheduled within the first week of being home – and his wife ensured he would make it.

On that day, Robert parked a bit down the road and decided to walk, as it was a nice clear morning. The office was located at the top of a hill, and he was determined to make it without stopping. Upon arrival, he opened the door, huffing and puffing, and sweat dripping from his forehead. He was glad to be in the air conditioning, and it felt very good after the walk. As he looked around the waiting room, he noticed a number of senior men staring at him. They, like him, were all there for their pacemaker and Coumadin checks. At this moment, his mortality overwhelmed him. Robert put his face in his hands and began to cry.

Although there are many young men and women like him, Robert doesn't like to talk about his condition, nor does he feel a need to engage in the dialogue surrounding his illness. The hope is this latest experience will change him. With two daughters and a lifetime ahead of him, it would be wrong for him to deny his own story. But there also needs to be a bridge to allow for more effective communication between clinicians and patients. Advocacy is a powerful thing, but it's most important for Robert to begin telling his story and working to maintain a consistent clinical dialogue, not only with his physician, but with his family and loved ones. I believe he will change. Effective delivery of care is not a one-way street. It is

dynamic, multidimensional and continuous. It affects patients, parents and physicians – it extends to families and friends-- and encompasses passion, heartache, loss, forgiveness and love. It can be shared, though, through storytelling and advocacy.

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