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Interrupting Sudden Death: Genes, Medicine, and the Healing Power of Love

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This autobiographical account centers on a life-threatening medical crisis—a sudden aortic dissection—and the dramatic story of how medical technology, the skill and care of medical professionals, and the support of family and friends all contributed to the interruption of sudden death and to the power of healing. Attention is given to the systemic nature of medical treatment—the challenge of collaboration among hospitals, outpatient facilities, an array of medical subspecialties, insurance companies, family members, friends, and the patient. The genetic roots and implications for at-risk family members are also examined. Ongoing life lessons are highlighted, especially the challenge of living confidently with a genetic disease that can kill while simultaneously generating a life of mindfulness and gratitude.

Keywords: aortic dissection, healing, geneticsryon

INVASION OF THE SUDDEN KILLER

The genetic code was set when sometime in the cold Indiana winter of early 1949, a sperm and an egg first united, sparking the cellular elegance that gave me life later in October of that same year. By all accounts, the genetic gods seemed to have shed benevolent light upon me. Rarely was I sick. I was not aware of any potentially debilitating genetic anomalies. I was free of chronic conditions and, I admit, had some presumptuous pride in my biomedical supremacy as I lived in my illusion of insulation. I ate right. I exercised faithfully. And I believed my psychological work, particularly my extensive family of origin projects (Framo, Weber, & Levine, 2003), would increase my resilience and facilitate my immunity against the imminent threat of death. In short, I was bolstering myself with the religion of biopsychosocial perfectionism. By scoring high on all the tests, I was engaging in what Ernst Becker termed "the denial of death" (1973) and projecting lethality and sickness outward, in the words of Kubler-Ross (1969), to "thee and thee, but not me (p. 274)."

The belief that I was prepared for anything changed without warning, swiftly and forever, in the early morning hours of Sunday, September 28, 2003, when the true nature of my previously unknown genetic makeup emerged and my life tumbled precipitously into the gaping jaws of death. On a typical morning I would have been running through the neighborhood for 3 miles between 5:30 a.m. and 6:00 a.m. For several weeks, however, I had been nursing a hamstring injury, which had slowed me

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to a walk. Had I been running that morning, I would probably have died on a dark side street. Surprisingly, my cursed injury probably saved my life. I returned home from the walk and all seemed well on this ordinary day.

Joan Didion (2005) writes in her memoir about her husband, John, who suffered a fatal coronary in front of her at the dinner table one night: "Life changes fast. Life changes in the instant. The ordinary instant. You sit down to dinner and life as you know it ends." I did not know it then, but an eruption in my internal tissue in this "ordinary instant" was about to end life as I knew it.

As I emerged from the shower and began to dress, without the warning of hints and whispers, the pain struck ferociously—lump in my throat, sharp pains ripping through my chest, shortness of breath, lightheadedness, nausea, weakening legs, limb paralysis, cold sweat—all within seconds. This moment brought some of the worst pain that I had ever experienced. As Elefteriades (2005, p. 66) describes it, "the knifelike tearing sensation . . . is described by patients as being orders of magnitude worse than the agony of childbirth or kidney stones." With this pain came an instantaneous feeling of catastrophic doom that led me to exclaim, "We must get to the hospital now!"

Misty, my wife, was deep in sleep in the adjacent bedroom. As I, in a cold sweat, tried to arouse her, I said in a calm and determined voice, "I think I'm having a heart attack." She slowly awoke and realized the implications of what was happening. We did not consider dialing 911 but quickly got into the car. I took the driver's seat in what now seems to have been a continuing protest against disability. I had driven only two blocks before Misty insisted that she drive: "I don't want you to kill us both!" We then traded seats and drove through the open Sunday morning roads to Evergreen Hospital, about 15 minutes away, aware of *a* crisis, but unaware of *the* explosion of the silent killer within. Stealthily doing its damage over many years, the genetic disease was now bursting forth.

Arriving at the Evergreen Hospital emergency room (ER), I walked in quickly, declared definitively that I was having a heart attack, and was led through an empty ER to bed 4. Curtains were drawn. The staff knew how to treat cardiac infarctions, and it seemed I was in good hands. I became a patient engulfed by medical paraphernalia and procedures—tubes, shots to help me relax, medications to lower blood pressure, blood draws, cardiac monitor, chest x-rays. As the protocol continued with Misty nearby, I felt a mixture of worry, relief, and annoyance. My routine was being interrupted. How long would I be here? I felt relieved to be in the ER but impatient. It was Sunday, and I had to work tomorrow. But I could not stop my shaking, and my back pain continued.

The ER medical report included the following:

> This is a 53-year-old male . . . who had a mother who died of heart disease at age 53. He otherwise does not smoke. No hypertension, diabetes, or hypercholesterolemia. No known risk factors for pulmonary embolism. The patient has no known connective tissue disorder. All other systems negative. Pulse 63. Respiratory rate 18. Blood pressure 106/50. Temperature 35.6. A well appearing although anxious male sitting upright in bed. Alert and oriented. Normal sinus rhythm with no ventricular ectopy. No signs of acute ischemia. Normal chest x-ray. Normal cardiac enzymes. Unremarkable CBC. Unremarkable comprehensive metabolic panel. Negative for myocardial injury.

The report did not mention (or did we forget to mention?) that my maternal grandfather also died of a heart attack . . . at the same age when his daughter, my mother, died of a heart attack: 53. I had

been aware of "age 53" for years and had felt ready for this year with my religion of biopsychosocial perfectionism. I knew about anniversary death and believed I was sufficiently armed to meet the threat of this year. Clearly, I was in shape, mentally, emotionally, physically. With only 2 weeks to go before I turned 54, I felt I was almost home free.

In the hospital, after all the tests, I appeared remarkably "unremarkable." With the appearance of normalcy in spite of the reported pain, Dr. W., the attending physician, suggested that I should be hospitalized about 3 days for observation. This moment was a critical juncture. Many patients presenting as I did are sent home with anxiety-reducing medications. For those with my disorder, this decision, based on typically negative or bewildering findings, can be lethal. Patients with this undetected silent killer within have a 37% chance of dying within 24 hours and a 90% chance within 48 hours. No wonder! The killer within me has been called "the great masquerader" because it mimics other critical coronary conditions but remains unidentified and therefore untreated as other conditions are ruled out. Usually the cause of death is discovered in the autopsy; if there is no autopsy, the death is usually attributed to myocardial infarction.

Fortunately, my body continued to cry out. My back pain increased, leading Dr. W. to consider the death of the actor John Ritter only 17 days before of a misdiagnosed ascending aortic aneurysm and dissection (ABC, 2004). Ritter had been at the ABC studios in Burbank filming his sitcom, *8 Rules for Dating My Teenage Daughter*. He had felt what seemed like food poisoning during the day. At 6:10 p.m. that night, he was complaining of "chest pain and tightness, nausea, vomiting, and dizziness" and was taken across the street from the ABC studios to Providence St. Joseph Medical Center. He was initially diagnosed with an acute myocardial infarction, then again misdiagnosed with pericardial tamponade. He was given an angiogram in the process. Ritter actually had a "large ascending aortic aneurysm and a massive dissection" and died at 10:48 that night. (On September 9, 2004, the Ritter family filed a wrongful death and medical malpractice lawsuit stating that the hospital and doctors "caused the untimely death of (Ritter) by misdiagnosing his condition and, as a consequence, failing to provide proper treatment in connection with an ascending aortic aneurysm that would have saved his life. . . . If proper procedures had been followed to diagnose and treat Mr. Ritter's symptoms, he would be alive and well today.")

John Ritter's death saved my life. Dr. W., recollecting Ritter's condition and the need to assess for an aortic dissection, ordered a CAT scan. Sure enough, he discovered just that within minutes. With a calm but urgent voice, he disclosed that I had what "John Ritter had" (not "what killed him") and said that I would need immediate surgery for my ascending aortic dissection. Evergreen Hospital did not have the resources for open-heart surgery, so I was quickly transported via ambulance to Overlake Hospital, about 15 minutes away. Strangely, with an aorta capable of bursting any second, I was not in a panic. Were the medications working? Was it the calm and determined direction of Dr. W.? Was it the loving presence of my wife, who also seemed grounded in her faith that I was in good hands? I said good-bye to her. She took my hand and kissed me, and that was the last time I saw her before entering Overlake's operating room (OR) for emergency open-heart surgery.

My life was dangling by a thin thread because, officially, I had a Type A (ascending) aortic dissection that was in progress and could rupture at any moment. Blood was seeping through a tear in the inner part of the three-layered aortic wall, causing the inner half of the wall to separate from the outer half, all the way from the aortic root (emerging from the heart) down

the aorta's entire length and through its branching arteries. The aortic valve had been severely damaged in the dissection that had, perhaps, been developing for years as an aortic aneurysm. Given that the aorta is the River Nile of blood vessels, the massive and extensive damage through the entire aorta usually kills instantly by blocking or diverting blood flow to the brain and essential organs. Survival is unlikely.

Aortic aneurysms, especially thoracic aneurysms arising from the heart, are deadly, killing between 15,000 and 25,000 people in the United States each year—more than die from AIDS and most kinds of cancer (Elefteriades, 2005; Helliker & Burton, 2003). Albert Einstein, actors Lucille Ball and George C. Scott, and several prominent athletes have been among the casualties. Individuals with well-defined genetic diseases (especially Marfan's, Ehlers-Danlos, and Turner syndromes) should be considered at risk for dissection. The male-to-female patient ratio is 3:1. Harsh blows to the chest and weightlifting may precipitate a dissection. Subtler forms of inherited metabolic disorders are also believed to be more significant than previously thought (Anderson, Rizzo, & Cohn, 2003).

New technology and awareness of aortic disease is improving the ability of physicians to detect and treat aortic aneurysms (Helliker & Burton, 2003). However, the misdiagnosis rate for aortic dissection is about 35%. The sudden onset of chest pain has been shown to have a sensitivity of 84% and warrants a CAT scan (Wiesenfarth, 2005). The pain of a dissection is differentiated from the pain of acute myocardial infarction by its abrupt onset.

SUDDEN DEATH

I arrived at Overlake Hospital in what seemed like a brief moment, was whisked upstairs to the OR, and met the surgeon, Dr. A. I did not have much time to review his credentials. I asked him from my flat position on the gurney what he knew about this kind of surgery. He informed me in a confident voice that he had written two chapters on aortic dissections (another miracle moment!). The anesthesiologist, Dr. P., also introduced herself, and I requested that she keep me alive. She smiled and said, "OK." The introductions were brief, crisp, and confident. Again, I felt I was in good hands because of the emotional presence, care, confidence, and touch of these physicians. The nurses also helped by distracting me with the biggest drama of the visit—how to get the rings off my fingers, especially one that I had not been able to remove for 28 years. Amazingly, these nurses, with years of expertise on ring removal using their special lubricant, a magical string, and nimble fingers, were able to do the job, perhaps giving me the reassurance that this was a place of special possibility. I was wheeled into the OR, saw about eight staff busily arranging the room, and then . . .

Open-heart surgery began about 9 a.m. on Sunday morning. The scheduled 3-hour surgery stretched into a 7-hour ordeal that lasted into the late afternoon on Sunday because of an emergency within an emergency. A cut was made in my femoral artery, which would receive blood from the heart–lung machine during the operation and circulate it backward through my body. My chest was cut open 12 inches with an oscillating saw, my heart was suspended in a pericardial cradle, and I was put on an open heart–lung machine. This surgery was as serious and invasive as procedures get. Because of the extent of the dissection, there was a significant mortality probability as well as the possibility of brain damage, paralysis, and other organ failures.

The area of the aorta just above the aortic root had been severely damaged (the surgeon later said it was like trying to suture wet tissue paper) and needed to be replaced by a highly durable, woven Dacron graft, introduced about 50 years ago by Dr. Michael DeBakey, who discovered

the material in a Houston department store (Anderson, Rizzo, & Cohn, 2003). The aortic valve, located at the base of the aorta, had been damaged, requiring repair or replacement. Dr. A., emerging from the OR, spoke with Misty in the waiting room and asked her if she wanted me to have a mechanical valve or a pig valve if he could not repair the valve. Feeling as if she were being asked, "paper or plastic?" in the grocery store and having no knowledge of aortic dissections, she immediately called her cardiac surgeon uncle in California, who was able to speak by phone with Dr. A. in the OR. Fortunately, the valve was not insufficient or stenotic and was thus eligible for a "valve-sparing" repair.

However, there was an emergency within *this* emergency: Dr. A. was not able to stop the bleeding. The graft was close to the three branching arteries that go to the brain at the aortic arch. The surgical team therefore had to use the highly risky procedure called "full circulatory arrest." I was placed in a "deep hypothermic circulatory arrest" (DHCA) for 30 minutes. My body was cooled from 37 to 18 °C to slow metabolism and lower the risk of death and brain damage. The heart and brain had to be put to sleep. An EKG would have reported no heart function. An EEG would have shown no brain waves. If the definition of life is based on heart and brain activity, there was none, and I was dead. The cells were being kept alive but were not making "noise." This critical step had to be done so that the blood would stop circulating and allow a successful suturing. This delicate process was an emergency within an emergency, a high-risk procedure designed to empty the aorta for exploration and repair as second by second the risk of death increases (Kaiser, 2004).

Through all this, Misty kept watch in the waiting room with increasing concern, going for hours without news from the surgical team, but she was well held by friends who came to sit with her. Two of our children, who lived in California and Indiana, flew in immediately. My three sisters, warned of the gravity of the procedure, were told, "You must come now." My father and stepmother arrived as well. A colleague and his wife also appeared later in the night. We were all supported by the power of the love of those gathered together and scattered across the miles who collectively held us in their hearts. Perhaps the community of mindfulness and compassion influenced the power of healing. Their beating hearts kept me alive while my own heart was stopped. Misty says she never believed that I would die, although she knew I was on the edge.

Those who waited worked much harder than I, lying deep in the darkness of surgery and beyond. Those who wait, not knowing for hours the status of the one they love, listen for clues in the footsteps, the phones, the voices, the beeps, the drips, the looks on the faces of the medical team who come through the doors. Those who wait, who suffer as they balance between hope and despair, need comfort as they comfort others.

The suture had been successful during the full circulatory arrest, but because of the 30-minute suspension of life, there was uncertainty about my neurological condition and how I might emerge and awake. Misty was warned. I was taken into the intensive care unit (ICU) on Sunday night and was slowly warmed. Through the night I was surrounded by a labyrinth of liquids and machines keeping me alive. The bleeding was difficult to stop, and I needed two blood transfusions. Those family members who dared to look said they saw a lifeless, waxy, bloated body that looked coffin bound. Some could not stand to look.

Family members were told not to touch me or talk to me while the sensitive recovery and degree-by-degree warming-up period was under way, but Misty disobeyed, remembering a critical story I had told her 26 years before. My mother was in the ICU in December 1978 after an acute heart incident (had she and her father, both at the

age of 53, had aortic dissections?). She was in a coma on the night of December 21, and I was with Misty in California, more than 1,000 miles from her bedside. I had told Misty that I had wanted so much to be by her side, to touch her, to talk with her. I had wanted to give her hope through touch and voice, but she died later that night from pulmonary edema.

So, on that night as I lay motionless in the ICU, Misty broke the rules, came to my side, touched me, and sang to me her prayers. She did for me what I could not do for my mother. I do believe it was in this moment that I felt her presence in the depths of darkness . . . and I heard, and felt, and breathed life. She remembered, was defiant, loved, and saved me.

The night was long for those who waited. And then, suddenly, my first memory was of a big ventilator being yanked from my mouth. My eyes opened, I felt disoriented and foggy, and then I remember saying, "My, these are nice people. They've brought all this equipment to our home!" Shortly thereafter, I also said, "Tell me the truth. Don't keep anything from me." I did not realize they were all standing around, ecstatic at what they were seeing and hearing—I could somewhat talk and somewhat think!

I then said to my sister, Jane, something that today is still hard to believe was on my mind. Earlier in the year (April 2003), Jim Framo, Felise Levine, and I had published a book, *Coming Home Again* (Brunner-Routledge), which features my family of-origin work. There I described the early deaths of my maternal grandfather and mother, both at age 53, and my commitment to do the family work of coming to terms with their lives and deaths in an effective manner so that I could bypass death at age 53. Doing this family work had felt like a coming of age, a movement beyond the legacy of early deaths. So, upon awaking, spontaneously and straight out of the depths of surgery I exclaimed, "Thank God I am alive because had I died, the book would be invalid!" What was I thinking? More than just an example of overworking, this strange outburst in the ICU pointed to the power of the family story, the family's emotional magnetism, and the drive to repair not only the aortic dissection but the family dissection.

RECOVERY, RELAPSE, AND REHOSPITALIZATION

At around midday on Monday, September 29, I was cleared to transfer briefly to the coronary care unit on my way to the progressive coronary care unit, hooked to monitors that gave continuous readings of my vital signs. Misty stayed with me and helped calm me during unfamiliar routines. I was searching for comfort in a new world.

The medical staff measured my progressive success by the raw basics-peeing and pooping. My output was being measured all the time. At one point during a period of low productivity, the nurse threatened to catheterize me and I suddenly became productive. Pooping was more difficult and would continue to be a problem over the next weeks. Postsurgical consequences and the side effects of pain medication such as Percocet were inhibiting. My world was tightly organized around this cycle of food and liquid intake and the exercising of elimination skills. These were the targets of attention, the barometers of success and failure, of pleasure and pain. My heart was not yet in sinus rhythm, affecting the capacity of my body to eliminate fluids. I had gained much fluid weight during surgery and had been put on diuretics.

I had trouble walking. My steps were small, shuffling. Others' arms supported me. A few steps in the room. An adventure leaving the room for the open hall. Arms held tighter. Soon, too soon, steps back to the room. The halls at first seemed too intimidating. I would lean against the wall to regain strength. "Try a little more," I was told. The risk of clotting was high. "Keep moving." I was a runner used to covering

from 3 to 5 miles a day, but these little steps seemed massive, and taking them, impossible and humiliating. My life was reduced to bare essentials—measuring "vitals" on schedule, blowing into an incentive spirometer to keep my lungs clear, tethered to the remote cardiac monitor.

Though I had had temporary experiences with being a patient through my life, I had tried to avoid them, as if the patient role was one diminished, impaired, weaker, infirm. I did not want to be dependent on others to lead, guide, instruct, and order me. I also did not want to be incapable. Now I was attending to breath, pain, and moving my body so that someone could have a piece of me for a moment to take, prick, test, extract, or infuse. I moved from one bed to another, from one exam to another exam, table to table, room to room, ward to ward, needle to needle. I would now be followed for life, dependent on medical care to stay safe and alive, a patient without a cure and with considerable uncertainty.

Langer (1997) has reported on research highlighting how "decision latitude"—inviting the "infirm" to take more initiative in shaping their experience—can enhance wellness. I was grateful to those on the medical staff who included me in this way. Simple things greatly mattered to me when it came to this kind of medical attention—simple things like people knocking on the door before entering; announcing their names and why they were in my room; warning me ahead of time how this or that might feel, sting, or hurt; asking me whether something was too much or too little; inquiring about my pain; adjusting their pace to match mine; answering my questions with respect instead of impatience; allowing me to make decisions about many little matters instead of ordering me here and there. All these aspects of caring in the small matters helped resurrect my sense of self.

It was now Tuesday. I had been in the progressive coronary unit only 24 hours. Visitors were kept away. Dr. A., the cardiac surgeon, visited me once to check on me and to present the findings from the pathologist's report. I was diagnosed with cystic medial necrosis, not so much a definition of the cause of the dissection as a description of the tissue condition. This was one of the first indications that I had a genetic connective tissue disorder that is common in Marfan patients but that can also occur in nonsyndrome-based patients such as myself for reasons only beginning to be understood genetically. I was also told that my aorta was split into two channels through the illiac arteries. The Dacron fiber replaced only about five inches of the aorta in the aortic arch. The words "your aorta is split in two" were frightening to me after hearing about my genetic condition, which catapulted me into a new risk category. Those who have had a dissection are, understandably, more at risk for further dissections. How, then, would I live knowing this? What could I do with something I could not cure, chase away?

Dr. A. was followed by one of his nurse practitioners who sat with both me and Misty and went over the "red book," a volume for outpatient cardiac care. Dr. K., the cardiologist, also met me for the first time. He was quiet and informative and checked on me periodically. He was especially interested in my weight, fluid retention, and heartbeat, which was out of sinus rhythm. How balanced did all this have to be before discharge, given the constraints of insurance companies, the risks of premature discharge, the risk of hospital infections (the fourth-leading cause of death in the United States), my desire to go home, and Misty's desire not to take responsibility at home for someone in such a precarious condition? I am sure the competing risks, the ethical obligations, and the multiple demands weighed heavily on the medical staff and on Misty. I seemed fogged up.

On Wednesday, October 1, less than 48 hours after waking up after open-heart surgery, I was declared fit to go home. Al-

though I considered this a momentary victory, a sign of progress, an affirmation of strength, I really did not feel well and could barely walk around the unit without being held. I did not question the premature discharge, looking forward to being home. Later, Misty told me that she had been worried, bothered by the early discharge, feeling as if the heavy burden of medical care was laid too soon upon her shoulders. She did not want me to die, especially on her watch.

The next seven days at home were painful. Misty had the assistance of her mother and Jenny, one of my sisters. I had difficulty breathing. I had gained more than 30 pounds in fluid weight since the surgery only days before. I was very tired and did not want anyone to visit. I slept often and for a long time, more than ever. Walking upstairs to the bedroom, exactly 12 steps, supported by the arms of others, was a grueling chore that I had to accomplish without using my hands and chest. Every inch of body movement could mean the difference between pain and rest. Once settled into a pain-free space, I did not want to move. No foods interested me, and I had a special aversion to sweets. I used the spirometer from time to time, primarily to satisfy those watching me. I took "rat walks" around the inside of our home that demanded shuffle steps from room to room. I was especially disappointed in my failure to lose weight despite the aggressive use of diuretics. Dr. A. wanted me to monitor my urine output and weight. I complied faithfully with all the instructions, but everything seemed like too much. Then, after only a few days at home, during one of my inside walks inside, I felt dizzy, collapsed, and could not get up. A quick call to Dr. A. was followed by a quick order: "Get to the hospital!"

THE CRISIS OF MEDICAL LOYALTY AND THE TRANSFER

This time, Misty took the wheel and drove swiftly to the Overlake ER. I was whisked into room 7, given a chest x-ray, and told that I had pericardial effusion—fluid surrounding the heart that was strangling it and keeping it from beating properly. Immediately I was rolled into another ER room, the surgical room, where I would go through procedures designed to drain fluid from the pericardial sack. With some vague idea of what might be ahead, I asked the staff to give me a gallon of whatever would numb me. I was awake for this surgery, which, I was told later, lasted about 3 hours. I was sufficiently aware to remember my gagging. The procedures failed to relieve pressure, so I was again hospitalized in the coronary care unit.

I was agitated and did not sleep much that night, awaking the next day unprepared for the encounter that was soon to take place. Despite the lack of any improvement, the medical staff was again ready to discharge me as Misty returned the next morning. I walked slowly with great labor. Because my heart was not beating in sinus rhythm, the cardiologist, Dr. K., was considering cardioversion, a slap on the chest with paddles to jump start my heart into rhythm before discharge. I was told it was not a pleasant procedure, so I tried to will my heart into rhythm, getting it to behave before it had to be slapped into shape.

Then the real war broke out. The nurse who had been working with me and who had befriended Misty mounted a protest with Dr. A. and Dr. K. regarding my discharge. She said I was not well enough to be released. Side conversations in other rooms with the nurses, doctors, and Misty unfolded. I was so exhausted that I simply sat there waiting to be taken somewhere, but I did recall wondering about this nurse who had been talking with us about how she disagreed with the discharge orders. "Had she grown up in a family in which she challenged her parents?" I wondered. I was struck by the overt absence of alignment in front of the patient and the family; this agitated Misty, who did not want me to come home. Then Christopher, my son, was

pressing for me to come home because he did not like to see me in the hospital. Our family was reflecting the division in the medical team, and I was growing more tired and resigned to whatever the outcome might be.

Calls were made to my cousin, Dr. L., chair of one of the departments at the University of Washington Medical Center (UWMC), who facilitated a transfer there. He had been away on September 28, but now that he was in town, he wanted me under his care. Aggravating this war on the unit were complex insurance coverage issues that eventually dictated that I could be admitted to UWMC if I came by ambulance. Fatigued as I was, I do remember feeling enormously disloyal to Dr. A. and Dr. K., who had been responsible for saving my life. I felt as if I were betraying them and Overlake, but I was also loyal to my cousin and trusted his opinion. Strangely, through this whole ordeal, I do not recall any medical staff asking me for an opinion, even though it was my life.

The transfer to UWMC on Thursday, October 9, did not take long and I was soon experiencing a new hospital, new floor, new room, new staff, new attending physician—Dr. V., head of cardiac-thoracic surgery at UWMC. Room 114 on the fifth floor of the cardiac unit was a single room overlooking the university campus. Within an hour after my arrival, D., Dr. V.'s nurse practitioner, came into my room, introduced herself, and asked one question and made one promise that I will never forget. She set the tone for collaboration and conversation in a world that had been feeling too fragile, uncertain, and isolating. D. first said, "Tell me your story. What have you been experiencing?" And then she listened for as long as it took. I felt "listened into speech," as Parker Palmer (1998) once said. I felt more complete as I told my story about emotional and physical fragmentation—being out of sinus rhythm, out of life rhythm. She wanted to know me, and being known is the connective tissue of life.

D. also made the promise, "You won't leave this place until you are ready." I felt confident that my needs would be elevated above all other considerations, including insurance demands. These simple statements by D. set the tone for healing and confidence, the oxygen breathed by healing. D.'s initial visit was followed by one by a committee of residents sent by my cousin, Dr. L., to welcome me to UWMC. Over the next week, they would occasionally stop in to check on me. My cousin also came by. The medical center had a strong heartbeat of compassion to complement the biomedical equipment and procedures. I felt in good hands.

I entered UWMC with weight loss, fluid discharge, and the return of my heart to sinus rhythm as top priorities. My stay had many of the same features as the previous hospitalization: daily blood draws, temperature measurements, pulse checks, respiration readings, wheelchair jaunts to radiology for x-rays, and missions to the echocardiogram unit. About 2 days after admission, I developed several blood clots in both legs below my knees, necessitating an IV of Heparin in conjunction with oral Warfarin, all aimed at thinning my blood to break up the clots and prevent further clotting. I would continue on Coumadin as an outpatient for 6 months with weekly outpatient checks and adjustments.

My urine was pouring forth. The diuretics seemed to be working, and periods of sustained sinus rhythm were increasing in number. I lost about 30 pounds in 4 days, measuring every cc of urine produced and achieving some personal bests, such as 2500 cc per day. Defecating was more challenging, pushing me to shift from pain medications to Tylenol as soon as possible.

The most comforting moment of each day came at night with the arrival of heated blankets cooked in the unit warmer. I requested several each night and crawled into a womb of warm comfort. These little moments mattered a lot. There were others, such as looking out the window and

seeing people crossing the intersection, and wanting to do just that, only that. My desires were dramatically reduced to the basics. The altered state of consciousness I experienced in the hospital was maintained by the fact that no visitors were allowed.

On my birthday, October 13, as I crossed from the age of 53 to 54, I was surrounded by my family and I felt grateful, but then I promptly experienced another deflation of ego. October 2003 was to have seen the official launch of *Coming Home Again,* the family of-origin book I had written with James Framo and Felise Levine. We had worked on this project for years. Jim had died 2 years before, making this month even more important as a time to honor him and his legacy in the field. Two events had been planned, one in Washington and one at a conference in California. Days before the Washington event, I realized I was too weak to make the trip and too fatigued for the California author signing.

What was the message? A "special professional" has a short shelf life. "Special" really means the privilege of being, of breathing—all grace and gift, not a given. I also began to think of 80% of all arguments in life as egocentric, having nothing to do with substance and justice, another inessential way of being "special." So many things that aroused my temper day after day were, in truth, quite nonsensical. These were sobering reflections that were beginning to infiltrate the deepest parts of me.

Dr. V. visited me twice on the unit and was pleased by my progress. On Thursday, October 16, 7 days after entering UWMC, I was ready to go home—slightly stronger, more in sync, and aware of the steep climb to recovery ahead.

GENES AND THE GENESIS OF A NEW LIFE

New life began at home with a small appetite, weight loss, fatigue, and extensive sleep. The most vexing problem was constipation that for weeks became the axis on which my life revolved, aggravated by postsurgical complications and pain medication. I experimented with an array of foods (prunes and raspberries), drinks, and over-the-counter remedies, finally succumbing three times over these weeks to a powerful soapsuds enema ritual at the Overlake ER. I yearned for a normal poop. I began outpatient visits with the cardiologist, Dr. S., with Misty in attendance. Dr. S. helped anchor me, often reassuring me, in his soft, informal style, that I was getting better. The echocardiogram kept looking better and sinus rhythm seemed intact. Cardiologist appointments were supplemented with weekly visits to the anticoagulation clinic for regular blood draws and both Coumadin and dietary monitoring.

I did not drive for 6 weeks because of the danger of an accident causing the steering wheel to jam my healing sternum. Misty drove me everywhere and actually loved my more dependent status because it gave us more opportunity to talk. We also walked more together with our dog during all hours of the day—hours when I would typically have been at work. This crisis of fragility was, at least momentarily, renewing for our marriage.

My academic department granted me a 3-month leave of absence, a great gift. I began seeing patients only briefly about 5 weeks after the dissection. I was impressed with the amount of energy required to sit erect for a couple of hours in meaningful conversation, but I wanted badly to resume some of my former life and, frankly, to be on the helping side for a change.

Patients had to be informed about my condition, and backup care needed to be provided. All patients received a general letter from my office, days after the dissection. I had never imagined that I would be disabled, driven out of clinical practice temporarily by an emergency, or dead. All clinicians should prepare for these possibilities.

I noticed that my emotions and conversations with patients shifted, especially with those who had suffered debilitating conditions such as Parkinson's disease. It is hard to admit, but I felt a new kinship with these patients. This link was more embodied, a feeling deep within me. They noticed a quieter, more pensive, and more reflective me. I greeted people differently. When asked, "How are you?" I responded, "I'm alive and I am grateful." I have continued that response because it is the simplest, clearest description of my life. Some patients would continue, "No, tell me how you really are," and I would repeat, "I'm alive," and then would add, "And when we're alive, anything can happen."

From time to time, I comment that in the most impossible situations that people face, there is possibility. I often use my experience to note many things, including the idea that the light of life can leak through the darkness of death. I bring a different lightness and playfulness into therapeutic conversations, not devaluing arguments, but setting them in the greater context of life itself and the shallow immaturity of ego. Carl Jung once said, "If people can be educated to see the lowly side of their own natures, it may be hoped that they will also learn to understand and to love their fellow men better" (Quindlen, 2005, pp. 40–41). From the lowly side, patients looked different.

Many times a day, day after day, I am amazed at the simple gift that I am here. All of a sudden, it will dawn on me. The obvious is startling and is illustrated in Pascal's musings in the *Pensees* (Bloom, 2004, pp. 131–132):

> When I consider the short duration of my life, swallowed up in the eternity before and after, the little space which I fill, and even can see, engulfed in the infinite immensity of spaces of which I am ignorant, and which know me not, I am frightened, and am astonished at being here rather than there; for there is no reason why here rather than there, why now rather than then. Who has put me here? By whose order and direction have this place and time been allotted to me?

As profound as these emotional changes were, there remained the practical questions of daily living. What could I do about sex, running, tennis, biking, swimming, skiing—anything that raises blood pressure? What about caffeine? What should I eat and not eat? What about medications? And even more speculative, but critical: What could I count on for my future? What was my prognosis? Would this happen again? What about my children and my siblings? Were they at risk? Was there genetic testing? If my aorta were to dissect again, how would I know and what would I do? My questions about genetics and the management of a chronic, potentially lethal disease left me more often confused than clear when I sought answers from my physicians and the literature.

Essentially, the pool of aortic dissection survivors is very limited. The longitudinal data is sparse. We do know that no patient should be considered cured of this disease. The 5-year survival rate is about 75%, whether the patient is treated medically (before the dissection) or surgically. The 10-year survival rate is between 40% and 69%, for surgically and medically treated dissections. Reoperation can be necessary (Wiesenfarth, 2005). What we know about how to live long and live well with an aortic dissection comes down to this: (1) Take a beta blocker daily. (2) Get a CAT scan once per year. (3) Listen to your body. (4) Visit the cardiologist once per year.

I was very concerned about exercise and potential damage to the aorta and elevation of blood pressure. There were many mixed opinions. Dr. S., the cardiologist who was a swimmer and biker, said, "I wouldn't advise you to run." Dr. V., the CT surgeon, a gutsy guy, said, "Do whatever you want, including playing tennis, since it's not re-

ally a sport." Dr. L, my cousin, said, "I'm worried about you. Watch it." When I asked Dr. V. what I should be aware of internally that could signal trouble, he told me to be aware of any pain, and if it continued, "Go to a major medical center." Overall, the final message on exercise was "do what feels right." I was also told to wear a medical bracelet so that, in case of collapse, medical personnel would know my history. I was told to take emergency phone numbers with me. I was told to continue traveling and that if I had another acute aortic event far away in the Alps, there were always medical helicopters that could fetch me. In other words, the advice was limited. I got the distinct feeling that, although I could be helped by good resources and high-tech medical equipment, I was really on my own. With no clear map of the world and directions to follow, how do you live?

At my first CAT scan after the dissection, I sat down with Dr. V. to review the pictures. When I kept pressing Dr. V. for more answers, he gave a most interesting response that was less than what I wanted, but actually more useful. He said, "We just don't know. Each person's body responds in different ways. Anything could happen. The body has marvelous healing properties." End of speech. I heard this as a hopeful message, possibilities in an open-ended universe, healing properties within. Whatever he intended, this was the message I received, blending the truth of the uncertainty with the possibility that much could happen because of healing powers within (Groopman, 2004).

Weeks later, I visited Dr. B., the genetic pathologist, at the Center for Human Development at UWMC. I had been looking forward to this visit and brought with me four questions: What caused my condition? What was the prognosis? What kind of testing would he recommend for my siblings and children? And what could be done, or what might be done in the future, to improve my situation? I had sent him some information in advance, including what I knew about my maternal grandfather's death and mother's death from cardiac incidents.

Dr. B. and his genetics counselor met with Misty and me for an hour, reviewing the information and addressing these questions. With only scant information about my grandfather's and mother's deaths, Dr. B. could not say for certain whether they had died of aortic dissections. He did not know why I had developed an aortic aneurysm/dissection and the cellular condition called cystic medial necrosis, because he had ruled out Marfan's syndrome. He reported that good progress was being made in genetic mapping and that genetic testing for this condition could be available in the near future. He thought that I could have been a victim of the "perfect genetic storm," the odd coming together of several genetic anomalies. The prognosis was uncertain, although victims of aortic dissections were reported to have been living for years beyond the incidents. There was no treatment, no genetic testing, and no specific recommendations other than taking beta-blockers, watching blood pressure, and having children and siblings checked for aortic anomalies. So many "I-don't-knows." No cure and continued risk in this open-ended universe.

The genetic search for the defective genes that could help identify those at risk continues. What is being learned? There seems to be a higher prevalence of thoracic aortic aneurysms (TAA) in first-degree relatives of TAA patients than in the general population. Up to 20% of TAA patients requiring surgery have other affected family members. This evidence strongly indicates that genetic factors influence the formation of nonsyndromic TAA. Furthermore, in the majority of families, TAA is inherited in an autosomal dominant manner, with marked variability in the age of onset. Age at death ranges from the teens to the 70s. Technically, this has been called "familial nonsyndromic TAA."

In early March 2006, I was stunned to

receive a call that shed new light on the question of genetic roots. Early on a Sunday morning, my first cousin Anne called and told me that her brother, Charles, the first-born child of my mother's younger sister and more than 4,000 miles away in South Carolina, had just suffered a thoracic aortic dissection at the age of 52 and was in recovery after a 7-hour emergency surgery. In a subsequent phone conversation with Charles several days later, he reported that another first cousin, David, more distantly related, had died suddenly the previous fall in Florida after calling 911 because of a medical emergency. He was 51 at the time of his death. So David had died suddenly of some unidentified medical emergency at age 51; Charles had sustained an aortic dissection at age 53; I had suffered my aortic dissection at 53; my mother had died of "heart failure" at 53; and her father had died of a "heart attack" at 53. The family's "genetic dots" were continuing to be connected. Upon hearing this news, Dr. B. responded, "Certainly this raises the ante on the likelihood that the other family members who have had cardiac 'incidents' actually had aortic dissections. . . . It also suggests that everyone who is potentially at risk for dissection should have an echocardiogram" (P. Byers, personal communication, 2006). We were now thinking more boldly about revising history, rediagnosing the earlier heart incidents as possible dissections and thereby raising the risk for living blood relatives.

Dr. Dianna Milewicz and her team at the University of Texas Medical School at Houston (Pannu et al., 2005) have begun to identify the molecular pathways and mutations that can be studied to find biological markers for the disease and to develop therapies. At-risk patients should undergo lifelong routine imaging of their aortas.

So we started recommending imaging (specifically transthoracic echocardiography, TTE) to my siblings and children. Two of my three children (still on our insurance) were scanned in 2004 with negative findings. Genetic testing is just now being made available to identify those individuals at risk for thoracic aortic aneurysm and dissection. Dr. B (P. Byers, personal communication, 2006) reported that a test called a TGF beta receptor study has been developed that will help in determining risk. If my TGF test is positive, then all my siblings and children will also be tested.

These advances in genomics, however, raise some obvious personal and political problems. In the case of aortic anomalies, knowing the genetic risk and being tracked could easily save one's life. On the other hand, being identified by third parties, such as health insurance companies and employers, as having this risk increases the chances of discrimination by them. Congress is now studying legislation that includes a myriad of considerations. My children want to know what they need to in order to decrease their risk, but they do not want to encounter potential discrimination as they launch their families and careers.

I know that the experience of my aortic dissection, which began with conception in 1949 and erupted in 2003, will continue unfolding in new ways until my last moment. I continue to feel the grief of losing the self I thought to be true but that is no more. Joan Didion (2005), commenting on grief, says that the shock tends to be "obliterative, dislocating to both body and mind." My dislocation continues into the vast unknown, as I must learn to integrate this experience into my emerging new self. Sometimes I think I know what I will die from. More important, however, I am grateful for being brought back from death, thankful for the lessons being learned, joyful because of simple breath in this short life, and renewed in using my best gifts in service to others.

REFERENCES

ABC (2004). *Accusations of malpractice, death of John Ritter*. Retrieved September 19, 2004, from http://abclocal.go.com/kabc/ontv/090904_ontv_John_ritter.html

Anderson, C. A., Rizzo, R. J., & Cohn, L. H. (2003). Ascending aortic aneurysms. In L. H. Cohn & L. H. Edmunds, Jr. (Eds.), *Cardiac surgery in the adult* (pp. 1123–1148). New York: McGraw-Hill.

Becker, E. (1973). *The denial of death.* New York: Free Press.

Bloom, H. (2004). *Where shall wisdom be found?* New York: Penguin.

Didion, J. (2005). *The year of magical thinking.* New York: Knopf.

Elefteriades, J. A. (2005). Beating a sudden killer. *Scientific American, 293*(2), 64–71.

Framo, J., Weber, T., & Levine, F. (2003). *Coming home again: A family of origin consultation.* New York: Brunner-Routledge.

Groopman, J. (2004). *The anatomy of hope: How people prevail in the face of illness.* New York: Random House.

Helliker, K., & Burton, T. M. (2003, November 4). Medical ignorance contributes to toll from aortic illness. *Wall Street Journal*, pp. A1, A12.

Kaiser, R. G. (2004, February 29). My telltale heart. *Washington Post,* W10.

Kubler-Ross, E. (1969). *On death and dying.* New York: Macmillan.

Langer, E. J. (1997). *The power of mindful learning.* Reading, MA; Addison Wesley.

Palmer, P. J. (1998). *The courage to teach: Exploring the inner landscape of a teacher's life.* San Francisco: Jossey-Bass.

Pannu, H., Fadulu, V. T., Chang, J., Lafont, A., Hasham, S. N., Sparks, E., et al. (2005). Mutations in transforming growth factor-β receptor type II cause familial thoracic aortic aneurysms and dissections. *Circulation, 112,* 513–520.

Quindlen, A. (2005). *Being perfect.* New York: Random House.

Wiesenfarth, J. (2005). *Dissection, aortic.* Retrieved November 24, 2005, from http://www.emedicine.com/emerg/topic28.htm