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The Grace of Denial

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I sat listening to the case presentation about a woman who waited far too long to seek care for advanced breast cancer. By the time she presented for medical evaluation, her right breast was twice the size of her left and hung like a misshapen butternut squash hidden under her blouse. The physical exam revealed that the tumor was breaking down her skin, which was ulcerated and excoriated, with the orange-peel texture common in advanced breast cancer. I listened quietly to the familiar conversation among the surgeons, oncologists, radiation oncologists, and presenting medical student. The "wonder why she waited so long" commentary was inevitable. The "what a shame" discussion followed. "She had health insurance," the well-meaning student added, as evidence that funding was not a barrier to care in her case. The tenor of the conference was familiar to me after 14 years in clinical practice. I am well versed in the concept of patients' denial in the face of a devastating diagnosis.

My mind drifted back to a lecture during my first year of medical school that had included a detailed discussion of the progressive symptoms and hopeless prognosis for patients with amyotrophic lateral sclerosis (ALS). Outlining the way in which function would decline until the patient was left with no ability to move even a single muscle, the professor described the disease as "a front-row seat to one's own death." "What a horrible fate," I thought, mentally cataloguing ALS as one of the worst diagnoses imaginable.

Then, during my second year of medical school, my father was diagnosed with ALS. To be honest, he wouldn't be formally diagnosed until my third year — a delay caused by my own denial. During a hurried call I made from a pay phone at the library, my dad mentioned that he was becoming slow to get to the ball on the tennis court. I laughed it off as his excuse for having lost a match to his cousin over the weekend. My dad was notoriously hypercompetitive in recreational sports — with his friends, his relatives, and especially his children. But despite my attempt to minimize his complaint, he insisted that his muscles felt weak. Odd — my dad rarely complained.

I don't know what prompted my next question. Perhaps it was the physiology test on motor neurons for which I was studying, but when I asked, "You don't have fasciculations, do you? You know — muscle twitching?" his affirmative answer stopped me cold. I had just heard another lecture on ALS, in which my neurophysiology professor had described lower motor neuron disease with dry, clinical detachment and opined that ALS is perhaps the worst of all diseases, because cognition remains intact while the body fails. A patient ultimately becomes "locked in" — fully aware but unable to communicate.

My father saw a neurologist within the next few weeks, and a full laboratory and imaging workup ensued, complete with a brain MRI, a lumbar puncture, electromyography, and a sural nerve biopsy. Meanwhile, I vigorously researched alternative diagnoses in the medical school library. In those days of photocopying of printed articles, before computer use became widespread, I pulled journal after journal from the bookshelves in a quest to find a better diagnosis to explain my father's symptoms. He had so much faith in me. I was determined to change his fate by doing an exhaustive search of the medical literature. This could not possibly be ALS. Dad was only 50 years old.

When I prodded him for more information, he told me he might have been bitten by a tick on a recent trip to the northeast. He said he'd developed a bull's-eye–shaped rash on his elbow less

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than a month ago. "Perfect," I thought. "It's Lyme disease." Everything seemed to fit.

I made a case to my father's neurologist that his weakness was not a symptom of ALS, but a rare neurologic manifestation of Lyme disease. I'm not sure whether I was truly convincing or whether the neurologist was merely allowing my family time to adjust to a diagnosis that he knew was a death sentence, with no effective treatment or cure. If he was indulging my denial, he was extra-indulgent in encouraging us to take a trip to New York to pursue exhaustive testing by experts in Lyme disease and starting my dad on empirical treatment for that condition. My father continued to decline clinically.

I got married that summer. My dad received his second daily dose of IV antibiotics for Lyme disease just before the ceremony began and then tucked the PICC catheter into his tuxedo shirt. He was determined to walk me down the aisle and to do the traditional father–daughter dance. He picked out the song for that dance, Whitney Houston's "Wind beneath My Wings." He'd always been the wind beneath my wings, supporting all my dreams, from college tennis through medical school, but on this occasion I was more literally the wind beneath his, as I physically held him up. I could feel the strain of his shaking muscles from the sheer exertion of dancing and the perspiration on his shoulders during what would be our last dance together. He was determined to defy the odds and prove the diagnosis wrong. This dance was one of his many self-tests, designed to convince himself that he could not possibly have something as devastating as ALS.

I was sitting with him on the wicker love seat on my parents' back patio early one evening, when he handed me the phone. His neurologist was on the line and had asked to speak to me. With no greeting or pleasantries, he said, "Heather, I'm afraid this diagnosis of Lyme disease is a red herring. It is time to come to terms with it. Please talk to your family."

I was not ready for this. I was 23 years old and didn't feel qualified or up to the task of relaying such terrible news to my parents. I hung up the phone and watched my parents' concerned faces as I conveyed the information. If I close my eyes, I can relive that moment as vividly as if I were there right now.

Over the course of the next year, my father went from a jovial, athletic man, the bedrock of our extended family, to a quadriplegic. He continued to decline. By the time he died, he had been quadriplegic for 10 years. He was on a ventilator. He was fed through a tube. And that all would have been tolerable, acceptable — if only he could talk. The inability to communicate is the most tragic part of ALS. My father could not smile or gesture with his face. He could not voluntarily blink. He could not wave a hand or move a finger. He could not express himself in any way to those who loved him. He suffered the living hell that my physiology professor had so accurately described in the lecture hall that day. But my dad never threw in the towel. He always had a will to live — for himself and for us.

So yes, I am familiar with denial. When I see patients who cannot face the prospect of a terrible diagnosis, I understand their delay, their reluctance, their trepidation on a deep level — a level that perhaps only someone who has witnessed a loved one's slow demise from a terminal illness can appreciate. In the face of a diagnosis for which there is no effective treatment and no cure, our denial allowed my family 6 months of relative peace before things became unbearable. We had a few extra months with my father without the constant awareness that his death was imminent. My medical inexperience, clouded clinical judgment, and desperate desire for more time with my dad extended our denial of medical reality for longer than is typical.

Today, when I hear detached descriptions of patients who've waited too long to address a devastating illness, I understand. "Denial helps us to pace our feelings of grief," Elisabeth Kübler-Ross explained. "There is a grace in denial. It is nature's way of letting in only as much as we can handle."

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