MIND TO MIND
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Living with Lymphangioleiomyomatosis: An Anesthesiologist's Experience

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Lymphangioleiomyomatosis (LAM) is a rare, under-diagnosed, progressive lung disease caused by uncontrolled smooth muscle cell proliferation. It results in pulmonary cyst formation, small airway obstruction, and eventual respiratory failure. LAM mostly affects women of childbearing age. There is no known cure. I am a current anesthesia resident at the Hospital of the University of Pennsylvania. I was diagnosed with LAM less than two years ago, at the onset of my training. I would like to share my story to raise LAM awareness.

My name is Lyndsay Hoy. I live in Philadelphia, PA. I am a resident physician pursuing a four-year residency in anesthesiology. One year into my training, I was diagnosed with lymphangioleiomyomatosis (LAM). At the time of my diagnosis, Wikipedia stated: "The majority of patients with lymphangioleiomyomatosis do not survive beyond seven to ten years after initial diagnosis."[1] I was twenty-eight years old.

As a wise man once said, "Sh*t happens." Apparently, I missed that memo. I hadn't penciled it into my ten-year plan and yet there it was - an unwelcome surprise that had tiptoed in while I was busy studying for boards, finding a husband, and re-certifying my ACLS. So what is one to do? *You keep on keeping on.* You wake up at five in the morning, you put on your scrubs, your white coat, your operating room mask. This uniform that we physicians don serves a dual purpose. It helps you trust me when we first meet before your surgery - which is probably good, as I am a towering five

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feet tall and look all of twenty-two. It also helps me remember my role. Today, I am not the scared girl undergoing yet another painful lung procedure. Today, I am the physician charged with safely putting you to sleep, placing a breathing tube in your throat, and keeping vigilant watch over you. I am a voice, a hand, a mask imploring you to open your eyes and return from the depths of unconsciousness you journeyed into. I will wake you up. I will deliver you back to your loved ones. We will probably not meet again. But I love what I do for you. For a finite period of time, you are in my care. I forget I am sick. Then at the end of my day, I remember - and it is sobering.

It is both a blessing and a curse to be a physician with an illness. On one hand, your medical knowledge provides you with a baseline to better understand the anatomy and pathophysiology of your disease. If you *have* to have bad luck and get sick, you might as well know what you're dealing with to the –nth degree, right? Not quite. In my experience, all rational medical/scientific thinking flies out the window the moment you are diagnosed (estimated time of return TBD). To this day, I still can't bring myself to read more than one or two medical journal articles about my disease. The gut wrenching statistics and objective terminology feel all too familiar yet eerily foreign and distinctly unfriendly; phase one clinical trials take on a whole new meaning when you are the guinea pig. I was however, very lucky to have an extremely expedited diagnosis. From the onset of vague respiratory symptoms to definitive diagnosis, my timeline was less than six days whereas I've read that patients with my disease have gone years without a diagnosis. It started with a simple chest x-ray taken during a lunch break between surgery cases, which showed a large pleural effusion. My fiancé, Vincent, an internal medicine resident at the time, promptly went to the hospital on his day off to discuss this worrisome, foreboding finding with colleagues. This, swiftly followed by an appointment with a pulmonologist, a diagnostic procedure, a chest CT scan, and finally - and unsurprisingly - bad news.

The curse of being a physician with a chronic illness: it feels like being haunted by the bloody Ghost of Christmas Yet to Come. Those worst-case scenarios that you try to push to the far corners of your mind can play out on a daily basis at work. You see the worst of the worst: intensive care patients on a ventilator battling infections, bleeding disorders, lung diseases, and cancers. You watch them languish day after day until there's barely a person left. You may even be taking care of someone with the same disease as yourself, as I have done. You mourn for them. You try to comfort their families. You fear for yourself.

Keep on keeping on.

The moment of my diagnosis lingers in my mind, seemingly suspended in time. The truth is, TV and movies have it all wrong. There is no emotional climax when you find out you have an incurable disease. It happens quietly, unassumingly. And then everything is the same, but everything is different. Vincent and I waited at home in the kitchen, both of us on our laptops anxiously willing the results of my CT scan to pop up on the screen. I nervously clicked between web pages - Pinterest, Facebook, Gmail (seeing flashes of normalcy while waiting for life altering news is a disquieting experience) - to make sure the browser was working. Within moments of pulling up the scan, I zeroed in on a foreign word: "Impression:

radiologic findings consistent with lymphangioleiomyomatosis." Vincent was silent and one look at his face told me everything even though I probed him in vain for a different reaction. I felt absolutely hysterical and yet overwhelmingly calm. Maybe when two extremes of emotion happen simultaneously, you're left feeling nothing. Vincent could only mutter: "There is no cure." The only thing worse than hearing that sentence was the tangible alarm and disbelief in my otherwise extremely composed cardiac surgeon father's voice when I called him.

Immediately after my diagnosis, Vincent, my parents, and I were plunged into a quagmire of sadness, denial, and confusion. Lymphangioleiomyomatosis, referred to as "LAM" and those afflicted as "Lammies" – (cue collective cringe) – in a word, sucks. Despite its cuddly, sorority-like acronym (my disease only affects women of childbearing age - sorority indeed), LAM is a vicious, unrelenting, and frankly terrifying disease to its victims. In the medical literature, LAM is described as one might characterize a beguiling femme fatale: "[A]n elegant, monogenic model of neoplasia, defying categorization as either benign or malignant."[2] It slowly but surely destroys your lungs, rendering you vulnerable to breathlessness, lung collapse, and eventual respiratory failure. My fate seemed grim, inescapable - signed, sealed, delivered.

Keep on keeping on.

Anger is certainly no stranger to any individual diagnosed with an incurable disease. Sometimes I wonder if it is just a phase or a new permanent fixture that flares occasionally like a stubborn pimple. Two days after I was diagnosed, I bought a non-fiction book written by a thirty four year old woman diagnosed with a rare stage IV liver cancer. I devoured two hundred pages in less than ninety minutes while sitting on a bench in Rittenhouse Square. I honed in on the sections that focused on anger and denial - it was transiently comforting to know I was not alone in this seemingly isolating roller coaster of emotions. While reading, I would glance occasionally at passers-by. I felt pure rage looking at the young couple eating Capogiro gelato and holding hands, or the group of Penn undergrad girls laying on blankets wearing Ray-Bans and wiling away the afternoon. I irrationally convinced myself that nothing was wrong in their lives, or anyone else's for that matter. On this sunny afternoon in the park, nobody else had to think about needing an eventual lung transplant or dying in the foreseeable future. In my mind, life for them felt seemingly endless while mine had already been stamped with a recommended expiration date, like the shelf life of a dairy product.

In my grief, I was incredibly desperate to retain a sense of normalcy. I was terrified that if I changed anything in my life, it meant I was succumbing and giving up. So I kept going. I kept going despite undergoing multiple extremely painful lung procedures where the total fluid output each time was on the magnitude of liters, and the discomfort so great that I required sedation. I kept going despite having a semi-permanent catheter placed in my right chest, and the initial pain of the catheter left me incapable of sitting up in bed for two days or walking to the bathroom on my own. I kept going despite feeling ridiculously short of breath after climbing half a flight of stairs or pushing a patient's stretcher. I kept going despite the fears and sadness I harbored day after day, and wept to Vincent about late at night. "Will this disease kill me?" "Did I become a doctor for nothing?" "What's going to happen to me and to us?"

Keep on keeping on.

It's been more than a year since my diagnosis. I eventually went on sabbatical from work. I saw a therapist, I spent time with friends and with Vincent. I read books on meditation and mindfulness, went outside more, cooked more, passed my anesthesiology boards. As LAM patients are often cautioned against becoming pregnant, Vincent and I underwent egg harvesting and froze embryos. Perhaps most significant of all, I started taking a medication - which I've now come to regard as my wonder drug and life line - sirolimus. Sirolimus eradicated my need for more agonizing lung procedures and seems to have halted the course of my lung destruction. It's truly amazing how much comfort a tiny pill with morning coffee can bring and the magnitude of what it represents. A few years ago, this treatment didn't exist for LAM patients but due in large part to the LAM Foundation and the promising research made possible through its efforts, I am incredibly blessed with this option. It is a medication but it is also hope.

I now try to live with a heightened sense of awareness. I try not to take as many things for granted. I try to breathe more fully and deeply. So maybe this new normalcy is okay. After all, nothing is a guarantee, in this life or the next. We all grapple with the uncertainties and struggles thrown at us - this ongoing experience is one of mine. And in the end, I am filled with so much gratitude. On my bad days, I try to remember that feeling. And to just breathe - happy that I am able to do so. The LAM Foundation enabled me to continue leading a fulfilling life - to function as a normal human being, a soon to be wife, a daughter, a friend, a physician. While there is still no known cure for LAM, I maintain a great deal of optimism for the future. The LAM Foundation gives me hope.

To all fellow LAM patients, LAM families, LAM physicians, LAM scientists, LAM researchers, LAM collaborators, LAM benefactors:

Keep on keeping on.

References

1. Information on Wikipedia has since been updated to more accurately reflect the prognosis of women afflicted with LAM; 10-year survival figures range from 49–79%. Lymphangioleiomyomatotis. (n.d.). In Wikipedia. Available at: https://en.wikipedia.org/wiki/lymphangioleiomyomatosis. Accessed July 24, 2015
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