Jenny Lefferts

LYMPHANGIOLEIOMYOMATOSIS (LAM)

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I'll never forget driving to the emergency room on a Saturday afternoon in April 2011. I hadn't been feeling well, and I was pretty certain I had pneumonia and didn't want to wait until Monday to start on a course of penicillin. The ER wasn't busy so the nurse told me to hop in a wheel chair and he would escort me to radiology. We laughed the whole way because I didn't feel it was necessary. The laughing stopped when my X-ray indicated a partial pneumothorax (collapse) of my left lung. I thought they were kidding. I started to laugh but this time they didn't join in. I said that I'd deal with it on Monday because I had stuff to do, and they explained that I was not allowed to leave the hospital and that they were immediately going to insert a Heimlich valve into my chest to inflate the lung and admit me.

Most people at risk for a pneumothorax are usually very tall, thin men. Since I am a middle-aged, 5'3" female it was strongly suggested upon discharge that I follow up with a doctor and have a high resolution CT scan with contrast. I complied and after my scan I followed up with the ordering physician. He simply stated that there was a long word, 24 characters to be exact, at the end of the report that he had never heard of—but not to worry! Although he did suggest that I should avoid any rock climbing or scuba diving plans in the future.

When I got home I searched the word Lymphangioleiomyomatosis (LAM). Wikipedia stated that I had a rare, incurable, and fatal lung disease with a life expectancy of 8 to 10 years after diagnosis. I was scared, angry, and upset. My biggest fear was that I had three children to raise. I immediately got online and searched for facilities that understood this disease. My location was a blessing as I quickly found out that the nearby Stanford Hospital had a LAM program that was studying the disease. I called them immediately and the staff there invited me to a LAM support group that was being held the following weekend.

LYMPHANGIOLEIOMYOMATOSIS (LAM)

Lymphangioleiomyomatosis (LAM) is a progressive cystic lung disease typically manifesting in women of reproductive age. LAM can be either sporadic or associated with tuberous sclerosis complex (TSC). LAM involves smooth muscle proliferation that contributes to parenchymal cysts formation in the lungs. While LAM is considered an interstitial lung disease, clinically, it is essentially a cystic lung disease and shares significant physiological features of emphysema including bilateral multiple cysts and airflow obstruction.

- Symptoms may include shortness of breath, collapsed lung, chest pain, cough, fatigue
- Women with LAM may be misdiagnosed with asthma, emphysema, or bronchitis.
- Median survival in patients with LAM has varied from 10 to 30 years.

Learn more from ATS Public Advisory Roundtable member The LAM Foundation. thelamfoundation.org

What happened next was nothing more than a miracle. LAM, a rare disease, with approximately 1500 documented cases globally, had formed a foundation in 1995. A mother of a LAM patient, Sue Byrnes and Dr. Frank McCormack founded the foundation together, and it is a driving force in finding effective treatments and hopefully one day a cure for the disease.

A representative called a LAM liaison for my region, and she met with me in person and told me about an NIH study. By September 2011 I was in Bethesda, Maryland being followed by one of the top physicians focused solely on the disease. I have been back three times.

Since my diagnosis in May 2011 lymphangioleiomyomatosis has an official ICD-10 code (diagnosis code), the approval of the use of Siralumus as the first ever drug to help treat the disease, and continued funding at the NIH.

My disease is progressing slowly. I am one of the fortunate ones. I still participate at the NIH and also follow up with my provider at UCSF. But if it hadn't been for the LAM Foundation I don't know how long I would have suffered as there was so little known about this disease.

I hope to give back however possible. One breath at a time.