

Audrey Knipe

LYMPHANGIOLEIOMYOMATOSIS
(LAM)



“LAM has changed my life in so many ways, but I won’t stop fighting back. The dedicated scientists and doctors who are researching the disease won’t stop either.”

I was diagnosed with a rare disease called lymphangioleiomyomatosis (LAM) when I was 35.

I never really suspected that my chronic cough was related to a lung disease because I did not smoke. By the time I was referred to a pulmonologist, I was miserable. I was coughing for about 18 hours a day. The workup included my very first PFTs and a chest X-ray. The results showed my FEV1 was 52 percent of the predicted amount, with a 16 percent improvement from a bronchodilator. The specialists found nothing in the X-ray, so I was diagnosed with asthma.

Fast forward five months: I'm in a hospital connected to four liters of oxygen and waking up from a Nissen fundoplication with severe, stabbing chest pain. A high-resolution CT clearly showed the LAM cysts all throughout my lungs. The pulmonologist told my family about my diagnosis before he told me, so when my family came into the room I could tell some of them had been crying. I was wondering, "What's going on? Am I dying?" The next day a nurse came into my room and said, "I'm sorry about your diagnosis, but look on the bright side, at least you'll get to have a double lung transplant."

I left the hospital a week later, on oxygen 24/7. With time, I was able to stop using oxygen during the day, but I needed it from then on when I slept or exerted myself. In retrospect, I was lucky to be diagnosed with LAM at the time that I was. Results of the MILES trial were published eight months later. Sirolimus was found to stabilize FEV1 and slow the progression of the disease. I started sirolimus shortly thereafter, and my FEV1 has been stable since. This has definitely staved off my need for a lung transplant—hopefully forever!

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LYMPHANGIOLEIOMYOMATOSIS

LAM is a progressive lung disease that usually strikes women during their childbearing years, which results in the destruction of healthy lung tissue caused by cyst formation and abnormal growth of smooth muscle cells not usually found in the lungs. Symptoms may include:

- Shortness of breath
- Collapsed lung
- Chest pain
- Cough
- Fatigue.

As many as 40 percent of women with LAM have a benign kidney tumor called angiomyolipoma. Women often go undiagnosed for years, and are frequently misdiagnosed with asthma, bronchitis, or emphysema. LAM does not usually appear on an X-ray. A high-resolution CT scan of the chest, and often the abdominal area, is required for accurate diagnosis. Lung capacity in LAM patients progressively declines, sometimes resulting in the need for supplemental oxygen.

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

Each day I learn to live with this life-sucking disease. It sucks away my energy and leaves me chronically fatigued. It also produces anxiety and insomnia, so when I'm seeking restful, restorative sleep, it will not come. To make matters more difficult, LAM is believed to be estrogen-mediated, and each month I'd have ten days where my shortness of breath worsens dramatically. I talked to my doctor about trying leuprolide in addition to sirolimus, just to see if that made any difference. It certainly did. I had wonderful success decreasing my shortness of breath for those ten days per month. Interestingly, my FEV1 also improved by about 10 percent over the next year. Ultimately, I chose to have an oophorectomy in the hopes of improving my lungs permanently.

I miss the old days when I felt good and had energy. I loved walking around the lake in the evenings, unencumbered and free. I now have to walk carrying a nine-pound concentrator. I used to love going to the mountains for a long weekend with my family. Now it is too much effort bringing all of my medical equipment with me. I take vacations at sea level instead, learning what I can and can't do in a day and not overdo it. LAM has changed my life in so many ways, but I won't stop fighting back. The dedicated scientists and doctors who are researching the disease won't stop either.