Mary Stojic

LYMPHANGIOLEIOMYOMATOSIS (LAM)

"I am active in advocacy, participate in regular visits to the NIH, and I volunteer… I care about giving other LAM patients hope, especially the young women starting their families."

Mary Stojic...

In 1985, at age 21, I suffered a spontaneous pneumothorax of my right lung, which required pleurodesis. I remember the thoracic surgeon saying that he performed aggressive mechanical pleurodesis and covered everything he could reach, removing cysts from the edge of my lung to prevent future problems. Many years and much research later, it is now known that cysts or blebs should not be removed for patients with LAM because we need the good lung surrounding the cysts for as long as possible.

This aside, even without a LAM diagnosis, the aggressive treatment I received granted me an incredible quality of life. I studied and traveled in Europe, married and had three children. Other than a small "nuisance" pneumothorax of only 10 percent during my second pregnancy and chronic bronchitis, I did well, until 1998. The flu vaccine was not effective that year, and six weeks after a minor flu, my other lung collapsed.

After a month of waiting for it to inflate, I returned to the doctor ready to demand surgery so that I could recover and resume my life. I was surprised to learn that my lung was fully collapsed. After the pleurodesis on the left lung, I anticipated I would resume my normal activities with no further problems. After two more bouts of bronchitis in the next six months, the pulmonologist requested a CT in October 1998.

While awaiting the results, I went to the library to do research online, read about LAM in the Rare Disease and Disorder Book and found the contact information for the LAM Foundation, which led to a doctor at the NIH and the beginning of my quest for a cure.

With the diagnosis came overwhelming fears for my three daughters, ages seven, six, and four. Although it seemed apparent I did not have the extremely fast type, as I had managed symptoms for a minimum of 13 years, my fear was that things would change. I could stumble upon a trigger, and my children would not have their mother. Since LAM seemed to affect only women, I was concerned that my daughters would inherit the disease. Later when we learned about tuberous sclerosis complex, more fears emerged.

LYMPHANGIO-LEIOMYOMATOSIS (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. www.thelamfoundation.org

I am now 53. My daughters have all graduated from college. My desire is to be here for their weddings and to hold their children.

Trying to stay well while raising active young children, with the endless ups and down of my health, was a challenge! In an effort to slow LAM, I took medroxyprogesterone for a year before the research clearly showed it was not effective. My husband deserves an award for seeing me through this trying phase.

I constantly fear lung collapse, and with every chest discomfort I ask myself, "Is it a pneumo or just a chest twinge? Is it necessary to visit the doctor or go to the hospital?"

I have some assurance that the pleurodesis will not allow a tension pneumo. Yet repeated "nuisance" pneumos (those I define as causing disruption to the family routine and requiring rest, but do not require medical intervention) have occurred on each side. In 2009 and 2010, after many disrupted family plans due to pneumos, both lungs had to be repleurodesised. As a consequence, I am a strong proponent of aggressive pleurodesis for a better quality of life, with longer periods of time between events.

Fatigue afflicts many LAM patients. It saps our energy, which can impact our career choices and finances. As a college graduate, I did not work outside the home other than the occasional mornings at the local nursery school to reserve my energy to be able participate in my children's lives after school. I continue to plan my schedule daily, weekly and monthly, so as not to have too many activities which would exhaust me and lead to bronchitis or a lung collapse.

Routine activities, like grocery shopping, have become events that need to be planned. What appears simple requires a lot of energy. Beyond the shopping, there's the loading and unloading of the car, sorting and storing food, and finally preparing the meals. Bronchodilators helped liberate me. Previously, I would have to nap most afternoons.

With my daughters grown, I decided to return to work. I needed a position with limited exposure to germs, so nothing in retail or working with children. Although my former career path was not an option as it would be too physically demanding, I found a sedentary job that allowed me to reserve some energy for my husband and our life together.

I have been taking an immunosuppressant for over five years to help slow the disease. I wonder about side-effects as I will likely be taking it for the rest of my life. My lungs have not collapsed since 2010. Yet I worry as I plan vacations, if I'll have to postpone or cancel trips, and I feel generally uneasy about being away from home and my doctors. My function has declined, and I am short of breath on exertion.

I am grateful to my pulmonologists. I am active in advocacy, participate in regular visits to the NIH, and I volunteer as a liaison for the LAM Foundation. I care about giving other LAM patients hope, especially the young women starting their families. Young women are given this diagnosis with too many unknowns, but this can change.

Where there's research, there is hope.

Mary Stojic was a patient speaker at the ATS 2017 International Conference in Washington, D.C.