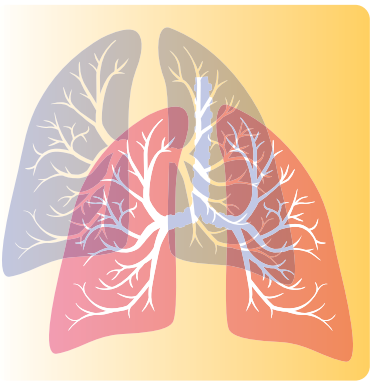
AMERICAN THORACIC SOCIETY
Patient Information *Series*

Lymphangioleiomyomatosis (LAM)

What is LAM?

Lymphangioleiomyomatosis (lim-FAN-jee-oh-ly-oh-my-oh-ma-TOE-sis), commonly known as LAM, is a rare lung disease that affects women almost exclusively, usually during their childbearing years. People with LAM have an unusual type of muscle cell that grows in the tissues of the lungs. Although these cells are not considered cancerous, they grow uncontrollably throughout the lungs. These muscle cells can block the lymph vessels, blood vessels, and airways, preventing the lungs from providing oxygen to the rest of the body.



An unusual kidney tumor called angiomyolipoma (an-jee-oh-my-oh-ly-PO-ma) is found in up to 50% of patients with LAM. It is rarely cancerous and usually does not cause problems.

Over 1,500 women have been identified with LAM worldwide, but now that we know of a genetic link to another disease called tuberous sclerosis (TS), scientists believe that there are over 250,000 women with LAM worldwide. Early symptoms of LAM are similar to those of other lung diseases, as a result, LAM is often misdiagnosed as asthma, emphysema, or pulmonary bronchitis.

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What are the symptoms of LAM?

Early symptoms of LAM are similar to those of other lung diseases. Symptoms include:

- Shortness of breath during physical activity
- Coughing
- Chest pain
- Fatigue
- Lung collapse
- Coughing up small amounts of blood

How does LAM progress?

LAM progresses at a different rate for different patients, so although we know that a patient's lung function will worsen over time, we cannot predict how rapidly this will happen. LAM is generally "slowly progressive," meaning that it will slowly worsen over time. As the disease advances, there can be more growth of muscle cells throughout the lung and repeated leakage of fluid into the chest cavity (pleural effusions). Blockage of lung tissue may cause small sacs (cysts) to be formed, and the lung takes on a "honeycomb" appearance on chest imaging. Cysts near or on the surface of the lungs can rupture. As air leaks from the lung into the chest cavity, the lung or part of the lung can collapse. In some cases, the lung can repair itself and re-expand. However, if air continues to leak into the chest cavity, the doctor may need to re-expand the collapsed part of the lung by removing the air using a tube inserted through the chest wall into the chest cavity. (See ATS Patient Information Series: Chest Tube Thoracostomy, http://www.thoracic.org/sections/education/patient-education/patient-education-materials/index.html). Eventually, LAM can lead to respiratory failure.

How is LAM diagnosed?

There are a number of tests a doctor can do to make a diagnosis of LAM and check for its progression. These include:

Chest X-rays can diagnose a collapsed lung (pneumothorax) or the presence of fluid in the chest cavity (pleural effusion). A routine chest X-ray often cannot pick up the lung cysts when they are small.

Pulmonary function tests can help show how much LAM has affected lung function. A person breathes into a machine called a spirometer to measure how much and how well air flows into and out of the lungs. (See ATS Patient Information Series piece: Pulmonary Function Tests, http://www.thoracic.org/sections/education/patient-education/patient-education-materials/index.html)

An arterial blood gas or pulse oximetry test helps show whether the lungs are providing an adequate supply of oxygen to the blood.

Computed tomography (CT) provides multiple two-dimensional images of the inside of the lungs and chest. It is considered the most useful imaging test for diagnosing LAM. A CT scan of the chest can show the presence of cysts in the lungs. A CT scan of the abdomen will

show whether a person with LAM has an angiomyolipoma.

In order to make a definite diagnosis of LAM, the doctor often needs to get a sample of lung tissue that can be looked at by a doctor. With a microscope, the doctor can see the abnormal muscle cells and cysts that indicate LAM. There are several ways that a piece of lung tissue can be removed:

Transbronchial biopsy. A long, narrow, flexible, lighted tube called a bronchoscope is passed through the nose down the windpipe and into the lungs (this test is called a bronchoscopy—see ATS Patient Information Sheet: Bronchoscopy http://www.thoracic.org/sections/education/patient-education/patient-education-materials/index.html). The doctor uses a tiny forceps to take bits of lung tissue. The transbronchial biopsy can be done in a hospital on an outpatient basis under local anesthesia. However, it may not get enough lung tissue to make the diagnosis of LAM.

Video-Assisted Thoracoscopic Lung biopsy. Tiny incisions are made in the chest wall to insert instruments and a viewing scope. The doctor removes small pieces of lung while watching on a video screen . Thoracoscopy is performed in the hospital under general anesthesia. It is a less invasive than an open lung biopsy.

Open Lung biopsy. A surgeon makes an incision in the chest wall between the ribs on one side. A few small pieces of lung tissue are removed. Open lung biopsies are performed in the hospital under general anesthesia.

How is LAM treated?

There is no cure for LAM at this time. Because LAM almost always affects women of childbearing age, doctors believe that the hormone estrogen may play a part in the abnormal muscle cell growth. Some LAM treatments focus on reducing the production or effects of estrogen, but no treatment has yet been proven effective. No one therapy has been found helpful for all patients; the response to treatment varies from person to person. Several new treatments for LAM are currently being studied.

A person with LAM eventually may need oxygen therapy if the disease continues to worsen (See ATS Patient Information Series: Oxygen Therapy, http://www.thoracic.org/sections/education/patient-education/patient-education-materials/index.html). Lung transplantation may be considered if a person has respiratory failure and is not responding to treatment.

How will LAM affect my lifestyle?

In the early stages of the disease, most people can live normally, and participate in daily activities such as school, work, and common physical activities. In more advanced stages, LAM patients may have very limited ability to move around and may need to use extra oxygen during sleep or all of the time.

Patients with LAM should follow the same healthy lifestyle recommended for the general population, including eating a healthy diet, getting as much exercise as they can tolerate, as well as plenty of rest. Like all lung disease, tobacco smoking can make symptoms worse. People with LAM should not smoke and should avoid being around smoke. Women with LAM should avoid estrogen products, such as birth control pills. Many doctors and LAM patients feel that pregnancy can make the disease progress faster.

LAM research programs

Some LAM patients may be eligible to participate in clinical studies at the National Heart, Lung and Blood Institute at the National Institutes of Health in Bethesda, Maryland. Participants must meet specific LAM Patient Protocol requirements. A new medication is beginning trials through a research program this summer. Contact The LAM Foundation at wwwthelamfoundation.org for further information.

Source: The LAM Foundation, "About Lymphangioleimyomatosis," http://www.thelamfoundation.org/LAM_aboutLAM.htm#patient

Additional Lung Health Information

American Thoracic Society
http://www.thoracic.org/sections/education/patient-education/patient-education-materials/index.html

ATS Public Advisory Roundtable
www.thoracic.org/aboutats/par/par.asp

National Heart Lung & Blood Institute
www.nhlbi.nih.gov/index.htm

American Lung Association
www.lungusa.org

Rx *What to do…*

- ✔ Let your doctor know if you are having problems with exercise, shortness of breath, or you cough up blood.
- ✔ Do not smoke and avoid being around smoke.
- ✔ Have your lung function checked regularly.
- ✔ Talk with your doctor about treatment to try to control LAM.

Doctor's Office Telephone:

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