



HEALTH FOR LIFE

Lymphangiomyomatosis

A rare lung disease, lymphangiomyomatosis (limf-an-geo-leo-my-o-ma-to-sis) is also called LAM. Lymph and angio refer to lymph and blood vessels. Leiomyomatosis refers to the bundles of unusual muscle cells that form.

With LAM, muscle cells invade the tissue of the lungs, including the airways, lymph and blood vessels near the lungs. Over time, these cells block the flow of air, blood and lymph to and from the lungs. The lungs are not able to get oxygen out to the rest of the body as a result.

Causes

The cause of LAM is not known. It most often occurs in women of childbearing age, but there have been several cases in women who were menopausal. It has become more common in the past 5-10 years but that may be because doctors are better able to diagnose it now than in the past.

Symptoms

- Shortness of breath with activity that worsens as the disease progresses
- Cough
- Chest pain
- Air-filled sacs like blisters, called blebs that form near or on the surface of the lungs
- Lung collapse from air leaking from blebs that rupture (pneumothorax)
- Fluid build up in the lungs (pleural effusion)

Treatment

No therapy has been found to work for all patients with LAM. Because most often LAM affects women of childbearing age, treatment has focused on reducing the production or effects of estrogen. This is done with medicines containing a hormone progesterone, or by surgery to remove the ovaries.

Oxygen therapy may be helpful, especially as the disease progresses. Eating a healthy diet, getting plenty of rest and regular exercise are recommended. Avoiding smoking and being around others who smoke is very important.

With severe LAM, lung transplant may be treatment option.