

Study finds treatment for rare lung disease

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An Oregon Health & Science University researcher has co-authored an international study that revealed a drug approved to prevent rejection in organ transplant patients helped treat a rare lung disease in women.

The life-threatening disease has no cure and, until now, no known treatment.

The clinical trial of the drug -- called sirolimus -- was the first randomized, controlled study designed to develop a therapy for the lung disease, [lymphangiomyomatosis](#), or LAM.

LAM is a progressive, cystic [lung disease](#) that occurs almost exclusively in women. In LAM, smooth muscle cells grow uncontrollably and spread to restricted areas in the body, including the lungs, lymph nodes and vessels and kidneys, limiting the flow of air, blood and immune system fluid, or lymph.

Shortness of breath and recurrent lung collapse are common in patients with LAM; until now, lung transplantation has been the only hope for patients who progress to respiratory failure. LAM affects about five per million people.

The positive results from the clinical trial of sirolimus, also known as rapamycin, were reported last month in the online edition of the *New England Journal of Medicine*.

The lead investigators in the study were from the University of

Cincinnati and Cincinnati Children's Hospital Medical Center. Alan Barker, M.D., an OHSU expert in rare lung diseases, was a principal investigator in the study.

The study was conducted at 13 institutions throughout the United States, Canada and Japan. The only West Coast institutions involved in the study were OHSU and the University of California at Los Angeles. Research subjects from the Northwest and northern California were enrolled in the study at OHSU.

Eighty-nine women with LAM, aged 18 or older, participated in the study. Some were treated with sirolimus; others were given a placebo.

The researchers found that sirolimus stabilized lung function and was associated with improvement in measures of functional performance and quality of life.

OHSU's Barker said that the study results point to a potential treatment for LAM. The results are also important because they could serve as a model for treating common types of cancers.

"Like cancer, LAM occurs when cells grow out of control; in LAM's case, cells in the blood vessels and breathing passages in the lung grow out of control," Barker said. "This drug stopped that abnormal growth of cells."

Barker said that OHSU also will be part of a follow-up study that examines the effectiveness of another drug in treating LAM.

Provided by Oregon Health & Science University

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