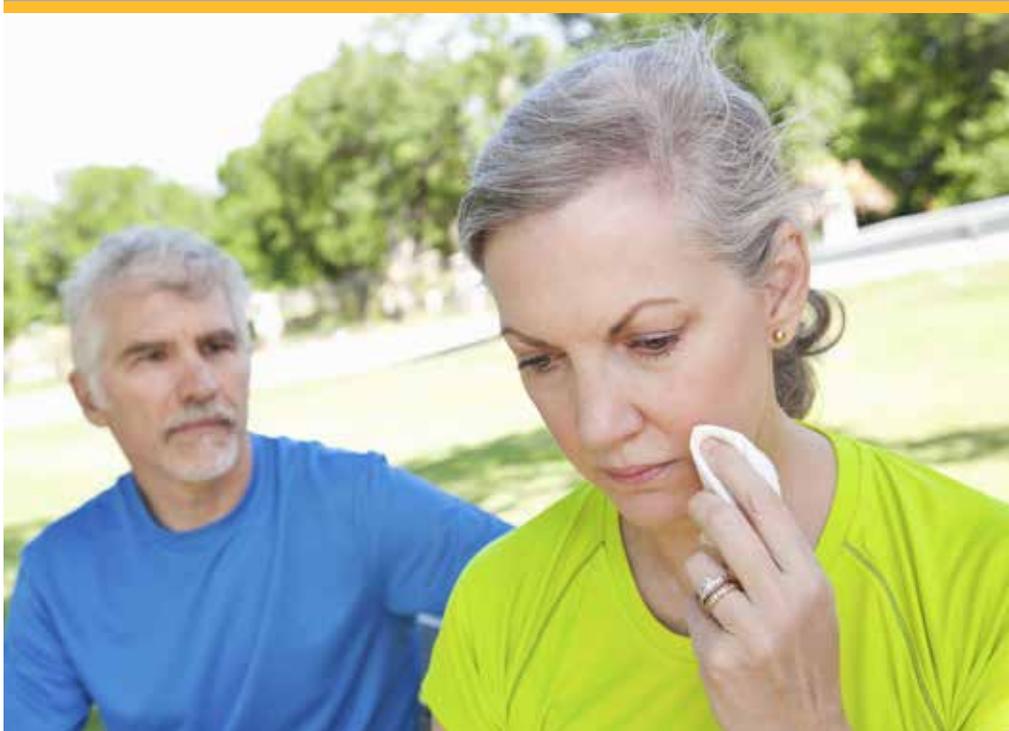


State-of-the-art treatment for pulmonary arterial hypertension



Pulmonary arterial hypertension (PAH), or high blood pressure in the lungs, is a deadly condition often diagnosed relatively late in the course of the disease. Increased research efforts have yielded significant advances in the field of pulmonary vascular disease and new therapies offer hope of better outcomes. Physicians at UCLA are quick to see patients referred for the condition and routinely utilize aggressive treatment regimens.

Individuals with PAH have narrowed blood vessels in the lungs, making it difficult for blood to circulate through the lungs and to deliver oxygen to other parts of the body. This “high pressure system” in the lungs places increased stress on the heart, and over time the right side of the heart becomes enlarged and thickened.

PAH symptoms often present as nonspecific and may include shortness of breath on exertion, generalized fatigue and lower extremity swelling. As the condition worsens, some patients experience chest pain and dizziness on exertion, which may result in loss of consciousness during routine physical activity. PAH symptoms are often attributed to other conditions such as asthma, depression, obesity and poor physical conditioning, factors that may contribute to why PAH is diagnosed late in many patients.

Quick consultation is key to improved survival

Since rapid diagnosis and treatment are critical, physicians at UCLA see PAH patients in a timely manner. The specialized Pulmonary Hypertension Program at UCLA offers a comprehensive approach to the care of PAH patients from initial diagnosis to aggressive therapeutics to long-term follow-up care, resulting in improved quality of life and overall survival for the majority of patients.

UCLA’s team of physicians and nurses include experts in pulmonary and critical care medicine, cardiology, congenital heart disease, rheumatology and thoracic surgery. Patients have access to numerous clinical trials, as well as to physicians in the Lung and Heart-Lung Transplant Program, one of the largest in the world.

“It’s a deadly disease — pulmonary hypertension of any origin is cause for serious concern,” says Rajan Saggur, MD, director of the Medical Care Intensive Unit at Ronald Reagan UCLA Medical Center. “PAH is often progressive and, left untreated, has a poor survival rate, akin to that of an untreated cancer. Seeing patients urgently is key.”

Causes and diagnosis

Pulmonary hypertension has several potential etiologies, including congestive heart failure, blood clots, HIV, drug abuse, cirrhosis, appetite suppressant medications, autoimmune disorders, sleep apnea, and lung diseases such as emphysema or pulmonary fibrosis. However, the condition often affects individuals without a significant medical history, particularly young to middle-age females. When PAH is being considered as the cause of a patient's symptoms, an echocardiogram, or ultrasound of the heart, is the screening test of choice.

An important cause of pulmonary hypertension that can easily be missed relates to patients with chronic thromboembolic pulmonary hypertension (CTEPH). There are approximately 2,500 new cases of CTEPH in the U.S. annually and 25-75 percent of patients have no prior history of an acute pulmonary embolism.

PAH patients are grouped into four functional classes. Class I patients display no symptoms under ordinary physical activity, while Class II patients show symptoms with ordinary activity and have slight limitation of overall physical activity.

In Class III PAH, patients exhibit symptoms with even less exertion and are more limited in their physical activity. Class IV patients have the least functional capacity, experiencing symptoms with any activity or even at rest. Patients seen at UCLA are most often advanced cases with Class III or IV symptoms.

Aggressive treatments can save lives and delay progression

Two decades ago, no treatment options existed for PAH. Now, more than a dozen FDA-approved therapies and modes of medication delivery are available and several exciting new treatments are in development.

Oral and inhaled medications can both be used to help delay disease progression. In more severe cases, a continuous infusion of medication may be needed, which can either be delivered subcutaneously through a small catheter placed in the abdominal area, or directly into a central vein using an intravenous catheter. Such parenteral therapies are being increasingly used by UCLA physicians to help PAH patients who are in advanced stages but who do not necessarily require hospitalization.

For CTEPH patients, pulmonary endarterectomy can significantly decrease mortality, while new medical therapies are available for CTEPH patients who are either not surgical candidates or have persistent pulmonary hypertension after surgery.

Modern therapies have significantly improved life expectancy among PAH patients. While the original NIH Registry showed a five-year survival rate of 34 percent for these patients, the more recent REVEAL Registry reports a five-year survival rate of 70 percent.

Some PAH patients may be candidates for lung transplantation, however improved medical therapy has reduced the need for the procedure. Currently, only five percent of lung transplantations are performed specifically to treat pulmonary hypertension.

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