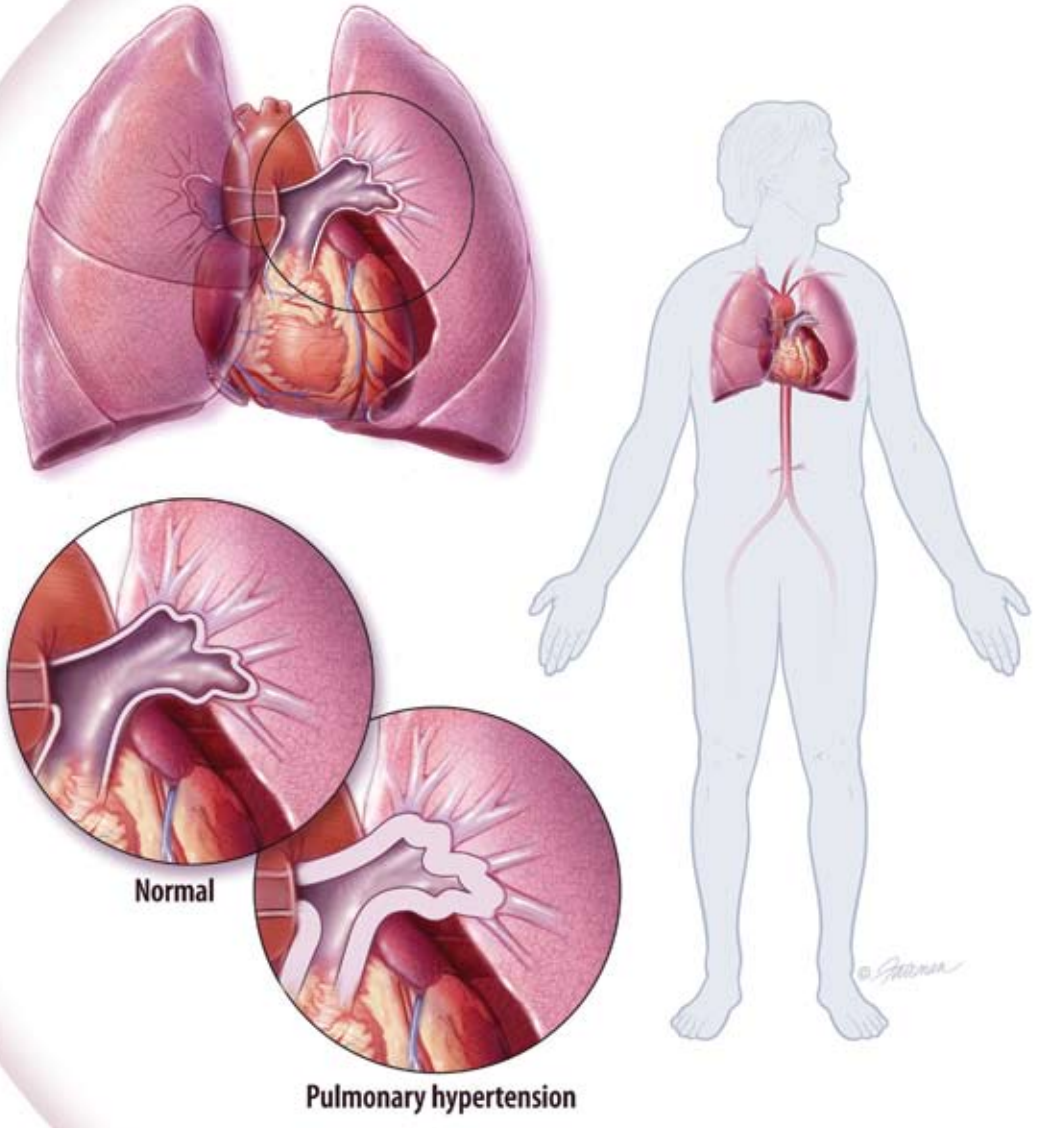
Helping the Heart-Lung Link

Lahey specialists collaborate to improve care for patients with a complex disease.

Healthy lungs work with the heart to provide oxygen to the blood. Normally, the right ventricle pumps blood from the heart through the pulmonary arteries to the lungs, where the blood receives oxygen. The newly oxygenated blood then flows back to the heart, where the left ventricle pumps it out to the body.

In patients with pulmonary arterial hypertension, the pulmonary arteries become thick, stiff or blocked. The arteries do not allow the blood to move efficiently, causing pressure to build in the arteries and blood to back up in the right ventricle. This, in turn, causes the right ventricle to work harder to pump blood, eventually leading to its enlargement and right heart failure.



As the year-end holidays approach, members of the Gilpatrick family will gather to help prepare ravioli for the family's celebrations. Eleanor Gilpatrick specializes in a style of ravioli passed down from her mother's family in Parma, Italy. It is she who supervises the production of the "secret" family recipe at her home in Arlington, Mass.

It's also a safe bet that, come Christmas morning, someone in the Gilpatrick family—perhaps a grandchild or great-grandchild—will unwrap a hand-made afghan as a present. After being hospitalized a few years ago, Eleanor Gilpatrick resumed a hobby she had put aside for a time: knitting cable-stitched afghans.

At 87, she accomplishes a lot, and she does so despite having pulmonary arterial hypertension (PAH), a serious chronic illness. PAH affects the lungs and the heart. It can be difficult to diagnose and extremely disabling in advanced stages. Eleanor Gilpatrick is benefiting from a new generation of treatment—and from multidisciplinary care that is helping her defy the odds.

A Diagnostic Challenge

In patients with PAH, the pulmonary arteries become thick or stiff, making them less efficient in allowing the movement of blood to the lungs (see illustration). As a result, pressure builds up in the arteries and the heart, and the heart has to work harder to push blood through. Over time, the extra work leads to an enlarged heart muscle and heart failure.

The condition is difficult to diagnose because the early symptoms—fatigue and shortness of breath—are so nonspecific. Many other conditions cause similar symptoms, and patients can easily be misdiagnosed.

"When a physician sees a patient with unexplained shortness of breath, he or she should suspect pulmonary arterial hypertension," says Lahey's Akmal Sarwar, MD, a pulmonary medicine specialist. "Even though it is a relatively rare condition, we believe it is significantly underdiagnosed."

PAH is classified as either primary or secondary. When no cause for the condition can be found, patients are said to have primary (or idiopathic) PAH. Secondary PAH, which is more common, is a long-term effect of another condition. Scleroderma (a connective tissue disease) and congenital heart defects are two of the most common causes of secondary PAH. Yet more than 70 other diseases can cause pulmonary hypertension, including emphysema, chronic obstructive pulmonary disease, heart diseases, liver disease, HIV, sleep apnea, pulmonary embolism and sickle cell disease.

"Identifying pulmonary hypertension doesn't give us the diagnosis; it only tells us the patient has abnormal pressure in the pulmonary arteries. Why that pressure builds up—that's where the diagnostic challenge comes in," says Sarwar. "It is a very broad category."

Even when PAH is suspected, physicians need to go through a process of elimination to rule out other conditions. Making such a diagnosis is not easy for primary care physicians in the community.

"There are so many conditions they need to consider," Sarwar says. "If the physician has done routine tests, such as a chest X-ray and pulmonary function tests, and hasn't come up with an answer, and the patient's shortness of breath remains unexplained, the physician should consider referring the patient to a specialty clinic like ours."

Sarwar is medical director of Lahey's Pulmonary Hypertension Clinic, a collaborative effort that brings together specialists from multiple disciplines. The new clinic combines the strengths of Pulmonary Medicine, Cardiology and other specialties in an effort to diagnose patients earlier and treat them before the disease becomes advanced.

A multispecialty focus on PAH is essential, because the lungs and the heart rely on each other to function properly. **"The heart and lungs are so intrinsically joined; what happens in one organ can really affect the other organ,"** says Lahey's Sergio Waxman, MD, a cardiologist who frequently consults with Sarwar. "There are initial, good screening methods, but many times these tests come back and don't reveal much. It takes a dedicated team to look at these patients and try to understand what is going on."

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"You really have to know your patient well, be very thorough in your evaluation of the patient, know the drug—what you're giving to the patient—and follow them very closely."

—Akmal Sarwar, MD

Above left:
Sarwar with Eleanor Gilpatrick.

Two of the most important diagnostic tests for PAH are evaluated jointly by pulmonologists and cardiologists.

- **Echocardiogram:** A noninvasive test, the echocardiogram can reveal heart problems such as heart valve disease or abnormalities in the right or left ventricles. In these cases, the normal blood flow can be disrupted, leading to pulmonary hypertension. If a significant heart problem is found early enough, it can often be repaired, possibly reversing the effects of pulmonary hypertension.
- **Right heart catheterization:** In this test, a catheter is threaded through an artery in the groin or the neck into the right ventricle and pulmonary arteries. With the catheter in place, physicians can get a true measure of the pressure in the different chambers of the heart and in the pulmonary arteries. This is considered the "gold standard" diagnostic test for PAH.

"The actual pressure and its response to a drug challenge are two of the metrics that can predict the patient's response to medication down the line," Waxman says. Physicians can actually administer certain medications during a right heart catheterization to see whether a particular drug helps reduce the pressure inside the pulmonary arteries.

Advances in Treatment

Sarwar credits newer medicines with helping to improve patients' quality of life and extending survival. These include blood vessel dilators, *prostacyclins*, *endothelin receptor antagonists*, and *phosphodiasterase enzyme inhibitors*. Though these drugs work through different mechanisms, they all serve to keep blood vessels open, relaxed and better able to move blood along its course.

"In the past 10 to 15 years, there has been significant interest and research in this field," says Sarwar. "As a result, researchers have been able to develop treatments that can alter the course of the condition."

Such has been the case for Eleanor Gilpatrick, who came to the hospital with signs of right heart failure. Her ordeal began when, she says, "I felt like my heart wasn't pumping right." She was having difficulty breathing and, according to her daughter, was sleeping most of the time.

When she refused to take a phone call from her son, the family knew something was very wrong. Her grandson notified his aunt, who lives nearby, and they called 9-1-1. Eleanor Gilpatrick went from her normal day to the Intensive Care Unit at Lahey Clinic Medical Center in a matter of 30 minutes.

Sarwar was the attending physician in the ICU that day. He suspected PAH. Once his patient's condition had been stabilized, he ordered a right heart catheterization, which confirmed his hunch. He further traced the origins of her pulmonary hypertension to scleroderma, a disease that commonly causes narrowing of blood vessels in the lungs and, therefore, PAH. Sarwar ordered oxygen and prescribed a relatively new drug that reduces the narrowing of blood vessels.

"Once Mrs. Gilpatrick got started on treatment, about four months later, we did an echocardiogram, and the symptoms were gone," Sarwar says. "The right side of her heart is good, and the pulmonary arterial pressure has improved significantly."

Still, he points out, Eleanor Gilpatrick is an exception. The vast majority of patients with advanced PAH do not respond as well to medication. Identifying the disease earlier is the key to helping people survive longer.

"In pulmonary hypertension clinics across the country, most patients are at advanced stages when they are diagnosed," Sarwar says. "The important point to make is for physicians to have a high index of suspicion for pulmonary hypertension. Early referral to a pulmonary clinic, and appropriate diagnostic evaluation and management can improve the quality of life for our patients, improve survival and prevent complications."

To make an appointment with a pulmonary medicine specialist at Lahey, call 781-744-3250.