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Packard Children's NEWS

Lucile Packard Foundation for Children's Health

Cover Story:

Diagnosis: Pulmonary Vascular Disease

The Vera Moulton Wall Center seeks causes and cures for this elusive condition.

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Ramona Doyle, M.D. and Jeffrey Feinstein, M.D.

Seven-month-old Robert Obelton Jr. suffers from severe pulmonary hypertension due to abnormal development of his pulmonary arteries. Here, his parents visit him in Packard's pediatric intensive care unit.

Diagnosis: Pulmonary Vascular Disease

By Lauren Cuthbert

Seeking Causes and Cures for this Elusive Condition

Michael, an athletic 15-year-old high school student, complained of decreased energy and shortness of breath while playing basketball. His pediatrician suspected asthma and began a course of treatment. The teen's condition worsened over the next few weeks and he found himself unable to climb a flight of stairs without becoming winded.

He was sent to Packard Children's Hospital for a lung function test, which showed an abnormally low level of oxygen in his blood. Concerned about possible heart disease, Michael's physicians ordered an echocardiogram (sonogram of the heart). Though the structure and function of the heart were normal, the blood pressure in Michael's right ventricle and pulmonary arteries was found to be nearly five times the normal level.

The diagnosis was pulmonary hypertension, which is characterized by high pressure in the blood vessels of the lungs. Michael was referred to Jeffrey Feinstein, M.D., director of the Pediatric Cardiac Catheterization Lab and assistant professor in Pediatric Cardiology, to determine what was causing high pressure within the lungs.

Dr. Feinstein explained to the family that pulmonary hypertension is a disease that impedes the heart's ability to pump blood through the lungs, resulting in an inadequate supply of oxygen and nutrients reaching the body's organs. The high pressure within the lungs can, in turn, lead to pulmonary vascular disease, in which the vessel walls become thick in response to the high pressure. Pulmonary hypertension may be due to a number of different causes, such as lupus, congenital heart defects, blood clots, or the use of certain appetite suppressants. Whatever the cause of this disease, the result can be debilitating.

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> ■ Jeffrey Feinstein, M.D.

"The longer this goes on without being detected, diagnosed, and treated, the harder the heart has to work," Feinstein says. "Over time, a person can develop heart failure."

In addition to referring Michael for evaluation by specialists in rheumatology and pulmonary medicine, Feinstein sent him for a ventilation/perfusion scan, which compares the air and blood supplies to the lungs. Ideally, both air and blood should travel the same route. In Michael's case,

The Vera Moulton Wall Center for Pulmonary Vascular Disease

Last fall, a $31.8 million anonymous gift was given to establish the Vera Moulton Wall Center for Pulmonary Vascular Disease at Lucile Packard Children's Hospital and Stanford University Medical Center. The gift, made through the Lucile Packard Foundation for Children's Health, will allow Stanford to become a national leader in the investigation and treatment of pulmonary vascular disease in children and adults.


Vera Moulton Wall

Jeffrey Feinstein, M.D., M.P.H., assistant professor in Pediatric Cardiology and director of Pediatric and Congenital Cardiac Catheterization at Packard Children's Hospital, is the Center's director. Ramona Doyle, M.D., assistant professor in Pulmonary and Critical Care Medicine and associate director of the Lung/Heart-Lung Transplantation Program at Stanford, is co-director.

The Center's aim is not only to improve diagnosis and treatment for pulmonary vascular disease through state-of-the-art research, but also to increase awareness among physicians of this complex and often overlooked disease. Collaboration among adult and pediatric specialists in cardiology, pulmonary medicine, and vascular biology, as well as between the Schools of Medicine and Engineering, will be central to the Center's efforts.

The Wall Center's overlap between medicine and engineering falls within Stanford's Bio-X program—an initiative to unite the biological sciences by encouraging collaboration between different disciplines. "The Wall Center fits the Bio-X model of using the strengths of Stanford's many schools to approach a very important problem," says Harvey Cohen, M.D., chief of staff at Packard Children's Hospital and chair of pediatrics at Stanford. "We're going to train the next generation of scientists—both physician scientists and engineering scientists—who may be the ones to come up with tomorrow's advances."

Vera Moulton Wall, for whom the Center was named, was born in Savannah, Georgia in 1927. After graduating from Mary Baldwin College in Virginia with a degree in biology, she pursued a career in biology and teaching, spending the majority of her adult life in Dallas, Texas. At the time of her unexpected death on Mother's Day 1988, she had been happily married for 40 years and had six grandchildren. Vera is most remembered for her generous spirit and her love of children. The Vera Moulton Wall Center honors her memory through the children, now and in the future, who will be helped by the Center's efforts to treat pulmonary vascular disease. ■

however, it was immediately clear that there were areas air reached but blood did not, due to a number of blood clots that were causing the high pulmonary blood pressure.

Diagnosing Pulmonary Hypertension

Statistics show pulmonary hypertension and pulmonary vascular disease to be rare, affecting only three of every million Americans. But Feinstein says that such numbers may be misleading because the disease often goes unrecognized or is misdiagnosed, especially in children. "Clearly," he says, "we're missing a lot of cases."

Fortunately for Michael, he wasn't one of them. Employing a novel angioplasty technique, Feinstein inserted a catheter through the large vein in Michael's neck to reach the blood vessels of his lungs, opening the areas blocked by clots. Feinstein is one of only two physicians in the nation currently doing this procedure. He says the teenager now appears fully recovered and is back to most of his favorite sports. "He was lucky," Feinstein says. "Thanks to some very observant physicians along the way, his disease was caught early on. That isn't always the case."

Currently, few physicians are familiar with the condition, instead attributing a patient's complaints to common ailments such as asthma (as in Michael's case), also manifested by shortness of breath, fatigue, and poor exercise tolerance.

"Given the dearth of expertise on pulmonary vascular disease, it has become increasingly important to get the word out."

■ Ramona Doyle, M.D.

"Given the dearth of expertise on pulmonary vascular disease, it has become increasingly important to get the word out," says Ramona Doyle, M.D., associate director of the Lung/Heart-Lung Transplantation Program and assistant professor in Pulmonary and Critical Care Medicine. Drs. Doyle and Feinstein have worked in partnership to diagnose and treat many patients. "I've been talking to hospitals and am available for consultation with referring physicians. Outreach education and physician training are key if we're going to identify patients with this disease early enough to treat it successfully," says Doyle.

Equally important is providing top-notch care to patients once they are diagnosed. "The bottom line," Feinstein says, "is that we still do not have cures for most causes of pulmonary hypertension and pulmonary vascular disease. The treatments available today are better than what we had some years ago, but are by no means good. A tremendous amount of work must be done before we can say we're winning the battle against this disease, and this will require new therapies."

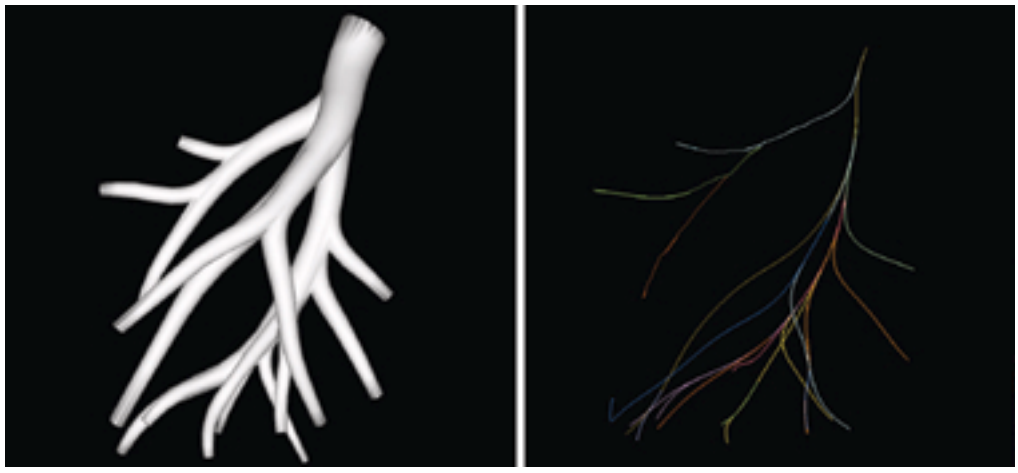
The mother of an 18-month-old Packard patient is hoping for such a breakthrough. Her son was born with a hole in his heart and pulmonary vascular disease. Since the boy's health is too tenuous to consider transplant as an option (the pressure in his lungs is so great that it is unlikely he would survive the surgery), the family's hopes rest on research advances. "We're praying for a miracle," said his mother.

Innovative Research Programs

Among the most innovative research programs today are investigations of the mechanics of blood flow in the lungs. Feinstein hopes to build on groundbreaking work he has been conducting for several years in collaboration with Stanford's School of Engineering. Funded by a grant from the Montgomery Street Foundation, the project allows researchers and physicians to use patient-specific computer models to determine which blockages are most important to relieve. "This research is light years ahead of its time and will likely change the way we practice medicine in the future," Feinstein says.

At present, the only recourse for the sickest patients is a lung or heart and lung transplant. "Currently, success means we are able to manage the sickest patients until they can get a transplant," Doyle says.

Adds Feinstein, "Ten years from now, success will mean eliminating the need for transplants in these patients." ●



A non-invasive CT scan of a patient's pulmonary artery (left) is converted to a computer model (right) which displays the mechanics of each patient's unique pulmonary flow. By using this model as a diagnostic and investigative tool, researchers can explore innovative treatment options without exposing the patient to risks.