

## UCLA's Cystic Fibrosis Program receives accreditation for pediatric and adult care



**The Cystic Fibrosis Program** at Mattel Children's Hospital UCLA offers comprehensive clinical care, research and educational support for infants, children, adolescents and adults with cystic fibrosis. The program was recently accredited by the Cystic Fibrosis Foundation. This designation recognizes high-quality, specialized care that leads to improved survival and quality of life.

Approximately 30,000 children and adults in the United States have cystic fibrosis (CF), an autosomal recessive disorder that affects one in about 3,000 Caucasian births. The discovery of the CF gene in 1989, isolation of the CFTR protein (cystic fibrosis transmembrane conductance regulator) and improved understanding of the molecular mechanisms of the disease are being translated into new therapies. Life expectancy for patients with CF has improved dramatically.

### New therapies to maintain lung function

Patient care has changed significantly in recent years largely due to two major avenues of progress:

- The publication of standardized care guidelines by the Cystic Fibrosis Foundation
- The emergence of oral medications that target the underlying cause of the disease and slow the decline of lung function

### Access to state-of-the-art treatments

"We know there are many different elements to excellent care for children with cystic fibrosis," says Marlyn S. Woo, MD, professor of pediatrics and Cystic Fibrosis Program pediatric director. "This is why accreditation will benefit our patients. All accredited CF centers get information on the latest developments and treatments first from the Cystic Fibrosis Foundation. We have access to newer treatments, research medications and new findings."

Having a nurse specialist, respiratory therapist, dietitian and social worker on the team is also beneficial for patients. "CF patients have specialized needs, they need a lot of treatments and have a lot of equipment they need to learn to use. We provide that support" Dr. Woo says.

Following a visit, notes and records are sent within 24 hours to the patient's primary care physician. Dr. Woo says. "Primary care physicians are vital partners in caring for these children."

In 2012, the Food and Drug Administration (FDA) approved ivacaftor, a cystic fibrosis transmembrane conductance regulator potentiator. The drug works by increasing the transfer of chloride into cells and is approved for patients ages 6 and older with the G551D mutation and nine other mutations associated with abnormal chloride-channel function. The FDA is currently reviewing a new drug application for ivacaftor for children ages 2 to 5 and the use of ivacaftor in combination with the investigational drug lumacaftor in people with two copies of the most common CF mutation, Del508.

## Multidisciplinary care at one location

Even with the availability of new medications, achieving optimal health outcomes relies on comprehensive care. Young children require meticulous medical oversight to maintain lung health, prevent infections and promote weight gain and growth. UCLA's program is designed to meet all patient needs. The multidisciplinary team includes physicians specializing in cystic fibrosis along with a CF nurse specialist, social worker, dietitian and respiratory therapist. In addition, appropriate consultation services and specialists are available in genetics, endocrinology, orthopaedic surgery and intensive care. Liver- and lung-transplant services are also available. Our laboratory specializes in detection of CF-related bacteria and meets stringent Cystic Fibrosis Foundation criteria for sweat chloride collection and testing.

## Specialized adult CF care and transitioning program

Today, more cystic-fibrosis patients reach adulthood. A dedicated adult cystic-fibrosis clinic is provided for these patients featuring oversight by an adult-pulmonology specialist trained in treating adult CF patients. Adult CF patients also benefit from having a full CF care team (CF nurse specialist, dietitian, social worker and respiratory therapist) available to them at every clinic visit.

UCLA's Transition Care Program was established to serve patients with chronic conditions who are transitioning from pediatric to adult programs. The network of resources includes CF Foundation-approved patient education and support programs to assist patients in assuming responsibility for their health as adults.

## Coordination with primary care physicians

All patients, adult and pediatric, see a team of specialists during a single visit at one location. UCLA's physician specialists collaborate with each patient's primary care provider to coordinate care. Patient records are provided to PCPs within 24 hours of a visit to the CF clinic.

## Pioneering research on neurocognitive function

The Cystic Fibrosis Program at Mattel Children's Hospital UCLA is a participant in the CF Foundation National Registry and is involved in cystic fibrosis clinical research through Mattel Children's Hospital UCLA's Children's Discovery and Innovation Institute. UCLA specialists are directing the first study using brain MRI to measure neurocognitive function and changes in the brains of CF patients.

## Participating Medical Team

### Marlyn S. Woo, MD

Pediatric Director, Cystic Fibrosis Program  
Professor of Pediatrics  
Pediatric Pulmonology

### Patricia H. Eshaghian, MD

Adult Director, Cystic Fibrosis Program  
Assistant Professor of Medicine  
Pulmonary and Critical Care Medicine

### Douglas Li, MD

Associate Director, Pediatric Cystic Fibrosis Program  
Assistant Professor of Pediatrics  
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### David Sayah, MD

Associate Director, Adult Cystic Fibrosis Program  
Assistant Professor of Medicine  
Pulmonary and Critical Care Medicine

### Elaine Harrington, RN, BSN, AE-C

Cystic Fibrosis Program Coordinator  
Pulmonary Nurse Specialist  
Pediatric Pulmonology

### Suzanne Hollander, MS, RDN

Registered Dietitian Nutritionist

### Sabah Akmal, LCSW

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