UCLA Health

Program focuses on unique needs of adults with cystic fibrosis

About half of all cystic fibrosis patients in the United States today are adults, according to the Cystic Fibrosis Patient Registry. This remarkable statistic represents significant progress in the care of patients with the autosomal recessive disorder. More effective therapies in childhood and the emergence of detailed treatment guidelines have extended the median predicted survival age to 40 years.

The growing number of adults with cystic fibrosis has created a need for specialized services unique to this population. UCLA has an adult CF clinic focused on patients 18 years and older. The clinic is part of UCLA's Cystic Fibrosis Program, which is accredited by the Cystic Fibrosis Foundation.

New oral medications

Progress in the care of cystic fibrosis patients is attributed to following best practices as well as the introduction of new oral medications that target the underlying cause of the disease and slow the decline of lung function. In 2012, the Food and Drug Administration approved Kalydeco® (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 years and older with the

Convenient care for adult CF patients on the Westside

The adult cystic fibrosis program at UCLA offers convenient and coordinated care to better serve patients on the Westside of Los Angeles, says Patricia H. Eshaghian, MD, adult director, Cystic Fibrosis Program, and assistant professor of medicine in Pulmonary and Critical Care Medicine.

"Because there are so many guidelines on how you manage cystic fibrosis, it's important that patients receive care at a specialized center," she says. "We have access to all the specialists a patient might need."

Careful attention is devoted to the seamless transition of pediatric CF patients to adult care.

"What used to be a pediatric disease has become a disease of adulthood," she says. "As these patients enter adulthood, they have to take more responsibility in caring for themselves. Education about the disease becomes very important. It can be hard to adhere to treatments. Patients need a medical team that understands and helps them with compliance."

G551D mutation and nine other mutations associated with abnormal chloride-channel function. The FDA recently approved the drug Orkambi® (lumacaftor and ivacaftor) in patients 12 years and older who have the F508del mutation, the most common mutation in CF.

The special needs of adults with CF

Cystic fibrosis typically progresses with age due to complications associated with multi-resistant bacteria, the onset of diabetes and poor nutrition. At UCLA's adult clinic, patients receive care from a multidisciplinary team of specialists with experience in this phase of the disease. Mental health and psychosocial support become increasingly important to help these patients simultaneously manage their disease and adult responsibilities, such as educational and job pursuits and family relationships. Patients considering pregnancy are referred to reproductive-health specialists. At UCLA, the adult CF team includes adult pulmonary and critical care medicine specialists, a CF nurse specialist, dietitian, social worker and respiratory therapist. Patients are referred to other adult specialists, such as endocrinologists, gastroenterologists and infectious-disease experts, as needed.

Care is coordinated so patients can see a number of specialists in one visit to the clinic. CF patients who become candidates for lung transplantation are referred to UCLA's outstanding lung-transplantation program, one of the largest in the region.

Transition from pediatric care

A major focus in the care of CF patients today centers on smoothing the transition from pediatric to adult care. The transition period, occurring in late adolescence or early adulthood, can be a particularly vulnerable time. Gaps in care or problems in adherence to therapies can result in setbacks to a patient's health. Nationwide, as many as 60 percent of patients experience gaps in medical care, health insurance coverage or both as they move from adolescence to adulthood.

Pediatric pulmonologists at Mattel Children's Hospital UCLA work with their adult pulmonology counterparts at Ronald Reagan UCLA Medical Center and UCLA Medical Center, Santa Monica to facilitate a seamless transition of care. The network of resources includes CF Foundation-approved patient education and support programs such as the CF-RISE program to assist patients in assuming responsibility for their health. Extensive additional educational opportunities are provided to UCLA patients beginning in adolescence. In addition, patients at UCLA are further assisted with insurance coverage issues, a significant factor in maintaining stable health into adulthood.

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