Cystic Fibrosis, An Issue of Pediatric Clinics of North America, 1e (The Clinics: Internal Medicine)

Susan G. Marshall MD, Drucy Borowitz MD

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Cystic Fibrosis (CF) is a multisystem disease whose symptoms and signs involve the gastrointestinal tract (thus affecting nutritional status), endocrine system, reproductive system and the respiratory tract (nose, sinuses and lungs). Despite new treatments, the median survival for patients with CF is less than optimal, primarily due to complications of obstructive lung disease. Currently there are approximately 60,000-80,000 people worldwide with CF. The clinical manifestations of CF are caused by dysfunction of CFTR (cystic fibrosis transmembrane conductance regulator), a multifunctional cyclic-AMP regulated ion channel protein. Over time, there has been dramatic improvement in CF patient life expectancy, in large part related to earlier diagnosis (newborn screening), better understanding of molecular genetics and underlying pathophysiology, the integrated and highly specialized Cystic Fibrosis Foundation Accredited Care Centers, and development of a wide range of new treatments and therapies, some of which target the basic CFTR defect. This edition of Pediatric Clinics of North America will offer general pediatricians and family physicians, as well as subspecialists, an update of the extraordinary progress made in the understanding and treatment of Cystic Fibrosis.

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