



Care of the Adult Patient With Cystic Fibrosis

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Recently published consensus statements and conference proceedings from the Cystic Fibrosis Foundation provide new direction with regard to the etiology, diagnosis, and treatment of the adult patient with cystic fibrosis (CF). Survival has improved through early diagnosis and improved treatment, and aggressive management of complications. The current state of knowledge, the most recent evidence-based treatment options, and essentials of nursing care for the adult patient with CF are outlined.

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Cystic fibrosis (CF) is an inherited, autosomal-recessive disease that disrupts ion transport in epithelial-lined organs. The CF gene was first discovered in 1989 and mapped to a single locus on the long arm of chromosome seven. The protein product of the CF gene has been identified as the cystic fibrosis transmembrane conductance regulator (CFTR). CFTR is involved in the transport of chloride and sodium ions across the membranes of cells in the epithelial lining of the ducts of exocrine glands (sweat glands, pancreas, liver, and reproductive systems). When the CFTR malfunctions in the patient with CF, the membrane is unable to transport electrolytes normally, leading to impaired water secretion, salty sweat, and abnormally thick exocrine secretions (Orenstein, Rosenstein, & Stern, 2000). More than 1,000 mutations have been identified on the CFTR gene, creating the innumerable variations in the clinical progression seen in this disease. Probably the most common mutation cited in research is Δ F508, which accounts for 66% of the CF mutated alleles worldwide (Yankaskas & Knowles, 1999). This classic CF clinical phenotype

is associated with severe pancreatic insufficiency, obstructive azoospermia, and a variety of pulmonary manifestations (Davis, Drumm, & Konstan, 1996).

Once considered to be only a disease of childhood, CF is now also a disease of adults. Before 1950, the gastrointestinal, nutritional, and pulmonary complications of CF resulted in a median survival of 1 year or less (Yankaskas & Knowles, 1999). Due to a number of basic scientific discoveries and improvements in clinical care, the median survival of CF patients in 1999 rose to 29.1 years (Cystic Fibrosis Foundation Patient Registry, 1999). Adults over 18 years of age now account for more than 37% of patients with CF, and survival past the age of 76 years has been documented (Yankaskas, 2001). CF is one of the most common genetic diseases in Caucasians, with an incidence ranging from 1 in 1,900 to 1 in 3,700 in the United States (Hamosh et al., 1998; Welsh, Ramsey, Accurso, & Cutting, 2001). CF is less frequently found in Hispanic, Asian, and African-American populations (Gibson, Burns, & Ramsey, 2003).

CF is a multi-system genetic disease with a highly variable presentation and course that require comprehensive, multidisciplinary care. Some individuals with CF

