Editorials

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Role of the Family Physician in the Management of Cystic Fibrosis

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Individuals with cystic fibrosis (CF) are living healthier, longer lives, and as a result, family physicians are more likely to encounter adult patients with CF in their practices. In the 1970 Cystic Fibrosis Foundation annual registry, 700 adult patients were documented as having CF, which was only 10% of the total CF population.[1] By comparison, the 2012 registry documented 13,651 adult patients with CF, comprising 49.1% of the total CF population of 27,804.[2] The median survival age is now older than 40 years with a birth prevalence of one in 2,500 to 3,500.[2,3] A better understanding of CF physiology, advances in medical therapy, and newborn screening programs nationwide all contribute to longer life expectancies for patients with CF.[3] As treatment options continue to improve, family physicians will increasingly be in a position to make valuable contributions in the care of these patients in conjunction with the specialized pulmonary care provided by CF centers.

The Cystic Fibrosis Foundation recommends that all patients with CF be evaluated quarterly in an accredited care center.[2] During these visits, a multidisciplinary team that includes physicians, nurses, social workers, dietitians, respiratory therapists, physical therapists, and psychologists evaluates each patient. The core objectives of the team are to ensure optimal care, facilitate access to pertinent medical resources, coordinate care among subspecialists and family physicians, and support quality of life and independence.[1,2] Although some centers function in a primary care role, most focus on CF and defer other management to the family physicians. As part of the health care team, the family physician is ideally positioned to have a significant impact on patients with CF by assisting them in four main areas: maintaining an adequate body mass index (BMI); screening for and managing CF-related diabetes mellitus; promoting general health, including exercise; and encouraging compliance with medications and therapies.[1,2,4]

Decades of registry data demonstrate a positive correlation between BMI and forced expiratory volume in one second (FEV_1). Maintaining a BMI of 22 kg per m^2 for females and 23 kg per m^2 for males puts patients in a position to maintain a maximal FEV_1.[2] Dropping below these benchmarks, patients experience a linear decline in FEV_1 as BMI declines. Nutritional supplements and pancreatic enzymes are often necessary to provide adequate quality and quantity of caloric intake to sustain BMI.[1,2]

As pancreatic sufficiency decreases over time, a significant percentage of adult patients will develop CF-related diabetes. Adequate blood glucose control, even before onset of full CF-related diabetes, is demonstrated to prevent and improve FEV_1 decline.[1] Vigilant monitoring for blood glucose elevation is essential, and often early low-dose basal insulin is enough to gain adequate control of blood glucose levels to maximize FEV_1. The growth hormone effect of insulin may also lead to weight gain and help maintain ideal BMI. Screening for diabetes is recommended annually for all adults with CF,[1,2] and the family physician can be instrumental in early detection and management of CF-related diabetes.

As patients with CF live longer, they share the same health risks as the general population.[1,2] Prevention of disease through risk factor modification is just as important in patients with CF. Standard cancer screening practices such as mammography, colonoscopy, and Papanicolaou smears are the same as for the general population.[1] Routine immunizations are advised, with special emphasis on pneumococcal and annual influenza vaccinations.[2]

Because CF affects multiple body systems,[1-6] the treatment burden can be significant.[1,2] Patients are often taking many medications and spending long hours every ▶

day administering aerosolized treatments, receiving chest physical therapy, and exercising. Of note, exercise is known to provide airway clearance of a quality equal to other therapies, attenuates FEV_1 decline, is recommended for all whose physical condition does not otherwise prevent it, and should be considered a standard treatment for all patients with CF.[4]

If CF-related diabetes is present, there is the additional burden of intensive blood glucose monitoring and insulin therapy. Sometimes patients can feel like they spend most of their time administering medications and treatments, and may become discouraged and noncompliant. The continuity and closeness that a family physician has with these patients has the potential to be a stabilizing and encouraging force in assisting with compliance and disease prevention, enabling patients with CF to maximize their quality and quantity of life.

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