Exercise's Effects on Nutritional Status in Cystic Fibrosis

Subjects: Rehabilitation

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Physical exercise is an important part of regular care for people with cystic fibrosis (CF). Exercise training can produce positive physiologic changes in children with CF without impairing their nutritional status. In fact, resistance exercise can help improve body mass.

Keywords: cystic fibrosis ; exercise ; nutritional status

1. Introduction

Cystic fibrosis (CF) is a relatively rare genetic disease affecting over 30,000 people in the United States and more than 70,000 people worldwide [1], with a prevalence varying from country to country but being as high as 1 in 900 in parts of Canada to as low as 1 in 25,000 in Finland [2]. CF is caused by a mutation in the gene responsible for the cystic fibrosis transmembrane conductance regulator (CFTR). This protein is expressed in epithelial cells and serves to directly transport chloride and indirectly affects sodium and water transport. CFTR dysfunction leads to sticky mucus, causing mucus obstruction in various organs including the lungs, pancreas, liver, and intestines. Therefore, cystic fibrosis is a multisystem disease, leading to a decreased life expectancy and significantly impaired quality of life.

CF care requires a multidisciplinary team. It not only focuses on preserving pulmonary function, but also the organ-specific and systemic manifestations of the disease as mentioned above. Malnutrition is a common problem among CF patients, and it is a consequence of multiple factors. Poor bicarbonate secretion from the pancreas, mucosal abnormalities leading to poor intestinal wall function, and poor gut transit time are all thought to contribute to decreased fat absorption [3]. Patients with more pulmonary disease manifestations have a higher concentration of circulating inflammatory markers, which has been linked to decreased fat-free mass (FFM) and bone mineral density (BMD) [4]. People with CF also have an increased resting expenditure rate at baseline [5].

Given that people with CF struggle with malnutrition, their nutritional status, assessed via anthropometric measures, most commonly body mass index (BMI, for adults) or BMI percentile (for children), is also a primary focus of CF care. BMI has been identified as an independent predictor of mortality in cystic fibrosis, with one study demonstrating a hazard ratio of 5.5 (CI 1.8–16.8) for adolescents 12 to 14 years old with a BMI of 15.8 or less [6]. BMI also has implications for morbidity in patients with CF; a cross-sectional study demonstrated decreased FEV1 in patients whose weight was less than 90% predicted [7]. Current CF guidelines recommend BMI goals for individuals with CF; children aged 2–20 are recommended to maintain a BMI ≥ 50th percentile, while adult women are recommended to maintain a BMI of 22–25 and adult men a BMI of 23–25 [8].

The morbidity and mortality of people with CF can also be predicted by their exercise capacity. It has been demonstrated that peak VO2 from cardiopulmonary exercise testing (CPET) can also serve as a predictor of mortality. Both Nixon et al. (1992) [9] and, more recently (2019), Hebestreit et al. [10] found a stepwise increase in survival for people with CF based on increased quantiles of percent predicted peak VO2. Another study examined the longitudinal relationship between habitual physical activity and FEV1, finding that those who were more physically active had a slower decline in FEV1 [11].

Thus, there appears to be a potential conflict between nutritional status and exercise. Patients have an increased resting energy expenditure [5], and exercise would further increase total energy expenditure, perhaps worsening their nutritional status by causing additional weight loss. However, since patients with CF can improve their aerobic capacity through exercise, it remains unclear how exercise may affect their nutritional status/body composition. Most people think of exercise as a way to maintain or lose weight; thus, some people with CF that are underweight or at their goal weight may be reluctant to begin an exercise program. On the contrary, with the advent of highly effective modulator therapies, some patients are now concerned about gaining too much weight [12][13].

2. Exercise's Effects on Nutritional Status in Cystic Fibrosis

Exercise, in the short term, in spite of a population that was mostly normal to underweight, does not negatively affect body composition in CF patients. In fact, Selvadurai, whose participants were the most malnourished (mean weight for age 16%) demonstrated that RET can improve body mass, body composition and muscle strength [14]; they were also able to demonstrate that AET led to larger increases in aerobic capacity and a slight, but statistically insignificant, increase in body mass compared to the control group; they ultimately suggested that a combined training program may be of most benefit to patients with CF. In initial study (2012) [15], Santana Sosa did not notice any significant difference in BMI or FFM with a combination of AET and RET, but in the later study (2014) [16], they did find a significant increase in FFM in the exercise training group.

More recently (2021), Van Biervliet reported on a prospective pre–post intervention study design for patients with CF (6 to 40 years old) to improve nutritional status and body composition; patients participated in a short-term (3 weeks), inpatient, physical exercise and nutritional intervention program [17]. Weight, BMI, and fat-free mass were improved in both children and adults; in addition, the number of adults classified as “malnourished” decreased from 41% to 24%, but was unchanged (24%) in children.

In fact, RET could help maintain or increase body mass and potentially lean body mass. Clinicians should counsel patients that are concerned about the speculative effects of exercise on their nutritional status and body composition that exercise is not detrimental and may even improve their nutritional status. The CF care team should continue to rely on the CF care team’s registered dietitian to provide appropriate individualized nutrition care plans that compliment exercise regimens to help patients meet their personal goals related to weight and body composition (e.g., the team reported by Van Biervliet included a physician, dietician, psychologist, social worker and physical therapist [17]). In addition, both aerobic exercise (AET) and resistance exercise training (RET) have additional benefits for patients with CF (increased aerobic capacity and strength), benefits which are associated with a positive prognosis.

Hommerding demonstrated an increase in physical activity level in patients that had frequent follow-up for their exercise regimen [18]. For those working in multidisciplinary settings, referral to a physical therapist or an exercise specialist with experience with CF that can guide exercise regimens over time and as their health waxes and wanes would be of more benefit. There are standard guidelines on exercise testing [19], exercise prescription [20], and physical activity assessment [21] for clinicians working with individuals with CF.

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