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A comparison of respiratory and peripheral muscle strength, functional exercise capacity, activities of daily living and physical fitness in patients with cystic fibrosis and healthy subjects

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ABSTRACT

There are limited reports that compare muscle strength, functional exercise capacity, activities of daily living (ADL) and parameters of physical fitness of cystic fibrosis (CF) patients with healthy peers in the literature. The purpose of this study was to assess and compare respiratory and peripheral muscle strength, functional exercise capacity, ADL and physical fitness in patients with CF and healthy subjects. Nineteen patients with CF (mean forced expiratory volume in one second-FEV_1: 86.56 ± 18.36%) and 20 healthy subjects were included in this study. Respiratory (maximal inspiratory pressure-MIP and maximal expiratory pressure-MEP) and peripheral muscle strength (quadriceps, shoulder abductors and hand grip strength) were evaluated. Functional exercise capacity was determined with 6 min walk test (6MWT). ADL was assessed with Glittre ADL test and physical fitness was assessed with Munich fitness test (MFT). There were not any statistically significant difference in MIP, %MIP, MEP and %MEP values between two groups ($p > 0.05$). %Peripheral muscle strength (% quadriceps and shoulder abductors strength), 6MWT distance and %6MWT distance were significantly lower in patients with CF than those of healthy subjects ($p < 0.05$). Glittre ADL-test time was significantly longer in patients with CF than healthy subjects ($p < 0.05$). According to Munich fitness test, the number of bouncing a ball, hanging score, distance of standing vertical jumping and standing vertical jumping score were significantly lower in patients with CF than those of healthy subjects ($p < 0.05$). Peripheral muscle strength, functional exercise capacity, ADL performance and speed, coordination, endurance and power components of physical fitness are adversely affected in mild-severe patients with CF compared to healthy peers. Evaluations must be done in comprehensive manner in patients with CF with all stages.

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1. Introduction

Cystic fibrosis (CF) is an important autosomal recessive genetic disease in terms of morbidity and mortality often seen in the white race (Mall & Boucher, 2014). CF is caused by a defect in the CF transmembrane conductance regulator (CFTR) gene, on the long arm of chromosome 7 and it leads to absence of normal CFTR protein, a cyclic adenosine monophosphate (cAMP)-activated ion channel (Davies, Ebdon, & Orchard, 2014). With increasing age, patients expectorate mucus that is highly viscous and hyperconcentrated, exhibit reduced mucociliary clearance, and develop intermittent to chronic polymicrobial infection with bacterial species. Chronic infection is accompanied by persistent neutrophilic inflammation leading to progressive irreversible lung damage (Kerem, Conway, Elborn, & Heijerman, Consensus Committee, 2005; Mall & Boucher, 2014).

It was shown that respiratory and peripheral muscle strength of patients with CF were significantly lower than healthy peers (Dassios, Katelari, Doudounakis, Mantagos, & Dimitriou, 2013; Lands, Desmond, Demizio, Pavilanis, & Coates, 1990; Pinet et al., 2003). Many factors as nutritional status (Arora & Rochester, 1982), hypoxia (Coates, Boyce, Muller, Mearns, & Godfrey, 1979), inactivity (DuBois & Almon, 1980) and corticosteroid use (Barry & Gallagher, 2003) affect skeletal muscle in CF patients. Therefore, fat-free mass, quadriceps muscle strength and quadriceps cross-sectional area is reduced and peripheral muscle strength is decreased in CF (Barry & Gallagher, 2003; Elkin, Williams, Moore, Hodson, & Rutherford, 2000; Hussey, Gormley, Leen, & Greally, 2002; Pinet et al., 2003). Respiratory muscle function is affected by chronic obstructive pulmonary disease, chronic pulmonary inflammation, genetic predisposition, corticosteroid therapy and chronic nutritional deficiency in patients with CF (Arora & Rochester, 1982; Dassios, 2014; Marks, Pasterkamp, Tal, & Leahy, 1986).

Some people with CF have reduced aerobic and/or anaerobic fitness compared with their healthy counterparts (Cabrera, Lough, Doershuk, & DeRivera, 1993; de Meer, Gulmans, & van Der Laag, 1999). Several factors such as limitation of ventilation and gas exchange, pneumothorax and haemoptysis, fluid and electrolyte losses, weight loss and hypoglycaemia may limit exercise capacity in patients with CF (Almajed & Lands, 2012; Coates et al., 1979; Hebestreit, 2014; Pastre et al., 2014; Selvadurai et al., 2002; Troosters et al., 2009).

Variable health status, ilness or infection, poor nutritional status, perception of barriers to participation, overprotective child syndrome and burden of disease may prevent patients with CF from participating to activities of daily living (ADL) regularly (Schneiderman Walker et al., 2005; Selvadurai et al., 2002; Wilkes et al., 2009). Most patients felt their lives were marked by a loss of freedom and opportunities because of their poor health and time-consuming treatments. Short stature, weakness, fatigue, and susceptibility to infections contributed to participants' frustration, social isolation, and feelings of being different (Jamieson et al., 2014; Wilkes et al., 2009). It was shown that CF children spent less time in moderate to vigorous activity compared to healthy peers (Aznar et al., 2014; Nixon, Orenstein, & Kelsey, 2001).

In children, who typically have very different activity profiles than adults, the concept of fitness has been widened to include strength, endurance, flexibility and coordination, amongst other factors (Almajed & Lands, 2012). There are limited studies in the literature that compares muscle strength, functional exercise capacity, ADL and parameters of physical fitness of CF patients with healthy peers (Dassios et al., 2013; de Meer et al., 1999; Gruber, Orenstein, Braumann, & Hüls, 2008; Johnson, Ferkol, & Shepherd, 2006). Therefore, the primary aim of this study was to compare respiratory and peripheral muscle strength, functional exercise capacity, ADL and physical fitness in patients with CF and healthy subjects. The secondary objective was to explore the practical utility of standardized Glittre-ADL test for evaluating ADL and show discriminative properties of this ADL test method for patients with CF.

2. Method

2.1. Participants

Nineteen patients with CF between the ages of 7-25 who don't have other systemic or acute illness, diagnosed by Hacettepe University, Faculty of Medicine, Department of Child Health and Diseases, Unit of Chest Diseases according to CF-compliant clinical manifestations and sweat chloride is above 60 mequiv./L in both measurements were enrolled in the study. The patients with a physical disability or who suffer from acute exacerbation and can not adapt to evaluations were not included in the study. Healthy group consisted of volunteer children who were received permission from their families and participants without any systemic disease and physical disability. The study was approved by the Ethical Committee of Hacettepe University and subjects and their families signed an informed consent form.

2.2. Assessments

The assesments were done by two physiotherapist which specialist in cardiopulmonary rehabilitation program. The physical and sociodemographic characteristics of all subjects were recorded. Body mass index (BMI) was calculated from the formula (body weight/height^2, kg/m^2). Subjects were classified as thin ($<$18.5 kg/m^2), normal (18.5-24.9 kg/m^2), overweight (25-29.9 kg/m^2) and obese (30-39.9 kg/m^2; World Health Organization, 1995).