Paediatric Respiratory Reviews 10 (2009) 105–109

Contents lists available at ScienceDirect

Paediatric Respiratory Reviews

Mini-symposium: The Clinical Applications of Exercise Testing in Children

Exercise and physical activity in children with cystic fibrosis

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ARTICLE INFO

Keywords:
exercise
habitual physical activity
training
paediatric
cystic fibrosis

SUMMARY

Regular exercise and habitual physical activity are important for patients with cystic fibrosis (CF). Research has demonstrated the benefits of aerobic, anaerobic, and strength exercise training programs for health and quality of life, however, the CF patient is faced with unique barriers and challenges to participation. Recently, increased levels of habitual physical activity have been shown to slow the decline in lung function in patients with CF, and regular participation in a variety of activities may result in greater adherence in the long term. Research is now available to justify the incorporation of exercise into the routine care of patients with CF. This paper provides the background and rationale for the implementation of exercise and habitual physical activity recommendations by the health care team. Education of health care providers regarding the importance of exercise and habitual physical activity for patients with CF is needed in order for exercise and physical activity to be incorporated as key components of clinical practice and into the lives of patients with CF.

INTRODUCTION

Despite advances in clinical care, life expectancy in patients with cystic fibrosis (CF) remains shortened.[1] It has been well documented that exercise capacity in this group is limited by lung function, peripheral skeletal muscle function[2], nutritional status[3–5] and the cardio-respiratory system's ability to meet the metabolic demands associated with exercise.[4] Interestingly, the physiological consequences that have been observed in patients with CF are similar to the effects of de-conditioning including poor cardiovascular function, reduced muscle mass and impaired strength and power.[6] Further, children with CF may be more physically inactive[7] due to the burden of their chronic disease[8] and therefore may be at risk of compounding the combined effects of chronic disease and physical inactivity.

The importance of exercise and habitual physical activity (HPA) for patients with chronic disease has been identified as an area of importance for health care providers, clinicians, researchers and most importantly, patients.[6] Higher levels of exercise capacity have been reported to be important for survival in patients with CF. Several studies have demonstrated an association between aerobic capacity and clinical status in pediatric patients with CF. Nixon and co-workers reported a significant correlation between aerobic fitness (VO_2) and survival[9], which remained intact after adjustment for other predictor variables such as age, sex, lung function, nutritional status and bacterial colonization. The authors suggested that VO_{2peak} could be used as an independent predictor of survival. This finding has been recently confirmed by Pianosi and colleagues[10] who studied 28 patients 8-17 years over five years to determine the effect of declining lung function and VO_2 as predictors of survival over the subsequent 7–8 years. They found that patients with a VO_{2peak} of greater than 45 ml $kg^{-1}min^{-1}$ had an improved survival compared to those with a VO_{2peak} less than 32 ml $kg^{-1}min^{-1}$. Klijn and co-workers[4] also reported a longitudinal relationship between changes in nutritional status, lung function, and VO_{2peak} in children with CF. Fat-free mass (FFM), lung function, and VO_{2peak} were assessed in 65 children with CF with a wide range of FEV_1 at baseline and two years later. They concluded that longitudinal changes in lung function, and to a lesser extent nutritional status, were positively associated with functional changes in the aerobic capacity of children with CF.

Whereas 'exercise' infers structured activity, typically prescribed according to intensity and duration, HPA emphasizes activity which is incorporated into daily life, and encompasses a broader range of options for being active. Activity has been shown to specifically contribute to the health of patients with cystic

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doi:10.1016/j.prrv.2009.04.001

fibrosis.[11–13] Therefore the importance of exercise and HPA in patients with cystic fibrosis is critical and presents an opportunity for members of the health care team to implement regular activity recommendations as a routine part of clinical practice. This review summarizes a growing body of research on exercise and habitual physical activity in CF which provides a rationale and justification for their inclusion into routine clinical therapy for paediatric patients and for identification of areas for future research.

CHALLENGES OF EXERCISE AND ACTIVITY PARTICIPATION FOR PATIENTS WITH CYSTIC FIBROSIS

Despite the positive benefits of exercise and habitual physical activity in patients with chronic disease[6], children with CF are not as active as their healthy peers.[7] The specific challenges associated with single exercise sessions and participation in HPA must be considered by both clinicians and researchers if successful recommendations are to be made and evaluated in this population. Research documenting these challenges in CF is summarized in the following sections.

Physiological limitations may negatively affect the exercise capability of patients with CF to perform single bouts of exercise. For example, it has been proposed that there may be inefficient aerobic oxidative metabolism in CF patients due to impaired oxygen delivery[4,14] or intrinsic abnormalities in muscle function.[15] Impaired anaerobic performance[16], abnormal anaerobic metabolism[17], and decreased maximal muscle strength and power have also been reported.[18–20] Taken together, these studies provide strong evidence for the pathophysiological impact of CF on muscle function and exercise capacity.

Significant barriers to regular participation in habitual physical activity exist for patients with CF. These challenges may include, but are not limited to, variable health status and illness or infection, poor nutritional status, the perception of barriers to participation, the vulnerable child syndrome[21] and the burden of disease.[22]

Children with CF have reported that their health problems interfered with physical activity more frequently than healthy controls.[23] Variable health status in addition to acute illness or infection may require aggressive treatment and/or hospitalization which creates major barriers to physical activity for children with CF. In addition to a daily routine of therapy and regular visits to CF clinics, many patients are hospitalized at least once per year for a period of time that interferes with regular activity and exercise routines. The time requirements of these intensive treatment regimes, as well as their physical and emotional demands, present barriers to physical activity participation for the child with CF.

Poor nutritional status may limit exercise capacity in children with CF which, in turn, may diminish activity levels. Boucher and colleagues demonstrated that activity level was related to nutritional status but not lung function in patients with significant air flow limitation.[8] Similarly, nutritional status has been identified as a major determinant of anaerobic exercise capacity in CF patients.[16] Therefore, poor general health and/or nutritional imbalance may lead to a reduction in activity, whether it is aerobic, anaerobic or a combination of the two.

Parents of children with CF may see their child as vulnerable or in 'at risk' circumstances. This 'vulnerable child' syndrome may limit the parents' perception about their child's physical abilities.[21] Boas and co-workers[21] demonstrated that parents of children with CF perceive fewer benefits of exercise and greater barriers to activity than the parents of healthy children and was more prevalent in parents of girls than boys. In addition, they found that less than half of the parents of children with CF knew that exercise performance was related to long term prognosis or that exercise could be beneficial for even the most severely affected child with CF.

The burden of disease may make compliance with recommended exercise activities more difficult.[22] In healthy children, perceptions of competency, enjoyment of activity, availability of a variety of activities, and social support are factors related to positive exercise compliance. In patients with CF, complications such as fatigue and time required for treatment(s) and co-morbidities associated with the disease such as Cystic Fibrosis Related Diabetes, may make compliance with exercise programs more challenging.

In summary, children with CF face significant barriers to regular participation in exercise, physical activity and training programs. These limitations may be primarily related to the CF disease itself, and secondarily to psycho-social factors related to chronic disease in general. These factors should be considered by health care providers to ensure that exercise and activity recommendations are delivered to patients and their families with the greatest chances for successful implementation and adherence.

Evidence of benefit for exercise training and habitual physical activity in patients with cystic fibrosis

Significant benefits of exercise and habitual activity have been documented for children with cystic fibrosis.[24,25] The effects of exercise training have been shown to provide the same positive benefits to children with CF as their healthy peers (see Fig. 1). These benefits include improvements in cardiovascular endurance[26,27], muscular strength[28,29], quality of life[30,31], and the added benefit of mucus clearance[32] which may specifically target the physiological challenges faced by the child with CF (see Fig. 2). For a complete review of the aerobic and anaerobic bioenergetic physiology please refer to "Bioenergetic provision of energy for muscular activity", also published in this symposium. However, the use of exercise as therapy is underutilized[33] and the role of exercise and its contribution to health may be unclear to clinical staff, parents of children with CF and the patients themselves. Clinicians in the CF clinic may encourage children to participate in regular activity to improve cardiovascular, respiratory and muscle function and mucus clearance. However, the specifics of the activity – type, duration, frequency and intensity are often less defined by the CF team, leaving the CF patient and family unclear regarding where to begin and how to continue activity. This section will summarize the benefits of exercise training programs in children with cystic fibrosis.

EVIDENCE OF BENEFIT OF EXERCISE TRAINING

The benefits of aerobic exercise training programs have been reported in the pediatric CF population. Several studies have demonstrated that patients with CF can improve aerobic capacity.[26,27] Recently, evidence for improvements in pulmonary function in relation to physical activity levels have been reported[13], with girls participating in higher levels of activity demonstrating a significantly slower rate of decline in FEV_1 compared to less active girls. Interestingly, comments from authors suggest that despite evidence of benefits to lung function and quality of life[28], adherence to training programs after the conclusion of the study is challenging.[26,13]

The effects of short term endurance and strength training on physical capacity and clinical indicators has also been evaluated.[28,29] In keeping with the principle of specificity of training, these studies demonstrated that children who received aerobic