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Review article

Exercise testing and training with the young cystic fibrosis patient

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Abstract
The purpose of the article is to review the literature related to exercise and Cystic fibrosis (CF), with particular focus on the young CF patient. Exercise intolerance is a characteristic of CF, however, recent studies in adults have advanced our understanding of how exercise can be used effectively as a prognostic marker and for rehabilitation purposes. New analyses from exercise testing have shown to have prognostic value, and different methods of exercise training have been reported to improve the functional capacity and quality of life of the young CF patient. There is a growing awareness and belief among clinicians of the benefits of exercise testing and training, however, recent work suggests that exercise is being underused in the healthcare management of the CF patient. More research is needed to identity which exercise tests and training programmes would be most feasible to incorporate into CF centres routine clinical procedures.

Key words: Cystic fibrosis, young patients, exercise testing, training programmes, physical activity.

Introduction

Cystic fibrosis (CF) is a genetic disease. It is caused by a mutation in a single gene on the long arm of chromosome seven. The gene encodes a protein, the cystic fibrosis transmembrane conductance regulator (CFTR), which functions as a chloride channel. Mutations to the CFTR cause abnormal chloride concentration across the apical membrane of epithelial cells, especially in the airways of the lung and the pancreas. Consequently, patients with CF experience progressive lung disease and malnutrition. The mutated gene responsible for CF was identified in 1989 (Riordan et al., 1989), however, no cure for the disease has been found. In the UK CF affects approximately 7,500 people, and each week five babies are born with the disease. The average life expectancy for a CF patient in the UK is 31 years.

Exercise intolerance is an established characteristic of CF and is dependent on the progression of the disease. When compared to healthy controls, young patients with CF exhibit a reduced muscle size and strength, and a deficiency in aerobic and anaerobic capacity. Research investigating the effects of exercise in CF began in the early 1970s, when Godfrey and Mearns (1971) began studying the physiological responses. During the late 1970s and early 1980s work by Keens et al. (1977) and Zach et al. (1981) showed that exercise training programmes and exercise can be used to improve ventilation and help clear respiratory mucus, respectively. Over the last three decades, however, extensive research into exercise and the young CF patient has been lacking. At present only 118 research articles have been published in this area since the work of Counahan and Mearns (1975), who to the best of the author's knowledge, were the first to specifically research young CF patients by examining the prevalence of exercise-induced bronchial constriction in their relatives.

Compared to the extensive amount of research that exists into exercise and the healthy child, research into exercise and the young CF patient has been neglected. By comparison many of the physiological responses and adaptations to exercise in the young CF patient remain unknown. Finding a clinical use for exercise, whether as a prognostic tool or therapeutic therapy, is an area of renewed interest to clinicians and physiotherapists. Nixon et al. (1992) reported an association between the aerobic capacity of young CF patients and survival over eight years. Recently, studies by Pianosi et al. (2005) and Javadpour et al. (2005) have shown the significance of exercise testing and the relationship between quality of life, life expectancy and functional capacity. Furthermore, Klijn et al. (2004) and Orenstein et al. (2004) have recently reported the benefits of anaerobic and strength training in young patients with CF. These methods of exercise training have been traditionally neglected in exercise prescriptions in favour of aerobic exercises. Such work has helped further the continuing interest among clinicians of the importance of exercise in the management of CF in young patients. Recent work by Barker et al. (2004) has shown that exercise is being incorporated into CF centres routine clinical procedures in Germany.

The purpose of the present review is two-fold. Firstly, the review aims to inform the reader of recent contributions to the exercise and CF literature, with particular attention on the young CF patient. Recent contributions to the literature will be categorised under the following headings; exercise testing and prognosis, exercise training, physical activity, and physiological comparisons between the young CF patient and health. Secondly, the review aims to provide future directions in exercise and CF research, and investigate the current census of opinion regarding the role of exercise and exercise testing in the young CF patient.

Exercise testing and prognosis

In a landmark study, Nixon et al. (1992) determined factors that are associated with an eight year survival rate of 109 young patients with CF. They found a significant correlation between peak oxygen uptake ($\dot{V}O_{2\ peak}$) and survival at eight years. Thus, becoming the first study to report an association between the aerobic capacity of children, adolescents and young adults with CF and sur-

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vival over an eight year period. Nixon and colleagues reported that $\dot{V}O_{2\,peak}$ was a strong predictor of survival, even after adjustment to other prognostic variables such as age, sex, lung function, bacterial colonisation and nutritional status. Patients with a high level of fitness (≥ 82 % predicted $\dot{V}O_{2\,peak}$) had a 83 % chance of survival at eight years, compared to a 51 % and 28 % chance of survival for patients with medium (59-81 % predicted $\dot{V}O_{2\,peak}$) and low fitness (≤ 58 % predicted $\dot{V}O_{2\,peak}$), respectively. These data, however, do not establish a cause and effect relationship. Aerobic capacity may not be an indictor of the patient's overall health. Nevertheless, the study showed that aerobic capacity could be used as a strong predictor of survival in these patients.

A recent study by Pianosi et al. (2005), however, is the first to investigate both the utility of $\dot{V}O_{2\,peak}$ and decline of $\dot{V}O_{2\,peak}$ as a predictor of survival. Twenty-eight children aged 8 to 17 years, over a five-year period performed annual pulmonary function and maximal exercise tests to determine FEV_1 and $\dot{V}O_{2\,peak}$. The magnitude of the change in the young patients' FEV_1 and $\dot{V}O_{2\,peak}$ over time, and survival over the subsequent 7 to 8 years were used to determine their significance as predictors of survival. Pianosi and colleagues reported that the measurement of $\dot{V}O_{2\,peak}$ from maximal exercise tests were significant predictors of patient survival. Furthermore, during the observation period a mean annual decline in $\dot{V}O_{2\,peak}$ of 2.1 $mL \cdot kg^{-1} \cdot min^{-1}$ was reported in 70 % of the patients. A dramatic increase in survival over the subsequent 7 to 8 years was seen in patients whose $\dot{V}O_{2\,peak}$ exceeded 45 $mL \cdot kg^{-1} \cdot min^{-1}$. In contrast, patients with a $\dot{V}O_{2\,peak}$ less than 32 $mL \cdot kg^{-1} \cdot min^{-1}$ showed a survival rate of 60 % over the subsequent 7 to 8 years. The FEV_1 was also reported as a significant predictor of survival. The patients' first and last measurement of FEV_1 over five years and rate of decline in FEV_1 over the time period were all significant predictors of mortality.

Previous studies investigating the prognostic value of exercise testing in young CF patients have placed much emphasis on the assessment of $\dot{V}O_{2\,peak}$ to predict outcomes such as functional capacity, disease severity and survival. Other physiological assessments derived through exercise testing have been neglected. The oxygen uptake ($\dot{V}O_2$) kinetics during exercise and recovery, carbon dioxide (CO_2) retention, oxygen desaturation and muscle strength can all be assessed through exercise testing and may all have prognostic value. Researchers are now focusing attention on other physiological assessments derived from exercise testing that may hold prognostic significance, other than $\dot{V}O_{2\,peak}$ alone. Furthermore, researchers are using more sophisticated techniques to model the $\dot{V}O_2$ kinetic responses from exercise in CF patients.

Hebestreit et al. (2005) has investigated the $\dot{V}O_2$ kinetics at the onset of exercise in patients with CF. Eighteen CF patients and 15 healthy controls aged 10-33 years completed two to four transitions from low-intensity cycling at 20 W to cycling at 1.3-1.4 $W \cdot kg^{-1}$ body weight. $\dot{V}O_2$ data from the submaximal exercise tasks were modelled interpolated second by second, time aligned and averaged. Monoexponential equations were used to describe phase II $\dot{V}O_2$ responses. Hebestreit and colleagues reported no significant differences in the amplitude in the model of the phase II response between the CF patients and healthy controls; however, the time constant tau was significantly prolonged in the CF patients. Thus, demonstrating a slower $\dot{V}O_2$ kinetic response in patients with CF.

Recent work by Pouliou et al. (2001) investigated the $\dot{V}O_2$ kinetics during early recovery from maximal exercise in adult patients with CF. Pouliou and colleagues reported that the $\dot{V}O_2$ kinetics following maximal exercise is prolonged in adult CF patients when compared to healthy adults, and that the prolonged recovery is closely related to the Schwachman score ($r = 0.81$; $P < 0.001$), a widely accepted system of clinical evaluation to assess disease severity. The $\dot{V}O_2$ kinetics following maximal exercise was also significantly correlated to other indexes of functional capacity such as FEV_1 ($r = 0.90$; $P < 0.001$) and $\dot{V}O_{2\,peak}$ ($r = 0.81$; $P < 0.001$). This novel approach to health assessment offers, from a clinical view, an exercise assessment that is independent from effort and physical fitness. Pouliou and colleagues used a linear regression model to evaluate the $\dot{V}O_2$ kinetics for the first minute of the recovery period. However, no rationale was given to suggest why a linear model would fit the response. The linear model used by Pouliou et al. does not identify the slow phase of the recover period and, furthermore, may only identify 40 % of the fast phase. Assessment of the $\dot{V}O_2$ kinetics during recovery can be used even with submaximal exercise, which is important for debilitated CF patients who cannot produce maximal exercise performances. Furthermore, data from maximal exercise testing may not be reproducible, is dependent on patient motivation and the criteria used by the clinician to terminate the test.

Javadpour et al. (2005) recently examined CO_2 retention during exercise testing, and discovered it had an association with a rapid decline in lung function. Children with CF aged 11 to 15 years performed annual pulmonary function and exercise testing over a three-year period. CO_2 retention was defined as a rise of ≥ 5 mm Hg end tidal CO_2 from the first work rate until the peak work rate, and a failure to reduce end CO_2 after the peak work rate by 3 mm Hg by the termination of exercise. Using this definition of CO_2 retention Javadpour and colleagues reported that children with CF who were found to have CO_2 retention on exercise testing showed a faster rate of decline in FEV_1 when compared to those who did not retain CO_2. The decline in FEV_1 between patients who retained CO_2 and patients who showed no CO_2 retention during exercise over a 12 month period was reported as –3.2 % (SD 1.1) and –2.3 % (SD 0.9), respectively. After 24 months the decline was reported as –6.3 % (SD 1.3) and –1.8 % (SD 1.1), respectively. Finally, after 36 months the decline in FEV_1 was –5.3 % (SD 1.2) and –2.6 % (SD 1.1), respectively. Both patients who retained CO_2

during exercise and those who showed no CO_2 retention started the study with similar baseline FEV_1, 62 % and 64 % predicted, respectively. The overall decline in FEV_1 was, however, 14.8 % (SD 2.1) and 6.7 % (SD 1.8), respectively. The study suggests that children with CF with a similar degree of pulmonary disease as measured by FEV_1, if found to have CO_2 retention on exercise testing, will have a greater decline in FEV_1 over a three year period than children with CF who do not retain CO_2. This shows the identification of CO_2 retention during exercise can be an additional prognostic marker of disease progression in children with CF. Furthermore, as FEV_1 is closely associated with survival in this patient group, CO_2 retention during exercise testing will help identify those patients who may require more intensive therapy to prevent this increased rate in pulmonary decline.

Exercise training

Aerobic fitness is associated with prolonged survival and quality of life. Therefore, most studies investigating the potential of exercise programmes have assessed the effects of aerobic training. In one training study, however, Selvadurai et al. (2002) compared the effects of both aerobic and strength training. CF inpatients aged 8 to 16 years were randomly allocated into an aerobic or strength training group, and exercised five times a week for 3 weeks. Selvadurai and colleagues reported that children who received aerobic training had significantly better $\dot{V}O_{2\,peak}$, activity levels and quality of life than children in the strength-training group. Children in the strength-training group when compared to the aerobic training group, however, had better weight gain (total mass, as well as fat-free mass), lung function and leg strength, which all have a high correlation with health status. The findings of this study support the proposition that a combination of aerobic and strength training may be the best training programme for young CF patients. Future studies assessing the potential of exercise programmes should assess the combination of different exercises to optimise training programmes for young CF patients.

Klijn et al. (2004) are the first authors to conduct a high intensity anaerobic training programme for children with CF. The authors suggest that children may be more suited to high intensity anaerobic training, as children's natural activity patterns are characterised by very short vigorous bouts of physical activity, interspersed with varying levels of low to moderate intensities. In a randomised, controlled study 11 children with CF participated in a 12-week anaerobic training programme. The children trained two days per week, with each session lasting 30 to 45 minutes. The training consisted of anaerobic activities lasting 20 to 30 seconds. The control group consisting of 9 young CF patients were asked not to change their normal daily activities. Participants of the exercise programme showed significant improvements in both anaerobic and aerobic performance and quality of life as measured by a disease specific health-related quality of life questionnaire. Children in the control group showed no improvements in any measured parameters. This study also identified the benefits of including different exercises into training programmes designed for children with CF. Furthermore, the inclusion of different types of exercises adds variation to training, which may help to improve adherence.

A later study by Orenstein et al. (2004) also supports the suggestion of incorporating both aerobic and anaerobic exercises into training programmes to optimise the functional capacity of the young CF patient. During a one-year randomised controlled trial, 67 young patients with CF, aged 8 to 18 years, participated in either an aerobic or upper-body strength-training programme. All children were asked to exercise at least three times per week for 12 consecutive months. Each patient in the aerobic training group was given a stair stepping machine and instructed to exercise five minutes per session, gradually increasing to 30 minutes per session. Young patients in the upper-body strength group were given a weight resistance machine, which they used to perform bicep curls, lateral pull-downs and bench presses. The exercises were individually tailored to each patient's strength, and the exercises increased gradually by the number of sets and repetitions as well as by the amount of resistance per bout over the year. Contrary to previous studies, Orenstein and colleagues reported that aerobic training did not, however, produce greater fitness or greater pulmonary function than strength training. Furthermore, both types of training increased upper-body strength and physical work capacity.

Physical activity

Maintaining high levels of physical activity is important in the management of CF. Clinicians encourage young patients with CF to take regular exercise to help mobilise mucous secretions and enhance or preserve breathing capacity. Exercise tolerance, however, in CF is limited and inactivity itself results in deconditioning. Thus, a negative feedback loop is formed, making physical activity even more difficult. A recent study by Selvadurai et al. (2004) measured the habitual activity levels in boys and girls with different degrees of CF lung disease and healthy controls. Each child completed an activity diary for two weeks and wore a validated accelerometer on the same days the activity diary was completed. What made this study unique from previous studies, however, was the authors sought to determine correlations between the young CF patients' physical activity and functional capacity, and quality of wellbeing. Selvadurai and colleagues reported significant correlations between activity levels and aerobic capacity ($r = 0.55$; $P < 0.01$), anaerobic power ($r = 0.63$; $P < 0.01$) and quality of wellbeing ($r = 0.61$; $P < 0.01$). In children with moderate to severe lung disease, body mass percentile correlated well with activity levels, implicating possible effects of poor nutrition on muscle mass and functional ability.

Selvadurai and colleagues further reported that there were no significant differences in habitual activity between prepubescent children with CF and controls, and regardless of disease severity there were no differences in habitual activity between prepubescent boys and girls with CF. After puberty, however, differences in habitual activity become evident. Pubescent boys with CF were

significantly more active than girls with the same degree of disease severity, and pubescent children with moderate to severe CF were less active than controls. Only pubescent children with mild CF were significantly more active than controls. This study highlights the importance of maintaining physical activity levels after puberty, when it is suggested that children with CF start becoming less active, particularly in girls.

The importance of maintaining physical activity in girls with CF is also emphasised in a recent study by Schneiderman-Walker et al. (2005). Schneiderman-Walker and colleagues reported that the annual rate of decline in FEV_1 was related to habitual physical activity in girls. Children with CF were divided into quartiles depending on activity levels derived from a Habitual Activity Estimation Scale; their FEV_1 was measured over a two-year period. In the lowest two activity quartiles girls had a more rapid rate of decline in FEV_1 (–3.40 and –3.05 % predicted, respectively), than girls in the two highest activity quartiles (-0.93 and +1.17 % predicted, respectively). In boys, however, the rate of decline of FEV_1 was similar in all activity quartiles (-0.93 % predicted). Preserving FEV_1 is central to the management of CF as it has a strong relationship with survival. An inactive lifestyle may partially explain the poorer survival of female patients with CF.

Physiological comparisons between the young CF patient and healthy children

CF is also characterised by diminished nutritional status caused by diseased gastrointestinal tracts and pancreatic insufficiency. This can lead to malnutrition affecting the growth and maturation of skeletal muscle through childhood and adolescence. Recent studies have investigated the effect of reduced muscle size on exercise performance, to determine if intrinsic pathophysiological defects exist in young CF muscle. In a study by de Meer et al. (1999) it was reported that children with mild to moderate CF had significantly weaker peripheral muscle force than healthy controls. Peripheral muscle weakness was associated with diminished work capacity and an increased oxygen cost of exercise. A disproportionate decrease in maximum cycle workload was observed in children with CF when compared to healthy controls, even in patients with normal pulmonary function and nutritional status. From these findings de Meer and colleagues implicated that impaired exercise performance in the young CF patient is at least partly due to a pathophysiological factor in skeletal muscle that cannot be readily attributed to nutritional factors. As a consequence, intrinsic abnormalities may exist in the muscle of young CF patients, which contribute to a reduced exercise performance.

Moser et al. (2000) hypothesised that exercise impairment in children with CF is primarily due to a reduction in muscle mass rather than abnormalities in muscle metabolism. Muscle size was estimated from midthigh muscle cross-sectional area (MTCSA) by magnetic resonance imaging, and exercise performance was determined by progressive cycle ergometer testing. Contrary to the hypothesis, the results showed that a reduction in $\dot{V}O_{2\,peak}$ was observed even when normalised to MTCSA, whereas MTCSA was only slightly smaller in children with CF than in control subjects. Therefore, a reduced muscle mass alone could not account for all of the impairment of $\dot{V}O_{2\,peak}$ observed in the young CF patients. In agreement with de Meer and colleagues, impaired oxygen delivery or intrinsic abnormalities of muscle function are also responsible for reduced exercise performance in children with CF. More studies are needed to identify specific intrinsic abnormalities of skeletal muscle in CF.

Future directions

There is a growing awareness among clinicians of the importance of exercise testing and prescription. Recent research by Pianosi et al. (2005) has reinforced the prognostic value of $\dot{V}O_{2\,peak}$ in young patients with CF, reporting a strong correlation between annual decline in $\dot{V}O_{2\,peak}$ and survival over an eight-year period. Work by Pouliou et al. (2001) and Javadpour et al. (2005) has started to focus attention on other physiological parameters derived from exercise testing, besides the assessment of $\dot{V}O_{2\,peak}$, to determine if they hold any prognostic value. Pouliou and colleagues found a relationship between the $\dot{V}O_2$ kinetics during recovery from maximal exercise in adult CF patients and disease severity. Whilst, Javadpour et al. reported that CO_2 retention on exercise testing was associated with a faster rate of decline in FEV_1 in children with CF. Further investigations of other physiological parameters which can be measured through exercise testing is needed, to determine if they hold greater prognostic value than established ones.

Advances in exercise prescription are being initiated. The benefits of including anaerobic and strength exercises into training programmes for young patients with CF have been reported (Selvadurai et al., 2002; Klijn et al., 2004; Orenstein et al., 2004). Furthermore, anaerobic and strength exercises can provide variety to training programmes, which have been generally dominated by aerobic exercises. Novel and exciting exercise programmes can now be prescribed to help maintain the enthusiasm of the young CF patient to continue and adhere to training programmes. The importance of adherence to exercise prescriptions and maintaining physical activity levels in children with CF has also been highlighted. Children with CF after puberty become less active than their healthy peers and this trend is particularly evident in girls (Selvadurai et al., 2004). A relationship between the annual rate of decline in FEV_1 and habitual physical activity in girls has been identified (Schneiderman-Walker et al., 2005). Preserving FEV_1 through exercise prescription and physical activity is central to the management of CF, and this observation may partially explain the poorer survival of female CF patients. More studies are needed to assess which training programmes are the most successful in promoting adherence. Consequently, studies would require a longer follow-up period to monitor patient attrition.

The use of exercise testing and prescription among clinicians appears, however, to be underused. Barker et al. (2004) conducted a national survey to characterise the

opinions and practises of CF centres in Germany. From the results of the survey Barker and colleagues reported that despite an overwhelming belief in both the benefits of exercise testing and prescription among clinicians, exercise remains underused. From the returned questionnaires 87 % of clinicians viewed physical exercise as being extremely important or very important, and expressed an interest in expanding the prognostic and therapeutic applications of exercise. The feasibility of conducting exercise tests was not viewed as a major problem, equipment was available and one physician plus an assistant could conduct the majority of tests. Only 60 % of specialised centres, however, performed some sort of exercise testing at an average frequency of 2.3 years for patients aged eight and above. Furthermore, protocols and indication criteria were often non-standardized or not specified at all. Only two thirds of caregivers advised their patients to engage in physical activity, but failed to discuss specific modalities and potential hazards.

Whether this survey presents an accurate reflection of the opinions and practises of exercise in CF centres across Europe and the developed world remains as yet unknown. The work of Barker and colleagues shows a belief among clinicians in the importance of exercise testing and prescription; however, these beliefs are often not being put into practise. Feasibility studies may be a future direction for research to determine if the logistics of exercise tests within public healthcare are practical. It is well established that exercise in children with CF is both safe and beneficial, and the value of exercise tests are recognised among clinicians. Informing and motivating clinicians may increase the use of exercise tests and programmes as a prognostic tool and therapy within the healthcare management of the young CF patient.

Conclusion

Exercise testing offers an integrated, objective assessment of cardiovascular, respiratory, muscular and metabolic function of the patient. This is not achieved by individual tests of lung function, radiological investigations or measures of nutritional status that are routinely used in the clinical evaluation of CF. Additional information from exercise testing on the overall functional capacity of the patient can provide the clinician with a more comprehensive clinical assessment. This assessment could then be used to prescribe medication, more intensive therapy and transplantation. Results from exercise tests can also be used to prescribe individualised exercise programmes designed to help preserve and maintain the functional capacity and overall health of the patient. Exercise should be central to the management of CF. There is a strong association between fitness and survival, and exercise programmes have shown to improve exercise tolerance and functional capacity. At present no other therapy is more effective, in maintaining health and an active lifestyle in young patients with CF, than exercise.

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Key points

- New methods of health assessment from exercise testing in the young CF patient have shown to have prognostic value.
- The introduction of new training methods into exercise programmes can improve the functional capacity and quality of life of young patients with CF.
- Exercise is still being underused in the healthcare management of the CF patient.

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