Physical activity and exercise training in young people with cystic fibrosis: Current recommendations and evidence

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Abstract

Despite the acceptance of physical activity (PA) being integral to a young person’s health, children with disability often exhibit low levels of PA. In young people with cystic fibrosis (CF) the importance of exercise and daily PA is acknowledged by clinicians and their support teams, however, there is a lack of knowledge related to its prescription. CF is a recessive genetic disorder affecting the lung, pancreas and sweat glands. CF is the most common life shortening genetic disease in the Caucasian population for which there is no cure. In the UK, CF affects over 9000 people, with 4000 under 16 years of age. Only about half of the CF population can expect to live beyond 40 years of age. Besides drug therapies, rehabilitative exercise programmes form an important component of treatment and long term exercise programmes are considered positive treatment strategies, but all lack any detailed prescriptive information. Several reviews and editorials have highlighted the lack of evidence based research in PA and exercise training in young people with CF; but advocate a greater need for understanding the role of exercise in therapeutic interventions. The purpose of this review is to update the reader on the current recommendations and evidence in PA and exercise training for young people with CF. These developments have extended our understanding of PA and exercise training in children and adolescents with CF, and its implementation in the management of this chronic disease.

1. Introduction

Cystic fibrosis (CF) is an autosomal recessive genetic disorder affecting a number of body systems, including the lung, pancreas and sweat glands. The basic defect in CF is a disruption in the CF transmembrane conductance regulator (CFTR).1 The mutated CFTR causes a decrease in chloride ion conductance across the apical membrane of the epithelial cells, resulting in sweat that is high in sodium and chloride levels, and mucus that is abnormally thick and viscous.2 Mucus in the pancreas and gastrointestinal tract results in insufficient secretion of digestive enzymes and malabsorption of nutrients.3 Nutritional deficiencies lead to decreased fat stores and, as a result of protein malnutrition, muscle wasting.4,5 In the lungs the increased viscosity of mucus and its retention causes bronchial obstruction, recurrent or chronic infections, and finally respiratory failure. In more than ∼95% of all CF patients impaired respiratory function with hypercapnia, chronic hypoxaemia, and exhaustion of the respiratory muscles is the eventual cause of death.6

In addition to the control of chronic infection, airway clearance techniques, such as chest physical therapy and postural drainage, have been conventionally used in the management of CF to release mucus from the airways of the
lung.7 However exercise is being used more commonly for mucus clearance, with the intention to prevent recurrent lung infection. Moreover, physical activity (PA) and exercise training in CF have been reported to improve or slow down the rate of decline of important prognostic indicators such as lung function and aerobic fitness. Research from our children’s centre, amongst other centres around the world, have shown that children and adolescents with CF have a reduced ability to tolerate exercise.8,9 This has serious implications for their well-being and quality of life (QoL) and limits the opportunities for their involvement in sports and recreational activities. This is critical as aerobic fitness has been shown to be a key predictor of disease prognosis and mortality in patients with CF, independent of factors such as age, sex, lung function, and nutritional status.10 Understanding the efficacy of using exercise training to promote PA, physical fitness, well-being, and longevity is therefore critical in young people with CF. However, in a recent survey it was shown that clinicians and their multidisciplinary teams, whilst valuing exercise prescription, knew little about how to implement it in practice.11 In the UK current recommendation by the Cystic Fibrosis Trust12 advocates the use of exercise, but it is poorly and sporadically implemented nationally and lacks detail concerning the most appropriate exercise intensity and duration, and how this can be implemented alongside other aspects of routine clinical support (i.e., nutritional and psychological).

The implementation of an improved evidence based exercise prescription programme will, undoubtedly, improve the quality of clinical care and practice.13 Therefore, several key publications highlight the timeliness of this type of work and its potential impact on a national and international stage.14,15 Adherence is a key outcome for the success of any training programme, and is one reason why many studies fail to report significant improvements in physiological and psychological performance. By including a psychosocial element to a training programme, adherence may be improved but this has yet to be tested within the CF population. A recent supervised physiotherapy outpatient exercise programme showed excellent improvements in QoL, reduction in antibiotic days and some improvements in exercise capacity, despite no significant changes in body mass index (BMI).16 Accounting for these factors, researchers will be able to monitor the combined effect of exercise, psychological and nutritional support, and should be able to be precise about the magnitude of the training response. The aim of this review paper is to present the reader with the current recommendations and evidence in PA and exercise training for young people with CF.

2. Recommendations and evidence for PA in young people with CF

PA is defined as a behavior trait and subject to multiple extraneous factors.17 In young people with CF the burden of lung disease can impact on PA levels. Indeed, patients with CF attend regular clinic appointments, comply with drug therapy and physiotherapy sessions, and may miss school due to increased illness and/or pulmonary exacerbation. The burden of lung disease may reduce the opportunities to be active on a regular basis, and cause the PA patterns of young CF patients to become irregular. In 2004, recommendations on PA and health for healthy children were re-affirmed by the UK’s Chief Medical Officer.18 These guidelines have since been further reviewed by researchers in the United States who concluded that school children should engage in at least 1 h of moderate to vigorous activity per day, and that activities should be enjoyable and developmentally suitable. More recent updates on the PA guidelines for healthy children in the UK have been issued.19 However, one key point the UK group commented on was the lack of PA data for children with chronic diseases. Therefore, as a minimum when data are being collected on physical fitness of patients with CF, PA data should also be collected. Preferably the PA measurements should include both questionnaire, as well as, more objective measures such as accelerometry.

Comprehensive reviews on the measurement and assessment procedures have broadly defined the PA patterns of healthy young people and adults.20,21 However, for children with CF the evidence is less well established. In healthy young people, irrespective of instrument or device to measure PA, similar results have been found. Firstly, boys are more active than girls; secondly, activity declines with age; and thirdly, the relationship between fitness and activity becomes stronger with increasing age. The latter finding is important because it has yet to be conclusively shown that PA is a strong predictor for fitness in healthy young people as it is in adults. Although it has yet to be definitively established in childhood diseases, it is likely that the relationship between fitness and activity is stronger because of the connection between the disease progression and physical function. In adults, the two measurable outcomes, activity and fitness, are linked to morbidity and mortality and the association is even stronger. For children and adolescents with CF, the associations between health, activity, and fitness are important to understand in order to establish how these variables impact on the disease processes. Similarly, CF clinicians and their support teams wish to know and understand more about fitness and PA as these two variables are often used as part of the management and care of their patients.11 In young CF patients it has been shown that maintaining high levels of fitness can have a significant impact on mortality.10

Children with mild to moderate CF can be active but often choose not to be. This is due to a range of reasons, including parental overprotection, fear on the part of the child, lack of understanding of the disease by teachers and healthcare professionals, self or peer imposed social isolation,22 as well as, perceptions of “no time to play”.23 For the CF child it is important to be as active as possible from the beginning of their lives because when the progression of the disease becomes severe, the disease will itself become a direct cause of their hypoactivity. This pattern creates a cycle of ensuing hypoactivity leading to deconditioning within the body systems, leading to further deterioration in physical function that causes further hypoactivity. Therefore, it is imperative that
Exercise and/or PA are utilised as much as possible in the management of the disease. Although the use of exercise will not affect the pathophysiological process of CF, it will bring significant benefits to the CF patient.

Longitudinal measures of PA in young people with CF are not common, but one study by Schneiderman-Walker and colleagues showed for females 7–17 years of age that those in the two bottom quartiles suffered a more rapid rate of decline in forced expiratory volume in 1 s (FEV1) than girls in the top two quartiles. In boys with CF, there was no significant relationship found between PA and FEV1 decline. It is generally known that healthy boys are more physically active than girls, and this is likely even in young males with CF and might explain poorer survival rates of female CF patients. Other studies have found lower and similar PA levels compared to healthy children. The study by Britto and colleagues noted that adolescents with CF did not differ significantly when asked to recall participating in three or more activities a week which made them “sweat or breathe hard”. When adjustments were made by gender and health status, adolescents aged <14 years of age and between 15 and 16 years of age were more likely to participate in activities three times per week compared to adolescents aged >17 years of age. This is not uncommon in normal healthy children, but does suggest that adolescents making the transition to adult care need to be reminded to increase their PA levels.

Apart from age effects, Selvadurai and colleagues found that, like healthy children, puberty in CF affects the level of PA. When matched with prepubertal controls, boys and girls with CF had similar levels of PA. However, after puberty PA was higher in boys with CF compared to girls, although, interestingly, children with mild CF had higher levels of PA when compared to controls. When the PA of moderate and severe CF was compared to controls, however, PA was lower. These observations no doubt reflect the importance of the management of exercise when the disease progression is minimal compared to the incidental affects of the disease as it progresses more rapidly and hypoactivity is increased.

To the best of our knowledge only one study has recommended a more vigorous promotion of PA. Using Kriska’s Modifiable Activity Questionnaire (MAQ) to measure habitual activity, in patients with the worst lung disease, vigorous activity was significantly and positively related to peak oxygen uptake \((\text{VO}_{2\text{peak}})\) \((r = 0.83, p < 0.05)\). Although vigorous activity was less than when compared to healthy controls, Nixon and colleagues suggested more vigorous activities should be encouraged for CF patients in order to promote aerobic fitness which may impact on survival rates. Despite PA not being measured objectively, the findings of this study do reflect the interlinked correlates between fitness, PA, and morbidity which become more prominent as the disease progresses from mild to severe status. However, caution must be exhibited when inferring these results at an individual patient level. Ruf and colleagues have shown when measuring PA by a variety of questionnaires (Habitual Activity Estimation Scale; 7-day Physical Activity Recall; Lipids Research Clinics Questionnaire), as well as via accelerometry and testing aerobic fitness, that all the questionnaires were unable to generate valid PA data and aerobic fitness on an individual basis. All the associations were, at best, moderate \((0.32 < r < 0.56)\). Thus, using the questionnaires to advise PA programmes for patients with CF would be imprecise and should, at the minimum, be based on objective measures captured through accelerometry. For those clinics unable to afford accelerometers, a recent study published in the Journal of Cystic Fibrosis has reported that less expensive pedometers could be used to indicate significant changes in health status and self reported activity. However, further research is required to validate these findings, as 12 of 30 (40%) CF participants had a decrease in their measured step rate when well, compared to when ill (as assessed by the high standard deviation of the mean step rates).

In summary, more data are required before PA recommendations for young people with CF can be supported. Whilst the potential benefits of habitual PA and exercise are known, the lack of guidelines and recommendations are holding back many young patients and their families enjoying these benefits. Given the relative inexpensive cost of the many opportunities to increase PA, exercise prescription needs to occupy a more central role in the therapeutic intervention of young CF patients. Therefore, the challenge to researchers is to acquire more evidence based practice of the cost-benefit of PA and exercise interventions.

3. Recommendations and evidence for exercise training in young people with CF

The use of exercise training programmes to maintain or improve physiological health outcomes in young people with CF requires further investigation. In this section we refer to exercise training in the context of PA which is above and beyond normal habitual movement. In this scenario, the increased levels of PA, or exercise, are completed with a purpose to improve function, whether physiological or psychological. At present, evidence to suggest that participation in exercise training programmes can improve the health of the young CF patient is limited, and there is a lack of the highest, or level 1, evidence (i.e., randomised controlled trials (RCTs)) to support empirical data and validate conclusions. However, there is some high level evidence to suggest that both aerobic and strength training programmes can have a positive impact (e.g., improve or maintain important clinical markers) on patient health. There are currently five robust RCTs that have investigated the impact of exercise training on pulmonary function, aerobic fitness, and muscular strength in young people with CF. These studies all included the following criteria: subjects aged 6–18 years of age all diagnosed with CF by a medical professional; the exercise intervention was at least 2 weeks in duration, and described in sufficient detail to be repeated; the randomised grouping of control and treatment subjects; and a control group receiving routine medical care and no additional formal exercise training, with the exception of the study by Orenstein et al. Any attempts to amalgamate the results of these studies,
however, are confounded by differences in the mode, frequency, intensity and duration of the exercise intervention and initial disease severity of the patients. Another aspect lacking to these RCTs was a very singular focus on either aerobic or strength training in their respective programmes. In reality for programmes to be successful and to increase adherence, programmes should look to be multi-faceted and should incorporate aspects of psychological and nutritional support so that a more holistic approach is provided. The results of these RCTs are summarised in Table 1.

From a clinical perspective, the studies by Sosa et al.34 and Selvadurai et al.33 provide evidence for the use of supervised in-hospital exercise training in the treatment of patients admitted for pulmonary exacerbation; which can be defined as a worsening of patient symptoms, including an increase in cough, sputum production, dyspnoea and fatigue, and more objective measures such as decline in pulmonary function, fluctuation in core temperature, and weight loss. Selvadurai and co-workers33 reported that, compared to the control group, both aerobic and resistance training resulted in significant improvements in FEV1 at discharge and 1 month following; subjects in the aerobic training arm also demonstrated markedly greater improvements in exercise capacity (i.e., VO2peak) at discharge and 1 month after, compared with resistance training and controls. This study provides support that exercise training, particularly aerobic, may have a useful role on the in-patient ward for the treatment/rehabilitation of pulmonary exacerbation.

The study by Sosa and colleagues34 assessed the effects of an 8-week in-hospital exercise programme that combined circuit weight and aerobic training. Significant increases in VO2peak were observed in the training group by 3.9 mL/kg/min; and improvements in five rep-max bench press, leg press and seated row were all significantly greater in the training group. A novel approach to the study by Sosa and co-workers34 is the inclusion a 4-week detraining period. During the detraining period, in the training group VO2peak decreased by a comparable magnitude to baseline, however, all significant improvements in five rep-max outcomes from the training period were retained. No significant changes in pulmonary function, BMI, fat-free mass and body mass were reported during the training and detraining period in either the training or control group. Maintaining or improving pulmonary function and aerobic capacity, and slowing down the annual rate of decline in FEV1 and VO2peak is important as both are strong predictors of survival for children with CF.10,35 Therefore, any intervention that can impact positively on these health outcomes would be of potential benefit to patient prognosis; however, it is still not known whether exercise training can improve survival.

Schneiderman-Walker et al.24 reported that a 3-year unsupervised aerobic training programme resulted in significantly less annual decline in forced vital capacity (FVC) than the control group receiving usual care, however, no difference in the annual rate of decline of VO2peak was observed. Klijn et al.31 has presented data to support the use of anaerobic training in children with CF to significantly improve both VO2peak and anaerobic power. This supervised 3-month anaerobic training program resulted in significant improvements in peak power (watts) which were maintained over a 12-week period post training. The longest exercise programme duration of the five RCTs was conducted by Orenstein et al.32 These investigators compared the impact of aerobic and strength training over a 6- and a 12-month period. The results of the RCT showed that both aerobic and strength training can significantly improve leg muscle strength, whilst pulmonary function is maintained. To control for changes in body mass of subjects over the 6- and 12-month period, muscular strength was adjusted for changes in body mass over time.

The only RCT that reported significant improvements in both pulmonary function and aerobic fitness was Selvadurai et al.33 study conducted in a supervised in-hospital setting. RCTs by Schneiderman-Walker et al.24 and Orenstein et al.32 reported no changes in aerobic fitness from exercise training; moreover, Orenstein et al.32 reported a significant reduction in aerobic fitness from 6 months of strength training. This might be attributed to low adherence reported by Orenstein and co-workers, and the unsupervised nature of the exercise programme of Schneiderman-Walker and colleagues.24 Furthermore, no changes in pulmonary function were observed by Orenstein et al.32 over the 6- and 12-month period, which, again, may be attributed to the lack of adherence. When compliance was reported to be high, however, a significant protective effect on FVC was shown.24 Klijn and co-workers31 reported from their 3-month anaerobic training programme no significant changes in pulmonary function or aerobic fitness; however, the control group demonstrated a significant fall in aerobic fitness over the training period.

Despite a wide divergence between the RCTs in duration, setting and disease severity of participants, the evidence shows that none of the exercise training programmes had a negative effect on pulmonary function or muscular strength, and the fall in aerobic fitness of the strength group at 6 months of a 12-month exercise programme reported by Orenstein et al.32 may have been due to poor compliance. The RCT by Selvadurai et al.33 showed great promise for the value of exercise training in children with CF, with significant improvements reported in pulmonary function, aerobic fitness and strength from the exercise groups. It must be taken into consideration that these subjects entered the study with pulmonary exacerbation, increasing the likelihood of a training affect, and masked by the medications received whilst in hospital. However, no significant improvements were reported in the control group, and, in fact a significant decline in FEV1 was shown over the 19-day training period in the control group.

It is encouraging that all exercise programmes, including a strength training component, reported increases in leg strength in the children with CF.24,31–34 Only in the study by Selvadurai et al.33 was this increase not reported as being statistically significant. These data show that improvements in leg strength can be achieved by both short- and long-term training and of moderate to high intensity. CF is characterised...
## Table 1
Exercise training programmes in young people with cystic fibrosis: randomised controlled trials.

<table>
<thead>
<tr>
<th>Study</th>
<th>Exercise training programme</th>
<th>Group ((n))</th>
<th>Frequency</th>
<th>Intensity</th>
<th>Duration</th>
<th>Pulmonary function</th>
<th>Aerobic fitness</th>
<th>Muscular strength</th>
<th>Anaerobic fitness</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>(\Delta) % (p)-value</td>
<td>(\Delta) mL/kg/min (p)-value</td>
<td>% (\Delta) p-value</td>
<td>% (\Delta) p-value</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>% (\Delta) pre-post</td>
<td>% (\Delta) pre-post</td>
<td>% (\Delta) pre-post</td>
<td>% (\Delta) pre-post</td>
</tr>
<tr>
<td>Schneiderman et al. (2000)</td>
<td>Control ((n = 36))</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>−3.47 (4.93) NR</td>
<td>−2.42 (4.15) NR</td>
<td>−1.85 (2.51) NR</td>
<td>−1.80 (2.21) NR</td>
</tr>
<tr>
<td></td>
<td>Aerobic ((n = 36))</td>
<td></td>
<td>3 × 30 min/week</td>
<td>70%–80% HR(_{\text{max}})</td>
<td>3 years</td>
<td>−1.46 (3.55) NR</td>
<td>−0.25 (2.81) NR</td>
<td>−1.85 (2.51) NR</td>
<td>−1.80 (2.21) NR</td>
</tr>
<tr>
<td>Selvadurai et al. (2002)</td>
<td>Control ((n = 21))</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>−4.51 (6.90) &lt; 0.05</td>
<td>2.28 (4.22) NS</td>
<td>−1.22 (6.15) NS</td>
<td>−6.30 (6.10) NS</td>
</tr>
<tr>
<td></td>
<td>Aerobic ((n = 21))</td>
<td></td>
<td>5 × 30 min/week</td>
<td>70% HR(_{\text{max}})</td>
<td>19 days</td>
<td>−6.54 (7.76) &lt; 0.05</td>
<td>2.34 (4.62) NS</td>
<td>7.31 (6.29) &lt; 0.01</td>
<td>1.83 (6.23) NS</td>
</tr>
<tr>
<td></td>
<td>Resistance ((n = 22))</td>
<td></td>
<td>5 × 10 reps</td>
<td>70% max resistance</td>
<td></td>
<td>10.09 (7.43) &lt; 0.01</td>
<td>2.45 (4.18) NS</td>
<td>0.73 (5.89) NS</td>
<td>18.32 (7.02) &lt; 0.01</td>
</tr>
<tr>
<td>Klijn et al. (2004)</td>
<td>Control ((n = 9))</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>−0.6 (1.9) NS</td>
<td>1.5 (2.6) NS</td>
<td>−11.7 &lt; 0.001</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Anaerobic ((n = 11))</td>
<td></td>
<td>2 × (30–40) min/week</td>
<td>High</td>
<td>12 weeks</td>
<td>−</td>
<td>−</td>
<td>−</td>
<td>−</td>
</tr>
<tr>
<td>Orenstein et al. (2004)</td>
<td>Aerobic ((n = 26))</td>
<td></td>
<td>3 × 30 min/week</td>
<td>70% HR(_{\text{max}})</td>
<td>6 months</td>
<td>−2.78 NS</td>
<td>−</td>
<td>−</td>
<td>−1.91 NS</td>
</tr>
<tr>
<td></td>
<td>Strength ((n = 30))</td>
<td></td>
<td>3 sessions/week</td>
<td>&lt;55% HR(_{\text{max}})</td>
<td></td>
<td>−1.3 NS</td>
<td>−</td>
<td>−</td>
<td>−2.16 &lt; 0.01</td>
</tr>
<tr>
<td></td>
<td>Aerobic ((n = 25))</td>
<td></td>
<td>3 × 30 min/week</td>
<td>70% HR(_{\text{max}})</td>
<td>12 months</td>
<td>−4.68 NS</td>
<td>−</td>
<td>−</td>
<td>−0.91 NS</td>
</tr>
<tr>
<td></td>
<td>Strength ((n = 28))</td>
<td></td>
<td>3 sessions/week</td>
<td>&lt;55% HR(_{\text{max}})</td>
<td></td>
<td>−0.97 NS</td>
<td>−</td>
<td>−</td>
<td>−1.73 NS</td>
</tr>
<tr>
<td>Sosa et al. (2012)</td>
<td>Control ((n = 11))</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>5.6 (L/s) NS</td>
<td>3.1 (L) NS</td>
<td>2.2 NS</td>
<td>−4 &lt; 0.01</td>
</tr>
<tr>
<td></td>
<td>Weight &amp; aerobic ((n = 11))</td>
<td></td>
<td>3 sessions/week</td>
<td>1 × 12–15 reps (11 exercises)</td>
<td>8 weeks</td>
<td>−</td>
<td>3.7 (L/s) NS</td>
<td>3.3 (L) NS</td>
<td>3.9 0.002</td>
</tr>
</tbody>
</table>

Abbreviations: FEV\(_1\) = forced expiratory volume in 1 s; FVC = forced vital capacity; VO\(_{2\text{peak}}\) = peak oxygen uptake; HR\(_{\text{max}}\) = heart rate maximum; NR = not reported; NS = not significant.
by pancreatic insufficiency and malabsorption of nutrients, leading to deterioration in fat and muscle mass. Therapeutic interventions, such as exercise training programmes, that can improve muscular strength and size, will not only improve the functional capacity of the young CF patient, but have been shown to correlate strongly with improved prognosis. It is also particularly encouraging that data from Sosa and co-workers showed that improvements in muscular strength can be maintained during a 4-week detraining period, and that continuous improvements in leg strength can be achieved over a 6- and a 12-month training period.

Improvements in QoL have been reported in children with CF participating in exercise training programmes. Selvadurai et al. used the Quality of Well Being Scale to measure QoL pre- and post-1-month following aerobic training, resistance training and in controls. A significant improvement in QoL was reported 1 month later in the patients who had participated in the aerobic exercise training programmes whilst in hospital. However, no significant improvements in QoL were reported from the resistance training or control groups. The findings of this study show that improvements in QoL only reached statistical significance in the aerobic training group, where VO2peak significantly increased, are consistent with the findings of Orenstein et al. in that, QoL correlates better with changes in aerobic fitness than changes in pulmonary function. This emphasises the importance of cardiopulmonary exercise testing, as not only does it provide important clinical information of the impact of the disease over time, but also the impact of the disease on the patient’s QoL. Klijn et al.31 used a disease specific QoL questionnaire, the CF questionnaire; which takes into account the different developmental stages, and makes it possible to measure the health status and QoL of patients aged 6 years of age through to adulthood. At the end of the 12-week anaerobic training programme, a significantly higher score was found in the domain of physical functioning on the CF questionnaire in the training group, but no change was found in the control group or in other QoL domains.

Despite the lack of RCTs designed to investigate the physiological effects of exercise training programmes, the reviewed RCTs provide good evidence that pulmonary function, exercise capacity, and muscular strength can be improved in young people with CF by participation in exercise training programmes. This has important implications for patient prognosis in young people with CF. The best improvements in these parameters have been derived from in-patient supervised exercise training programmes. Indeed, this in-patient hospital study was the only study to report significant improvements in both pulmonary function and exercise capacity. Patient adherence to the exercise training programme may have been an important factor in these studies. Aerobic training has been shown to improve pulmonary function, and slow down its annual rate of decline; however, improvements in VO2peak from exercise training are unclear. Aerobic fitness improved during a short duration of aerobic training and an 8-week programme that combined both weight training and aerobic exercise, but not with any other form of aerobic training. This is likely due to the typical error of VO2peak testing for which at least a 10% change from pre- to post-test score is required for it to be of a sufficient magnitude to declare a clinically meaningful change. Changes in muscular strength from exercise training showed the most improvement, and showed increases with strength/resistance, anaerobic and aerobic training.

In summary, there is great potential for further research in this area to determine the most effective exercise programme for children with CF. Indeed, large improvements in muscle function have been shown with strength training; and the study of weight and resistance programmes regarding intensity and dose response relationships hold much promise.

To date there have been very few attempts to combine physiological, psychological, and nutritional interventions in training studies of young CF patients. The likely benefits to the patients include clinical improvements in health status. Clinical improvements in exercise capacity, lung function, self-efficacy, competence, and self-confidence and thus QoL are all accompanied by improvements brought about by training. This has important implications to the cost-benefit of exercise prescription in CF management. Indeed, in one study an antibiotics saving of £66,000 over 1 year was attained. As a consequence of these clinical improvements hospital admissions and antibiotic prescriptions are reduced, school attendance increases, self confidence when exercising increases, the young patient is empowered to participate in physical education and games with their peers on a more regular basis, and their social life can be significantly enhanced. However, more empirical data, followed by RCTs, will begin to help the clinical team assess the cost-benefit of the long term exercise interventions. In other countries, lessons from Chronic Obstructive Pulmonary Disease (COPD) training models have also been transferred to CF programmes. These analyses will help determine the sustainability of the programme and the clinical and economic benefits.

Several studies have highlighted that healthcare providers need to be better educated in PA promotion and exercise training prescription, and implementing both in clinical practice. A new integrated exercise training model of care would significantly improve the quality of clinical practice, by improving clinical outcomes and translating into longer term survival and QoL prospects, is needed in the management of young people with CF.

4. Conclusion

Clinicians and their support teams recognise the value of PA and exercises training in the therapeutic intervention for young people with CF. However, these teams are unsure about how best to implement its use amongst their paediatric patients. There is currently not enough detailed physiological information to prescribe programmes of sufficient duration, intensity and frequency in which to affect clinical outcomes. Additionally, little is known about the effects of exercise programmes on bone health and CF related diabetes, and more
work should be directed at investigating how exercise can improve and control these outcomes. The impact of exercise training programmes on other physiological outcomes such as patient symptoms, nutritional status, bacteria colonisation and QoL requires more attention. Determining the effects of exercise training post lung transplantation would also be an important area of future study. Furthermore, strategies to improve adherence to exercise training in children with CF are needed to realise the full potential of exercise programmes as therapeutic interventions.

Currently, a broad approach to increasing or maintaining PA of an aerobic and strength basis is the best that can be prescribed. However, there are some convincing data which show savings related to hospital admissions and reduced costs in medication, as well as, improvements in current disease status. As more of this type of evidence is provided, the relative low cost of implementing exercise programmes will be seen even more favourably. The provision of this type of evidence should act as a driver to investigate more comprehensively the efficacy of exercise prescription tailored to each patient’s needs.

References


