Pulmonary rehabilitation in children and adolescents with cystic fibrosis

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Abstract

The increase in survival of children affected by cystic fibrosis (CF) has generated new needs, paramount of which is for them to be able lead, as far as possible, a normal life and that will include involvement in sports activities. Several authors have developed sports activity programmes for children with CF – at home, in hospital, supervised and unsupervised, and studies with these have shown a minor increase in lung function test, but no significant increase in maximal oxygen consumption (VO_2 max). VO_2 max is an integrated measure of cardio-respiratory-muscular function and has been demonstrated to correlate with longer survival. Training for sports activities increases VO_2 max and, as CF can slow down the reduction of this parameter, special training programmes tailored to the needs of children with should be considered.

Keywords cystic fibrosis; sport

Introduction

The involvement of the sweat glands, lung and gastrointestinal tract in cystic fibrosis (CF) is well known. The challenge for the third millennium is to find a genetic solution. However, while waiting for this solution, the increase in survival time of patients with CF due to the prevention of infection with more efficient antibiotic therapy and more effective physiotherapy, has generated new needs. The principal need is for these patients to be able to lead a 'normal' life and it is difficult to how a 'normal' life could exclude physical activity.

Clinical findings and discussion

Rehabilitation is generally thought of as the recovery of abilities lost due to heart or cerebral ischaemic disease, or as a result of orthopaedic injury. In children with CF, rehabilitation is more a matter of maintaining and increasing abilities, for example through exercise and sport, which could gradually improve the efficiency of the whole body, in particular the heart and lungs.

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Some key points concerning physical exercise in CF have been studied, in particular the cardiovascular and ventilatory response, the behaviour of gas exchange and, if present, desaturation.[1] These points have been of interest since 1986 when Bar Or, writing about sport for sick children, emphasized the fact that the major problem in CF was the low O2 content in the arterial blood.[2] In 1987, Canny and Levison pointed out that exercise can be performed safely in CF, and provides a simple and reproducible indication of overall health.[3] Exercise in CF is limited by the degree of lung disease and, to a lesser extent, by compromised nutritional status. Patients therefore should be provided with individual exercise prescriptions. In Canny and Levison's study, pulmonary function generally remained unchanged.[3] In 1987, it was not known whether such rehabilitation would improve long term prognosis.

Since 1987, several studies have been carried out on exercise programmes in CF. Emphasis was mainly on the intensity of programmes (if exercise was maximal or submaximal or dictated by heart rate); the frequency of sessions (from two per day to two per week); the location (if in a clinical setting or at home, supervised or unsupervised); and the duration (generally 3 months). The results of these studies have been similar: little or no changes in lung function test (LFT) but improvement in maximal oxygen consumption (VO2 max).[4–6] VO_2 max is an integrated measurement of the performance of the heart–lung–muscle system. As expressed in the Fick equation, it is the product of cardiac output and the difference between the O_2 content in the arterial and mixed venous blood. It is expressed in L/min and, to correlate better with body size, in ml/kg/min. A normally fit healthy child has a VO_2 max of about 40 ml/kg/min, and the higher a child's VO_2 max, the higher their performance.

Sporting activities could have some potential adverse effects in CF. Some patients with more severe lung disease may experience low oxygen levels in the blood as a result of exercise. In most cases, blood oxygen levels will normalize within minutes of stopping exercise. However, to avoid this problem, patients should be tested for aptitude before being enrolled in sports activity programmes. Many patients will cough during exercise and this may be quite distressing. The coughing is not dangerous and may even be helpful in clearing mucus. Shortness of breath, present in some children, is due to exercise-induced bronchoconstriction and may be prevented in the same way as exercise-induced asthma is. Particular attention should be paid to salt and fluid loss as patients with CF are at greater risk for this sport-related problem than their healthy non-CF peers.

Physical training in patients with CF may improve exercise tolerance, even in the post-training period. De Jong et al investigated the effects of a 3-month home sub-maximal exercise training (15 min, twice a week, supervised) for 2 months. During the last month, 10 adolescent patients, mean age 20 years, were advised to continue the programme without supervision. Significant improvement in maximal exercise capacity and VO_2 max was observed, and was maintained after the unsupervised exercise period.[4] Schneiderman-Walker et al, in a 3-year home exercise programme for 72 patients aged 7–19 years, showed that pulmonary function declined more slowly in the exercise than in the control group, suggesting that patients with CF benefited from participating in regular aerobic exercise.[5] These

authors stressed the necessity for compliance and an appropriate attitude to sport activities before including an exercise programme in the conventional care regimen for patients with CF.

Orestein and Nixon studied the usual physical activity of patients with CF and found those children with CF, even those with good lung function, engaged in less vigorous activities than their non-CF peers.[6]

We studied a group of 12 CF patients, mean age 15 years, using LFT and maximal exercise stress test before and after a 3-month, twice weekly, supervised exercise period of 30 min of running on a treadmill: at 60% of the maximal baseline heart rate (HR) for the first 4 weeks, at 70% for the following 4 weeks, and at 80–85% for the last 4 weeks. The results showed a significant improvement after the training period in duration of exercise, VO_2 max and pulmonary ventilation, and with no changes in LFT.

Strength and anaerobic training have measurable effects on VO_2 max, aerobic performance and on health-related quality of life.[7] Data on oxygen consumption from the study of Orenstein et al show that this VO_2 max value could predict early mortality.[8] Pianosi, in a recent paper, reported a study on 28 CF patients, aged 8–17 years, followed for 5 years with LFT and stress test. VO_2 fell during the observation period in 70% of patients. Initial peak VO_2 was not a predictor of mortality, but the rate of decline and final VO_2 max were significant predictors.[9]

Conclusion

High VO_2 max is a marker for longer survival in CF patients. With physical training, it is possible to increase or at least slow down the decrease in this parameter. We should therefore offer children with CF the possibility to participate in sports programmes and physical activities that could help them to obtain a better physical condition and better quality of life.

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