Are exercise programs an effective treatment for children with cystic fibrosis?

Why could exercise be beneficial in cystic fibrosis? Cystic fibrosis (CF) is a genetically inherited, autosomal recessive disease that is caused by a mutation in the gene encoding the cystic fibrosis transmembrane conductance regulator protein. As a result of this deficit, multiple organs, primarily the lungs, pancreas and digestive system, become congested with thick, sticky mucus resulting in infections and inflammation. Despite advances in medical care, life expectancy remains shortened, with 85% of CF-related deaths caused by lung disease [1]. Exercise capacity in this population may be limited by reduced lung function, nutritional status, metabolic deficits and peripheral muscle function.

Extensive research into the effect of exercise training and physical activity in the healthy child has demonstrated physiological and quality-of-life benefits. Guidelines for exercise testing, prescription and training for healthy children are also well documented. However, by comparison, there is much less evidence demonstrating that children with CF achieve the same level of benefits as their healthy peers, and guidelines for exercise are still being developed. Most available evidence relates to short-term benefits of exercise, with a particular dearth of good-quality longitudinal studies.

Current guidelines for the physiotherapy management of children and adults with CF suggest that regular exercise and physical activity be prescribed in conjunction with airway clearance and inhalation therapy (ACT) [2]. The rationale is that exercise may enhance sputum clearance [3], improve or maintain lung function [4], reduce breathlessness [5], increase aerobic capacity and muscle strength [6], and improve bone health [7] and quality of life [8]. Regular adherence to exercise and ACT is variable in both adults [9,10] and children [11], with time required to complete ACT, nutritional status and progression of the disease impacting uptake.

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"...recent pilot studies in subjects with more advanced disease, where the exercise intervention runs over a longer period, have implied marked improvements in a variety of outcomes..."
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How should exercise be prescribed in CF?

Exercise programs by design should include three core components: exercise testing, exercise prescription and exercise training. Comprehensive maximal and/or submaximal exercise tests, such as a cardiopulmonary exercise test (CPET) or functional field-based incremental walk/run tests (ISWT), should be performed to monitor for exercise-induced limitations before exercise is prescribed, and exercise training is implemented in children with CF[15]. However, a survey of exercise testing and training programs in UK-based CF centers reported that exercise tests, such as CPET, are limited to a few specialist centers, and even they are underused[16].

The advantage of using CPET to determine a safe, individualized exercise prescription in children with CF is that the test measures gas flow, VO_{2\text{peak}}, CO_{2} production, respiratory exchange ratio, pulse oximetry and heart rate during exercise. When combined with the estimated work rate achieved during the test, variables such as anaerobic threshold and minute ventilation relative to CO_{2} exhalation can be calculated[17]. These outcomes are useful in determining exercise limitations such as pulmonary hypertension, exercise induced hypoxemia and cardiac arrhythmias. The results should then be used to determine target heart rates and/or ventilatory threshold exercise training ranges for the child. In the absence of CPET, field-based test results should be performed.

Based on the results of exercise tests, frequency, intensity, time and type exercise principles should be used to guide an age-appropriate, individualized exercise training program for the child. Dependent on the aims of the training program, clinicians could prescribe aerobic, anaerobic or muscle strength training or a combination of these for the child[14]. It should be noted that guidelines for exercise prescription and training in children with CF require further development.

What is new in children with more severe disease, and where do we go from here?

In the last 5 years there have been three novel, 12-month observational studies that have explored an alternative model of physiotherapy care in small groups of sicker children with moderate-to-severe CF. Children who have required >40 days of intravenous (iv.) antibiotic treatment in the previous year were included in the programs, and data from the intervention year were compared with the previous year’s data. Each program provided regular supervised, individualized exercise training in an outpatient[18] or outreach setting[19,20], in addition to current specialist CF care.

In the 2008 Brisbane study, Black et al. (n = 10) demonstrated a 48% reduction in iv. antibiotic requirement, and a significant increase in lung function and 20 m ISWT scores[18]. Urquhart et al. (n = 12) demonstrated a 17%
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“...the aim of these studies is to quantify both health improvements and the overall cost impact of extending such initiatives more widely across pediatric cystic fibrosis care.”

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