Exercise programs for children with cystic fibrosis: a systematic review of randomized controlled trials
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CRD summary
The review concluded that both aerobic exercise and strength training may benefit pulmonary function, aerobic fitness and strength among children with cystic fibrosis. Factors that limited the review included the small amount of evidence available and inconsistencies between studies. Therefore, the author's conclusions may require cautious interpretation.

Authors' objectives
To assess the effect of exercise programmes on pulmonary function and fitness in children with cystic fibrosis.

Searching
MEDLINE, SPORTDiscus, CINAHL, AMED, EMBASE and Cochrane Central Register of Controlled Trials (CENTRAL) were searched from inception to October 2008. Search terms were reported. The search was restricted to published studies in English.

Study selection
Randomised controlled trials (RCTs) of exercise training for six to 18 year olds with medically diagnosed cystic fibrosis were eligible for inclusion. The intervention was required to last for at least two weeks, be structured and described in sufficient detail to be replicated. Studies were required to compare exercise training versus a differing modality of exercise and/or usual care (without additional formal exercise). Studies that were of inspiratory muscle training only or a single session of exercise were excluded.

Participants in the included studies had mild, moderate or severe cystic fibrosis. Mean age was 11 to 14 years. Mean height was 143cm to 160cm. Mean weight was 36kg to 48kg. Schwachman scores ranged from 67 to 90. Interventions included aerobic and anaerobic exercise and resistance training, which varied in intensity, duration, length of sessions and setting. Controls received usual care and activities. Outcomes reported in the review were pulmonary function and fitness (aerobic, strength and anaerobic). Measures used included forced vital capacity (FVC), forced expired volume in one second (FEV1%), maximum volume of oxygen consumed, change in muscle force during leg extension and change in peak power output.

A single reviewer selected the studies.

Assessment of study quality
Published criteria were used to assess components of study validity and of the quality of the exercise programme. Criteria included: randomisation; allocation concealment; baseline comparability of groups; eligibility criteria; blinding; reporting of effect estimates; use of intention-to-treat (ITT) analysis; compliance; drop-outs; and description of intervention.

The author did not state how many reviewers conducted the assessment.

Data extraction
Data on changes from baseline in each group were extracted for each study and p values for differences between the groups (where reported) in the primary studies.

The author did not state how many reviewers conducted the assessment.

Methods of synthesis
The studies were combined in a narrative synthesis organised by outcome. Differences between the studies (such as population, type of intervention and setting) were discussed in the text.

**Results of the review**

Four RCTs were included (n=212, range 20 to 72). All studies met criteria for randomisation, allocation concealment, baseline comparability and reporting of effect estimates, drop-outs and intervention details. Eligibility criteria and compliance rate were reported in three RCTs each. Blinding and ITT analysis were reported in two RCTs each.

**Pulmonary function (four RCTs):** One RCT found that FEV1% was significantly improved from baseline by short-term aerobic exercise (p<0.05) and resistance training (p<0.01); function significantly decreased in controls (p<0.05). The other three RCTs had longer follow-up and none reported statistically significant findings for FEV1%. One RCT of aerobic exercise reported a significantly smaller decline in FVC over long-term follow-up in the intervention group than in controls (p value not reported). Other findings for FVC (one short-term RCT) were not statistically significant.

**Fitness (four RCTs):** Both relevant RCTs (one short term and one long term) reported significantly improvement from baseline in leg strength in one or more intervention group (p≤0.03); improvements lasted up to 12 months (one RCT). Findings for aerobic fitness were inconsistent (four RCTs). Exercise training was associated with significant improvement from baseline in one intervention group of one short-term RCT (p<0.01) and with significant deterioration from baseline in one intervention group of one long-term RCT (p<0.01). Anaerobic fitness was significantly improved in the intervention group compared to controls (p<0.0001) in the only RCT that reporting this outcome (short term).

**Authors' conclusions**

Both aerobic exercise and strength training may benefit pulmonary function, aerobic fitness and strength among children with cystic fibrosis.

**CRD commentary**

The objectives and inclusion criteria of the review were clear and relevant sources were searched for studies. The search restriction by language and publication status meant that some studies may have been missed. It did not appear that efforts were made to minimise error and bias by having more than one reviewer undertake review processes. It was unclear whether statistical pooling of studies was considered. In view of heterogeneity between studies it appeared that narrative synthesis was probably the most appropriate method of combining the studies. As the author noted, inconsistent findings across studies could have related to differences in the duration of the intervention, study setting, disease severity, compliance, outcome measures and/or cointerventions.

The review was limited by a number of factors, including the small amount of evidence available and inconsistencies between study methods and findings. Therefore, the author's conclusions may require cautious interpretation.

**Implications of the review for practice and research**

**Practice:** The author stated that children with mild to severe cystic fibrosis may benefit from exercise training, especially in an in-patient setting. Both short-term and long-term strength programmes were effective, even in prepubertal children.

**Research:** The author stated that research was needed into the most effective exercise programme for children with cystic fibrosis. Research needed to include which exercise modalities (or combinations) and what intensity, duration and dose of exercise were most effective. The author recommended research (RCTs included) of home-delivered exercise, strength training and anaerobic exercise that reported outcomes such as quality of life, need for chest physiotherapy, recurrent infection rate, nutritional status and physiological end points.

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