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Cystic fibrosis

What is cystic fibrosis?

Cystic fibrosis (CF) is an autosomal recessive genetic disorder where the balance of substances in the body, especially salt and water, is not properly controlled. Many body systems are affected, but especially the lungs and digestive system. For example, thickened mucus in the airways causes persistent coughing, high phlegm production, chronic infections and changes in the airways (bronchiectasis). Thickened mucus can also block ducts in the digestive system, preventing food digestion and absorption, and causing malnutrition. Mucus blockage of the bile duct may cause liver cirrhosis and portal hypertension (high blood pressure). Excessive salt is lost in sweat and many organs also become inflamed, including the bones and the pancreas, which may lead to diabetes or bone diseases.

People with CF may have lung and bowel problems, fatigue, low fat and muscle mass, muscle weakness, problems with metabolism, osteoporosis and bone fractures. The functions of all the affected body systems must be regularly tested to monitor the progress of the disease. Unfortunately, the effects of the disease on the body organs often lead to poor health, with a median survival age of 37 in people with CF.

How is it monitored?

There are a number of outcome markers in use to monitor the progression of CF; such as: lung function, bone mineral density, inflammatory markers, VO2max, number of hospitalisations, number of chest infections and chronic illness severity scores.

Why is exercise important?

Research on exercise for people with CF has shown variable results, but the positive effects of exercise on the lives of people with CF include:

- reducing mucus thickness and stickiness;
- increasing exercise tolerance;
- increasing endurance;
- improving cell functioning;
- increasing muscle mass and strength; and
- increasing quality and length of life.

Research also shows that there is:

- an association between the aerobic fitness of young CF patients and survival over eight years;
- a significant link between activity levels and quality of wellbeing; and
- a good link between physical activity levels and body mass percentile in children with moderate to severe lung disease (suggesting possible effects of poor nutrition on muscle mass and functional ability).

Exercise is an important part of the daily treatment routine for people with CF. Despite this, children with CF have trouble doing as much vigorous physical activity as healthy children because of the debilitating effects of the disease and the frequent bouts

of illness. Participation in regular exercise and training programs is often challenging for many CF children, their parents and their healthcare providers.

What type of exercise is best for people with CF?

Unfortunately, the optimal type and amount of exercise for people with CF is not yet established. However, based on CF training studies and the general principles of exercise programming in healthy and clinical populations, a rounded program of aerobic, resistance and flexibility exercises, and sport are recommended. For example:

- aerobic exercise (e.g. running, swimming) improves heart and lung fitness and endurance, and insulin function;
- resistance exercise (e.g. lifting weights) improves muscle mass, strength, bone mass and density, and insulin sensitivity;
- impact exercises (e.g. jumping) improve bone density; and
- flexibility exercises (e.g. stretching, yoga) improve general flexibility and joint function.

Exercise programs for children should emphasise regular, varied and fun-filled physical activities. The table below provides a guide to exercise programs for people with different severities of CF signs and symptoms. The programs may need to be individually tailored according to the results of monitoring tests.

Exercise recommended for people with CF

Exercise characteristic	CF severity	Aerobic	Resistance	Impact
Intensity	Mild–moderate	70–85% HRmax 60–80% VO2peak	70–80% 1RM 3 sets of 8–12 repetitions	10–50 jumps/day Each jump 8 cm high
	Severe	60–80% HRmax 50–70% VO2peak	50–70% 1RM 2–3 sets of 8–12 repetitions	---
Duration	Mild–moderate	30–60 min	---	---
	Severe	20–30 min	---	---
Frequency	Mild–moderate	3–5 days / week	3 days/week	3–6 days/week
	Severe	2–5 days / week	2–3 days/week	
Mode		Walking, running, cycling, swimming, rowing, tennis	Weights	Jumping

Note: HRmax = Maximum heart rate; VO2peak = peak oxygen consumption; 1RM = maximum weight you can lift for 1 repetition.

Related information and references

Exercise & Sports Science Australia www.essa.org.au
Cystic Fibrosis Australia www.cysticfibrosis.org.au

1. Selvadurai, HC, Blimkie CJ, Meyers N et al. Randomized controlled study of in-hsopital exercise programs in children with cystic fibrosis. *Pediatr Pulmonol* 2002; 33(3):194–200.
2. Williams CA, Benden C, Stevens D et al. Exercise training in children and adolescents with cystic fibrosis: theory into practice. *Int J Pediatr* 2010; doi: 10.1155/2010/670640
3. Elborn JS. How can we prevent multisystem complications of cystic fibrosis? *Semin Respir Crit Care Med* 2007; 28(3):303–11.