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Cystic Fibrosis: Symptom of Destructive Lower Respiratory Infections

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Opinion Article

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DESCRITPION

Cystic fibrosis is an autosomal recessive disorder characterized by repeated and destructive lower respiratory infections, resulting in the gradual destruction of the lung tissue.

The cause of CF is due to mutations in the cystic fibrosis transmembrane conductance regulator gene on chromosome 7. The CFTR gene regulates chloride and sodium transport in the epithelial surfaces of the airway, pancreatic and biliary ducts, the gastrointestinal tract, sweat ducts and the vas deferens. Pathogenic mutations either remove or reduce the function of the CFTR gene. This results in the production of sticky mucus build up and blockages that affect mainly the lungs, pancreas and sweat glands.

CF is an inherited condition that affects 1 in 2500 male or female pediatrics. All Australian babies are screened at birth for CF. Blood tests are carried out for the genetic testing of the CFTR gene and sweat tests are carried out to measure the amount of salt in the sweat. There is currently no cure for CF, there are over 1500 CFTR gene mutations but not all are associated with CF. CF patients have a reduced lung capacity, tidal volume, peak aerobic capacity and poor pulmonary function due to the thick and sticky mucus build up in the lungs. The mucus clogs the small air passages and encourages bacteria to grow resulting in repeated infections and blockages causing irreversible lung damage.

CF also affects nutrition as the ducts from the pancreas to the intestine can become blocked preventing enzymes reaching the intestine for digestion, physical weakness, poor growth, delayed puberty and weight loss even though someone with CF may have a healthy appetite. The sweat glands also secrete sweat that is high in salt, when the body losses too much salt it causes dehydration, low blood pressure, tiredness and an increased heart rate.

CF can severely affect quality of life and reduce the ability to participate in day-to-day activities. Daily physiotherapy air clearance techniques such as chest percussion or positive expiration pressure therapy are used to clear secretions out of the lungs. Antibiotics are taken to treat and control persistent infections. Bronchodilators are used to make it easier to breathe by relaxing the muscles in the lungs and widening the airways. Steroids are also used to reduce inflammation in the airways and improve lung function. Pancreatic enzyme replacement capsules and nutrient supplements are consumed along with a high energy and salt diet. Exercise is a key factor in treatment for CF, it helps to loosen secretions, increase lung capacity, improve airway clearance, increase energy, bone density, strength, endurance and life expectancy.

20–30 minutes of aerobic exercise (swimming, cycling, walking, jogging) 3-5 times a week. Aim to keep the heart rate between 140-160 beats/minute. To encourage children to adhere to exercise programs use fun activities and games that meet the above heart rate and duration. Anaerobic exercise and strength training have been shown to have benefits for Cystic Fibrosis sufferers, especially when combined with aerobic activity. 30-45 minutes of anaerobic activity lasting for 20-30 seconds 2 times a week has been proven to increase anaerobic and aerobic performance. Whilst some studies have looked at the possibility of regular physical activity replacing conventional Cystic Fibrosis treatment, none have safely proven that it should be considered complimentary to the treatment already being received. Participants first beginning an exercise regime should do a pre-exercise screening to check their suitability for exercise and any associated risks. It is not recommended that patients in acute stages of the disease participate in exercise. When exercising it's important to monitor hydration status and the environmental conditions. Due to issues with sweating, exercising in hot and humid conditions may cause problems with temperature regulation and dehydration.