CYSTIC FIBROSIS

CF Fact
cystic fibrosis fact sheet

Bronchiectasis

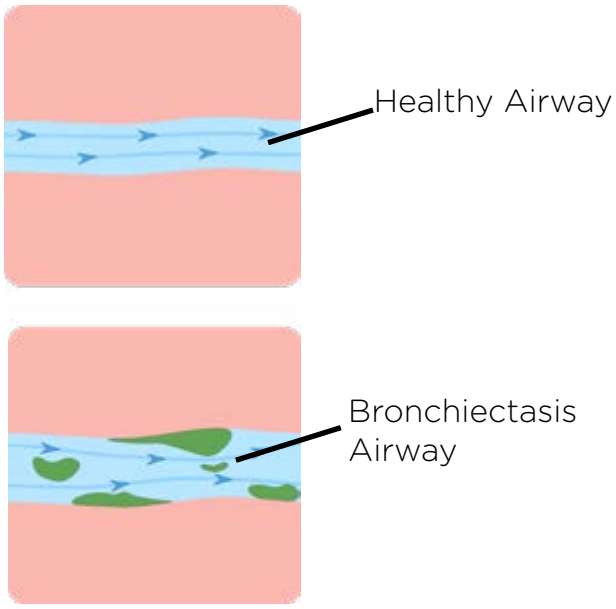
Cystic fibrosis (CF) lung disease is on-going and bronchiectasis occurs as a result of chronic infection, inflammation and mucus obstruction in the CF lung.

What is Bronchiectasis?

Bronchiectasis is defined as abnormal widening of the airways of the lung (bronchi). This is due to repeated infection and inflammation. The damaged airways are not able to clear mucus which builds up and becomes infected. This infected mucus blocks the airways and leads to more infection which damage the airways and impair the removal of secretions, resulting in airway obstruction.

The muscular and elastic components of the airways are also destroyed, and the tissues may be scarred. The excessive mucus that is produced leads to cough and recurrent respiratory infections. The widening of the airways and problems with cilia function in CF make it difficult to bring up secretions from the lower airways. Cilia are tiny hairlike structures that line the airways. They have a rhythmic beating motion that helps move the mucus towards the mouth so it can be cleared.

In CF, damage to the airways occurs early and babies often have no symptoms. Diagnosis of bronchiectasis can only be made with high resolution CT scan.



Management

Treatment aims at optimising well-being, lung function and reducing exacerbations to prevent lung function decline.

It is important to:

- Recognise symptoms of exacerbation (increased cough, fever, change in colour of sputum, weight loss, fatigue) and seek advice from your clinic.
- Avoid irritants e.g. cigarette smoke (including passive smoke inhalation), noxious fumes/gases and airborne pollutants.
- Avoid contact with those who have respiratory infections.
- Immunise according to immunisation schedule, including annual influenza vaccine.

- Eat well and use enzyme replacement according to fat intake. Good nutrition helps the immune system adapt to infections.

Airway Clearance

Airway clearance is an essential part of managing CF. It will improve gas exchange in the lungs and reduce excess mucus that provides a good environment for infection, thus reducing the inflammation.

- Take the medications as prescribed. Follow the correct order and technique when using inhalers and nebulisers.
- Drink adequate fluids to maintain hydration and keep mucus thinner to help with mucus clearance from the lung.
- Daily aerobic exercise in addition to airway clearance.
- Airway clearance routine as recommended by your physio.

Useful Resources

- Bronchiectasis Toolbox
- CFWA Factsheets

Cystic Fibrosis WA
The Niche
11 Aberdare Road
Nedlands WA 6009

T: +61 8 6457 7333
F: +61 6457 7344
E: admin@cfwa.org.au