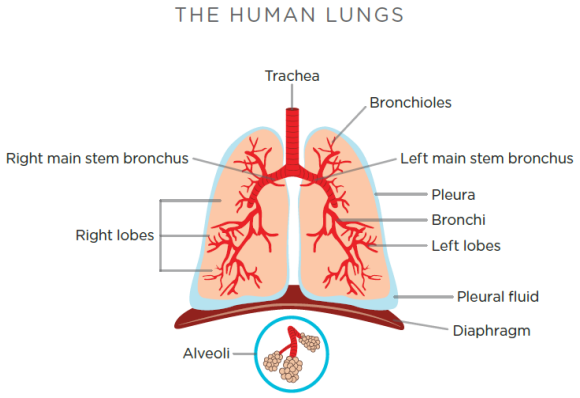
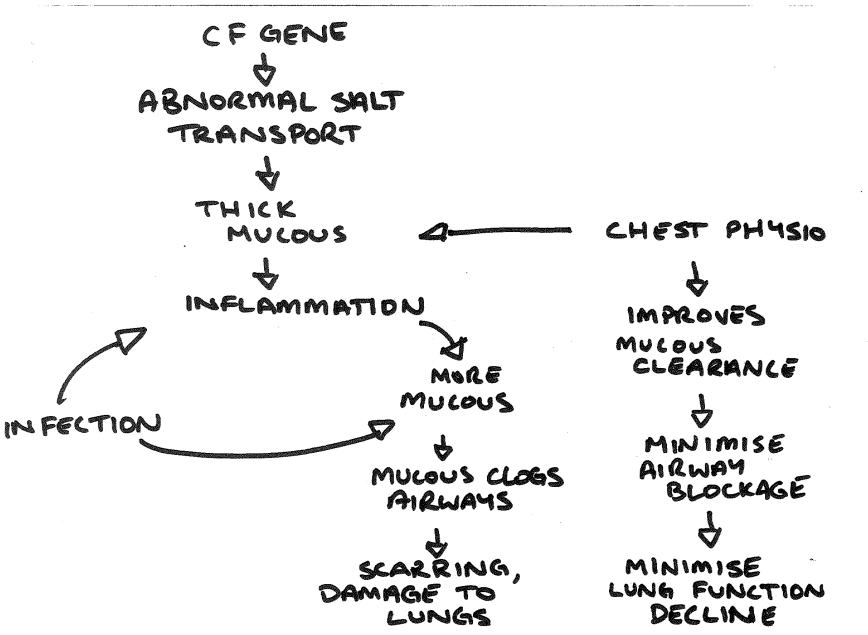
How CF Affects the Lungs

Your lungs are normal at birth, but because of the abnormal CF protein, the thin fluid secretions that are normally present in the airways, (bronchi), of the lungs become think and sticky. This makes it hard for the hairs on the bronchi, (cilia) that line the airways to move unwanted germs and particles out of the lungs. This can then cause lung inflammation with an infection, causing stickier, thick mucus, resulting in a vicious cycle. Frequent infections can cause irreversible lung damage. Chest physiotherapy is incorporated into the management of CF to help promote sputum clearance to prevent a buildup of sticky mucous blocking the airways.



How to manage mucous?

It is very important to keep your child's chest clear. At diagnosis, you will be taught how to assess your child's lungs and perform chest physiotherapy. It is recommended to do chest physiotherapy sessions once a day for 15-25 minutes per session if your baby is well, but it may need to be more frequent if they are unwell.



What is Chest Physiotherapy?

Chest Physiotherapy, also known as airway clearance, is a way to clear sputum from the lungs. Your physiotherapist at the hospital will teach you how to perform airway clearance techniques for your baby or child. We strongly advise children to lead active lives and participate in physical activities and sports to assist with airway clearance, lung function, bone density and global strength and conditioning.

Your child's CF physiotherapist will help create an individualized physiotherapy program for your child. As your child grows their physiotherapy program will change to suit their needs. If you have any questions talk to your child's CF physiotherapist.

Procedures

Lung Function Test

A lung Function test, (also called spirometry), is a non-invasive test that is performed to assess and monitor someone's lung capacity and function. It is performed using a portable machine in clinic or in the Monash Lung and Sleep Lung Function Laboratory. It is the measurement of forced expiration to assess the volume of air in the lungs (forced vital capacity) and if there is any blockage of the airways (forced expired volume in 1 second)

Chest X-ray or Chest CT scan

These are diagnostic imaging techniques that are performed to detect areas of increased mucous and infection. All children with CF will have yearly chest imaging to monitor lung health. Imaging may be done more frequently if your child is unwell or needs admission to hospital.

Pharmacy

Medications help with the management and treatment of CF. Each child will have a specific medication plan that best suits their treatment plan. Oral, nebulized and inhaled medications are the most common. The following information provides a brief

overview of each medication that may be prescribed for the treatment of CF lung disease. Medications need to be prescribed by your respiratory physician.

Salbutamol (Ventolin)

Salbutamol is a bronchodilator that is inhaled. It helps to relax the airway muscles which help enlarge the airways. This then will assist in mucous movement and generally taken before exercise, airway clearance and hypertonic saline.

Hypertonic Saline

Hypertonic saline is sterile, extra salty water that is inhaled through a nebuliser. It helps to draw moisture into the airways to rehydrate the lungs and helps to loosen and clear mucous. Before taking hypertonic saline, it is important to have a bronchodilator, (such as salbutamol), to open up the airways.

Dornase Alfa (Pulmozyme)

Dornase Alfa more commonly known as Pulmozyme is a mucous changing drug that helps to loosen and clear the mucous to assist mucous clearance and help prevent lung damage and reduce exacerbations. Pulmozyme should not be mixed with any other medication.

Bronchitol

Bronchitol is a dry powder in capsule form that is inhaled via a special inhaler twice a day. It helps to hydrate the airways to make mucous clearing easier. It is advised to use a bronchodilator prior to using Bronchitol.

Antibiotics

Antibiotics kill bacteria and are given to CF children to help prevent as well as treat new lung infections. Various antibiotics may be prescribed orally at home or intravenously which need to be given in hospital. Inhaled antibiotics can also be given such as TOBI to administer the medication directly to the lungs.

Anti-inflammatory

Reduce the inflammation in the lungs. Azithromycin is an antibiotic tablet that also has an anti-inflammatory effect and is used for patients with chronic lung infection with *Pseudomonas*.

CFTR Modifiers

These medications have recently been developed to help treat the underlying cause of the CF symptoms. **Kalydeco** (**ivacaftor**) is a tablet that was approved in 2016 for use in some children who have the specific "gating" gene mutation known as G551D. Kalydeco helps the defective CFTR protein stay open to allow the movement of salt and water in and out of the cells restoring the normal salt and water balance.
Orkambi is a new drug that combines **Ivacaftor** with **Lumacaftor**, a drug that helps more CFTR proteins come to the surface of the cell to normalize CFTR function. It is used for patients with two copies of the ΔF_{508} mutation.
It is likely that several more 'mutation-specific CFTR modifier drugs will become available for use in Australia in the near future.

Notes

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Photographs by Paige Emmett (2019)
Flow chart by Paige Emmett (2019)
Image logo by Melinda Kant (2019)

FAQ

What is **Kalydeco?**
Kalydeco is the brand name for a drug called Ivacaftor. Kalydeco helps the defective CFTR protein stay open to allow the movement of salt and water in and out of the cells restoring the normal salt and water balance. It is used only for patients with at least one copy of the G551D mutation

What is **Orkambi?**
Orkambi is the brand name for a drug that combines Ivacaftor and Lumacaftor that helps more CFTR proteins come to the surface of the cell to normalize CFTR function. It is used for patients with two copies of the ΔF_{508} mutation.

What is the **CFTR protein**?
A protein called Cystic Fibrosis Transmembrane Conductance Regulator. The CFTR protein acts as a channel that transports salt and water between the inside and outside of the cell.