

Medications

Medications help people with cystic fibrosis (CF) keep healthy and maintain a good lifestyle. The following information should increase your understanding of the medications that you or your child may be prescribed.

Common Types of Medications for CF Lung Disease

Bronchodilators

Relax the airway muscles which help enlarge the airways so mucus can be coughed up more easily. Ventolin (salbutamol) is an inhaled bronchodilator and may be used before exercise or airway clearance and before taking mucus thinning treatments such as Hypertonic Saline (HTS) nebulisers or Bronchitol.

Mucus Changing Drugs

Are inhaled medications that help breakdown the mucus in the airways and lungs so it can be coughed up more easily.

- Dornase alfa (Pulmozyme®) is a mucolytic (a type of mucus changing drug) that helps thin mucus. Pulmozyme® acts like “scissors” to cut the strands of DNA in the mucus, making it easier to clear. This helps to reduce lung infections and slow the lung damage caused by CF. You should not mix any other medications with Pulmozyme®.

Airway Surface Liquid Restorers

Increase the fluid in the airway to help thin mucus and make it easier to clear.



A bronchodilator like Ventolin® may be needed before you have this medication.

- Hypertonic Saline (HTS) is a sterile solution of extra salty water inhaled through a nebuliser. The salty mist solution draws water into the airways and makes it easier to clear mucus from the lungs. The first dose of HTS will need to be given in hospital or the clinic, and so lung function tests will be done to see if it is the right treatment for you or your child.
- Bronchitol® is a dry powder in a capsule which is inhaled using a special inhaler twice a day. Bronchitol® increases the hydration in the airway helping clear mucus from the lungs.

Antibiotics

Are medications that kill bacteria, however they are not effective against viral infections. They are prescribed to treat new infections and exacerbations

or may be used as a preventative treatment or as long-term therapy for those who have persistent or recurrent infections. The dose, length of treatment and type of antibiotic is different for each person depending on the infection.

- Oral antibiotics - liquid or tablets to fight bacteria causing lung infection.
- IV antibiotics - liquid medication given directly into the bloodstream through an intravenous catheter. A hospital admission is usually required to start the course which is sometimes completed at home.
- Inhaled antibiotics - a nebuliser may be used to change liquid medication into an aerosol or mist which is inhaled into the lungs. Dry powder inhalers deliver medication from a capsule into the lungs in powder form, such as the Tobi Podhaler®.

Anti-Inflammatory Medications

Reduce inflammation in the lungs. Some medications e.g. Azithromycin® can be given in special doses to reduce inflammation in the airways with the aim to prevent lung damage in childhood.

CFTR Modulators

Are designed to target the underlying defect in the CFTR protein that regulates the flow of salt in and out of the cells in the lungs and other organs of people with CF. These medications are only effective for specific gene mutations but with ongoing research it is hoped that in the future all people with CF will benefit. See [CFTR Modulator Therapy factsheet](#).

Pancreatic Enzymes

Are used in those who have pancreatic dysfunction to improve digestion. They contain enzymes normally produced by the pancreas which are needed to digest fats, carbohydrates and proteins. Doses of pancreatic enzymes are different for each individual depending on body weight and are sometimes adjusted

depending on symptoms. Creon® and Panzytrat® are the most common enzymes used in Australia.

Salt

Requirements vary depending on symptoms. People with CF lose large amounts of salt in their sweat which increases the risk of dehydration. Salt supplements will be recommended by your dietician and you may be advised to add extra salt to food.

Vitamins

Are needed for people with CF due to maldigestion and malabsorption of nutrients, in particular fat. This means that fat soluble vitamins are less readily absorbed by the body. It is important to take Vitamins ABDEK regularly as prescribed.

Gastric Acid Reducing Medications

Are used for those with Gastro-Oesophageal Reflux Disease (GORD). Drugs e.g. Nexium® and Losec® reduce the production of acid in the stomach reducing the irritation and indigestion. See [Reflux and CF factsheet](#).

Useful Resources

- [CFWA Medications Factsheets](#)
- [Drug Development Pipeline \(CF Foundation\)](#)
- [CFTR Modulator Therapies \(CF Foundation\)](#)
- [Medicines for Children With CF \(CF Conference 2015\)](#)

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