CYSTIC FIBROSIS

CFfacts
cystic fibrosis fact sheet

Medications

Medications help people with cystic fibrosis (CF) keep healthy and have a good lifestyle. Every medication treatment plan is individualised, however oral, nebulised and inhaled medications are most often in the treatment plan. 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. They are usually taken before airway clearance so mucus can be coughed up more easily. Ventolin is an inhaled bronchodilator and may be used before exercise, airway clearance and Hypertonic Saline (HTS) nebulisers.

Mucous 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 and loosen the mucus. It acts like "scissors" to cut the strands of DNA outside the cells in the CF mucus. The DNA is from white blood cells that are produced to fight infections. The DNA builds up and makes the mucus thick. Even if the person with CF is not aware of the lung infection the DNA is still outside the cells in the airways. Being able to clear the mucus helps slow the lung damage caused by CF and reduce exacerbations (lung infections). You should not mix other medications with Pulmozyme. Nebulisers for Pulmozyme: E-flow, Pariboy SX, Aero-neb.
- Airway surface liquid restorers increase the fluid in the airway to help accelerate mucus clearance.
- Hypertonic Saline (HTS) is a sterile solution of extra salty water inhaled via a nebuliser. The salty mist solution draws water into the airways to rehydrated the airway and make it easier to loosen and clear mucus from the lungs. Before HTS nebuliser it is important to use a bronchodilator as ordered by your doctor. The first dose of HTS will need to be given in hospital or the clinic, and some lung function tests will be done to see if it is the right treatment for you or your child. HTS is not suitable for all people. You should not mix other medications with HTS.

- Bronchitol is a dry powder in capsule form and inhaled using a special inhaler twice a day. Bronchitol increases the hydration in the airway helping clear mucus from the lungs.
    - A bronchodilator like Ventolin will be needed before you have the HTS or Bronchitol.

Antibiotics are drugs that kill bacteria, however they are not effective against viral infections. They are given to people with CF to treat new infections, as a preventative treatment, to treat exacerbations, and as long-term therapy for those who have persistent or recurrent infections. The dose, length of treatment and type of drug is different for each person depending on the infection. Antibiotics can be given orally, intravenously (IV) or inhaled.

- Oral antibiotics - liquid or tablets to fight certain bacteria causing lung exacerbations
- IV antibiotics - liquid medication given directly into the bloodstream through an intravenous catheter. A hospital admission may be required to start the course. Sometimes the course is completed at home.
- Inhaled antibiotics - an aerosol or mist via a nebuliser such as TOBI or dry powder inhaler, such as Tobi podhaler, that acts on the airways directly.

Anti-inflammatories 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 Modifiers are designed to tackle the defective CFTR (Cystic Fibrosis Transmembrane Conductance Regulator) protein that regulates the flow of chloride and sodium (salt) in and out of the cells in the lungs and other organs.

- Kalydeco is a tablet that has been approved in Australia for some children and adults who have specific CF gene mutations. This drug helps the defective CFTR work at the surface of the cell to move salt and water into the airway making the mucus thinner and easier to cough out.
- Orkambi (Lumacaftor and Kalydeco) is a combination drug being trialled in some people with the F508del mutation. Its action is again to help the defective CFTR to work which helps thin the mucus in lungs and other organs. This is not yet approved for general use amongst those with CF.

Pancreatic Enzymes are used in those with CF who have pancreatic dysfunction. The enzymes improve food digestion and contain enzymes normally produced by the pancreas (lipase, protease and amylase). These enzymes are needed to digest fats, carbohydrates and proteins and the doses are different for each individual depending on body weight. Doses are sometimes adjusted depending on symptoms. Creon and Panzytrat are the most common enzymes used in Australia.

Salt requirements vary depending on CF symptoms. All those with CF lose large amounts of sodium and chloride (salt) in their sweat. The CF sweat gland is not able to absorb salt back into the blood and leads to a higher risk of dehydration. To prevent dehydration salt replacement is necessary. Salt supplements will be recommended by your dietician and it is recommended that you also add extra salt to your food.

Vitamins are needed for all 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 ADEK regularly as prescribed.

Gastric acid reducing medications for those with Gastro-Oesophageal Reflux (GORD), drugs called proton pump inhibitors (Nexium and Losec) reduce the production of acid in the stomach. This leaves little acid in the stomach juice so that if stomach juice backs up into the oesophagus it is less irritating and is not as painful.

Useful Resources

- CF Conference 2015 "Medicines for Children With CF" by Courtney Munro www.youtube.com/watch?v=g7QDLixfPhs
- Cystic Fibrosis Foundation: Drug Development Pipeline www.cff.org/Our-Research/Drug-Development-Pipeline/
- Pill Swallowing Factsheet www.cysticfibrosis.org.au/media/wysiwyg/CF-Australia/Fact_Sheets/CF_Aust_Fact_Sheet_Pill_Swallowing_Techniques.pdf

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