CHOC
Specialty Pharmacy Services

Treatment Overview for Cystic Fibrosis

When a child or adolescent is diagnosed with cystic fibrosis (CF), the entire family is affected. That's why at CHOC, we specialize in providing the very best CF treatment and care to ensure your child lives a long, healthy life.

CF is an inherited disease in which the body makes very thick, sticky mucus. The mucus causes problems in the lungs, pancreas and other organs. In people with CF, changes in the cystic fibrosis transmembrane conductance regulator (CFTR) gene prevent water from getting to certain cells. This causes the mucus in some organs to become thick and sticky.

While there is no cure for CF, the goals of treatment are to ease symptoms, prevent and treat complications, and slow the progress of the disease.

The main treatments of CF are:

- Airway clearance to help loosen and get rid of the thick mucus that can build up in the lungs.
- Inhaled medicines to open the airways or thin the mucus. These are liquid medicines that are made into a mist or aerosol and then inhaled through a nebulizer.
- Pancreatic enzyme supplement capsules to improve the absorption of vital nutrients. These supplements are taken with every meal and most snacks. People with CF also usually take multivitamins.
- An individualized fitness plan to help improve energy, lung function, and overall health.
- Medications called CFTR modulators to target the underlying cause of cystic fibrosis. There can be different kinds of changes to the CFTR gene and the medications that have been developed so far are effective only in people with specific changes

Patient Resources

CHOC Pediatric Pulmonology: www.choc.org/programs-services/pulmonology/cystic-fibrosis-center/
Cystic Fibrosis Foundation: https://www.cff.org/
KidsHealth: https://kidshealth.org/en/parents/cf.html