

Cystic Fibrosis (CF)

What is cystic fibrosis?

Cystic fibrosis (CF) is an inherited disease affecting 30,000 people of almost all races in the United States. In people with cystic fibrosis, glands in many parts of the body don't handle salt and water normally. Glands in the lung plug airways with thick mucus causing chronic lung infections, inflammation, and breathing problems. Digestive enzymes secreted by the pancreas aren't delivered to the intestines, and thick mucus in the intestines causes problems with growth and nutrition. Sweat glands in the skin lose too much salt causing problems with dehydration in hot weather.

How does a person get cystic fibrosis?

CF is a genetic disease, that is, a disease children are born with that is inherited from their parents. One person in every 30 in the United States has one CF gene. Each parent passes down a CF gene they inherited from their own parents. You need two CF genes to actually have the disease. Because each parent usually has only one CF gene, they don't have CF themselves, and usually don't know they carry the gene.

CF is not contagious, and it is not caused by anything the parents did or didn't do during conception or pregnancy.

How is CF diagnosed?

- Medical history: Doctors ask about problems with digestion, such as poor growth, abdominal pain and loose, oily bowel movements. They also ask about productive cough, wheezing, frequent lung or sinus infections, or difficulty breathing.
- Physical exam: Doctors look for signs of CF, including poor growth or weight gain, and noisy lungs.
- Laboratory tests: The sweat test, which looks for excessive salt in the sweat, is the standard test for CF. Doctors also look for abnormalities in the gene that causes CF, and look for bacteria in mucus that people with CF often have.

Not all people with CF have all the usual signs and symptoms, and some are sicker than others when they are diagnosed. Most are diagnosed during infancy, but some are not recognized until adulthood.

Can CF be cured?

There is no cure for CF at this time, but with treatment at a CF Center most people with CF grow up to lead active, full lives. Doctors, scientists, nurses and people with CF are working together to discover and test new treatments, and to find a cure.

How is CF treated?

CF centers across North America have specialists to treat all the problems caused by CF. Doctors prescribe digestive enzymes, vitamins and nutritional supplements to help dietitians promote normal growth. Doctors prescribe treatments to help respiratory therapists teach people with CF

how to keep mucus and infections from building up in their lungs. Social workers help families meet the challenges of caring for individuals with CF. Nurses work with families and other team members to make sure that each person with CF gets the treatments they need.

How can I find out more?

If you think you or someone you know has CF, ask your doctor about your concerns. The staff of the Children's Hospital and Health Center/University of California, San Diego Cystic Fibrosis Center can be reached at 858-966-6790. Call the Adult CF Program at 858-657-8628 for questions about adults with CF. The Children's Hospital and Health Center/University of California, San Diego Cystic Fibrosis Center has been accredited by the Cystic Fibrosis Foundation as a patient care, teaching, and research center, and has been designated a Therapeutic Development Center for research of new treatments. For more information, visit the Cystic Fibrosis Foundation website at <http://www.cff.org>.