

Cystic fibrosis

Cystic fibrosis (CF) is the most common life-threatening genetic disorder among Caucasians. It primarily affects the respiratory system (lungs), the digestive system (pancreas and sometimes liver) and the reproductive system.

When a person has CF their mucus glands secrete very thick sticky mucus. In the lungs, the mucus clogs the tiny air passages and traps bacteria. Repeated infections and blockages can cause irreversible lung damage and a shortened life.

The pancreas is also affected, preventing the release of enzymes needed to digest food. This means that people with CF can have problems with nutrition and must consume a diet high in kilojoules, fats, sugar and salts.

People with CF produce abnormal mucus

CF affects the exocrine glands, which secrete body fluids such as sweat, mucus and enzymes. People with CF produce abnormally thick, sticky mucus which blocks small air passages in the lungs. This causes difficulty clearing infections and can result in lung damage over a period of time.

A range of symptoms

People with CF may have the following symptoms:

- Persistent cough, with great physical effort
- Some difficulty breathing
- Tiredness, lethargy or an impaired exercise ability
- Frequent visits to the toilet
- Salt loss in hot weather which may produce muscle cramps or weakness
- Poor appetite.

How common is CF?

Cystic fibrosis is the most common genetically-inherited life-shortening chronic illness affecting young Australians today. A baby is born with cystic fibrosis every four days.

In Australia, one in 25 people are carriers of the CF gene. Carriers of the CF gene do not have any symptoms of the condition. If two people carry the gene and have a child, each pregnancy will have:

- A one in four chance that the child will have CF
- A two in four chance that the child will not have CF but will carry the gene
- A one in four chance that the child will not have CF and will not be a carrier.

One in every 2,500 births produces a child with CF. Approximately 3,000 people in Australia have CF.

CF is usually diagnosed at birth

In most Australian States all babies are screened at birth for CF through the newborn screening test. This involves collection of a blood sample. If the results of the screening test reveal very high levels of a substance called immunoreactive trypsin (IRT), CF is suspected and the DNA in the blood is then analysed for the most common mutation-causing CF.

A sweat test may be done to measure the amount of salt (sodium chloride) in the sweat and confirm the diagnosis.

Some babies may also be diagnosed shortly after birth as a result of an intestinal blockage called meconium ileus. Most babies who have CF are now diagnosed within the first two months of life.

Treatment aims to slow progress

Treatment for CF can be intensive and time consuming. At present there is no cure for CF. Treatment aims to slow progression of the condition and includes:

- Chest physiotherapy
- Antibiotics
- Inhalations via a compressed air pump and nebuliser
- Enzyme replacement capsules with meals and snacks
- A well balanced diet high in protein, fat and kilojoules
- Supplementary vitamins
- Salt supplements
- Regular exercise.

Regular attendance at a major CF clinic is beneficial and recommended.

CF gene testing before becoming pregnant

Carriers of a CF gene are healthy and often not aware that they are carriers. If you are planning a pregnancy you can now have testing to clarify your status for cystic fibrosis. Victorian Clinical Genetics Services has produced cystic fibrosis carrier testing packs.

Where to get help

- Your doctor or obstetrician
- Cystic Fibrosis Victoria offers a range of services for people living with CF, their families, friends and carers Tel. (03) 9686 1811 or 1800 633 685
- Cystic fibrosis clinics at Monash Medical Centre, The Alfred or the Royal Children's Hospital.
- Cystic fibrosis carrier screening program

Things to remember

- There is no cure for CF but treatment can slow progression of the disease
- One in 25 people carry the gene but will have no symptoms
- Cystic fibrosis is usually diagnosed at birth
- Cystic fibrosis is not contagious
- Cystic fibrosis occurs in males and females
- Testing before pregnancy can now identify if you are a CF gene carrier.

This page has been produced in consultation with, and approved by:

Cystic Fibrosis Victoria

Content on this website is provided for education and information purposes only. Information about a therapy, service, product or treatment does not imply endorsement and is not intended to replace advice from your doctor or other registered health professional. Content has been prepared for Victorian residents and wider Australian audiences, and was accurate at the time of publication. Readers should note that, over time, currency and completeness of the information may change. All users are urged to always seek advice from a registered health care professional for diagnosis and answers to their medical questions.

For the latest updates and more information, visit www.betterhealth.vic.gov.au

Copyright © 1999/2012 State of Victoria. Reproduced from the Better Health Channel (www.betterhealth.vic.gov.au) at no cost with permission of the Victorian Minister for Health. Unauthorised reproduction and other uses comprised in the copyright are prohibited without permission.