

Cystic fibrosis

Cystic fibrosis (CF) is the most common life threatening genetic disorder amongst 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?

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 using a heel prick test (Guthrie Test) which involves collection of a blood sample. If the results of the blood spot 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 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

Where to get help

- Your doctor
- Cystic Fibrosis Victoria Inc.
- CF Clinics at Monash Medical Centre, The Alfred or the Royal Children's Hospital.

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
- CF is usually diagnosed at birth
- CF is not contagious
- CF occurs in males and females.

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

Cystic Fibrosis Victoria

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