

## Lungs

People with CF produce abnormally thick, sticky mucus which blocks small air passages in the lungs, causing difficulty in clearing infections and can result in lung damage over a period of time.

## Digestion

The pancreas is a gland just below the stomach and one of its functions is to produce enzymes which break down food so that it can be absorbed by the digestive system. In CF the sticky mucus can make it difficult for the enzymes to reach the digestive system and as a result the food eaten cannot be fully digested. Children with CF may therefore have difficulty in gaining weight and the undigested food results in large bulky bowel actions.

## Sweat Glands

People with CF do not sweat more than other people but they do lose more salt and potassium in their sweat. The main diagnostic test for CF is the Sweat Test which measures the level of salt in the sweat.

## CF Associations

### Cystic Fibrosis Australia Inc.

51 Wicks Road, North Ryde NSW 2113  
PO Box 254, North Ryde NSW 1670  
phone (02) 9878 5250 freecall 1800 635 008  
facsimile (02) 9878 5058  
email [general@cysticfibrosisaustralia.org.au](mailto:general@cysticfibrosisaustralia.org.au)  
[www.cysticfibrosisaustralia.org.au](http://www.cysticfibrosisaustralia.org.au)

### Cystic Fibrosis NSW

51 Wicks Road, North Ryde NSW 2113  
PO Box 149, North Ryde NSW 1670  
phone (02) 9878 2075 facsimile (02) 9878 4890  
email [general@cysticfibrosisnsw.org.au](mailto:general@cysticfibrosisnsw.org.au)

### Cystic Fibrosis Victoria Inc

80 Dodds Street, Southbank VIC 3006  
PO Box 3036, South Melbourne VIC 3205  
phone (03) 9686 1811 facsimile (03) 9686 3437  
email [admin@cfv.org.au](mailto:admin@cfv.org.au)

### Cystic Fibrosis WA

PO Box 959, Nedlands WA 6909  
phone (08) 9389 8766 facsimile (08) 9389 8768  
email [info@cysticfibrosiswa.org](mailto:info@cysticfibrosiswa.org)

### Cystic Fibrosis Tasmania Inc

PO Box 245, Hobart TAS 7001  
phone (03) 6334 8754 facsimile (03) 6334 8755  
email [cftas@optusnet.com.au](mailto:cftas@optusnet.com.au)

### Cystic Fibrosis Queensland Ltd

31 Kate Street, Kedron QLD 4031  
PO Box 2245, Chermside Centre QLD 4032  
phone (07) 3359 8000 facsimile (07) 3359 3380  
email [admin@cysticfibrosisqld.org.au](mailto:admin@cysticfibrosisqld.org.au)

### Cystic Fibrosis SA Inc

143-145 Sturt Street, Adelaide SA 5000  
phone (08) 8221 5595 facsimile (08) 8221 5596  
email [cfsa@cfsa.org.au](mailto:cfsa@cfsa.org.au)

### CF Association of ACT Inc

PO Box 909, Civic Square ACT 2608  
phone / facsimile (02) 6259 7922

### Australian CF Research Trust

PO Box 254, North Ryde NSW 1670  
phone (02) 9878 5250 freecall 1800 635 008  
facsimile (02) 9878 5058  
email [general@cysticfibrosisaustralia.org.au](mailto:general@cysticfibrosisaustralia.org.au)

*CFA gratefully acknowledges the support of LJ Hooker franchises across Australia.*



## Question & Answer

# Understanding Cystic Fibrosis



## What is Cystic Fibrosis?

Cystic Fibrosis (CF) is an inherited recessive genetic condition which mainly affects the lungs, digestive system and the sweat glands. It is the most common life-threatening, recessive genetic condition affecting Australian children.

Cystic Fibrosis is a condition in which there is a considerable variation in the severity of symptoms. CF was first recognised as a specific condition in the 1930s at which time the outlook for babies born with the condition was not good. Today with earlier diagnosis, greater understanding of the condition, improvements in treatment and better management the majority of children live into adulthood.

## How does CF affect the body?

In CF there is a problem with the protein that controls the movement of salt in and out of the cells. Too much salt in the cells causes mucus to be very thick and sticky and to build up in organs like the lungs.

## What are the symptoms of CF?

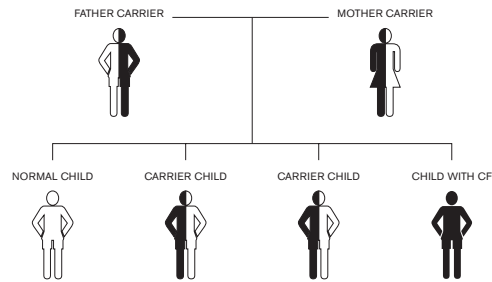
People with CF may have the following symptoms:

- Persistent cough, particularly with physical effort
- Some difficulty in breathing or wheezing with effort
- 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?

Amongst people of Caucasian ancestry 1 out of 25 are genetic carriers for CF. If two people are genetic carriers for CF and they have a child there is (with every pregnancy) –

- a 1 out of 4 (25%) chance that the child will have CF
- a 2 out of 4 (50%) chance that the child will be a genetic carrier
- a 1 out of 4 (25%) chance that the child will not have CF and will not be a genetic carrier for CF



In Australia there are approximately 80 babies born with CF each year. The incidence is 1 in every 2500 births. There are approximately 2500 people with CF in Australia. CF is most common amongst Caucasian races.

## How is CF diagnosed?

In Australia, all babies are screened at birth for CF along with several other conditions. A blood test is done 3-5 days after birth and if this test proves to be positive a sweat test will be done to measure the amount of salt in the sweat. Most babies who have CF are now diagnosed within the first two months of life.

## How is CF treated?

Treatment for CF can be intensive and time consuming. At present there is no cure for CF and treatment is therefore aimed at slowing progression of the condition.

### Lungs

Treatment for lung problems includes:

- chest physiotherapy
- antibiotics
- inhalations via compressed air pump and nebuliser

Most people will require this treatment on a regular daily basis and some may require aerosol medications to assist breathing.

Exercise is encouraged on a regular basis to maintain a healthy lung capacity. Sport and aerobic exercise are recommended for all age groups of people with CF.

### Digestion

To improve absorption of food most people with CF require enzyme replacement capsules with meals and snacks.

To improve nutrition and maintain body weight a well balanced diet high in protein, fat and calories is required.

Some people may also require supplementary vitamins.

### Sweat Glands

Salt supplements and additional fluids are required in hot weather, during strenuous exercise or in cases of fever.

Treatment for CF has improved greatly in recent years and advances continue to be made. Studies have proved that regular attendance at a major CF centre or clinic is beneficial. This enables people to experience the expertise of the CF team which includes doctors, physiotherapists, dietitians, social workers and nurses.

## COMMON QUESTIONS

### Is there a cure for CF?

No, not yet. CF is a life shortening condition and in the past most children with CF did not survive childhood. However today with improved treatment most people with CF are living into adulthood and leading normal and productive lives. At present there are promising research developments in the area of gene therapy.

### Is CF contagious?

NO. CF is a condition which is present at birth. Coughing is a frequent symptom of CF and some people worry they may "catch it". You cannot catch CF and you cannot give it to anyone else.

## Can CF be diagnosed before birth?

Yes, if there is already a child with CF in the family then testing can be carried out at 10 weeks of pregnancy. Parents of a child with CF are encouraged to have genetic counselling before planning a further pregnancy.

## Is CF carrier testing available?

Following the discovery of the CF gene in 1989 it is now possible to offer carrier testing. However, this is not a simple matter as whilst there is one most common gene for CF there are over 900 mutations of the gene. Because of this carrier testing is usually offered only to those people who have a family history of CF. However the availability of testing varies from state to state in Australia. Further information is available from CF Associations or genetic counsellors.

## Is diabetes related to CF?

A small percentage of people with CF may also develop diabetes (another function of the pancreas is the production of insulin) however this is not common before the teenage years.

## Will CF affect a child's schooling?

- CF does not impair intellectual ability in any way.
- Most students with CF have a good attendance record but will require time off for clinic appointments and possible hospital admissions.
- If you require more information please ask for the special pamphlet for teachers from your local CF Association.

