Understanding Cystic Fibrosis

A guide to CF
What is Cystic Fibrosis?

Cystic fibrosis (CF) is an inherited, genetic condition that alters the way salt is transferred in and out of cells. Too much salt in the cells causes mucus to be very thick and sticky and to build up in the organs. It mainly affects the lungs, digestive system and the sweat glands. It is the most common life-shortening, recessive genetic condition in Australia.

There are more than 1,000 mutations of the CF gene causing considerable variation in the severity of symptoms. CF was first recognised in the 1930s and at the time the outlook was very poor. Today, with earlier diagnosis, improvements in treatment, greater understanding, and better management most children live well into adulthood.

How does CF affect the body?

People with CF produce abnormally thick, sticky mucus which blocks small air passages in the lungs and other organs. This causes difficulty breathing and clearing infections and results in lung damage over time.

The pancreas is a gland just below the stomach. 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. As a result, people with CF may have difficulty gaining weight and the undigested food results in bowel problems.

People with CF do not sweat more than other people but they do lose more salt and potassium when they sweat. This increases the risk of dehydration and extra salt and fluid supplements may be necessary.
What are the symptoms of CF?

People with CF may have the following symptoms:

- Persistent cough, particularly following physical effort.
- Difficulty breathing.
- Tiredness, lethargy or impaired exercise ability.
- Poor growth rate, being underweight or a failure to thrive.
- Late onset of puberty.
- Salt loss and dehydration, especially in hot weather or after exercise.
- Poor appetite.
- Cramps, diarrhoea or constipation, wind and greasy stools.
- Some people with CF also develop specific types of diabetes, arthritis and brittle bones.
How is CF diagnosed?

In Australia, all babies are screened at birth for the most common forms of CF and several other conditions. A heel prick blood test is done three to five days after birth and if the test proves positive, a sweat test will be done to measure the amount of salt in the sweat. It’s this test that usually confirms the diagnosis. Most babies who have CF are now diagnosed within the first two months of life, however, mild or rare forms may not be diagnosed until later in life.

More than a million people carry the CF gene. Nationally that’s one in 25 Australians but the incidence is even higher in Tasmania where it’s one in 20 people. A baby is born with CF every four days and more than 3,000 people currently have a diagnosis. If two carriers have children every pregnancy has a:

- one in four (25%) chance that the child will have CF.
- two in four (50%) chance that the child will be a genetic carrier.
- one in four (25%) chance that the child will not have CF and will not be a genetic carrier for CF.
How is CF treated?

Living with CF is relentless and time consuming. There is no cure, yet, and treatment is aimed at slowing progression of the condition.

**Treatment for the lungs** includes:
- air-way clearance, including chest physiotherapy to clear secretions and build strength.
- antibiotics to fight infection.
- inhalations of medicines to thin the mucus and aid breathing.

**Daily exercise** is essential to maintain healthy lung capacity. Some people may require aerosol medications to assist their breathing during exercise. Sport and aerobic exercise is recommended for all age groups - even babies.

To improve the nutritional absorption from food most people with CF require enzyme replacement capsules with meals and snacks that contain fat, carbohydrates and protein.

To provide optimal nutrition and maintain body weight, a well-balanced diet high in protein, fat and calories is necessary. Many people will also require supplementary vitamins.

**Salt tablets and extra fluids** may be required in hot weather, during exercise or in cases of fever. Most people with CF have a reduced tolerance to heat as they lose a high level of salt through their sweat and are at risk of dehydration.
The treatment of CF has improved greatly in recent years and advances continue to be made. Studies prove that regular attendance at a CF centre or clinic is beneficial. Health and treatment plans are coordinated by expert, multidisciplinary CF teams which include doctors, physiotherapists, dieticians, social workers, nurses and pharmacists.

**COMMON QUESTIONS**

**Is there a cure for CF?**

No, not yet. But with an active, healthy lifestyle and medical treatment most people with CF live well into adulthood and have full, productive lives. Research is continuing and there are promising new developments in treatments and genetics.

**Is it contagious?**

No. You're either born with CF or you aren't. Coughing is a common symptom but isn't anything to be concerned about. You can't catch CF.

**Is CF carrier testing available?**

The CF gene was identified in 1989 and it's possible to test to see if you're a carrier. However, this is not a simple matter as there are more than 1,000 gene mutations. Carrier testing is usually only offered to people who have a family history of CF and availability varies from state to state. Further information is available from CF organisations and genetic counsellors.

**Can CF be diagnosed during pregnancy?**

Yes. If someone in the family has CF a test can be done at 10 weeks of pregnancy. Parents of a child with CF are also encouraged to have genetic counselling before planning another pregnancy.

**Will CF affect a child's schooling?**

CF does not impair intellectual ability. Most students with CF have good school attendance but can require time off for clinic appointments and hospital admissions. Exercise is important so children with CF should be encouraged to participate in sport as well as all the other school activities.
CF Organisations

National Cystic Fibrosis website
www.cysticfibrosis.org.au

Cystic Fibrosis Australia
Rose Cottage
2 Richardson Place
North Ryde NSW 2113
PO Box 268
North Ryde NSW 1670
T: +61 (02) 9889 5171
F: +61 (02) 9888 6276
Freecall: 1800 232 823
E: general@cfa.org.au

Cystic Fibrosis ACT
3/30-36 Woolley Street
Dickson ACT 2602
PO Box 909
Civic Square ACT 2608
T: +61 (0)4 3748 5454
E: info@cfact.org.au

Cystic Fibrosis NSW
Unit 46, 11-21 Underwood Rd
Homebush, NSW 2140
PO Box 4113
Homebush South
NSW 2140
T: (02) 8732 5700
F: (02) 8732 5799
Freecall: 1800 650 614
E: general@cfnsw.org.au

Cystic Fibrosis Victoria
80 Dodds Street
Southbank VIC 3006
T: (03) 9686 1811
F: (03) 9686 3437
Freecall: 1800 633 685
E: admin@cfv.org.au

Cystic Fibrosis WA
‘The Niche’, Suite C
11 Aberdare Road
Nedlands WA 6009
PO Box 959
Nedlands WA 6909
T: (08) 9346 7333
F: (08) 9346 7344
Freecall: 1800 678 766
E: info@cysticfibrosiswa.org

Cystic Fibrosis Tasmania
Level 2, 38 Montpelier Retreat, Battery Point
TAS 7004
GPO Box 245
Hobart TAS 7001
T: (03) 6234 6085
Freecall: 1800 232 823
E: general@cftas.org.au

Cystic Fibrosis Queensland
31 Kate Street
Kedron QLD 4031
PO Box 2245
Chermside Centre
QLD 4032
T: (07) 3359 8000
F: (07) 3359 3380
Freecall: 1800 670 990
E: admin@cfqld.org.au

Cystic Fibrosis SA
143 Sturt Street
Adelaide SA 5000
T: (08) 8221 5595
F: (08) 8221 5596
Freecall: 1800 232 823
E: cfsa@cfsa.org.au

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