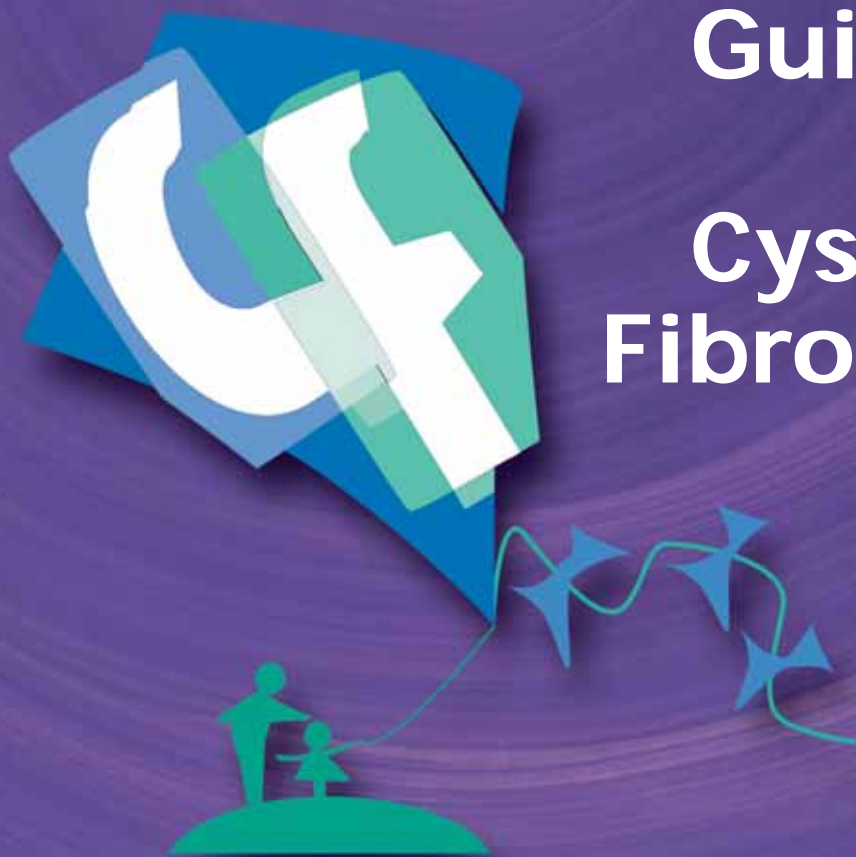


The Cystic Fibrosis Association of Ireland

Your Guide to Cystic Fibrosis



What is Cystic Fibrosis?

Cystic Fibrosis (CF) is an inherited disease, affecting mainly the lungs and digestive system. It is Ireland's most common life threatening genetically inherited illness.

Approximately 1 in 19 people are carriers of the CF gene giving people a 1 in 4 chance of having a child with CF.

CF affects the secretory glands, damaging many organs including the lungs, the pancreas, the digestive tract, the liver and the reproductive system. It causes a build up of mucus to be produced, blocking the bronchial tubes and preventing the body's natural enzymes from digesting food.

CF affects primarily the lungs and digestive system. A build up of mucus can make it difficult to clear bacteria which leads to cycles of lung infection and inflammation. Mucus can also block the ducts of the pancreas making it difficult to digest and absorb adequate nutrients from food. The result is that people with CF can be prone to constant chest infections and malnutrition.

Persons with CF must consume pancreatic enzymes with food to absorb nutrients and must also perform daily chest physiotherapy to ensure they stay fit and healthy.

Not all children are affected in the same way or to exactly the same degree– some are affected more or less than others.

As a parent of a child with CF you will meet many people involved in the care of children with Cystic Fibrosis but you will be carrying out some of the treatment yourself. To do this effectively, you will need to understand as much as you can about the illness.

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This booklet has been written to assist you.

It is not intended to replace any advice you may receive from your doctor or CF Clinic.

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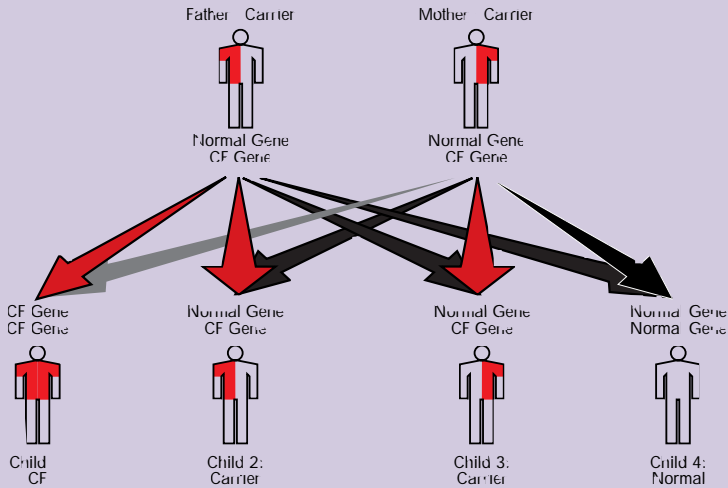
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Why does my child have CF?



A baby may be born with CF only if BOTH parents are carriers of the recessive Cystic Fibrosis gene

If both parents are carriers, a child has:

- a one in four chance of being born with Cystic Fibrosis
- a two in four chance of being a carrier but not having the disease
- a one in four chance of not having the condition or being a carrier of the faulty CF gene.

All this means that CF is a *genetic* disorder. Most carriers of the defective *gene* have no idea that they are carriers, because they are completely healthy. Most people find out only when they

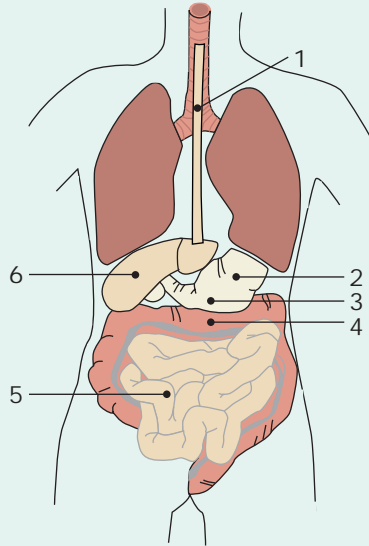
have a child with CF, or a close relative is affected and they are tested for the genes.

In Ireland, one person in 19 is a carrier and one in approximately every 2,000 babies will have CF.

The small family illustrated above is imaginary and shows that there's a 1 in 4 chance of having a CF child and, therefore, a 3 in 4 chance of having an unaffected child (carrier or child without CF). But each baby is conceived separately (apart from identical twins), so the risks are exactly the same each time and what happened in the last pregnancy doesn't increase, or decrease, the risks next time. They'll be exactly the same.

How does CF affect the digestive system?

The pancreas is a gland in the abdomen; one of the functions of the pancreas is to produce digestive juices, or enzymes, which pass into the intestine, where they help us digest and absorb the food we eat.



In those with CF, the small channels down which the digestive juices flow become blocked with *mucus*. The *enzymes* then build up in the pancreas, which becomes inflamed. This causes the formation of *cysts* and *fibrosis* = CYSTIC FIBROSIS. The effect of CF on the *pancreas* and digestion varies very much between people. At one extreme, some newborn babies cannot digest milk and fail to gain weight and have loose stools.

Children with CF need a higher intake of calories and protein than children without CF.

1. Gullet (oesophagus)
2. Stomach
3. Pancreas (behind the stomach)
4. Large intestine
5. Small intestine
6. Liver and gall bladder

When it comes to nutrition, it is necessary for parents to consult their CF clinic and dietician to construct an effective dietary plan for their children.

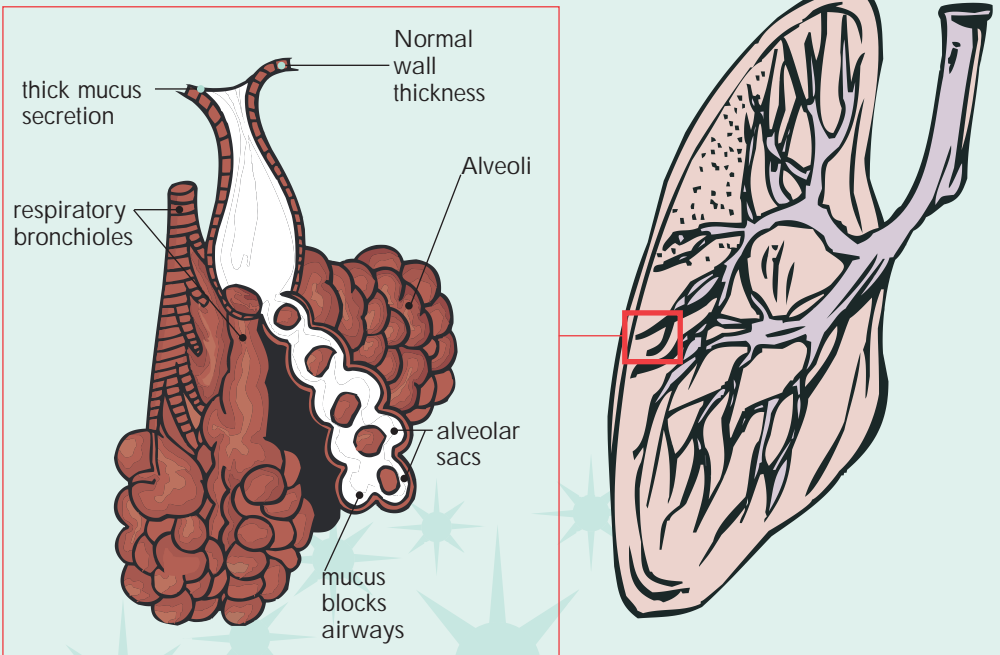
A high fat, high calorie diet is necessary to keep well nourished – your dietician will be able to give you relevant recipes for nourishing meals.

How does CF affect the chest?

We all have *mucus* in our lungs but in children with CF the *mucus* produced is abnormally thick. This can block some of the smaller airways and this leads to infection and, later, damage to the lungs.

In the early years the infections are usually caused by viruses and certain bacteria eg *Staphylococcus aureus* and *Haemophilus influenzae*. Later on, infections are caused by other bacteria called *Pseudomonas aeruginosa*.

Much of the damage caused can be prevented by early treatment and physiotherapy.



In cystic fibrosis, thick mucus blocks the narrow airways of the lungs

How CF is treated?

- Physiotherapy

The main aims of physiotherapy are twofold:

To keep the lungs clear

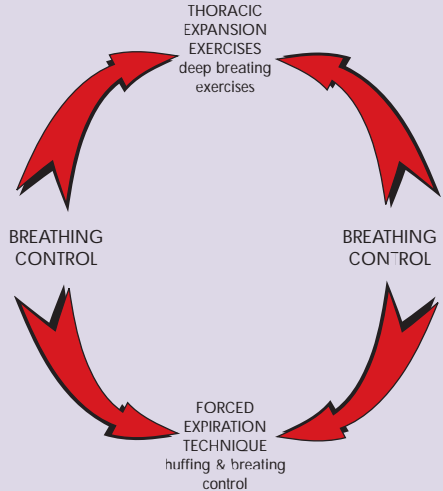
To maintain and improve aerobic fitness

Keeping the lungs clear:

Airway clearance techniques are performed daily to keep the lungs as clear as possible, and so minimise the risk of a chest infection developing. The range of techniques used is wide and varied, which allows treatment programmes to be tailor-made to suit the age and specific needs of each person. Techniques which may be used include Active Cycle of Breathing Technique (A.C.B.T.), Autogenic Drainage, PEP mask, Flutter, Acapella, positioning and percussion, among others.

Maintaining and improving aerobic fitness:

While airway clearance techniques work more specifically on clearing secretions from the airways, general fitness must also be addressed. Exercise plays an important role in keeping the lungs and heart healthy and strong. Activities such as bouncing on a trampoline, cycling, running or swimming are among those which can be fun while also being of benefit to the lungs.



- Antibiotic Treatment

Antibiotics can be administered in three ways

1. Orally (i.e. tablet form)
2. Nebulised (i.e. aerosol inhaler)
3. Intravenously (I.V.) (through a vein)

Intravenous antibiotic treatment can be administered at home provided that your doctor and family are comfortable doing this. Taking home IV's may lessen disruption to one's lifestyle while still applying the full rigour of a hospital treatment. Home IV's may also reduce the risks of cross infection.

When Should I take my child to see a Doctor?

The answer is a simple one – when you are worried about him or her. The first port of call should be contacting your local GP - your local GP may identify the problem and contact the CF team accordingly. The members of the CF team (i.e. Doctor or Nurse) can guide your GP on antibiotic treatment and on the most recent sputum result and sensitivities. Your GP may ask you to attend your CF clinic if he/ she has further concerns.

It is recommended that all people with CF have regular contact with the staff of a CF Clinic, visits every three months are the norm.

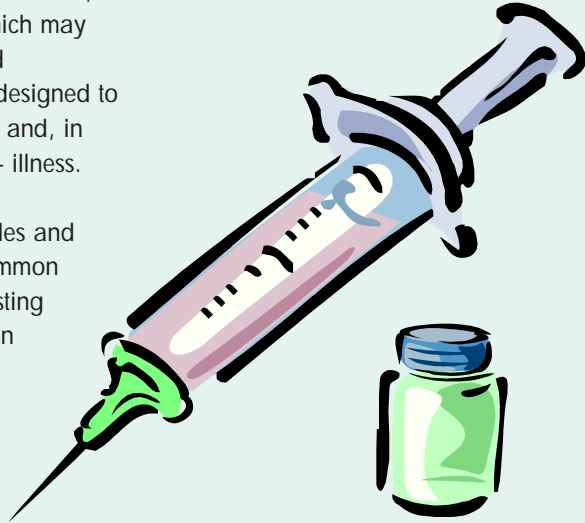
Here are some signs to watch out for:

- cold symptoms
- increased or frequent cough
- increased sputum
- increase or change in colour of sputum
- breathlessness
- fever
- decreased or poor appetite
- weight loss
- tummy aches
- frequent or loose stools
- vomiting
- decreased ability or unwillingness to exercise

Are immunisations important?

Children with CF are particularly at risk from the common childhood diseases, especially those infections which may affect the lungs. The standard immunisation programme is designed to protect babies from serious – and, in some cases, life threatening – illness.

Some illnesses, such as measles and whooping cough, are still common and may have severe and lasting effects on the lungs of children with CF. They must be protected at an early stage, before they come into regular contact with other children at nursery or school. Flu can cause an especially nasty illness in children with CF and it is recommended that every child over six months old is immunised each year at the beginning of the winter season. Children with CF respond just as other children to immunisation and are no more likely to have reactions to the injections. The normal immunisation schedule is appropriate for children who have CF and injections should be postponed only in very exceptional circumstances and after consultation with your clinic. Just



having a cough or cold is not enough reason to delay having an immunisation. There are very, very few medical reasons to avoid immunisation.

Can CF affect other parts of the body?

Yes – but the degree of effect varies widely from person to person.

Ears, nose and sinuses

People with CF can be prone to *sinusitis* and hay fever, which may need to be treated with nasal sprays or antibiotics. Some older children and adults can have nasal *polyps*, which may need to be removed by a small operation if they become troublesome.

Liver

Some people with CF get *cirrhosis*. All children and adults will have their liver function tests checked annually in the form of a blood test in all CF centres.

Diabetes mellitus

Diabetes mellitus can occur in adults with CF and results in an abnormally high level of sugar in the blood. Treatment is by some alterations to the diet and injections of insulin.

Asthma

About 30% of children with CF wheeze from time to time. This happens when muscles surrounding the small airways contract and cause them to narrow slightly. The children find it more difficult to catch their breath and often have a feeling of tightness in their chest. Wheezing responds well to medicines used for children with asthma.

Joints

Some older children develop a form of arthritis, usually in one or two large joints, such as the knee. In most cases this improves with time and treatment.

Delayed Puberty

In some children with CF, particularly those who are underweight, puberty is often later than usual but children do develop normally in time.

The Future

Since CF was first recognised, many new treatments have been developed to help people with CF live normal and active lives. Medical science research has greatly improved the treatment of CF thereby increasing life expectancy. Research is ongoing in the endeavour to find a cure for CF.

The future is without doubt an optimistic one and we must ensure that until a cure is found that all children and adults with CF are given optimum care and encouragement to fulfil their lives.

Glossary

Alveolus (Alveoli) - The specialised part of the lung where oxygen can enter the blood and carbon dioxide can leave.

Bronchus (Bronchi) - Small airways in the lung.

Cirrhosis - A term which is used in a general sense to mean progressive fibrous tissue overgrowth in an organ.

Cyst - A fluid or air filled space. In CF these are usually in the pancreas or lung.

DNA - The commonly used abbreviation for deoxyribonucleic acid, the principal molecule carrying genetic information.

Enzyme - A chemical that can help another one to change in some way. In CF this usually refers to digestive enzymes which digest food before it can be absorbed and used by the body.

Fibrosis - Fibrous tissues is useless "gristle" which replaces normal tissue when it is damaged. In CF this occurs in the lungs and pancreas.

Gene / Genetic - Every cell has thousands of genes which are made up of DNA (*see above*) and are passed on from parent to child. Genes are responsible for a person's individual characteristics such as eye colour, blood group and whether or not they have certain genetic diseases such as CF.

Haemophilus - Bacteria commonly known as Haemophilus influenza which causes respiratory tract infections.

Haemoptysis - Coughing up blood

Intravenous - Sometimes antibiotics or other medicines are given into a vein rather than by mouth. If this is needed for a course of treatment a small plastic cannula (tube) can be left in the vein so that the drug can be put in through it

rather than a fresh injection each time. There is also a cream available to numb the skin before the cannula is put into the vein.

Meconium Ileus - An obstruction of the small intestine at birth.

Mucoviscidosis - Another name for CF, literally it means that the mucus is thick or viscid. It may be understood in other countries.

Mucus - A slimy fluid secreted by mucous membranes. Mucus lubricates and protects parts of the body particularly the lungs and digestive system.

Nebuliser - A small machine which converts a liquid medication to a fine mist which can be breathed in to work directly in the lungs.

Paediatrician - A doctor who specialises in the treatment of children.

Pancreas - A gland which lies behind the stomach and makes digestive juices or enzymes and insulin.

Polyps - A small growth of mucous membrane that can grow from the sinuses on the lining of the nose.

Pseudomonas Aeruginosa - A bacteria infection that can affect the lungs.

Sinusitis - Inflammation of the membrane lining the facial sinuses (the air-filled cavities in the bones surrounding the nose).

Sputum - Mucous material produced by the cells lining the respiratory tract.

Staphylococcus Aureus - A bacteria infection that can affect the lungs.

BOOKLETS

THE FACTS & FINDING OUT

An introduction to the causes and effects of Cystic Fibrosis. A guide for parents of newly diagnosed children with Cystic Fibrosis.

NUTRITION

Eating well with Cystic Fibrosis –
A guide for children and parents.

TREATMENT

Physiotherapy for Cystic Fibrosis. Illustrates methods of physiotherapy in CF.

RIGHTS & ENTITLEMENTS

A guide to rights and entitlements for people with CF and their families.

LEAFLETS

CFAI GENERAL LEAFLET

Outlines the structures and help available from the Cystic Fibrosis Association.

CYBER CAMPUS LEAFLET

Description of online computer course for young adults with CF.

THE FACTS

General introduction to causes and effects of CF.

MAGAZINES

ANNUAL NEWSLETTER

Yearly magazine which looks at the work and achievements of the Association.

FUTURE FORCE

Magazine designed and written by CF adults covering large range of topics and issues affecting CF adults.



Further enquiries about literature, including booklets produced by the Association can be obtained from

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