CYSTIC FIBROSIS TRUST

CYSTIC FIBROSIS & YOU

A guide for children with CF aged 8-12 years

Cystic Fibrosis Trust. Registered Charity No. 1079049. Registered Company No. 3880213.

The Cystic Fibrosis Trust funds medical and scientific research aimed at understanding, treating and curing Cystic Fibrosis. It also aims to ensure that people with Cystic Fibrosis receive the best possible care and support in all aspects of their lives.

Contact details for information and other literature published by the Cystic Fibrosis Trust are given on the back cover.

Published by the Cystic Fibrosis Trust with a grant from Marks & Spencer.

November 2000

CYSTIC FIBROSIS TRUST

CONTENTS

This booklet has answers to some of your questions about Cystic Fibrosis, or for short "CF" – we use both names in this booklet. It is based on one written by the Cystic Fibrosis Association of New South Wales (Australia) for children with CF aged between 8 and 12 and their parents.

When you were young, your parents had to take charge of finding out about CF and giving you the right treatment. Now you are older, you will want to find out for yourself and be responsible for your own treatment. Cystic Fibrosis is your illness and you have a right to know as much as possible about it, what treatment is available, why you need it and what happens if you do not have it.

It is not possible to answer all your questions. These are some of the people you could contact if you want to find out more –

- From your CF Clinic: your doctor, nurse, social worker, physiotherapist or dietitian.

- From the Cystic Fibrosis Trust: Support Service. Our address and telephone is given at the back of this booklet.

What is Cystic Fibrosis (CF)?

CF is a disease which affects how your lungs and digestive system works.

Why do I have CF?

You were born with CF. By chance, you received two copies of the 'CF gene' from your parents – one copy from your mother and one from your father.

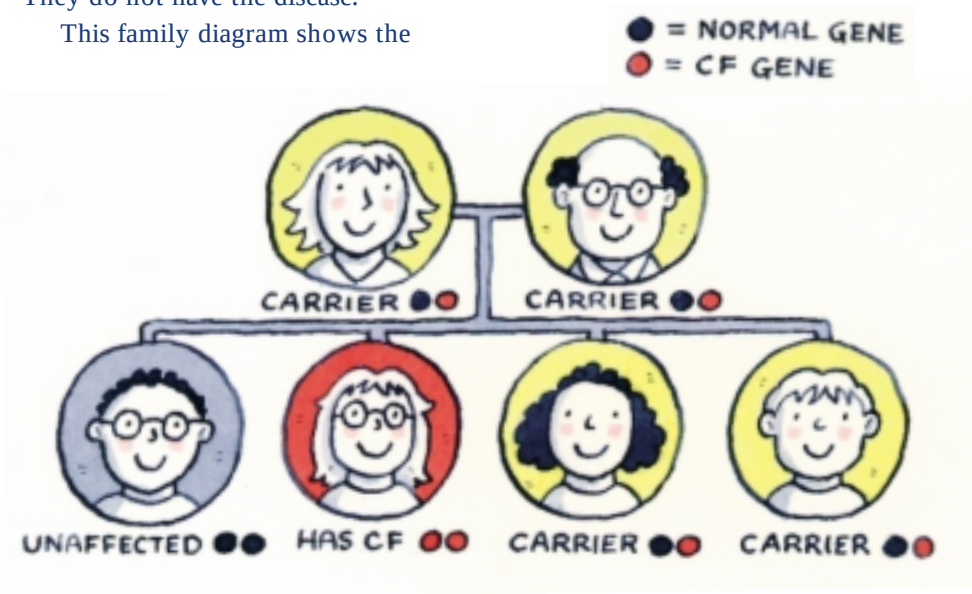
Genes are chemical instructions passed onto us by our parents. They control how our bodies develop. Some genes control whether we have blue eyes, some whether we have brown hair and some whether we have certain diseases – like CF. People who have only one copy of the 'CF gene' are called carriers. They do not have the disease.

This family diagram shows the chances of being born with CF. Both the mother and the father are carriers of CF with one copy each of the 'CF gene'.

Their genes are passed onto their children by chance. One child did not get any copies. Two children got one copy each and are carriers like their parents. The other child got two copies and has Cystic Fibrosis.

In every pregnancy where both parents are carriers of the 'CF gene' there is a one-in-four chance that their child will be born with Cystic Fibrosis.

In the UK, about one person in 25 carries the CF gene. Scientists now know what the gene looks like, so people can be tested to see if they are carrying it.



How can I be sure that I have CF?

Have you noticed how your skin tastes salty, especially when you have been working hard? Well, this is normal for people with CF. They have more salt in their sweat than other people.

Your doctor will have given you a special test. This measures the amount of salt in your sweat and is called the SWEAT TEST. The sweat test shows whether people have CF. You may also have had a test to find out what sort of CF gene you have.

Can CF be cured?

Unfortunately, not at the moment. But doctors all over the world are working on ways to find a cure and on better treatments to fight Cystic Fibrosis.

Your doctor will know what the best treatment is for you. Everyone is different, so your treatment may not be the same as someone else's. It might also have to change from time to time as you change.

Can I lead a normal life?

Yes. You should be able to do what most other children your age can do.

You just have to remember three things:

- Physiotherapy and exercise
- Enzymes
- Antibiotics

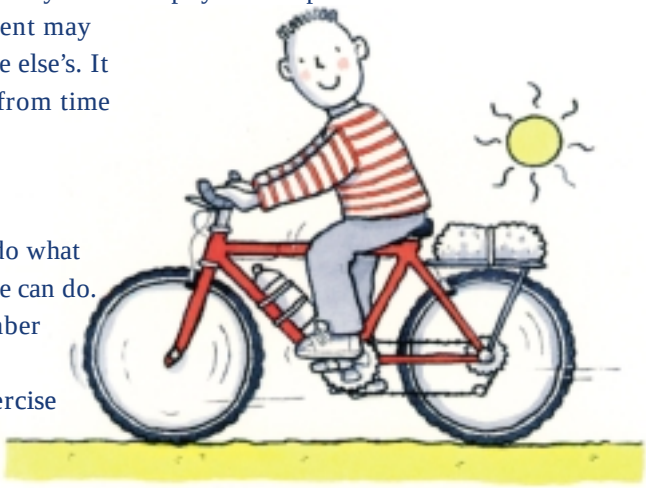
Why do I need chest physio?

People with CF have extra thick and sticky mucus. You need to keep your lungs clear of mucus to stop infections.

The best ways to keep your lungs clear and healthy are:

- Daily physiotherapy such as breathing exercises and huffing to help clear mucus.
- Inhalations to help open the tiny air passages in the lungs and to loosen mucus.
- Exercising as much as possible such as playing sport, riding your bicycle.
- Staying away from smoky and dusty places and unnecessary exposure to "colds".

If you would like to know more about your lungs, or different ways of clearing the mucus, ask your physiotherapist at the CF clinic.



Why do I need antibiotics?

The sticky mucus in the air passages in the chest are easily infected with germs. People with CF have difficulty clearing these infections without the help of antibiotics to kill the germs and physiotherapy to clear the mucus.

Why do I have to take enzymes?

An enzyme is a chemical which breaks down food into the fuel your body needs for growth and energy. The enzymes which help break down fats, proteins and carbohydrates are produced in the PANCREAS.

When you have CF, the tiny tubes in the pancreas can get blocked with mucus. This stops the enzymes getting to where they are needed.

The enzymes you take as capsules during meals make up for this. They make sure your body uses the food you eat to help you grow, give you energy, keep you fit and reduce "tummy aches".

What happens if I don't take my enzymes?

- Your body won't be able to use the food you eat; meaning you will not be properly nourished.
- You will not have the energy to do the things you want to do.
- You will get a pain in your stomach.
- You will have to go to the toilet a lot.

Can I eat a normal diet?

Yes. You should be able to eat the same food as anyone else, although you will need to eat more. That's because even with the enzymes your food isn't being digested as efficiently as theirs and you will probably need more energy than they do. Just remember, the more fatty food you eat, the more enzymes you may need. So talk to your doctor and dietitian at your CF clinic about the best diet for you and exactly how many enzymes you need.

Do I need extra salt?

When it is very hot, you may lose more salt in your sweat than other people. Salt tablets are not usually necessary but are advised in hot climates or if you are undertaking severe exercise in hot weather in this country. Your doctor will advise the dose.

Why do I need regular check-ups?

To make sure you are well. It is easier to treat an infection when it begins rather than to wait until it starts to make you ill.

When you have a check-up, you will have a throat swab culture to identify any infection, a lung function test ("blows") or a sputum test.

Lung-function tests check how well your lungs are working. If they are not working properly, your doctor will tell you what you can do to help them work better.

Throat cultures and specimens (your "spit") are tested for germs that cause infection. This helps your doctor know what antibiotic treatment you need to clear the germs.

Why might I need to go into hospital?

Sometimes your doctor will decide that you need special treatment that can only be given in hospital.

Why do I get tired easily at times?

Your body has to work harder than other people's bodies to keep you feeling healthy and fight off infections.

Sometimes it cannot keep up with this work and you will feel tired. This might happen if you:

- have an infection
- are not digesting enough food
- are very busy
- are doing a lot of exercise

Why do I need exercise?

You are more likely to get lung infections than other people. To make sure you have a good supply of oxygen to keep you healthy, you need to do exercises which get your lungs and heart working.

Good exercises are running, skipping, swimming, cycling… Choose those that suit you best.

By being fitter, you will feel great when you are well and your body will cope better when you are sick.

Will I miss much school?

No, you should not.

If you know you are going to be in hospital, it is a good idea to talk to your teacher about it so you can take work with you.

If you are away from school for a while, there are teachers in hospital and even some who can help you at home.

What can I do if people make rude comments?

Some people do not try to understand. They might call you names or give you a hard time because you often have a bad cough or have to take pills.

If you are having trouble – and there is no reason why you should be – talk it over with your family or friends or someone else that you feel comfortable with.

Should I tell other people that I have got CF?

Everyone is different, so this has to be up to you. Some people find it helps to talk to one or two close friends about their Cystic Fibrosis.

If you are not sure, talk about it first with your:

- Family
- Doctor
- CF Nurse
- Social Worker
- Teacher
- CF Trust Support Service, where there are staff you can talk things over with in private.

Maybe you could give your friends this book to read.

Why do I sometimes feel angry or depressed?

It is normal to feel depressed or angry from time to time. With CF, what sometimes gets you down is:

- thinking that people are rejecting you because of your CF; you feel alone and even sometimes that you do not need friends
- being completely fed up with having to do your treatment every day and sick and tired of your parents nagging you to do it
- feeling there is no point in making plans for the future
- a really bad spell of being ill
- when you want to do something special and having CF gets in the way.

You will not be the only one feeling down, but if it keeps on, try finding someone you can talk to. It could be your parents, a brother or sister, a close friend, staff at the CF Clinic or at the CF Trust's Support Service, or someone at school.

What about death?

Cystic Fibrosis is one of those subjects that makes a good news story, often with pretty dramatic headlines. When these stories come out and other kids who know you have got CF read them or see them on television, there are bound to be comments. So what are you to make of it?

Remember, news stories have to be unusual or they would not be news. CF affects people in a lot of different ways. Some have it severely, but many have it mildly or moderately but the papers do not write about them. No one can tell you how long you are going to live. Treatment is getting better and people are living longer and longer.

Do not, whatever you do, think to yourself, "Oh well, I will not be making it to 40, 50, 60, or 70, so what is the use of bothering…"

If this really does bug you, find someone to talk to NOW!

Some questions for the future.

Can I smoke?
Are you serious?! Smoking is bad for everyone. For someone with lung problems, the answer is definitely NO!

What about drinking alcohol?
Drinking too much alcohol is bad for anyone. For people with CF, there is also the matter of your liver and how well it is working. Some people with CF have liver problems. Others could be taking medicines that should not be mixed with alcohol – always check with your doctor.

Can I take any job?
Within reason. What is important is to make sure you are not working where, for instance, there is a lot of air pollution from dust and smoke. To be safe, talk it over with staff at the CF clinic.

Will I be able to have children?
Fertility varies from person to person, as it does for everyone else.

Women with CF may be slightly less fertile. However, several have had children. It is important that women with CF thinking of having a baby should check that they are healthy enough to do so. Your doctor will be able to give you advice when the time comes.

Men with CF are often not fertile. Sadly, the tubes that the

sperm goes down are not formed properly. This does not mean problems with having sex, but having children can be difficult for many. However, there are new techniques being developed to allow some men with CF to have children.

Will my children have CF?
They will carry the 'CF gene'. Only if your partner also carries the 'CF gene' will your children have a very high chance (1 in 2) of having CF.

Before having a baby your partner should be checked to see if he or she is a carrier.

Further information

In this booklet we have been able to answer only some of the questions people often ask. However if you feel you would like to talk with someone or that you still have some unanswered questions you can speak to staff, in private, at the CF Trust's Support Service by telephoning 020 8464 7211.

The CF Trust Support Service has a range of information leaflets on Cystic Fibrosis and their trained staff will be pleased to hear from you about anything you may be unsure of.

Staff at your CF Clinic will be able to offer you support and help – so why not make a note now of your contacts, either at the CF Trust or your CF Clinic.

I N F O R M A T I O N

For further information and literature published by the CF Trust, or how to make a donation, please contact:

Cystic Fibrosis Trust
11 London Road
Bromley
Kent BR1 1BY

Tel: 020 8464 7211
Fax: 020 8313 0472

enquiries@cftrust.org.uk

www.cftrust.org.uk