

FINDING OUT

A guide for parents of newly diagnosed children with Cystic Fibrosis

Published by the Cystic Fibrosis Trust (updated September 2010) with assistance from Dr James Littlewood OBE, Dr Michael Green and Dr Wendy Stannard.

This booklet has been written to help you to get a better understanding of Cystic Fibrosis . It is not intended to replace any advice you may receive from your Specialist CF Centre or CF Clinic.

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.

*Any words appearing in bold italic type in this factsheet can be found and are explained in the **glossary** (page 19).*

© Cystic Fibrosis Trust 2010. This factsheet may be copied in whole or in part without prior permission being sought from the copyright holders, provided the purpose of copying is not commercial gain and provided due acknowledgement is given.

www.cftrust.org.uk

11 London Road, Bromley, Kent BR1 1BY Tel: 020 8464 7211 Fax 020 8313 0472 enquiries@cftrust.org.uk

What is Cystic Fibrosis?

Cystic Fibrosis (CF) is an inherited disease, affecting mainly the lungs and digestion.

Normally, the liquids and mucus that line many of our organs are clear, lubricating and helping to protect them from infection. In CF there is a fault in a *gene* (the 'CF gene') that controls the amount and composition of fluid lining the airways and other organs. The fluid lining the airways is reduced, resulting in an excess of sticky secretions that are prone to infection and difficult to cough up. In the pancreas, the sticky secretions block the flow of digestive juices into the gut leading to impaired digestion and poor absorption of food.

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

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 asked to carry out some of the treatment yourself. To do this effectively, you will need to understand as much as you can about the disease.

All the questions, which are answered in this factsheet, have been asked by other parents coming to terms with the news that their child has Cystic Fibrosis.

How do you feel?

If you have just been told that your child has CF, this has probably come as a considerable shock. You may well be feeling a sense of loss or grief. These reactions are quite normal and other parents have experienced them before you. You may find it helpful to find people to talk to about CF, perhaps a member of the CF team at the hospital, the Cystic Fibrosis Trust or other families. Make sure you talk to people who have up to date knowledge of CF, as treatment has improved markedly over recent years and out of date information could give you an entirely wrong impression.

Disbelief

Many parents find it difficult to believe that the diagnosis is correct, especially if the child seems well and the diagnosis has been made through *screening* in the newborn period. It is important at an early stage to be referred to a Specialist CF Centre to have the diagnosis confirmed and where CF can be fully explained. Write down questions you want to ask as you think of them in case you forget later.

Anger and blame

When you first hear the diagnosis you may be angry, you may feel guilty or you may want to blame somebody. Remember – no-one is to blame. These feelings are understandable but don't help.

Strained relationships

There can be few greater strains on a relationship than having a child with a chronic illness which will be with him or her and you for life. Make sure that you discuss everything with your partner, try to avoid misunderstandings and don't be frightened to seek help from others if conflicts arise. It is much easier to be open with family members and friends than to hide your fears and worries.

Bewilderment

You will be given a huge amount of information and advice from various sources – some of it will be conflicting. Cystic Fibrosis is a complicated condition and each child is affected slightly differently, so everyone's experience differs somewhat.

You cannot expect to know everything about the disease immediately and no-one expects you to. It will be a long time before you understand CF in general and only experience will tell you exactly how it is affecting your child. It is useful to remember that CF affects only about 1 in every 2,500 children born in this country, so some ***health-care professionals*** will have little experience of it – in some cases, much less than you will.

Never be afraid to ask questions and try not to be frustrated if the person you are talking to appears to know less than you. Even among the 'experts', opinions can differ quite widely about various aspects of treatment and how important they are.

It is important that if something is worrying you, you ask someone for help. Never wait for things to get worse.

Positive approach

It is natural to feel especially close to a child with an illness. It is important, though, to try not to over-protect your child – remember that they are normal children who happen to have Cystic Fibrosis. Consequently they will be naughty sometimes, just the same as other children. There is no reason to treat them differently in relation to behaviour, education or even most physical activities. If you do treat them differently, you will not only be doing them a disservice but may also be creating problems for yourselves as parents in the long run. Children with CF want to be the same as other children, not different.

The life expectancy and lifestyle for children with CF has improved beyond recognition in recent years. With the latest advances in research, there is every chance that new therapies will be developed to further limit the harm from the disease.

It is important not to forget the impact there may be on any brothers and sisters of the child with Cystic Fibrosis. They are just as likely to be upset and anxious and may even feel either guilty, because they are well, or left out because of all the attention the child with CF is receiving.

Getting help

As with all things, life is easier if there is someone or somewhere to turn to for practical advice, emotional and financial support. That is exactly where the CF Trust comes in. It is a source of information and experience which parents can draw on about any aspect of life with Cystic Fibrosis.

Cystic Fibrosis affects the whole family including brothers, sisters, aunts, uncles, grandparents and other relatives. They are all welcome to use the facilities and resources of the Cystic Fibrosis Trust. Contact details are shown on the back page.

Why does my child have CF?

CF is a **genetic** disease. A baby may be born with CF only if BOTH parents are carriers of the faulty Cystic Fibrosis **gene**. Even then both parents having the faulty CF **gene** won't automatically mean that every baby they have will have Cystic Fibrosis.

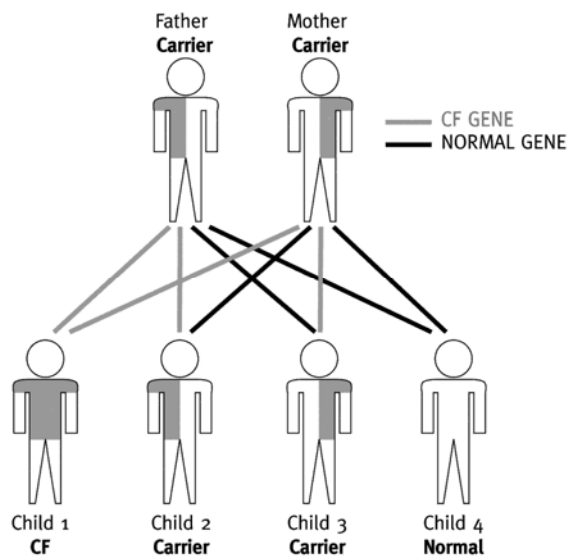
If both parents are carriers, a child has:

- 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 being completely free of it, i.e. not having CF nor being a carrier of the faulty CF **gene**.

Most carriers of the faulty **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
- they are identified in a **screening** programme during pregnancy.

In the UK, one person in 25 is a carrier and one in every 2,500 babies will have CF – which means that there is a baby born with CF nearly every day.



The diagram opposite shows that if both parents are carriers there is a one in four chance of having a child with CF and, a three in four chance of having an unaffected child (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. The chances are the same for each pregnancy.

Two carrier parents might have several affected children, or only unaffected children or a mixture of both affected and unaffected children. It is impossible to predict what will happen for any particular pregnancy, although it is possible to test an unborn child to see if it has CF or not.

So if carrier parents already have a child with CF, this does not change the risks in the next pregnancy. Each pregnancy has exactly the same chances as the one before: one in four of an affected child and three in four of an unaffected child (e.g., carrier or child without CF). Both boys and girls have an equal chance of being affected.

Genetics is a complicated subject – keep asking questions until you are happy that you understand.

How is CF diagnosed?

Screening tests

Your child may have been unwell before CF was diagnosed, or the diagnosis may have been made after a routine newborn **screening test** which is now performed by all health authorities in the UK. Sometimes CF is diagnosed in pregnancy, by a special antenatal test or sometimes because a routine scan identifies a problem.

Heel-prick test

In the first week a simple heel-prick blood sample is taken from all newborn babies (Guthrie Test). The blood sample is tested in the laboratory for signs of several diseases. The CF Trust campaigned for CF testing to be routinely included in the Guthrie Test because late diagnosis can result in more severe disease. Now, all newborn babies in the UK are screened for Cystic Fibrosis, meaning they will be able to receive appropriate treatment immediately, which will help to keep them healthy.

A few of the babies who are tested for CF will need a second sample taken. This is usually because the results were borderline and need to be confirmed. If the test is positive the child will be referred to a **paediatrician** who will arrange other tests, including a **sweat test** and a **genetic test**. This should be carried out in a Specialist CF Centre.

Sweat test

In the 1950s it was recognised that children with Cystic Fibrosis have more salt in their sweat than normal. Some parents comment that their child tastes salty when they kiss him or her. However, children with CF do not sweat more than other children.

The **sweat test** measures the amount of salt in the sweat. There are a number of methods for collecting sweat, none of which is painful or dangerous.

First the skin, usually on an arm or a leg, is cleaned and two discs of a special jelly are placed on the skin a few inches apart. The discs of jelly are connected to a battery, which passes a tiny electric current between them – this does not hurt. After about five minutes the skin under one of the discs should be sweating nicely. The discs are removed and the skin is dried.

A paper disc or special device (Macroduct) is put over the place that was sweating and the new sweat produced flows into the tubing. Collecting enough sweat for the laboratory to be able to measure the salt level takes anything from 10 to 30 minutes. Occasionally not enough sweat is

produced and the test has to be repeated.

If the salt level is abnormally high, the child has Cystic Fibrosis. Sometimes the result will be borderline and the test may need to be repeated to be sure of the result.

A sweat test is required in the following situations:

- When the *screening tests* on a newborn baby are abnormal.
- When there are other symptoms that can indicate CF, such as:

a troublesome cough
repeated chest infections
prolonged diarrhoea/abnormal stools
poor weight gain

- If your baby is born with a blocked bowel Cystic Fibrosis may be suspected. Of every ten babies born with CF one is very ill in the first few days of life with an obstruction of the bowel. There are several types of obstruction but the most common in CF is *meconium ileus*. All newborn babies have *meconium* in their bowel – this is the thick, black material they pass the first time their bowels are open. In CF the *meconium* can be so thick and sticky that it blocks the bowel. Babies with *meconium ileus* often need an urgent operation to relieve and bypass the blockage to allow the bowel to recover. When the baby has recovered from this, a *sweat test* and *genetic test* will be done to see if the blockage was due to Cystic Fibrosis.
- If you have a child with CF, other children in your family should have a *sweat* and/or *genetic test*.

Genetic testing

Nowadays, a sample of blood or cells obtained by gently rubbing the inside of the cheek with a little brush will be taken. These specimens are used to look for the CF *gene* and can be useful if the result of the *sweat test* is borderline. They are also useful for testing which members of a family may be carriers of the CF *gene* but not affected by the illness themselves.

What are the main problems with Cystic Fibrosis?

Because Cystic Fibrosis results in the production of thick, sticky *mucus*, the organs which are particularly affected are those where the *mucus* has an important job to do, particularly the digestive system and the lungs.

How does CF affect the digestion?

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

In those with CF, the small channels down which the digestive juices flow become blocked with sticky *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 from person to person. The majority of newborn babies with CF cannot digest milk and without treatment, fail to gain weight and have very loose stools. At the other end of the spectrum are the 5 to 10% or so of people with CF whose pancreas retains some useful function all their lives.

How can I help my child's digestion?

It is now possible to replace most of the missing *enzymes* with *pancreatin* – this is a general name given to all *pancreatic enzyme* medicines. They come in the form of powder, granules or capsules, but for most children, capsules are preferable. They contain many *enteric coated microspheres*. The outer capsule dissolves in the stomach, releasing all the *microspheres*. They then pass into the upper part of the small intestine, mixing with the food, allowing digestion to take place. Because the *microspheres* are protected from the acid in the stomach by a special coating, they are not released in the stomach but in the upper small intestine where they are needed to do their job. Your CF Centre will advise you which preparation is appropriate for your child and how to use it best. For infants a smaller version *microspheres* called *minimicrospheres* (Creon Micro) is available.

Do all babies with CF need enzymes?

A few babies do not need *enzymes* at first but may need them later on. It is important to watch that their bowel function and growth remain normal. Most babies with CF need them from birth and most will need to take them for the rest of their lives. This is not normally a problem. It becomes routine and children soon learn how to take them themselves.

Which type of *pancreatin* should my baby have?

This often depends on age. Your Specialist CF Centre will advise you.

How many enzymes will my child require?

This varies widely and is very much a matter of trial and error. In time, you will be taught how to vary the dose according to the type of food your child is having and when a change of dose may be needed. **Always obtain advice from the CF Centre dietitian and Doctor.** The *enzymes* help to digest fat and protein, so meals with a lot of fat and protein need more enzymes than low fat/low calorie meals.

What if I forget to give the enzymes?

Forgetting a single dose is not likely to be important, although your child may have looser stools afterwards. However, if *enzymes* are missed regularly, the digestion will be poor and the baby's growth is likely to suffer.

How do I give enzymes to a baby?

Most young babies will take the *microspheres* removed from the capsules or the *minimicrospheres*. They can be mixed with fruit puree or a little milk and given to the baby before each feed from a teaspoon. Do not mix them with a bottle of milk – the milk will curdle if it is in contact with the *enzymes* for too long.

Are there any problems giving enzymes to a baby?

Pancreatin given in the correct dose and swallowed will do no harm at all. However, if it stays in contact with the baby's skin it can make it sore, especially around the mouth and in the 'dribble area'. A breast-feeding mother may become sore around the nipples. It is helpful to put

a little Vaseline on the skin in these places before giving the *pancreatin* and to rinse the breasts with water after the feed. It can be a good idea to use a nipple shield to protect breasts when feeding.

Breast or bottle?

Most babies with CF thrive on breast milk. If you are able to feed yourself, this is generally the best idea but formula milk will do just as well.

Whichever way your baby is fed, his/her weight gain will be monitored at CF Clinic visits and should be normal. If this is not the case, a change of milk or the addition of an extra-calorie supplement may be advised. Your Specialist CF Centre and the CF dietitian will help you.

You can find more details about different types of milk and food that may be used in the CF Trust booklet *Eating well with Cystic Fibrosis: A guide for feeding infants*.

Do babies need more enzymes if they are re-fed in a short time?

There is usually no need to repeat the dose within one to two hours of the last dose. This is especially important if the baby is being breast fed on demand.

What about weaning?

This is the same as with any baby. If a baby appears exceptionally hungry, weaning a little earlier than usual at approximately three months should be advised. Remember that children with CF may need more calories than other children to grow at the same rate. Your CF Centre and dietitian will advise you.

What if my child does not eat a meal after having the enzymes?

Remember that your child is no different from any other. All toddlers refuse to eat at some time (some more than others) and your child will be no different.

However, your child has CF and toddlers with CF may not feel very hungry when they have an infection. They will come to no harm if occasionally nothing is eaten after a full dose of *enzymes*. If this happens frequently then it may be helpful to give half the dose at the beginning of the meal and the remainder halfway through. Nutrition is very important in CF but as with all children, you do neither yourself nor your child a favour in the long run if each mealtime ends in a battle because of untouched or unfinished meals. If mealtimes are becoming difficult, talk to the staff in your CF Clinic as soon as possible. Do remember that most children go through periods when they do not seem to eat much.

What should a child with CF eat?

In general, children with CF should eat whatever the rest of the family is having. However, even with *pancreatin* a child with CF may not absorb all the nourishment needed to grow normally, and may therefore, need more calories. So, extra calories added to meals and additional milky drinks may be helpful. Children with CF are also encouraged to have small high calorie snacks in between meals, not to replace meals but supplement them. Your CF Clinic and the CF Trust booklet on nutrition will help you. (*see back cover on how to obtain a copy.*)

Does my child need extra vitamins?

Children with CF do not absorb vitamins well, especially those vitamins which are dissolved in fat (vitamins A, D and E). Your child will usually need additional supplements of these vitamins in the form of drops or tablets each day. This is not the same as the infant vitamin drops

sometimes given to other babies. Extra minerals, such as iron, are not usually needed.

Why is a child with CF weighed and measured so often?

It is important to be sure that a child with CF is growing well, so your CF Clinic will plot your child's measurements on a growth chart.

Does my child need extra salt?

In this country the answer is generally 'no'. You may be specifically advised to give salt supplements by your CF team and they will tell you when and how to give them. If the weather is exceptionally hot here or if you are going abroad to a hot climate people with CF may need extra salt.

IT IS VERY DANGEROUS TO GIVE A BABY EXTRA SALT WITHOUT MEDICAL ADVICE.

Does CF affect the teeth?

Cystic Fibrosis does not affect teeth directly, although poor nutrition may affect the teeth's growth. Most antibiotic medicines are now sugar-free but a lot of the foods that are full of calories are very sweet, and therefore although very helpful for CF, may not be so good for the teeth.

You should encourage your child to brush his or her teeth every morning and before going to bed, where possible after eating or taking medicine – and, of course, to visit the dentist regularly.

What about fluoride supplements?

Your dentist will tell you whether these are necessary. Do tell him or her that your child has Cystic Fibrosis.

Other problems in the digestive system or the gut

There are several other ways in which CF can affect the gut:

Tummy aches

Many children without CF have tummy aches for no apparent reason. They usually go as quickly and as mysteriously as they come, and children with CF will occasionally have these types of tummy aches too. Children with CF may complain of tummy aches after a bout of coughing. If it is happening regularly, then you should seek advice from your CF team who may arrange further investigation or a change in *pancreatin* dose. If you are worried about tummy aches, do speak to your CF doctor.

If your child has severe, acute abdominal pain **seek medical advice immediately.**

Distal Intestinal Obstruction Syndrome (DIOS) or Meconium Ileus Equivalent (MIE)

This has nothing to do with the *meconium ileus* seen in newborn babies but occurs in older children and adults. The bowel becomes blocked by sticky, mucousy motions and food, causing recurrent pain and, sometimes, vomiting. The cause of this condition is not fully understood but

it requires investigation and treatment with special medicines which your CF Centre will tell you about.

How does CF affect the chest?

In the lungs there are lots of tiny tubes, called *bronchi*. Air passes down these tubes to reach the specialised parts (*alveoli*), where oxygen enters the bloodstream and carbon dioxide leaves, to be breathed out of the body.

We all have liquid and *mucus* in our lungs which help them to function, but in children with CF the *mucus* produced is abnormally thick due to there being too little liquid. This can block some of the smaller airways and this leads to infection. If not controlled, infections can lead to damage to the lungs. In the early years infections are usually caused by viruses and certain bacteria e.g. *Staphylococcus aureus* and *Haemophilus influenzae*. Later on, infections are caused by other bacteria, including one called *Pseudomonas aeruginosa*. Much of the damage these infections can cause can be prevented by proper treatment, as described below.

How is the chest treated in CF?

The aim is to keep the lungs as clear of mucus and infection as possible. There are two main ways in which this is done, both of which are important.

- prevention and treatment of chest infections, usually with antibiotics.
- clearing the sticky **mucus** from the lungs by **physiotherapy**, breathing exercises and regular physical exercise.

Prevention and treatment of chest infections

Should my child be kept away from other children?

It is impossible to prevent your child picking up infections from other children and adults and so there is no point in isolating him or her for that reason. It is sensible, though, to avoid close contact with people who have streaming colds.

Although the risk of picking up CF-related infections from other children with CF is low, it is advisable to avoid contact with others with CF where possible. Clinic attendance is important though and necessary arrangements will have been made at the hospital to ensure children with different infections are kept apart from each other. Your CF team will be able to explain how they do this.

It is **very important** that children with CF avoid smokers; smoke particles cling to everything and there is now good evidence that 'passive smoking' can affect the lungs; those with Cystic Fibrosis are particularly vulnerable. Pets are not a problem unless your child has an obvious allergy to them. This includes horses, although it is important to avoid the stables which are commonly contaminated with fungal spores. So children with CF can ride horses, but should not be allowed to "muck out".

Can antibiotics prevent chest infections?

There is increasing evidence that early, frequent, and in some cases, continuous antibiotics can prevent or delay the lung damage in Cystic Fibrosis. The staff at your CF Centre will discuss the

most suitable approach for your child. Whereas the general population is advised to be cautious in the use of antibiotics, for those with CF, antibiotics on a regular basis when necessary are invaluable.

How will coughs and colds affect my child?

All children, whether they have CF or not, suffer numerous colds in their first few years. In children with CF the symptoms often last longer because of the increase in lung secretions. Colds are caused by viruses and the only cure is time but children with CF may need antibiotics to prevent an infection with bacteria following straight on. Extra **physiotherapy** is often needed if there is a lot of extra **mucus**.

Are cough medicines useful?

Cough medicines suppress the cough. If a child with CF is coughing it is important to find out what is causing it; there may be an infection which requires antibiotic treatment. Cough medicines should not be given to children with CF without discussing them with your Specialist CF Centre first. Vaporisers and ionisers may be useful but they should not be used instead of **physiotherapy** and antibiotics.

How are chest infections treated?

- A child with a chest infection needs extra **physiotherapy** (longer and more often) to clear the **mucus** and infected material from the lungs.
- S/he also needs antibiotics. The doctor will decide which antibiotics to use by knowing which bacteria are likely to be present and by taking a **sputum** sample or swab. The antibiotics will usually be given by mouth as liquid medicine, tablets or capsules. Sometimes they are inhaled as a mist from a **nebuliser** or given by the **intravenous** route, i.e. directly into a vein.

Are any special tests needed?

If possible a **sputum** sample (or swab, cough swab or **nasopharyngeal aspirate [PNA or NPA]** from a younger child) is taken to identify the bacteria (if any) causing the infection. A chest X-ray and, occasionally, blood tests, may be helpful. Older children may be asked to blow into a tube or machine to see how much 'puff' they have. This is particularly useful if it is done from time to time in between infections, because the levels often fall before the infection becomes obvious, enabling it to be caught early.

Will my child have to go into hospital every time s/he has a chest infection?

Most chest infections can be treated at home but if the infection is severe and persistent s/he may need intensive treatment, including **intravenous** antibiotics. If so, s/he may need to be admitted to hospital, although with the help and supervision of the CF Nurse Specialist many children can now have **intravenous** antibiotics at home.

DNase (Pulmozyme®)

There is now a drug, DNase, which breaks down the **sputum** and makes it thinner; this should make it much easier to clear by **physiotherapy** and coughing. The drug is taken by inhalation from a **nebuliser** once a day. It does not help all children but your doctor may suggest a trial of treatment if your child has particularly thick and troublesome **sputum**.

Physiotherapy

Physiotherapy, breathing exercises and regular physical exercise help to clear the sticky *mucus* from the lungs of someone with Cystic Fibrosis.

What is chest physiotherapy?

Chest *physiotherapy* is a way of clearing the excess *mucus* from the lungs. There are different ways that chest *physiotherapy* can be given to your baby or child. It is important that you learn the correct technique and your CF Centre will advise and teach you the best method for your baby or child.

Do not be afraid to ask the physiotherapist to watch you doing it from time to time to make sure that you are still doing it as effectively as possible for your child's chest. The CF Trust produces several factsheets on physiotherapy,

When should I start to do physiotherapy?

You will be taught to do physiotherapy soon after your baby or child is diagnosed as having Cystic Fibrosis. Your physiotherapist will advise you on when and how to do it.

When should I do physiotherapy and for how long?

It is very important that you learn the correct form of *physiotherapy* soon after the diagnosis of Cystic Fibrosis. How well your child is will depend on how often you need to give your baby (or child) *physiotherapy*. Traditionally, it has been carried out routinely twice a day and more often when babies/children are unwell with a chest infection. However, nowadays many babies and children with CF are very well and may have clear chests. Therefore, it may not always be necessary to give twice daily treatment when they are very well, particularly if they are very active and have plenty of exercise.

When your baby needs chest *physiotherapy* it is generally performed for about ten minutes at a time, *before* a feed. This may need to be increased if they are unwell. The physiotherapist at your CF Centre will be able to advise you.

Who should do the physiotherapy?

To begin with, those adults who care for the child on a daily basis should do it, usually the parents. However, later on other relatives or friends should learn, so that no one person becomes indispensable to the child, and that the person who normally does it can have a break from time to time.

Breathing exercises can be introduced in the form of a game from the age of two or three and, as the child gets older, s/he learns to do his or her *physiotherapy*. From about the age of nine, most children can start doing some *physiotherapy* themselves without help from the family. Most teenagers become completely independent and only require help from time to time.

Will physiotherapy hurt?

When it is done correctly it does not hurt, although small children may try to avoid it by complaining that "it hurts".

Do we need special equipment?

There are lots of different *physiotherapy* techniques. Some need special devices and these are usually provided by your CF Centre.

In babies and small children chest *physiotherapy* is usually carried out on the adult's lap but as they grow, if they continue to require tipping for their treatment, a special foam wedge or supportive frame can be useful. Your CF Physiotherapist will advise you which is the best method of chest *physiotherapy* for your child and whether any specialist equipment is needed.

Will my child be able to run, play, swim etc like other children?

Regular exercise is an important part of care for children (and adults) with Cystic Fibrosis. It helps prevent deterioration of the lungs. It also improves physical strength and is very good for keeping bones healthy.

Toddlers often like running, jumping and trampolining, all of which are very good for them. When at school, children with CF should take part in P.E. and games just like other children in their class and you should also strongly encourage them to do plenty of physical exercise out of school, such as cycling, football, swimming, tennis, etc. It is often more fun to have company when exercising, therefore plan some of these activities with the whole family and/or friends.

Other problems in the chest

There are several other ways in which CF can affect the chest.

Asthma

About 30% of children with CF wheeze from time to time. This happens when the muscles surrounding the small airways contract and cause them to narrow slightly. They may find it more difficult to catch their breath and sometimes have a feeling of tightness in the chest.

Wheezing responds well to the medicines used for children with asthma such as bronchodilators and steroids. They are usually inhaled from an inhaler device which can be easily carried around, or a *nebuliser*.

Haemoptysis

Haemoptysis is coughing up blood and is rare in children with Cystic Fibrosis. In adults it is quite common for streaks of blood to be in the *sputum*. It can be a sign of infection and you should see your CF doctor if it occurs.

When should you take your child to see the doctor?

The answer is a simple one – when you are worried about him or her.

Here are some useful pointers:

- *cold symptoms*
- *increased or frequent cough*
- *increased sputum*
- *change in colour of sputum*
- *breathlessness*
- *fever*
- *decreased or poor appetite*
- *weight loss*
- *tummy aches*
- *frequent or loose stools*

- *vomiting* *exercise*
- *decreased ability or unwillingness to*

Children with CF should attend a hospital Specialist CF Centre or Clinic. There should be someone at the hospital you can ring if you are worried. Alternatively, you can contact your GP, who will send you to the hospital if s/he thinks you need to go. But remember, GPs are not experts on CF, so if you are worried, ensure you see your CF doctor. It is important that you find out what the local arrangements are.

Although many children attend the CF Clinic at their local hospital, it is strongly recommended that all people with CF should have regular contact with the staff of a Specialist CF Centre.

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. Ultimately, if every child is immunised, we will be able to eradicate these diseases from the community, just as smallpox has virtually disappeared from the world. However, these illnesses, such as measles, German measles, mumps, diphtheria, whooping cough, tetanus, polio and other serious infections, including meningitis caused by bacteria called *Haemophilus Influenzae Type B (HIB)*, will disappear only if everyone takes up the opportunity of vaccination.

Some of these infections, such as measles and whooping cough, still occur and may have severe and lasting effects on the lungs of children with Cystic Fibrosis. 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 against flu each year at the beginning of the winter season. Some CF doctors recommend vaccination against pneumococcal infection.

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 Specialist CF Clinic. Just having a cough or cold is not enough reason to delay having an immunisation – there are very few medical reasons to avoid immunisation.

Please discuss the pros and cons with your Specialist CF Centre which will be familiar with the latest immunisation recommendations and will be able to give you advice on what is best for your child.

Can CF affect other parts of the body?

Yes – but this 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 develop nasal *polyps*, which if troublesome, may need to be removed by a small operation .

Liver

Some people with CF get a kind of *cirrhosis*. Older children and adults will have their liver function checked from time to time by a blood test, as treatment is now available to prevent progression of any liver problems.

Diabetes mellitus

This develops in about 30% of adults with CF and results in an abnormally high level of sugar in the blood. Treatment is usually by insulin.

Bones and 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. Older adolescents and adults can also be prone to osteoporosis (thin brittle bones). As children born now with CF are better than today's adults, we do not expect them to develop osteoporosis to the same degree that adults with CF now experience it.

Puberty

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

Fertility

Fertility, or the ability to have children, is usually normal or only slightly reduced in women but most men with CF are infertile. This means that their sexual function is entirely normal but they can seldom father children naturally. However, recent advances in *in vitro* fertilisation and aspiration of sperm has allowed some men with CF to father children with clinical assistance. For further information, see the CF Trust's booklet *Growing up with cystic fibrosis: A guide for young people*. (Contact the CF Trust – details on back page).

Some other questions parents have asked

Will my GP and health visitor know about CF?

As we mentioned earlier, people in your family surgery may not have seen many children with CF but the hospital CF Clinic will keep them closely informed.

What about the usual baby clinics?

If you are attending the hospital frequently, you may feel that there is no need to attend the normal baby clinics as well. However, local baby clinics deal with all sorts of things, including immunisations, development checks, and hearing and vision tests. This makes them just as important but for different reasons. It is also nice to meet other parents with young children in your area.

Is any financial help available?

One of the benefits available from the Department for Work and Pensions (DWP) is the Disability Living Allowance (DLA). The Cystic Fibrosis Trust or the social worker at your CF Centre will be able to advise you, providing information on benefits and a detailed guide on how to apply for DLA, with trained staff to advise you as to how to complete the application form, which is rather daunting. For further details contact the CF Trust Benefits Helpline on 0300 373 1010.

The CF Trust also provides some financial grants to families affected by CF who are experiencing hardship. Details are contained in a factsheet called *Cystic Fibrosis A Guide to Financial Help*, which also gives details of other sources of financial help. For further details contact the CF Trust Welfare Grants Helpline on 0300 373 1020.

What should I tell other people about my child's CF?

Only you can decide but you may wish to discuss it with your CF Centre and with other people with relevant experience. You can also call the Cystic Fibrosis Trust Helpline on 0300 373 1000. You should also consider how and when to tell any other children you may have. In general it is better not to be secretive.

Unfortunately, some people may make unintentional, but hurtful, comments about your child, usually out of ignorance, so it is best to be prepared. These ignorant comments include: "Isn't he small." "He hasn't grown very much." "Fancy taking out a child with a cough like that." "Fancy giving a child medicine (*enzymes*) like that in public."

Try to take such remarks in your stride and, if possible, be prepared with explanations. People may also be ill-informed or years out of date in their understanding of Cystic Fibrosis. They are usually willing to understand and are, if anything, over-sympathetic when the situation is explained to them.

Will complementary medicine help?

There is no scientific evidence that any complementary medicine can do anything to help with CF and it is essential for the future health of your child that the conventional treatments your CF Centre recommends are given in the prescribed way. However, provided this is the case, the addition of complementary treatments should do no harm and some families report a benefit. Always consult your CF doctor first.

School years

Cystic Fibrosis does not affect intelligence. Children with CF attend pre-school playgroups and nurseries in just the same way as any other children. Most children with CF attend normal schools and join in all the normal activities. It is important that teachers and other staff in the school know about CF; the school doctor and nurse may be able to help with this.

The details of your particular child's treatment though, will come best from yourself. You know your child, so talk to the school before term begins and keep them informed of new developments or changes in treatment. This is especially important when your child changes teacher or school. Your CF nurse will sometimes visit your child's school to explain the nature of Cystic Fibrosis. Discuss this with the nurse if you think it might be helpful.

Most children with CF attend normal schools and join in all normal activities.

The CF Trust publishes a range of publications to help with school including a booklet called *School and Cystic Fibrosis: A guide for teachers and parents*. Your child, if aged between 8 and 12 years, might like to read *CF and You*, a CF Trust booklet which may help him or her answer some of the questions which naturally inquisitive friends are bound to ask. For further information or advice please contact the CF Trust – details are shown on the back page of this

factsheet.

Teenagers and leaving school

Adolescence is a challenging period for anyone. It is especially so for young people with CF, particularly if the disease causes them to mature later than their peers. Nevertheless, the full range of further education and employment opportunities should be available to any young adult with CF, depending on their intellectual and physical capabilities. There are some occupations which are less suitable, of course, but these considerations are for the future and decisions can be made at the time with help from the doctor and career specialists.

As children get older, most will also want to know more about the disease and how it will affect them. The CF Trust has published a booklet entitled *Growing up with CF: a guide for young people with CF aged 12-18 years*. This booklet answers many questions asked by teenagers with Cystic Fibrosis.

The future

Only thirty years ago, the outlook for a baby born with CF was very poor. Today, young adults with CF are living into their thirties, forties, and beyond and these people are leading active and fulfilling lives.

It is likely that the quality and length of life will continue to improve as a result of current research. New treatments which were supported by the CF Trust, and made available over ten years ago, are now bearing fruit.

The future is without doubt an optimistic one but, until a cure is found, we must aim to keep every child and adult with CF as fit and well as possible by controlling their symptoms, to maximise both the quality and the length of their lives.

Facts and figures

- Cystic Fibrosis was fully recognised only as recently as the 1930s.
- In the past the disease was known as 'fibrocystic disease of the *pancreas*' – the effects of the disease on the lungs was recognised later.
- Cystic Fibrosis is also known as *mucoviscidosis*, because the *mucus* is thick and sticky.
- Cystic Fibrosis is one of the UK's most common life-threatening inherited diseases.
- The disease affects approximately 1 in every 2,500 children born – that's roughly one child with CF born in this country every day.
- In the 1930s children with CF didn't live very long because there were no antibiotics to

- fight chest infections. Today antibiotics and other treatments are more sophisticated and most children do well and live into adulthood.
- The part of our *DNA* make-up (the *gene*) which is responsible for CF was discovered in 1989 and already clinical trials of '*gene* therapy' (trying to replace the faulty CF *gene* with a healthy version) are making considerable progress. We hope to be able to offer gene therapy treatment for CF in the foreseeable future.

Glossary

Alveolus (Alveoli)

The specialised part of the lung where oxygen enters 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.

DIOS

Distal Intestinal Obstruction Syndrome. A blockage of the gut which occurs in older children adults with Cystic Fibrosis.

DNA

The commonly used abbreviation for deoxyribonucleic acid, the principal molecule carrying genetic information in almost all organisms.

Enteric Coated

Covered with a coating which protects against acid in the stomach. This is useful for pancreatin.

Enzyme

A chemical that effects change in some way. In CF this usually refers to digestive enzymes which digest food so 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 overall health and functioning, as well as for individual characteristics such as eye colour, blood group. Faulty genes cause certain genetic

diseases such as Cystic Fibrosis.

Genetic Testing

The method of detecting certain genes, for example tests can determine when a person carries the gene for Cystic Fibrosis.

Haemophilus Influenzae

Bacteria which is a common cause of respiratory infection in Cystic Fibrosis. See page 9 and *Are Immunisations Important?*.

Haemoptysis

Coughing up blood.

Health Care Professionals

Doctors, nurses, physiotherapists, dietitians, social workers and pharmacists.

Intravenous

Sometimes antibiotics or other medicines are given into a vein rather than by mouth. To make it easier, a small plastic cannula (tube) can be left in the vein so that the drug can be put in through it rather than by a fresh injection each time. There is a cream available to numb the skin before the cannula is put in it.

Meconium Ileus

An obstruction of the small intestine at birth.

Microspheres

Enzyme granules contained within a pancreatin capsule.

MIE

Meconium Ileus Equivalent – See DIOS opposite:

Mucoviscidosis

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

Mucus

An essential 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 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.

Pancreatin

An extract of animal pancreas; the general name for all pancreatic enzymes.

Physiotherapy

Part of the treatment for Cystic Fibrosis.

PNA or NPA

Per Nasal Aspirate or Naso Pharyngeal Aspirate – a special way of getting a sample of sputum from a child too young to be able to cough it up.

Polyps

A small growth of mucous membrane that

can grow on the lining of the nose.

Pseudomonas aeruginosa

A bacterial infection which affects the lungs.

Screening tests

A test carried out to diagnose and treat a disease before it causes problems.

Sinusitis

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

Sputum

Mucus material produced by the cells lining the respiratory tract.

Staphylococcus aureus

A bacteria infection that can affect the lungs. *See page 9. How CF Affects the Chest.*

Sweat Test

The test used to diagnose Cystic Fibrosis.

For further general information and literature published by the Cystic Fibrosis Trust:

**CF Trust Helpline
0300 373 1000
For information, advice and support (including
contents of this booklet).**

Cystic Fibrosis Trust
11 London Road
Bromley
Kent BR1 1BY
Tel: 020 8464 7211
Fax 020 8313 0472
Email: enquiries@cftrust.org.uk
www.cftrust.org.uk