I N F O R M A T I O N

FINDING OUT

CYSTIC FIBROSIS TRUST

A guide for parents of newly diagnosed children with Cystic Fibrosis

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

Contents

This booklet has been written to assist you and your medical advisers. It isn't intended to replace any advice you may receive from your CF clinic.

Published by the Cystic Fibrosis Trust with assistance from Dr Michael Green.

Photography by Camilla Jessel. Page 8 photograph courtesy of Lewisham Hospital.

Published with the generous support of

SOLVAY HEALTHCARE

April 2000

What is Cystic Fibrosis?

CYSTIC FIBROSIS TRUST

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

All the questions, which are answered in this booklet, have been asked by other parents going through the same stages you are now.

Any words appearing in red italic type in this brochure can be found and are explained in the Glossary on pages 39 to 41.

If you have just been told that your child has CF, you may be finding the diagnosis quite a shock. You may even be feeling a sense of loss or grief. These reactions are quite normal and other parents have experienced them before you. This is why it maybe helpful for you to talk to other parents of children with CF from an early stage.

CYSTIC FIBROSIS TRUST

How do you feel?

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 CF specialist clinic where CF can be fully explained. Write down questions you want to ask as you think of them in case you forget later.

Anger

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 emotions are understandable but could do great harm to relationships in the family.

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.

The most important piece of advice is this – if something is worrying 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 CF. Consequently they will be naughty sometimes and will have all the same emotions as other children. There is no reason to treat them differently in relation to discipline, 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 and, with the latest advances in research, there is every chance that new therapies will be developed to 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 CF. 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 concern the child with CF is receiving.

Getting help

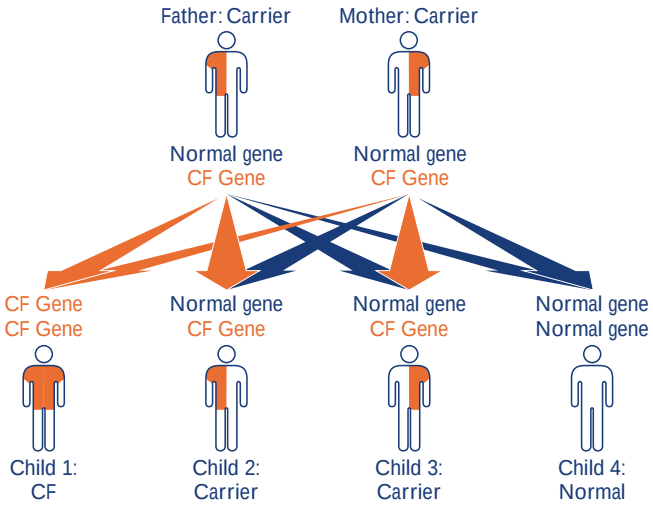
As with all things, life is easier if there is someone or somewhere to turn to for practical advice, emotional and financial support. That is exactly where the CF Trust Support Service comes in. It is a source of knowledge and experience which parents can draw on about any aspect of family life. To help you the CF Trust Support Service has four Regional Support Co-ordinators, each with a team of local volunteer Support Workers. To contact your nearest Co-ordinator ring the CF Trust on 020 8464 7211.

Cystic Fibrosis has some impact on the whole family including brothers, sisters, aunts uncles, grandparents and other relatives. The CF Trust Support Service is there for all of them.

A baby may be born with CF only if BOTH parents are carriers of the defective Cystic Fibrosis *gene*. Even then both parents having the defective CF *gene* won't mean that the baby will necessarily have CF.

CYSTIC FIBROSIS TRUST

Why does my child have CF?



If both parents are carriers, a child has:

- *a one in four chance of being born with CF*
- *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, ie not having CF nor being a carrier of the defective CF gene*

All this means that CF is a *genetic* disease. 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
- 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 small family opposite 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.

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.

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: 1 in 4 of an affected child and 3 in 4 of an unaffected child (ie carrier or child without CF). Both boys and girls have an equal chance of being affected.

The CF Trust publishes a booklet called Genetics, Carrier Tests and Tests during Pregnancy *which explains this in more detail. It also gives information about some of the tests available to families where there is a history of CF and when either you or another relative is considering having children.*

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

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 performed by some health authorities.

CYSTIC FIBROSIS TRUST

How is CF diagnosed?

A simple heel-prick blood sample is taken from all new born babies (Guthrie Test). The sample could be used to screen babies for CF providing invaluable early diagnosis. The CF Trust has campaigned for CF testing to be routinely included in the Guthrie Test because late diagnosis can result in more severe disease.

Screening tests

These days we prefer to prevent diseases or to treat them before they become serious.

All babies born in the UK have a sample of blood taken when they are 1-2 weeks old, usually from a prick in the heel. These spots of blood are tested in the laboratory for signs of several diseases. In some areas this will include CF but at present most health authorities do not include CF in their routine testing.

A few of the babies who are tested for CF 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*.

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 disk or special device (Macroduct) shown in the picture (on previous page) 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 CF. Sometimes the result will be borderline and the test may need repeating several times to be sure of the result.

When is a sweat test required?

- When the *screening tests* on a newborn baby are abnormal.
- Other symptoms which can indicate CF include:
  a troublesome cough
  repeated chest infections
  prolonged diarrhoea
  poor weight gain
- CF may be suspected when a baby is ill. In every 10 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* will be done so see if the blockage was due to CF.

- If you have a child with CF, the other children in your family should have a *sweat* and/or *genetic test*.

Genetic testing

Nowadays, a sample of blood or material 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.

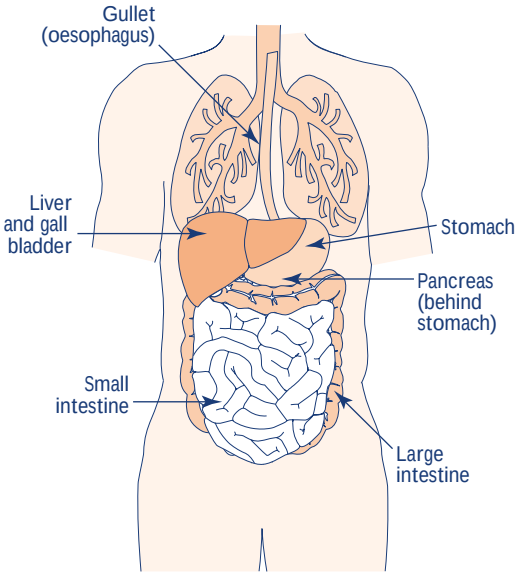
The *pancreas* is a gland in the abdomen; one of the functions of the *pancreas* is to produce digestive juices, or *enzymes*, which pass into the intestine, where they help us digest and absorb the food we eat.

CYSTIC FIBROSIS TRUST

How does CF affect the digestion?

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 very much between people. At one extreme, some newborn babies cannot digest milk and fail to gain weight and have very loose stools. Others 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 possible to replace most of the missing *enzymes* with *pancreatin* – this is a general name given to all *enzyme* medicines. They come in the form of powder or capsules. For most children, capsules are preferable. They contain granules called *enteric coated microspheres*. The outer capsule dissolves in the stomach, releasing all the granules or *microspheres*, which are then mixed well with the food.

Because the *enzymes* in the *microspheres* are protected from the acid in the stomach by a special coating, they can be released in the intestine, where they work best. Your CF clinic will advise you which medicine is appropriate for your child and how to use it best.

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.

Which type of pancreatin should my baby have?

This often depends on age. Your CF clinic will advise you.

How much enzyme will my child require?

This varies widely and is very much a matter of trial and error. In time, you will learn for yourself when a change of dosage is needed and how to vary the dose according to the meal your child is having. Always obtain advice from the clinic dietitian and doctor.

continued

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 will suffer.

How do I give enzymes to a baby?

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

Are there any problems giving enzymes to a baby?

Pancreatin given in the correct dose will do no harm. 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.

You can find more details about different types of feed that may be used in the CF Trust's booklet *NUTRITION: Eating well with Cystic Fibrosis. A guide for feeding infants.* Your CF clinic and the dietitian will help you.

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

There is usually no need to repeat the dose within 1 –2 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. However, if your baby's weight gain starts to tail off, then it may be useful to wean slightly earlier, eg at 3 months rather than waiting until 4 months. Remember that children with CF may need more calories than other children to grow at the same rate.

What if my child doesn't eat after having 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 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. This is as much the case with children who do not have CF.

If mealtimes are becoming difficult, talk to the staff in your CF clinic as soon as possible. It must be accepted that all children go through periods when they do not seem to eat much.

What should a child with CF eat?

In general, the diet should be 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. Your child, therefore, may need more calories. So, extra calories added to meals and milky drinks may be helpful. Children are also encouraged to have small high calorie snacks in between meals; but these must never replace meals. Your clinic and the CF Trust's booklets 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 need a dose of vitamin drops or tablets each day. Extra minerals, such as iron, are not often needed.

Why is a child with CF weighed and measured?

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, shown overleaf. The central line printed on the graph show a child of average size; the lines above and below are for larger and smaller children. Generally speaking, the better children's growth, the better they will be able to fight chest infections.

Does my child need extra salt?

In this country the answer is 'No', unless the weather is exceptionally hot. If you are going abroad, or if the weather becomes unusually hot here, your CF clinic will advise you about salt supplements.

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

Does CF affect the teeth?

CF 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, and therefore very helpful in CF, are very sweet.

You should encourage your child to brush his or her teeth morning and night and after eating or taking any medications – and, of course, visit the dentist regularly.

What about fluoride supplements?

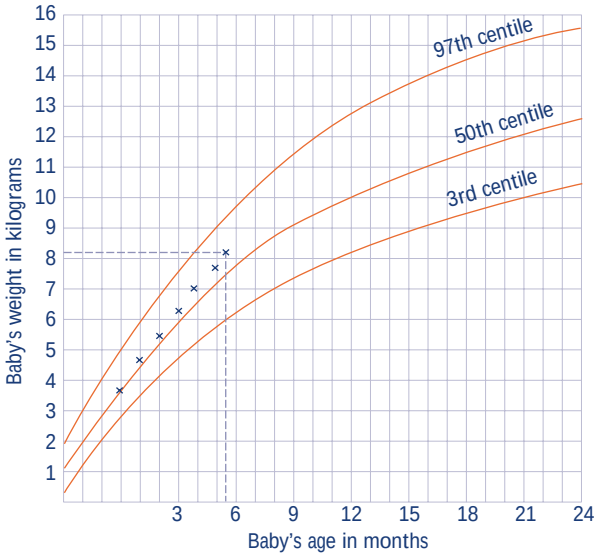
Your dentist will tell you whether these are necessary.

An example growth chart

This is a typical weight chart with age along the bottom and weight up the side. The three preprinted lines are called the centile lines and your baby's growth is compared to these. If you draw imaginary lines (see dotted lines) upwards from your baby's age and across from his or her weight, the point where they cross shows the point your child has reached.

For each age, on average 50 out of 100 babies will be above the 50th centile line and 50 below. Similarly, on average three babies will be above the 97th line and three below the 3rd line.

It is important to check that you baby's growth is following these lines like the example.



Other problems in 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 came. 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 clinic doctor who may arrange further investigation or a change in *pancreatin* dose.

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

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

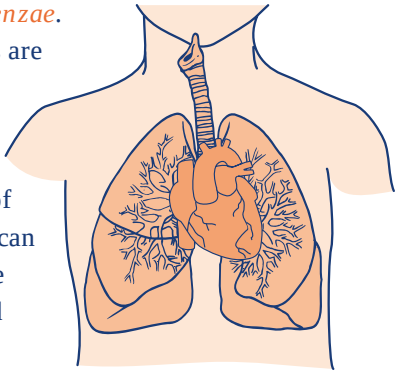
This has nothing to do with the *meconium ileus* seen in newborn babies but occurs in older children and adults. The bowel becomes blocked by sticky, mucusy motions and food, causing recurrent pain and, sometimes, vomiting. The cause of this condition is not fully understood but it requires investigation and treatment with special medicines which your CF clinic will tell you about.

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

CYSTIC FIBROSIS TRUST

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 the treatment described opposite.



How is the chest treated in CF?

The aim is to keep the lungs as clear as possible. There are two components to this part of the therapy:

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

Physiotherapy

Clearing the sticky *mucus* from the lungs by physiotherapy, breathing exercises and regular physical exercise

What is physiotherapy?

Physiotherapy is a way of clearing the thick, sticky *mucus* from the lungs. The pictures on the right give you a clue as to how it is done but you can only learn properly from the physiotherapist in your CF clinic and from lots of practice. Do not be afraid to ask the physiotherapist to watch you doing it from time to time to make sure that you are still performing the most efficient actions for your child's chest. The CF Trust booklet *TREATMENT: Physiotherapy for Cystic Fibrosis* will also help. *(See back cover on how to obtain a copy.)*

When should I start to do physiotherapy?

From the time of diagnosis.

When should I do physiotherapy and for how long?

It is important to get into a routine early on. Preferably, it should be done twice a day when your child is well and more often when s/he has a chest infection, because more *mucus* will be produced. In young babies 5-10 minutes at a time (and before the feed) is probably enough, although this will need to be increased as the child gets older. You should talk to your CF clinic, where you will be given the most appropriate advice.

Who should do the physiotherapy?

To begin with, the adults who care for the child should do it. However, later on some other relatives or friends should learn, so that no one person becomes indispensable to the child.

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 own postural drainage (see *TREATMENT: Physiotherapy for Cystic Fibrosis*). From about the age of nine, most children can start doing part of the treatment themselves without help from the family. Most teenagers become completely independent and only require help if they have increased secretions.

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".

Does it need special equipment?

A baby can be placed on the adult's lap but, for an older child, a special foam wedge or frame can be useful. Your physiotherapist will help you.

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

Regular exercise is very important for all children, whether they have CF or not. It should be encouraged in a form the child enjoys. Toddlers often like running, jumping and trampolining, all of which are very good for them. Swimming, too, is excellent exercise. When at school, children with CF should take part in PE and games as much as possible and you should encourage them to participate in out of school activities, such as cycling, football, tennis etc. The physiotherapist can show you exercises to help with posture.

Prevention & 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 risks of picking up CF related infections from other children with CF is very low, some parents choose to avoid unnecessary contact with others with CF where possible. Clinic attendance is, however, important and necessary arrangements will have been made to avoid cross infection. You may confirm these with your clinic.

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 health, particularly those with CF. Pets are not a problem unless your child has an obvious allergy to them.

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 CF. The staff at your CF clinic will discuss the most suitable approach for your child.

How will coughs and colds affect my child?

All children, whether they have CF or not, suffer numerous colds in the first few years. In children with CF the symptoms often last longer because of the increase in 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 are a lot of secretions.

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 CF clinic 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* 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.

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 may be needed and, occasionally, a blood test. 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.

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 will probably need to be admitted to hospital, although 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. It seems to be most useful in older children and 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*.

Inhaling 'nebulised' bronchodilator drug through mouthpiece

Other problems in the chest

There are 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. The children find it more difficult to catch their breath and often have a feeling of tightness in the chest.

Wheezing responds well to the medicines used for children with asthma. They are usually inhaled from a *nebuliser*. Other children can use the same medicines in inhalers, which they can carry around with them in their pockets.

Haemoptysis

Haemoptysis is coughing up blood and is rare in childhood. 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 the 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.

Most children with CF attend a hospital CF clinic and there will 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. It is important that you find out what the local arrangements are. You can keep a note of telephone numbers at the back of this booklet.

It is recommended that all people with CF should have some regular contact with the staff of a specialist CF Centre.

CYSTIC FIBROSIS TRUST

Here are some useful pointers:

- *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. Ultimately, if every child is immunised, we will be able to eradicate these diseases from the community, just as smallpox has disappeared from the world. However, these illnesses, such as measles, German measles, mumps, diptheria, whooping cough, tetanus, polio and the 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, 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.

Please discuss the pros and cons with your clinic, which will be familiar with the latest immunisation recommendation and will be able to give you advice which is best for your child.

Can CF affect other parts of the body?

CYSTIC FIBROSIS TRUST

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 a kind of *cirrhosis*. Older children and adults will probably have their liver function checked occasionally by doing a blood test, as treatment is now available to prevent progression of any liver involvement.

Diabetes mellitus

This eventually occurs in about 15% of 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. It is usually much milder and much less troublesome than in someone without CF.

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.

Fertility

This is the ability to have children. It 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 assistance. For further information, see the CF Trust's booklet *Growing up with Cystic Fibrosis: A guide for young people*.

Some other questions parents have asked

CYSTIC FIBROSIS TRUST

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, these 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 useful to meet other parents with young children and realise that you are not the only one with problems.

Is any financial help available?

One of the benefits available from the Department of Social Security (DSS) is the Disability Living Allowance (DLA). The CF Trust Support Service will be able to help you – providing advice on benefits and a detailed guide on how to apply for DLA, with trained staff to help you complete the application form.

The CF Trust Support Service also provides 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.

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

Only you can decide but you should discuss it with your CF clinic and with other people who have helpful personal experiences, such as the CF Trust Support Service local Support Worker. 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 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 are usually willing to understand and are, if anything, over sympathetic when the situation is explained to them.

Will complementary medicine help?

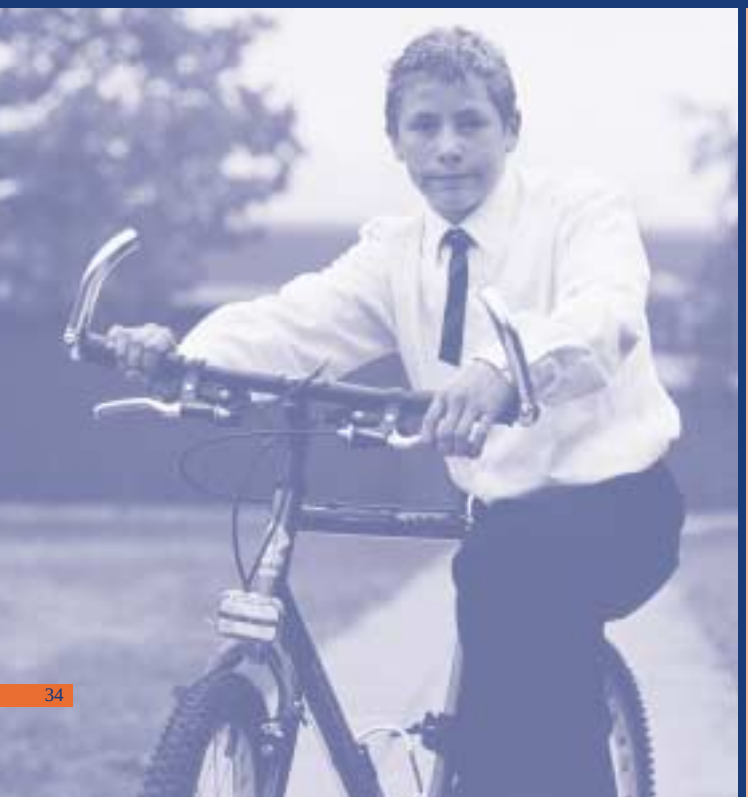
There is no scientific evidence that any complementary medicine can do anything to help with CF and it is essential for the future health of your child that the conventional treatments are given in the prescribed way. However, provided this is the case, the addition of complementary treatments should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

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 can help with this.

The details of your child's particular treatment, though, will come best from yourself. You know your child and his or her individual likes and dislikes, 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.

CYSTIC FIBROSIS TRUST

School Years



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

The CF Trust Support Service has produced a *Secondary School Pack* for use by pupils moving from primary to secondary education. It is intended to be used as a tool to enable pupils to provide information to various people within their new school about how Cystic Fibrosis affects them.

The CF Trust also publishes a range of publications to help with school. 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 inquisitive friends are bound to ask. For further information or advice please contact the CF Trust Support Service.

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, such as heavy manual work in a smoky or dusty atmosphere. These considerations are for the future and decisions can be made at the time with help from the doctor and career specialists.

Like everyone else, young people with CF need activities for their leisure time and realistic aims for the future. In addition to the continued treatment of their CF, they will also want to know more about the disease and how it will affect them.

The CF Trust Support Service has an Adolescent Liaison Officer offering advice and information on issues affecting adolescents with CF; she can be contacted on 01228 597405. In addition there is a free Helpline for adolescents with CF where advice is available on a confidential basis on Mondays, Wednesdays and Fridays from 2.00 pm to 5.00 pm on 0800 454 482.

An Adult Liaison Officer at the CF Trust can be contacted on 020 8464 7211 for information, help or advice on issues affecting adults. The Association of CF Adults can also be contacted through the Adult Liaison Officer.

The Future

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

It is likely that the quality and length of life will continue improving as a result of current research. New treatments which were supported by the CF Trust, and applied carefully 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, so that they may benefit from new therapies.

CYSTIC FIBROSIS TRUST

Facts & Figures

- CF 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.
- CF is also known as *mucoviscidosis*, because the *mucus* is thick and sticky.
- CF is the UK's most common life-threatening inherited disease.
- 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 you can expect your child to 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 defective CF *gene* with a healthy version) are under way. There is every hope of achieving a solution for CF by the time your child reaches adulthood.

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, see page 20.
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.
Cystic Fibrosis	See page 1.
DIOS	Distal Intestinal Obstruction Syndrome, see page 19.
DNA	The commonly used abbreviation for deoxyribonucleic acid, the principal molecule carrying genetic information.
Enteric Coated	Covered with a coating which protects against acid in the stomach. This is useful for pancreatin, see page 13.
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 characterstics such as eye colour, blood group and whether or not they have certain genetic diseases such as CF.

Genetic Testing	The method of detecting certain genes, for example tests can be made to determine when a person carries the gene for Cystic Fibrosis. See page 11.
Haemophilus Influenzae	Bacteria commonly known as 'flu. See page 20 *Are Immunisations Important*.
Haemoptysis	Coughing up blood, see page 26.
Health Care Professionals	Doctors, Nurses and hospital staff.
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.
Microspheres	Enzyme granules contained within a pancreatin capsule. See page 13.
MIE	*Meconium Ileus* Equivalent – a blockage of the gut which occurs in older children or adults with CF, see page 19.
Mucoviscidosis	Another name for CF, literally it means that the mucus is thick or vicid. It may be understood in other countries.
Mucus	A slimy fluid screted 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, see page 25.

Paediatrician	A doctor who specalises in the treatment of children.
Pancreas	A gland which lies behind the stomach and makes digestive juices or enzymes and insulin, see page 12.
Pancreatin	An extract of animal pancreas; the general name for all pancreatic enzymes.
Physiotherapy	Part of the treatment for CF, see page 21.
PNA or NPA	Per Nasal Aspirate or Naso Pharyingeal 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 bacteria infection that can affect the lungs. See page 20 *How CF Affects the Chest*.
Screening Test	A test carried out to find a disease before it causes problems, see page 9.
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. See page 20 *How CF Affects the Chest*.
Sweat Test	The test to used to diagnose CF, see page 9.

Index

Notes

Further enquiries about literature, including booklets produced by the Association of CF Adults, and donations should be sent to

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

Publications

AUDIO TAPES

Finding out about cystic fibrosis
A guide for parents and other relatives of newly-diagnosed children.

Growing up with cystic fibrosis
Advice and information for 12-16 year olds with CF and those who care for them. Useful for brothers, sisters and friends too.

Living with cystic fibrosis
Adults discuss some of the challenges of living with CF.

Cystic fibrosis and school
Advice for teachers, parents and people with CF starting or changing schools.

† **Diagnosis** A guide for parents and other relatives of newly-diagnosed children in the Asian community.

† **Adolescence** Information for Asian families about growing up with CF.

† *These tapes are available in Urdu and Gujarati only.*

THE FACTS
An introduction to the causes and effects of Cystic Fibrosis.

GENETICS
Genetics, carrier tests and tests during pregnancy.

A PATIENTS' CHARTER
The care of patients with Cystic Fibrosis. Describes the essential health care people with CF should expect. Drawn up by directors of specialist UK CF Centres in the UK with 50 or more patients.

FINDING OUT
A guide for parents of newly diagnosed children with Cystic Fibrosis.

NUTRITION
Eating well with Cystic Fibrosis – A guide for feeding infants (from birth to one year of age).

CYSTIC FIBROSIS AND YOU
For children up to the age of about 12.

NUTRITION
Eating well with Cystic Fibrosis – A guide for children and parents (from age one to 16 years).

CYSTIC FIBROSIS AND SCHOOL
A guide for teachers and parents of children with CF starting or changing school.

GROWING UP
A guide for young people with Cystic Fibrosis (12 to 18 year olds).

TRANSITION
A guide for young people moving from paediatric to adult care.

NUTRITION
Eating well with Cystic Fibrosis – A guide for adults.

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

TREATMENT
Home Intravenous Therapy and Cystic Fibrosis. A guide for patients, parents and carers.

DIABETES MELLITUS AND CYSTIC FIBROSIS
Describes the incidence and combined treatment.

TRANSPLANTS
Lung and heart-lung transplantation for patients with Cystic Fibrosis.

SUPPORT SERVICE
Outlines the help available from the Cystic Fibrosis Trust Support Service.