

# INTRODUCTION

## A GUIDE FOR PARENTS OF NEWLY DIAGNOSED CHILDREN WITH CYSTIC FIBROSIS

This booklet has been written to help you get a better understanding of Cystic (CF) Fibrosis. It is not intended to replace any advice you may receive from your Specialist CF Centre or CF Clinic. The Cystic Fibrosis Association funds medical and scientific research aimed at understanding, treating and curing Cystic Fibrosis. It also aims to ensure that people with Cystic Fibrosis (PWCF) receive the best possible care and support in all aspects of their lives.

### THE CYSTIC FIBROSIS ASSOCIATION OF IRELAND

The Cystic Fibrosis Association of Ireland was set up in 1963 to increase knowledge and awareness of CF and to give advice and support to people with Cystic Fibrosis and to their families.

The Association:

- Provides support and assistance to people with CF and their families.
  - There are 22 branches of the Association throughout the country, with approximately 2,500 members nationally. The membership base comprises of people with CF, parents, family members and friends of the Association.
- Funds Medical Research:
  - The Association is currently funding four Research Projects which are helping in the vital search for a greater understanding of Cystic Fibrosis. The ultimate aim is to find a cure for CF but in the interim, it is imperative that we promote research that will improve on current methods of treatment.
- Supports specialist CF Multi-Disciplinary posts in hospitals around the country.
- Provides a domiciliary physiotherapy service to people with CF (PWCF). This home-based service is provided to young children with CF and promotes training for parents in new breathing techniques.
- CFAI campaigns both locally and nationally for increased and improved services for people with Cystic Fibrosis. Our aim is to ensure that the best possible services are available to all of our members.



# CONTENTS

What is Cystic Fibrosis?	4
How do you feel?	4
Why does my child have CF?	5
What are the main problems with Cystic Fibrosis	6
How does CF affect the digestion?	7
Other problems in the digestive system or the gut	10
How does CF affect the chest?	10
Prevention and treatment of chest infections	11
Physiotherapy	12
Other problems in the chest	12
Can CF affect other parts of the body?	14
Some other questions parents have asked	14
School years	15
Teenagers and leaving school	16
The future	16
Facts and figures	16
Glossary	17



## WHAT IS CYSTIC FIBROSIS?

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

Normally, the liquids and mucus that line many of our organs are clear, lubricating and helping to protect them from infection.

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 booklet, 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 Association or

other families. Make sure you talk to people who have up to date knowledge of CF, as treatment has improved considerably 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 Respiratory Consultant, possibly in your local hospital, 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 illness 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 1,600 children born in Ireland, 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 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 and, with the latest advances in research, there is every chance that new

therapies will be developed to limit the harm from the disease.

Many people with CF live well into adulthood leading normal lives so it is of utmost importance that your child with CF follows the normal developmental milestones of their peers to prepare them for independence in later life. Independence is something you should speak to your specialist CF team about as your child with CF grows. Independence should be introduced from a very young age.

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. Both your CF team and the Cystic Fibrosis Association are sources 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 and it is important for them to be included in the learning process also.

## 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.

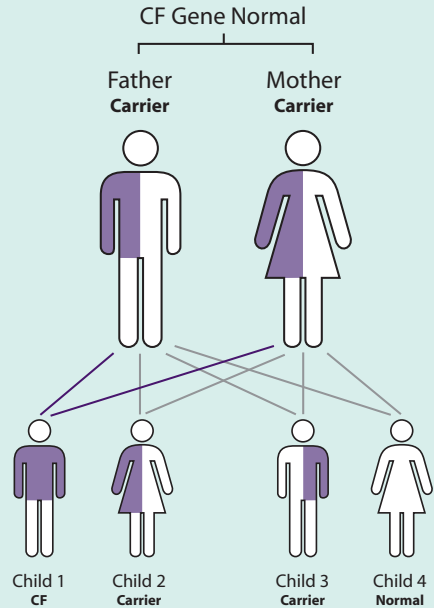
Most carriers of the faulty gene have no idea that they are carriers, because they are completely healthy. Most people find out only when:

- they have a child with CF, or
- a close relative is affected and they are tested

In Ireland, one person in 19 is a carrier and one in every 1,600 babies will have CF.

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.

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. You should ask to speak to your genetic counsellor if you have any more questions in relation to the risks of conceiving a child with CF.

## 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. As a result, the enzymes produced by the pancreas that are needed to digest food do not reach the food. Food will pass through the system undigested. This is often referred to as pancreatic insufficiency and will require enzyme replacement therapy.

This 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 enzyme replacement 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 enough useful function all their lives that they remain pancreatic sufficient (do not require enzyme replacement therapy).

## 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 team 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 and should be encouraged to do so from a young age.

## Which type of pancreatin should my baby have?

This often depends on age. A member of your specialist CF team 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 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 Clinic Dietician 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. It is important that your child with CF follows a diet full of high calories, fat and protein and takes the appropriate enzymes to manage this diet. A low fat/low calorie diet is not recommended for people with CF. Speak to your CF Dietician and Doctor about the appropriate food groups for your child with CF.

## 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 dietician will help you.

You can find more details about different types of milk and food that may be used in the CF Association booklet Paediatric Nutrition & Cystic Fibrosis.

## 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 team and dietician 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 team 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 team and the CF Association booklet on nutrition will help you.

### **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 Clinic 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. Your CF Clinic will advise you about salt supplements.

////////////////////////////////////  
**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, 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 consultant who may arrange further investigation or a change in pancreatin dose. If you are worried about tummy aches, do speak to your consultant.

Extremely loose stools or mild constipation are frequent in CF and may be the cause of stomach cramps. Speak to your CF Dietician and review the enzyme therapy if this persists.

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 Clinic 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 risks 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 to ensure children with different infections are kept apart from each other. Your CF Clinic 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 and 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 team at your CF Clinic 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. It may be worthwhile discussing influenza vaccination with your CF Team for your child with CF.

### 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 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 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.

## **PHYSIOTHERAPY**

### **What is chest physiotherapy?**

Chest physiotherapy is the process of clearing the sticky mucus from the lungs by physiotherapy, breathing exercises and regular physical exercise

### **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 PE 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
- weight loss
- Increased or frequent cough
- tummy aches
- increased sputum
- frequent or loose stools
- change in colour of sputum
- vomiting
- breathlessness
- decreased ability or unwillingness to exercise
- fever
- decreased or poor appetite increased fatigue

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.

## 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 Influenza 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 team. 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 Specialist CF team, which will be familiar with the latest immunisation recommendation 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 as 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 have allowed some men with CF to father children with clinical assistance.

## 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 of Social Welfare is the Domiciliary Care Allowance – an allowance paid to children with a long term disability. This allowance is paid in their own right is not means tested on the family income. All necessary prescription medication and

hospital treatment will be covered under the Long Term Illness Scheme (LTI). You will also qualify for incapacitated child tax credits and you may qualify for a carer's allowance. You may qualify for other schemes such as home improvement grants and diet allowance.

The Cystic Fibrosis Association, the medical social worker at your CF Clinic or the Citizens Information Centre will be able to advise you - providing information on benefits which are available and a guide on how to apply for these. A specific publication is available from CF House with further details on entitlements.

### **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 Clinic and with other people with relevant experience. You should also consider how and when to tell any other children you may have. In general it is better not to be secretive as this may cause unwelcome psychological obstacles for your child with CF as s/he grows older. It is most important to tell people who need to know such as child care staff and teachers.

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 clinic 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 Consultant 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 and that they know how CF affects your child – your child with CF will need to eat more frequently and will use the bathroom more often than their peers. Your child's teachers should be aware of the signs of an infection.

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.

### **Most children with CF attend normal schools and partake in all normal activities.**

The CF Association has introduced a publication to help, called School and Cystic

Fibrosis. For further information or advice please contact the CF Association.

## TEENAGERS AND LEAVING SCHOOL

Adolescence is a challenging period for anyone. It is especially so for young people with CF, particularly if the illness causes them to mature later than their peers. Nevertheless, the full range of further education and employment opportunities should be available to any young person 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 guidance specialists.

As children get older, most will also want to know more about the illness and how it will affect them. It is important to allow them to mature, to educate themselves around CF, and to become responsible for their own selves. This responsibility and independence must be encouraged from a very early age. Encourage your child to manage their own medication and pharmacy orders for example.

## THE FUTURE

Only thirty years ago, the outlook for a baby born with CF was very poor. Today, young persons with CF are living into their thirties, forties, and beyond and these people are leading active and fulfilling lives, studying, working, having relationships and having children of their own.

It is likely that the quality and length of life will continue to improve as a result of current research. New treatments are having a

significant impact on the lives of people living with cystic fibrosis.

The future is without doubt an optimistic one. We must, in the interim, aim to keep every person with CF, both child and adult, as fit and well as possible by controlling their symptoms, to maximise both the quality and the length of their lives.

## FACTS AND FIGURES

- CF was fully recognised only as recently as the 1930's.
- In the past, Cystic Fibrosis was known as 'fibrocystic disease of the pancreas' – the effect of the disease on the lungs was recognised later.
- CF is also known as mucoviscidosis, because the mucus is thick and sticky.
- CF is Ireland's most common life-threatening inherited disease.
- The disease affects approximately 1 in every 1,600 children born in Ireland.
- In the 1930's 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 gene with a healthy version) are underway.

# 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.

## **Cystic Fibrosis**

### **DIOS**

Distal Intestinal Obstruction Syndrome. A blockage of the gut which occurs in older children and 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 are 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 Influenza**

Bacteria which is a common cause of respiratory infection in Cystic Fibrosis.

## **Haemoptysis**

Coughing up blood.

## **Health Care Professionals**

Doctors, nurses, physiotherapists, dieticians, 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

### **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. In the treatment of CF this is a process of clearing the airways of congested mucous by various techniques

### **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.

### **Polyyps**

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**

Mucous material produced by the cells lining the respiratory tract.

### **Staphylococcus aureus**

A bacteria infection that can affect the lungs.  
See: How CF Affects the Chest.

### **Sweat Test**

The test used to diagnose Cystic Fibrosis.



