

A GUIDE FOR PARENTS

of Children Diagnosed with Cystic Fibrosis



**Cystic
Fibrosis**
Ireland





This booklet is not intended to replace any advice you may receive from your Specialist CF Centre or CF Clinic. CFI has sought to ensure the information in this booklet is accurate at the time of publication and will not be held liable for inaccuracies or omissions.

Published by Cystic Fibrosis Ireland, September 2014.

A GUIDE FOR PARENTS OF CHILDREN DIAGNOSED WITH CYSTIC FIBROSIS

This guide aims to give you a better understanding of Cystic Fibrosis and answer some of the questions you may have about how it will affect your child.

SECTION	PAGE
WHAT IS CYSTIC FIBROSIS?	3
How are you feeling?	3
WHY DOES MY CHILD HAVE CYSTIC FIBROSIS?	6
Genetics of Cystic Fibrosis – a closer look	6
HOW WILL CYSTIC FIBROSIS AFFECT MY CHILD?	8
How does CF affect the digestive system?	8
Other problems in the digestive system and gut	12
HOW DOES CYSTIC FIBROSIS AFFECT THE LUNGS?	13
Prevention and treatment of chest infections	13
Physiotherapy	15
Other problems in the chest	17
CAN CYSTIC FIBROSIS AFFECT OTHER PARTS OF THE BODY?	19
COMMONLY ASKED QUESTIONS	20
School years	21
Teenagers and leaving school	21
The future	21
GLOSSARY	22
FOR MORE INFORMATION & USEFUL LINKS	24
NOTES	25

This booklet is intended as a guide for parents whose child or children have been recently diagnosed with cystic fibrosis (CF). It contains information about CF and coming to terms with a new diagnosis. It also includes information on how CF affects the body and what treatments are available, and provides answers to some questions other parents have asked.

WHAT IS CYSTIC FIBROSIS?

Cystic Fibrosis is an inherited disease that primarily affects the lungs and the digestive system. Ireland has the highest incidence of Cystic Fibrosis in the world.

Cystic fibrosis (CF) is a genetic condition that mainly affects the lungs and digestive system.

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

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

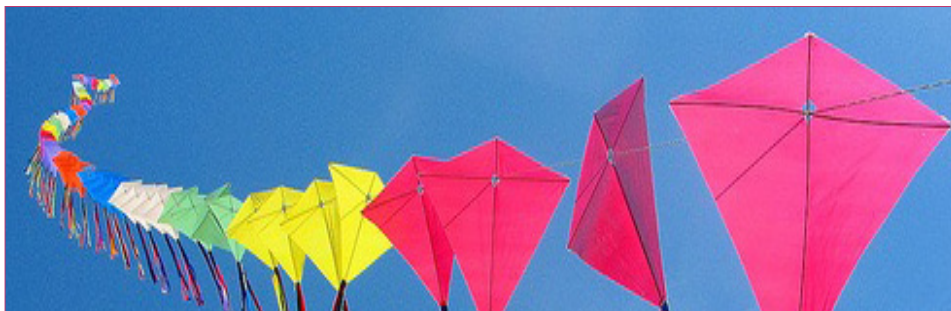
As a parent of a child with CF you will meet many people involved in the care of children with cystic fibrosis, but you will be asked to carry out some of the treatments yourself. To do this effectively, you will need to understand as much as you can about the condition.

All the questions that are answered in this guide have been asked by other parents coming to terms with the news that their child has CF.

HOW ARE YOU FEELING?

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 talk about CF, perhaps with a member of the CF team at the hospital, Cystic Fibrosis Ireland or other families. Make sure you talk to people who have up to date knowledge of CF, because treatment has improved markedly in 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 their child seems well and the diagnosis has been made through newborn screening. It is important at an early stage to be referred to a specialist CF centre to have the diagnosis confirmed and where CF can be fully explained. Write down questions you want to ask as you think of them in case you forget later.

Anger and blame

When you first hear the diagnosis you may be angry, you may feel guilty or you may want to blame somebody. Remember – nobody 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 them, 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 CF immediately and no-one expects you to. It will be a long time before you fully understand CF 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 healthcare professionals will have little experience of it – in some cases, 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.

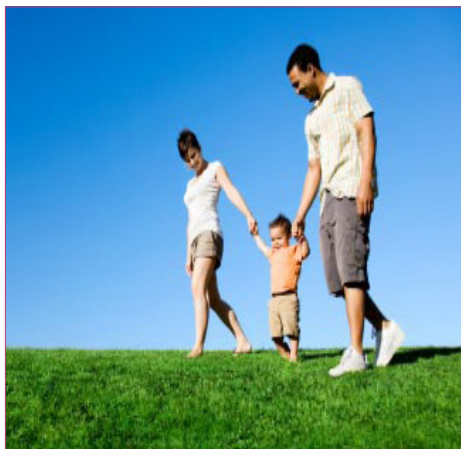
Positive approach

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. With the latest advances in research, there is every chance that new therapies will be developed to further improve the outlook for those with CF.

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 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 Cystic Fibrosis Ireland 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 CYSTIC FIBROSIS?

In this section we will take a closer look at the genetics of Cystic Fibrosis, how it is inherited and what it means to be a carrier of the altered gene.

GENETICS OF CYSTIC FIBROSIS – A CLOSER LOOK

Cystic fibrosis is a genetically inherited condition. A person with Cystic Fibrosis has two copies of an altered CF gene (CFTR), one inherited from each parent.

This gene (CFTR) normally makes a protein that controls the movement of salt and water in and out of our cells. For people with Cystic Fibrosis, this protein is faulty because the altered gene does not function properly.

Approximately 1 in 19 Irish people are said to 'carry' one copy of the altered gene that causes Cystic Fibrosis – these people do not have Cystic Fibrosis.

Both copies of the gene must be altered in order to have CF.

Most carriers of the altered 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 with each pregnancy 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.

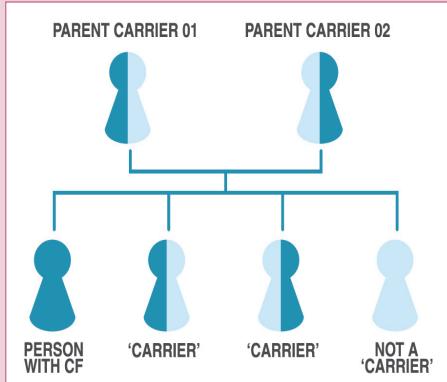
Two carrier parents might have several affected children, or only unaffected children or a mixture of both affected and unaffected children. It is impossible to predict what will happen for any particular pregnancy.

So if carrier parents already have a child with CF, this does not change the risks in the next pregnancy. Each pregnancy



If both parents 'carry' the faulty or defective gene, their child has:

- 25% chance of being born with Cystic Fibrosis
- 50% chance of being a healthy carrier of the altered gene, but not having the disease
- 25% chance of not having Cystic Fibrosis, and not being a carrier of the altered gene



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.

For more information on genetics and CF, check out the CFI Genetics Factsheet.

HOW WILL CYSTIC FIBROSIS AFFECT MY CHILD?

The symptoms of Cystic Fibrosis can vary from person to person. This section outlines some of the more common effects that this illness has on a person with CF.

As cystic fibrosis results in the production of thick, sticky mucus, the ducted organs which are especially affected are those where the mucus has an important job to do, particularly the digestive system and the lungs.

HOW DOES CF AFFECT THE DIGESTIVE SYSTEM?

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

In people 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, hence the name cystic fibrosis.

The effect of cystic fibrosis on the pancreas and digestion varies from person to person. The majority of newborn babies with CF cannot digest milk and without treatment, fail to gain weight and have very loose stools.

At the other end of the spectrum are the 5–10% or so of people with CF whose pancreas retains some useful function all their lives.

Pancreatic Enzymes

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 Dietitian will advise you which preparation is appropriate for your child and how to use it best.

How do I give enzymes to a baby?

Most young babies will take the microspheres removed from the capsules or the mini-microspheres. Enzymes should be sprinkled either directly (Creon for Children) or capsules (Creon 10,000) opened and contents sprinkled onto fruit puree and taken from a spoon. This will hold the granules into a gel and make them easier for the baby to swallow. This can be done from birth. Do not add enzymes to baby's bottle of milk.

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.

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 breastfeeding 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.

Which milk should I feed my baby?

Breast milk or standard infant formula is suitable milk for most babies with CF. Breast milk contains everything needed for growth and development during the first 6 months of life. It also contains antibodies which offer some protection against certain infections such as coughs and colds, ear infections and tummy upsets.

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 Dietician will help you.

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, but talk to your CF Dietitian who will give you specific advice for your baby's requirements.



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 dietitian will advise you.

You can find more details about feeding your baby in the CFI 'Nutrition for Your Baby' information sheet .

How many enzymes will my child need?

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

more enzymes than low fat/low calorie meals.

What if I forget to give the enzymes?

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

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 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 Centre 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 AND 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 Dietitian 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)

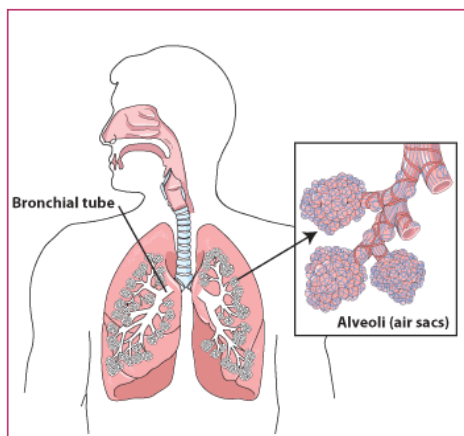
This has nothing to do with the meconium ileus seen in newborn babies but occurs in older children and adults. The bowel becomes blocked by sticky, mucousy motions and food, causing recurrent pain and, sometimes, vomiting.

The cause of this condition is not fully understood but it requires investigation and treatment with special medicines which your CF Centre will tell you about.

HOW DOES CYSTIC FIBROSIS AFFECT THE LUNGS?

This section explains how CF affects the lungs and what you can do to prevent and treat chest infections.

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 which leads to infection.

If not controlled, infections can lead to damage of the lungs. In the early years infections are usually caused by viruses and certain bacteria e.g.,

Staphylococcus aureus and *Haemophilus influenza*. Later on, infections are caused by other bacteria, including one called *Pseudomonas aeruginosa*. You can read more about these bugs in the CFI Information sheet 'Respiratory Bugs Common in People with CF'. 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 as possible from mucus and infection. 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 & 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 other people 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 Centre will be able to explain how they do this.

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

Can antibiotics prevent chest infections?

There is increasing evidence that early, frequent, and in some cases continuous antibiotics can prevent or delay the lung damage in Cystic Fibrosis. The team at your CF Centre will discuss the most suitable approach for your child. Whereas the general population is advised to be

cautious in the use of antibiotics, for those with CF, antibiotics on a regular basis when necessary are invaluable.

How will coughs and colds affect my child?

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

Are cough medicines useful?

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

How are chest infections treated?

A child with a chest infection needs extra physiotherapy (longer and more often) to clear the mucus and infected material from the lungs. S/he also needs antibiotics. The CF Consultant will decide which antibiotics to use by knowing which bacteria are likely to be present and by

taking a sputum sample or swab. The antibiotics will usually be given by mouth as liquid medicine, tablets or capsules. Sometimes they are inhaled as a mist from a nebuliser or given by the intravenous route, i.e., directly into a vein.

Are any special tests needed?

If possible a sputum sample (or swab, cough swab or nasopharyngeal aspirate [PNA or NPA] from a younger child) is taken to identify the bacteria (if any) causing the infection. A chest X-ray and, occasionally, blood tests, may be helpful. Older children may be asked to blow into a tube or machine to see how much 'puff' they have.

This is particularly useful if it is done from time to time in between infections, because the levels often fall before the infection becomes obvious, enabling it to be caught early.

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

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

DNase (Pulmozyme®)

There is now a drug, DNase, which breaks down the sputum and makes it thinner;

this should make it much easier to clear by physiotherapy and coughing. The drug is taken by inhalation from a nebuliser once a day. It does not help all children but your doctor may suggest a trial of treatment if your child has particularly thick and troublesome sputum.

PHYSIOTHERAPY

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

What is chest physiotherapy?

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

Do not be afraid to ask the physiotherapist to watch you doing it from time to time to make sure that you are still doing it as effectively as possible.

When should I start to do physiotherapy?

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

When should I do physiotherapy and for how long?

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

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

Who should do the physiotherapy?

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

Breathing exercises can be introduced in the form of a game from the age of two or

three and, as the child gets older, s/he learns to do his or her own physiotherapy. From about the age of nine, most children can start doing some physiotherapy themselves without help from the family. Most teenagers become completely independent and only require help from time to time.

Will physiotherapy hurt?

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

Do we need special equipment?

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

In babies and small children chest physiotherapy is usually carried out on the adult's lap.

Your CF physiotherapist will advise you which is the best method of chest physiotherapy for your child and whether any specialist equipment is needed.

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

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

Toddlers often like running, jumping and trampolining, all of which are very good for them. When at school, children with CF should take part in 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 Consultant if it occurs.

When should my child see a 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 Consultant.

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 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 CYSTIC FIBROSIS AFFECT OTHER PARTS OF THE BODY?

Yes – other parts of the body can be affected by Cystic Fibrosis, 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 cystic fibrosis 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.

COMMONLY ASKED QUESTIONS

You will probably have lots of questions following the diagnosis of your child. Some of the most common of these are included below.

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 Centre 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 household benefits package. Cystic Fibrosis Ireland, the medical social worker at your CF Centre or your local Citizens Information Centre will be able to advise you. The booklet 'Entitlements for Parents of Children Newly Diagnosed with CF' is available on www.cfireland.ie.

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

Only you can decide but you may wish to discuss it with your CF Centre and with other people with relevant experience. You should also consider how and when to tell any other children you may have. In general it is better not to be secretive.

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

Try to take such remarks in your stride and, if possible, be prepared with explanations. People may also be ill informed or years

out of date in their understanding of CF. They are usually willing to understand and are, if anything, over sympathetic when the situation is explained to them.

Will complementary medicine help?

It is essential for the future health of your child that the conventional treatments your CF Centre recommends are given in the prescribed way. However, provided this is the case, the addition of complementary treatments should do no harm and some families report a benefit. Always consult your CF Consultant and CF Dietitian first.

SCHOOL YEARS

CF 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. The details of your particular child's treatment however 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.

CF Ireland have a booklet called 'School and Cystic Fibrosis' which is available at www.cfireland.ie.

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 adult with CF, depending on their intellectual and physical capabilities. There are some occupations which are less suitable, of course, but these considerations are for the future and decisions can be made at the time with help from the doctor and career guidance specialists.

THE FUTURE

Only 30 years ago, the outlook for a baby born with CF was very poor. Today, young adults with CF are living into their 30s, 40s 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 CF. The future is without doubt an optimistic one. In the interim, we aim to keep every person with CF as fit and well as possible by controlling their symptoms, to maximise both the quality and the length of their lives.

GLOSSARY

Alveolus (Alveoli)

The specialised part of the lung where oxygen enters the blood and carbon dioxide can leave.

Bronchus (Bronchi)

Small airways in the lung.

Cirrhosis

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

Cyst

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

DIOS

Distal Intestinal Obstruction Syndrome. A blockage of the gut which occurs in older children 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 as 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 altered CF gene.

Haemophilus Influenza

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

Haemoptysis

Coughing up blood.

Health Care Professionals

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

Intravenous

Sometimes antibiotics or other medicines are given into a vein rather than by mouth. To make it easier, a small plastic cannula (tube) can be left in the vein so that the drug can be put in through it rather than

by a fresh injection each time. There is a cream available to numb the skin before the cannula is put in it.

Meconium Ileus

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.

Pancreas

A gland that 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

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.

Sweat Test

The test used to diagnose Cystic Fibrosis.

For More Information:

- CFI Parents Information Pack, which includes:
 - Booklet on Entitlements for New Parents of Children with Cystic Fibrosis,
 - Information sheets including Cystic Fibrosis Care, How CF Affects Daily Life, Nutrition for Your Baby, Grants and Support Services for Members, CF Advocate service, Respiratory Bugs Common in people with CF, Genetics Factsheet (CFI)
- Genetic Carrier Testing for CF (CFI)
- Link in with the Social Worker at your CF centre
- Refer to the Citizens Information Website **w:** www.citizensinformation.ie, freephone 0761 07 4000 or call into your local Citizens Information Centre
- Call our CFI Patient Advocates Tomas Thompson or Caroline Heffernan
- Contact the CFI National Office

Useful Links:

- 'Can You See What I See?' and 'School is Fun!' are two books written by Louise Byrne, parent of a little girl with CF. **w:** <http://louisebyrnebooks.com/>
- Getting Nosey about CF with Oli and Nush – A short film made by the CF Trust to help children with Cystic Fibrosis understand their condition and to explain to other children what Cystic Fibrosis is **w:** <http://youtu.be/Wu172eMrIQI>
- CF Voice – An online community for people of all ages living with CF. A place for motivation, inspiration and connection to the CF community **w:** www.cfvoice.com
- ECORN-CF : European Centres of Reference Network for Cystic Fibrosis provides expert advice on Cystic Fibrosis **w:** <http://ecorn-cf.eu>
- The Cystic Fibrosis Registry of Ireland (CFRI) collects and analyses information relating to cystic fibrosis in order to improve the quality of care for all of the people with cystic fibrosis in the Republic of Ireland. **w:** www.cfri.ie
- CF Trust, UK **w:** www.cftrust.co.uk | CF Canada www.cysticfibrosis.ca | CF Foundaiton **w:** www.cff.org |

NOTES

About Cystic Fibrosis Ireland (CFI)

CFI is a registered charity CHY 6350 that was set up by parents in 1963 to improve the treatment and facilities for people with CF in Ireland. It is a national organisation with many Branches around the country.

CFI is committed to working to improve CF services in Ireland and our recent progress includes:

- Lobbying to ensure that the new national adult CF centre in St Vincent's University Hospital was completed with up to 34 in-patient beds for people with CF
- Providing funding towards new CF Units around the country including Crumlin, Drogheda, Galway, Mayo, Waterford, Beaumont and Limerick Hospitals
- Funding research in Cork University Hospital and University College Dublin
- Campaigning to improve the rate of double lung transplantation in Ireland
- Providing advice and expertise

Cystic Fibrosis Ireland
CF House
24 Lower Rathmines Road
Dublin 6
Ireland

t: +353 1 496 2433
f: +353 1 496 2201
e: info@cfireland.ie
w: www.cfireland.ie
Company Reg: 449954
Charity: CHY6350

