

# Shared Care Service for Children and Young People with Cystic Fibrosis

Important information for patients

Mid Cheshire Hospitals NHS Foundation Trust  
Leighton Hospital  
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This leaflet is available in audio, Braille, large print and other languages.  
To request a copy, please telephone 01270 612071.

Produced with kind acknowledgement to Cystic Fibrosis Team, Alder Hey Children's Hospital



## Introduction

If you have just found out that your child/young person has Cystic Fibrosis (CF), or you have recently moved to our CF service from another hospital, we hope this booklet will provide you with information that you need about our team and the service we provide.

This booklet is designed to introduce you to the Leighton Hospital CF service which is part of Mid Cheshire Hospitals NHS Foundation Trust (MCHFT). It will inform you about how we care for children/young people with CF and their families who attend this hospital. If you need more information about CF there are information leaflets available from the CF Trust which explains what CF is and how it is diagnosed and treated. We can give you these leaflets, and we can show you how to register with the CF Trust so that you can be added to their mailing list and receive their regular newsletter.

### Cystic Fibrosis Trust contact details:


Telephone: 020 3795 1555

E-mail: [enquiries@cysticfibrosis.org.uk](mailto:enquiries@cysticfibrosis.org.uk)

Web: [www.cftrust.org.uk](http://www.cftrust.org.uk)

Confidential helpline: 0300 373 1000 or 0203 795 2184

This booklet will repeat some of the information that you have already heard at the time of diagnosis but much may also be new to you. As well as introducing you to the team of professionals who will be involved in your child/young person's care, it explains some of the medicines your child/young person will need to take and the outpatient and inpatient services. Some words or phrases which may be unfamiliar have been underlined and then explained further in a glossary at the end of the booklet.

It also offers some general advice and tips from other parents, and where these are included, you will see this symbol: 

We know that it cannot answer all your questions but please use it as a starting point. Any member of the team will be happy to discuss questions or concerns that you have. If you do not know which team member to ask, the CF Nurse is your first point of contact.

### Who we are?

We are a multidisciplinary team made up of the following professionals:

- Paediatric Consultant
- CF Nurse
- CF Link Nurse
- Dietitian
- Physiotherapist
- Pharmacist
- Children's Outpatient Department
- Paediatric Secretaries

### How to contact us

This booklet is intended to complement the information you will be given by members of the CF Team. Please use it as a reference but also discuss with us any questions or concerns that you have.

Leighton Hospital Switchboard: 01270 255141
Ward 17 for direct access outside office hours: 01270 612073
Alder Hey Children's Hospital Switchboard: 01512 284811
CF Nurse: 01270 612071
Medical secretaries (if you need to contact a doctor during office hours): 01270 612289
Dietitian: 01270 273625
Physiotherapist: 01270 273308
Paediatric Pharmacist: 01270 255141 and ask for Bleep 3236

## What we do

Leighton Hospital has a 'shared care' arrangement with Alder Hey Children's Hospital in Liverpool, to provide care for children/young people with CF.

Our aim is to help you look after your child/young person's CF and to provide you with the information and support that you need. The members of the CF team follow the UK National Guidelines for CF care. We meet regularly to discuss new information on the treatment of CF and how to develop and improve the service to our patients.

There are other people who work closely with the CF team, such as Paediatric Doctors, Ward Nursing staff and Play Specialists. Your child/young person may also receive care from other teams in the hospital depending on his/her particular needs.

The team also works closely with the hospital Pharmacy Department, the Radiology Department where your child/young person will have any necessary X-rays and the Pathology Department where blood from blood tests and sputum go to be analysed. The Paediatric Pharmacist will be available to liaise between the GP surgery and the Community Pharmacist.

Shortly after your child/young person's diagnosis the CF Nurse will arrange to visit you at home. They will provide advice and help you to get the daily treatment underway. The Dietitian and Physiotherapist will either arrange to see you at home, or at the hospital as soon as possible after your child/young person's diagnosis. Thereafter you will be given appointments to attend the CF outpatient clinic in the Children's Outpatient Department at Leighton Hospital. These clinics are held on Tuesday afternoons, where you will usually see all members of the team at each appointment.



**Feel free to contact the CF Nurse at any time. If no-one is available to take your call you can leave a message on the answerphone and one of the CF Nurses will call you back.**

## Key people in the community

The CF team work in the hospital but alongside them are many other professionals who will need to know about your child/young person's CF.

### General Practitioner (GP)

Your GP and surgery staff are very important in the management of your child/young person's health and well-being. Following a new diagnosis of CF, a letter will be written to the GP by the Paediatric Consultant. This will explain how the diagnosis was made and what treatment is required. The GP will be asked to prescribe all the regular medicines your child/young person will need.

After each CF clinic a letter will be sent to your GP to keep him/her informed of your child/young person's progress and any changes in treatment or medication.



**Make an appointment to see your GP as soon as you can and take a list of your child/young person's medicines to discuss with him/her. If you have changed your child/young person's GP you need to let him/her know of your child/young person's CF diagnosis.**


### Health Visitor

If your child/young person is under five years of age they will have a Health Visitor responsible for their health and development. Your Health Visitor can also give support and information about health promotion, immunisation, parenting, local services and amenities available, nurseries and schools.

## Pharmacist

You may or may not have met your local Pharmacist (chemist). He/she will play a key role in ensuring that you have a regular supply of the medication your child/young person needs.

Your local pharmacist will need to have a prescription from your GP to provide the correct medication. Some of the medicines you need may have to be specially ordered. This can take a few days, so always allow plenty of time to get replacement medicines.

 **Introduce yourself to your chemist and take a list of your child/young person's medicines to discuss. If you keep to the same chemist, they will become familiar with you and your child/young person's medicines.**

## Nursery, school and college

If your child/young person is at nursery, school or college it is important that the teachers and staff are aware that your child/young person has CF. The CF Nurse can visit the nursery, school or college with you, and explain about CF to the staff. This will make them more aware of the special needs your child/young person will have, and more confident about working with your child/young person. Every school has a School Nurse who will also be invited to this meeting if you wish.


If children/young people with CF are admitted to Leighton Hospital the play specialists will discuss the most appropriate way of keeping up with school work. Whenever possible, children/young people are encouraged to attend school between IV antibiotics if they are well enough to do so.


## Friends and family

The diagnosis of CF can affect all family members. Individuals react in different ways and this can cause problems initially. Brothers and sisters may be confused and upset, especially if admission to hospital is necessary. They may also be a little jealous of the attention they see their brother or sister getting. It is important to keep them informed and involved, and to keep to family routines as much as possible. Try to make some special time for you and your other children/young people and remind them how much you care for them.

If there are times when you or other family members find your child/young person's CF too difficult to cope with, please let us know and we will do what we can to help. Speak to any member of the CF team and they will try to help you through the ups and downs.

If you wish, other family members or friends can be taught about the care your child/young person needs. Sharing the care can take off some of the pressure from parents and give them time to themselves and the other members of the family.

 **Consider accepting offers of help, particularly in the early days. Friends and family usually want to help, and often it is the small practical matters that are most helpful, like ironing, cooking or collecting others from school.**

 **Have some of the CF Trust information about CF and this booklet available for close friends and family to read. In this way you may not have to explain CF so many times to others.**

## The Cystic Fibrosis clinic

Children/young people with CF are seen on a regular basis, usually every eight weeks as recommended by the CF consensus guidelines (CF Trust – Standard of Care 2001). Children/young people will be seen more often immediately after diagnosis, or if they have been unwell.

**At Leighton Hospital the clinics are held on a Tuesday afternoon in the Children's Outpatient Department. Occasionally, some clinics may be virtual. For these clinics, you will be emailed a link to join the virtual clinic – please remember to check your junk folder in your email. Your child should be present during a virtual clinic.**

If the appointment is in the Children's Outpatient Department, you will be given an appointment time and when you arrive you will be allocated a room, which you will stay in throughout the clinic. This is to reduce any risk of cross infection by mixing with other children/young people who have CF. As your child/young person will be seen by several members of the CF team the appointment will last approximately two hours.

The Play Specialists will provide play materials for your child/young person to use during the clinic.


One of the CF Nurses will be available at each clinic to co-ordinate your child/young person's visit and provide information and support.

 **Remember to bring any feeds, enzymes and nappies that your child/young person will need during the clinic.**

Your child/young person will first have their weight and height done before being seen by the Dietitian, then the Physiotherapist, followed by the Nurse and the Consultant.

The dietitian will see your child/young person at every clinic. Normal weight gain and height are important factors in keeping your child/young person well. The Dietitian will ask you about appetite, pancreatic enzyme use, bowel habits and eating behaviours. She will then be able to give advice to help keep your child/young person well nourished.

The physiotherapist will see your child/young person at every clinic. Regular physiotherapy is an important factor to keep your child/young person well. The Physiotherapist will do lung function tests if your child/young person is old enough to do them and will also take a cough swab or a specimen of sputum.

 **If possible, it is better to collect a cough swab or sputum specimen two weeks before clinic. You can also be taught how to do this when you are ready to do so.**

**The CF Nurse is able to do this at home.**

The Consultant will see your child/young person with the CF Nurse. During the clinic a record sheet moves around with your child/young person, and the notes from the Physiotherapist and Dietitian will be looked at. This allows the Consultant to see all aspects of your child/young person's care and review their treatment with you.

After all of the children/young people have been seen in the clinic, the CF team will get together and review everyone's progress and have all information that has been discussed.

### **Important things to remember about clinic:**

- If you cannot keep an appointment, please telephone the CF Nurse on 01270 612071 or the Children's Outpatient Department on 01270 612288 to make another appointment. Please do not telephone the hospital appointments number, because the appointment times need to consider which order children/young people are seen in clinics. This depends on the organisms (bugs) that are in their sputum/cough swab.
- Please do not come to the clinic without an appointment, as this may pose a cross infection risk to your child/young person or another.
- If your child/young person is unwell do not hesitate to contact us. We can arrange for them to be seen by the Paediatric team in the Children's Assessment clinic on a separate day.

- If you have an emergency, telephone the Children's Ward on 01270 612073 and arrange for a review.

## **Keeping your child/young person well**

### **Annual Review**

Each year your child/young person will have extra tests as a general review of their condition and how they are progressing. The 'Pre annual review clinic' will be held in Children's Outpatient Department from 8.30am to 12.00noon. School age children/young people will need to be absent from school for the whole morning. The CF Nurse will arrange the clinic and co-ordinate the activities of the morning.

### **This clinic includes:**

- Blood tests and chest X-ray

**Your child/young person will also have an abdominal ultrasound scan at Alder Hey Children's Hospital on another day. You will receive an appointment letter for this in advance.**

At the annual review clinic you will see the CF team from Alder Hey Children's Hospital and Leighton Hospital. They will go over the results with you and make a plan for the next year. This is a very important clinic visit as it gives us the opportunity to talk together about the year's progress. Following the clinic visit you will be sent a letter with all the results that have been discussed with you. A copy of this letter is also sent to your GP. Children/young people over the age of 13 will receive a copy of this letter for themselves.

### **Growing older with Cystic Fibrosis**

The CF team believes in encouraging children/young people to take increasing responsibility for their CF management. As part of this, all young people from the age of 14 are invited to see the Doctor on their own for the first part of the clinic consultation. Parent/s then join their child/young person for further discussion and the physical examination. We expect your child/young person to develop a sound knowledge of CF and an understanding of their treatment by the age of 17, when they will move to the adult services.

### **What if my child/young person becomes ill?**

If you are worried about any aspect of your child/young person's CF, please telephone the relevant person i.e. Dietitian, Physiotherapist, Nurse or Doctor listed at the front of this booklet.

If it is a general question, please contact the CF Nurse or leave a message on their answering machine. Someone will get back to you as soon as possible, usually within 24 hours. If you would like an extra cough swab taken, please telephone the CF Nurse who will arrange a time for this. You can contact the CF Nurse three or four days later for the result if you are worried.

If your child/young person is unwell outside normal working hours (9.00am – 5.00pm, Monday to Friday, including Bank Holidays) you can use your "direct access" letter and contact the Children's Ward on 01270 612073.

Sometimes your child/young person will need a back-up antibiotic. This is a second antibiotic which we suggest you have in the house to start if your child/young person develops extra symptoms such as an increase in cough, phlegm or a runny nose. At first we would like you to telephone us for advice about starting a back-up antibiotic but, in time, you will feel confident enough to know when to start this treatment. Before you start this back up antibiotic it is best to collect a cough swab or sputum specimen and send it to the Pathology lab at Leighton Hospital.



Recurrent abdominal pain (tummy ache) is a common problem. Occasionally this can be due to a sticky mucus mass in the bowel called DIOS (distal obstructive intestinal syndrome) and will require extra treatment. Please let us know if your child/young person has abdominal pain and we can investigate this and provide appropriate medication if necessary. If this is out of normal working hours, then please contact Ward 17 to arrange a review.

## Coming into hospital

There will be times when your child/young person may need to be admitted to hospital. Children/young people are admitted to the Children's Ward, which is on Wards 16 and 17. In order to help prevent cross-infection we ensure that each child/young person has an individual room (cubicle). On admission to a ward your child/young person will have a number of tests. These will be explained at the time.



**Please remember to bring your child/young person's medicines.**

If your child/young person is in hospital he/she will be seen by a Doctor every day as part of a daily ward round. Different members of the CF team will visit regularly and other specialist teams may also need to see your child/young person.

Parents are allowed to be with their child/young person at any time. One parent will be able to stay overnight with their child/young person in the cubicle.

There are Play Specialists on the Wards from Monday to Friday. They will help keep your child/young person occupied during the day.



**Many children/young people like to bring their own bedding to make their room feel more 'homely'.**

In time, parents may be taught how to do treatment that is usually given in hospital, such as **intravenous antibiotics** (IVs). However, home IV antibiotics are only possible with certain antibiotics.

## Living with Cystic Fibrosis

### School

When children/young people first start school or nursery the CF Nurses are happy to discuss CF with the staff and reassure them that there is nothing in a school day that will be affected by the CF treatment. Visits can also be made when children/young people move on to high school. The only difference for most children/young people with CF is that they need pancreatic enzymes when they have their lunch or snack.



**If your child/young person has a packed lunch, tape a small sandwich bag to the inside of the lid of the lunch box and put the enzymes inside. Check that this is acceptable to the school beforehand.**

### Holidays

Holidays are important for the whole family. The CF team can advise about insurance, flying, sun protection and extra medicines, such as salt which your child/young person must take if he/she going abroad to a hot country. The team will provide you with a letter explaining your child/young person's medicines and information about caring for children/young people with CF in other countries.

### Genetic Counselling

A genetic counselling service is available from Alder Hey Children's Hospital. If you would like an appointment with one of the genetic counsellors we can arrange this for you. Other family members may also wish to discuss the option for a test to find out if they are carriers of the CF gene.

## Smoking



A smoky atmosphere is very bad for a child/young person with CF, so we ask parents, family, and friends not to smoke.

## Keeping up-to-date with knowledge about Cystic Fibrosis and treatments

If you are a member of the CF Trust you will receive regular newsletters and information about progress in the understanding and treatment of CF.

The CF team at Leighton Hospital occasionally produce a short newsletter called the CF Link. If you are not happy to receive a copy of the newsletter please let us know at clinic.

Generally we find that with regular treatment, medication and reviews, children/young people with CF can remain healthy and maintain a good quality of life. We encourage children/young people to participate in all normal activities and to view CF as something they are living with rather than something they are. As children/young people grow older we will aim to help parents prepare their children/young people for more independent life and self-care with respect to their CF.

## A – Z of Medication

**If you have any concerns about whether a medication is safe for your child/young person, please contact us and we can discuss this with you.**

**Antibiotics (daily):** From the time of diagnosis we may ask you to give your child/young person a daily antibiotic to protect against a germ (bacteria or bug) called *Staphylococcus Aureus* (Staph. for short). This is a germ which is everywhere and cannot be avoided. It usually does not affect healthy people but can cause problems for people with CF. Your child/young person will not become immune to this antibiotic and it is safe to give long-term. However, as with all antibiotics, side effects, such as a rash or loose stools, can occasionally occur. If this happens there are other medicines we can use. Antibiotics are available in a liquid for young children/young people and as capsules when they are able to swallow these with a drink.



**We can help you with ways to give your child/young person's medication, if they cannot tolerate it, we may be able to suggest alternatives.**

**Back-up antibiotics:** This is an extra antibiotic which we suggest you have in the house to start promptly if your child/young person develops extra symptoms, such as an increase in cough, runny nose, wheeze etc. We will ask your GP to prescribe this. It can be kept in a powder form and made up with water when needed. Once the water is added it has to be used within a certain length of time. This information will be found on the bottle under 'Expiry once reconstituted'.

At first we would like you to telephone us for advice about starting a back-up antibiotic. In time, you will feel confident enough to know when to start this treatment. Remember to give the usual daily antibiotic as well.

**Cough medicines:** We suggest that you do not use any cough medicines. These can dry secretions which would prevent sputum being coughed up. If your child/young person has a cough that is not clearing with a back-up antibiotic, let us know. We will discuss alternative treatments.

**Enzymes:** To digest food we rely on our pancreas to release digestive enzymes. Most people with CF need enzyme supplements. This is because they cannot produce enough themselves to



properly break down, digest and take-in all the nutrients from their food. If food is not digested and absorbed properly then some of the valuable nutrition will be lost in the faeces (poo).

To help your child/young person digest their food properly we will ask you to give enzymes with all meals and most snacks. The enzymes are called Creon Micro, Creon 10,000 or Creon 25,000. The Dietitian will give you advice about how much enzyme to give with different foods and which foods your child/young person can have without enzymes as well as practical suggestions on how best to give the enzymes.



**The Creon granules or opened Creon capsules can be given with a spoonful of fruit puree or yoghurt to help them slide down easily.**

**Fluoride:** If you live in an area where there is little fluoride in the tap water, a fluoride supplement may be necessary. Discuss this with your child/young person's Dentist. It is important for a Dentist to see your child/young person regularly.

**Flu vaccine:** We recommend that all children/young people with CF get the flu vaccine each year. This can be arranged with your GP who will organise special flu vaccine clinics in October each year. The first time the vaccine is given there will be two doses at four to six weeks intervals. After that it will be one dose each year.

**Immunisation:** Normal full immunisation is essential and safe for children/young people with CF, even while taking antibiotics. Your Health Visitor will organise the immunisations. Immunisation against influenza (flu) is recommended too.

**Lactulose:** Some children/young people with CF need to use this medicine to soften their faeces and help them open their bowels regularly.

**Nebulised antibiotics:** As well as taking antibiotics by mouth, some children/young people with CF need to have nebulised antibiotics regularly at home. This means having a special small electrical pump which produces a fine spray of the antibiotic which is then breathed into the lungs through a soft mask or mouthpiece. If your child/young person needs nebulised antibiotics the Physiotherapist will show you how to do this. This treatment can be very effective for certain chest infections.

**Pain killers:** From time to time your child/young person, like any other child/young person, may require pain killers such as Paracetamol. It is safe for your child/young person to take these at the normal dose.

**Salt supplements:** People with CF can lose more salt in their sweat than those who do not have CF. This is of most importance for babies under one year and for older children/young people during the summer months. We suggest a salt supplement be taken daily until babies are one year old and from then on, between May and September each year. Anyone taking a winter sun holiday should take salt supplements while they are away.



**The taste of salt can be disguised in a small amount of milk.**

**Vitamins (Paravit-CF):** Vitamins A, D and E are fat soluble vitamins because they are absorbed with fat, and people with CF may have low levels of these vitamins in their blood because fatty foods are not well digested. For this reason we will give your child/young person vitamin supplements to keep his/her vitamins at the right level. This is available as a liquid or tablets for those able to swallow them. Once a year, at your child/young person's annual review, we will take blood to measure the vitamin levels.

**CFTR Modulators** – these medications are aimed at targeting specific gene mutations (therefore are not suitable for all of our CF patients). If your child is eligible for them, then we will discuss this with you when they are at an age that they can start them. Trials are still ongoing for other (rarer) mutations!

**Most medicines should be collected from your local chemist. If you need to collect some from Leighton Hospital Pharmacy, the opening hours are 9.00am - 5.00pm weekdays (excluding Bank Holidays), and 9.00 am - 12 noon Saturday.**

## **Glossary**

**Cough swab:** The Physiotherapist or CF Nurse will use a swab, which looks like a large cotton bud, to take a sample of mucus from the back of your child/young person's throat. This may feel uncomfortable but is not painful.

**Exercise tolerance:** This is a measure of the amount of exercise or activity your child/young person can do before he/she gets tired or out of breath.

**Intravenous antibiotics** (also called IVs): Antibiotics which are given directly into the blood via a cannula in the vein rather than taken by mouth (orally).

**Mucus** (also called spit, sputum, phlegm, secretions): Mucus is produced by everyone's lungs to keep them moist and clean. CF causes the mucus to be thick and sticky and more difficult to move.

**Nebuliser / Neb:** A small machine which pumps air through a container holding medicine and turning it into a mist. The mist is then breathed into the lungs via a mask or mouthpiece.

**Oxygen saturation** (or 'sats'): A machine that measures how much oxygen your child/young person is breathing in via a sensor which is clipped to your child/young person's finger. This test is sometimes carried out overnight to see if extra oxygen is needed.

**Pathology department:** This department carries out the tests on blood samples, cough swabs and sputum samples.

**Pulmonary function test** (also called lung function, spirometry or 'blows'): From the age of five years, each time your child/young person comes to clinic they will be asked to blow into a machine which measures how much air your child/young person's lungs can hold.

**Radiology department:** This department is responsible for taking X-rays and ultrasound tests which can give helpful information about your child/young person's lungs and abdomen. Your child/young person will have an X-ray every year at his/her annual assessment and at others times as the Doctor feels necessary.

**Ultrasound scan:** An ultrasound scan is a painless procedure that uses high frequency sound waves to create an image of an organ in the body.