

## THE CARE OF PATIENTS WITH CYSTIC FIBROSIS

**This factsheet describes the essential health care which children and adults with Cystic Fibrosis (CF) should expect to receive. It is based on guidelines drawn up by directors of Specialist CF Centres in the UK and is based on current best practice.**

### Comprehensive care

*Patients should have access to comprehensive care from a specialist front line team which consists of:*

- a consultant with a special interest in Cystic Fibrosis (paediatrician or adult physician)
- a junior doctor (usually a registrar)
- dietitian
- CF nurse specialist
- physiotherapist
- social worker

This team should be supported by a number of other specialists.

Children who live some way away from a recognised Specialist CF Centre can sometimes receive some CF care from their local district hospital, but this is only advisable if the local hospital has a proper shared care clinic with well recognised links with the CF Specialist Centre. When this arrangement is agreed, sometimes children with CF will visit the CF Centre but on other occasions they will see their local CF team. In some places the Specialist CF Centre team will attend appointments at your hospital on a regular basis, so children with CF get the benefit of both teams.

***Adults with CF should attend a major Adult Specialist CF Centre for all their hospital care.***

### Diagnosis and information

When the diagnosis of CF has been confirmed (usually by two positive sweat tests and a blood test to determine CF gene), and a full assessment of the patient's condition has been completed, the family should receive a full explanation of Cystic Fibrosis, together with information about treatment from the multidisciplinary CF team. At this stage the family should be offered genetic counselling. Suitable written information should be provided for the family to take away and absorb.

Members of the team should always be prepared to answer questions and to give families or patients whatever further information, explanation and support they need. Information should be given about the Disability Living Allowance and how to contact the Cystic Fibrosis Trust, as well as any local branches or groups. Details of the Cystic Fibrosis Trust support helplines should also be given. There are three helplines. For further information and a listening ear, the helpline number is ☎0845 859 1000, the Benefits Helpline is ☎0845 859 1010 and the Welfare Grants Helpline is ☎0845 859 1020. You will be offered a new parent pack, or a newly diagnosed adults factsheet, from the Trust that will give you further information and sources of support.

## Physiotherapy and diet

Patients and parents should be taught techniques by an experienced CF or specialist physiotherapist and be given advice on diet by an experienced CF or specialist dietitian.

## Your GP

The general practitioner (GP) should be given full details of the diagnosis and treatment by the Specialist team and should, where appropriate, prescribe the necessary medicines.

## Emergency treatment

Patients and parents should be given a telephone number for direct access to the hospital CF team for urgent treatment or advice.

## Outpatients

Outpatient supervision should be regular, usually at two or three monthly intervals in a recognised CF clinic. It will be more frequent with the babies and young children, with ample time for discussion and advice.

Patients should always see the consultant or another doctor experienced in Cystic Fibrosis. They should also regularly see their CF specialist nurse, physiotherapist and dietitian.

## What should happen at your clinic visit

### Every visit

- Measurement of weight and, in children, height.
- Clinical update and examination.
- Dietary and pancreatic enzyme review.
- Culture of the sputum or cough swab.
- Respiratory function tests in children from 6 years, and adults for example, FEV<sub>1</sub>, FVC, Oximetry.

### **At least once a year**

- Physiotherapy review.
- Nutritional review.
- Chest X-ray.
- Blood tests.
- Respiratory function tests.

### **As and when appropriate**

- Instruction and continuing support when carrying out home intravenous (IV) antibiotic therapy and additional or new treatment.
- Further genetic counselling.
- Interview with a social worker.
- Advice on education and schooling and for adult patients, on further education, employment, pregnancy, fertility and entitlement to allowances.

## **Hospital admission**

If patients need to be admitted to hospital, it should be to a ward where the staff are familiar with the problems of Cystic Fibrosis. Cubicle accommodation is recommended to prevent cross-infection.

## **Paediatric to adult care**

There should be joint clinic, or some other form of liaison, to make the transfer of a patient at around the age of 16 from the children's clinic to the care of an adult physician as easy as possible.

*If you have any queries whatsoever about the care you are receiving, you should raise them with your CF consultant or other members of the CF team.*

*For further help or for more detailed information, please ring the Cystic Fibrosis Trust on ☎ 020 8464 7211 or check our website, [www.cftrust.org.uk](http://www.cftrust.org.uk)*

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