The Leeds Teaching Hospitals NHS Trust

Information for Community Pharmacists about Cystic Fibrosis (CF)

This leaflet is intended to help you with dispensing prescriptions for patients newly diagnosed with CF.

It provides some limited information about the drugs that a newly diagnosed patient is likely to be taking and some other information that will help your pharmacy provide a comprehensive service to the patient and support the family.

The Paediatric Regional Cystic Fibrosis Unit is based at Leeds General Infirmary.

The Adult Unit is based at St. James's University Hospital.

The Leeds Unit is internationally renowned as a centre of excellence, providing care for over 600 patients in total.

CF is a multisystem disease characterised by:

- Recurrent lower respiratory tract infections;
- Pancreatic insufficiency (90% of CF patients);
- High sweat chloride.

CF is caused by a single gene defect on chromosome number seven. Many different genetic mutations have been described (currently over 2000), and increasingly, these have shown how variable the clinical presentation of CF can be. The most frequent mutation among CF patients is deletion of phenylalanine 508 (p.Phe508del.)

- It is the commonest life-threatening hereditary disease in Caucasians.
- It is an autosomal recessive disease. It requires both parents to be carriers of the defective gene. There is a one in four chance of every child they have having CF and a one in two chance of having a child who is a carrier of the defective gene.
- In the UK, the carrier frequency is 1/25 and the incidence is about 1/2500 live births.
- In the UK, there are over 10,000 patients with CF and more than half are adults.

Pathology

The genetic defect leads to the production of an abnormal protein called the CF transmembrane regulating protein (CFTR). CFTR controls chloride ion transport across cell surfaces. In CF, chloride ion transport is impaired, resulting in an abnormally concentrated fluid on epithelial surfaces in the lung and elsewhere. This makes the airway secretions very viscous and difficult to clear, predisposing the lung to chronic pulmonary infections and consequent lung damage. The presence of CFTR throughout the body explains the multisystem nature of CF, also affecting the sweat and salivary glands, the pancreas, gastrointestinal tract, liver and male reproductive system.

Pancreatic insufficiency affects 90% of patients with CF, resulting in steatorrhoea (fatty, offensive stools) and faltering growth, due to malabsorption, predominantly of fat. In the teenage years patients may develop CF related diabetes due to further pancreatic damage. Liver dysfunction is also seen in some patients.

Prognosis

Significant improvements in early diagnosis and treatment have increased the life expectancy of patients with CF. Currently half of the CF population will live past 47 years of age, and improvements in treatments mean a baby born today is expected to live even longer.

Therapy consists of three major components; drug therapy, nutrition and chest physiotherapy.

In recent years there has been a huge advance in the treatment of cystic fibrosis with the availability of CFTR modulator therapies. These drugs dramatically improve the quality of life and life expectancy for those patients eligible for treatment, which is currently 90% of people with cystic fibrosis. Choice of drug depends on the specific CF mutations and drug licence. It is worth noting that these drugs have significant drug interactions, therefore always check when dispensing additional extra medications.

The role of the community pharmacist

Patients with CF will be on long-term medication and when diagnosed will be started on the drugs listed on page 6-7. As the child becomes older, and if the disease progresses, it is likely that the medication will increase. Parents are advised to find a pharmacy that is able to provide a service that will help ease the burden of the daily routine of medication and develop a working relationship with the pharmacy.

Parents of newly diagnosed patients, understandably, have a lot to contend with, and anything that can help with the practical issues is appreciated. Experience shows that a mutual

understanding between parents and pharmacy helps to smooth the drug supply process, particularly when drugs are unlicensed, difficult to obtain or required as soon as possible.

Although patients with CF will usually have new drug therapy initiated at the hospital, community pharmacists have a very important role in ensuring that drug therapy and obtaining supplies within the community is uncomplicated.

If your pharmacy offers a 'Repeat Prescription Scheme' or a delivery service, it would be appreciated if this could be discussed with the parents.

Other things that can help:

- Please supply oral syringes with all liquids, as with repeated use and washing the markings on the syringes are removed and replacements are often required.
- Encourage GPs to prescribe adequate amounts, sufficient for the full month.

Additional treatment

When patients with CF get a chest infection, they are treated promptly with oral antibiotics e.g. amoxicillin, co-amoxiclav. The dose used is likely to be higher than in a non-CF patient, but the dose can usually be found in the BNFc.

If a patient becomes unwell and is found to be growing pseudomonas in their sputum, they will be prescribed oral ciprofloxacin and nebulised antibiotics (e.g. Colomycin®, Promixin® or Tobi®). The course of this treatment is likely to be three months initially. If you require further information about these drugs please contact the CF Unit/Pharmacist.

As the disease progresses it is likely that the list of medications will increase. This is not covered within the scope of this information but further information can be obtained from the contact numbers below.

Paediatric Cystic Fibrosis Unit: **0113 392 7125**

Outpatient Pharmacy: **0113 392 3308**

Paediatric Cystic Fibrosis Pharmacist: Helen Cunliffe, direct line: **0113 392 5250** or **0113 243 2799 bleep 80-2534**

Newly diagnosed babies will routinely be started on the following medicines:

Drug	Dose	Notes
Abidec® or Dalivit® This is prescribed as a vitamin A&D supplementation	0.6mL-1.2mL od Dose adjusted according to annual blood levels	These are not routinely interchangeable, as Dalivit® contains more vitamin A. When Abidec® is unavailable due to manufacturing problems Dalivit® may be prescribed instead by the CF team who will check whether the dose needs to be reduced. Once opened it should be used within 4 weeks.
Vitamin E (alpha-tocopheryl acetate) Vitamin E supplementation	25mg-100mg daily Dose adjusted according to annual blood levels	Suspension available from wholesalers. Alliance Pharmaceuticals recommend discarding the suspension one month after opening. Please ensure 500mg/5mL alpha-tocopheryl acetate suspension is prescribed.

Paravit-CF® A multivitamin preparation containing the fat-soluble vitamins, A, D, E & K, specifically designed for people with CF.	0.1mL-0.25mL daily. Dose adjusted according to annual blood levels	This preparation reduces medicines burden but is not suitable for all patients.
Creon Micro® Pancreatic enzyme to assist the digestion of starch, fat and protein. Taken with feeds/ meals/snacks	The dose is variable between patients and is dependent on the fat content of the feed/snack	Parents will be advised by the dietitian at the CF Unit on the dose and how to administer the enzymes. Please ensure patients are dispensed sufficient for one month.
Flucloxacillin Prophylactic antibiotic against Staph. aureus infection, preventing respiratory infections	Dose based on child's weight and usually given twice a day. 3-5kg: 125mg bd 6-8kg: 175mg bd 9-15kg: 250mg bd 15-20kg: 250-500mg bd Over 20kg: 500-1000mg bd	Once reconstituted some formulations expire after seven days. The parents may wish to obtain the flucloxacillin as 'dry powder', and reconstitute it themselves. The CF Unit provides the parents with information on how to reconstitute an antibiotic, but you may wish to advise on the required volume of water. Please ensure sufficient is provided for one month i.e. 4 bottles, if supplied as dry powder. The expiry is 7 days once reconstituted.

Sodium chloride oral solution 1mmol/mL (292.2mg/5ml) Sodium supplement for some babies with low urinary sodium levels. Patients with CF can be deficient in sodium, which may delay growth in babies	1-2mmol/kg/day in divided doses	Please advise parents about the expiry and storage conditions of the product as it depends on the manufacturer.

These medications are all long-term, with the exception of sodium chloride solution, which may be stopped when the child is weaned with sufficient quantities of foods that contain sodium. It would therefore be helpful if sufficient stock of the above is maintained within the pharmacy.

For a general overview about CF visit the Cystic Fibrosis Trust website, **www.cysticfibrosis.org.uk**

What did you think of your care?

Scan the QR code or visit **bit.ly/nhsleedsfft**

Your views matter

Developed by: Helen Cunliffe, Pharmacist.
Produced by: Medical Illustration Services • MID code: 20210715_004/NR

LN004521
Publication date 08/2021
Review date 08/2023