- Inform the CF team if repeat prescriptions are not being collected
- Arrange an annual flu vaccination
- Inform Diane Kingswood, the paediatric hospital pharmacist (023 9228 6000 bleep 1499) if you have difficulty in obtaining medications such as vitamins
- Send any cough swabs or sputum that a family obtain to Portsmouth Pathology department on the same day as collection – Please ensure the form has 'Cystic fibrosis' marked on it as samples are processed in a specific way in patients with CF
- Prescribe and dispense antibiotic requests for chest infections from the CF team as soon as possible preferably within 24hours
- Refer any children/young people with CF presenting with upper and lower respiratory or GI signs and symptoms to the CF team (023 9228 3330) or if urgent or outside normal working hours to CAU (023 9228 3344)
- Liaise with the CF team should you discover irregular pathology results relating to the child/young person
- Encourage all GP practice & pharmacy staff to read this leaflet
- Please let the CF team know if there is anymore information that you feel would be useful in your interaction with children and young people who have CF

Further information can be found at:
https://www.cysticfibrosis.org.uk/

Author: Paediatric CF Team - Version 1
Produced: Sept 2013
Review: Sept 2015
Ref: PAE/064
Medical Illustration ref: 13/2288

Produced by
Medical Photography
& Illustration
mpandi@porthosp.nhs.uk
mp+i

CYSTIC FIBROSIS KIDS OF PORTSMOUTH
REGISTERED CHARITY NUMBER 1115580

Portsmouth Hospitals NHS
NHS Trust

Cystic Fibrosis

Information for Health Professionals

This leaflet has been produced by the Portsmouth Hospitals NHS Trust Paediatric Cystic Fibrosis Team. It aims to provide health professionals based in the community setting (including GPs, GP reception staff, pharmacists, school nurses and health visitors) with an introduction to cystic fibrosis, it's management and the various roles of both the specialist and the community teams when supporting individuals with CF and their families.

CYSTIC FIBROSIS

Cystic fibrosis (CF) is a life limiting and life long inherited disease. Many body systems/organs can be affected by the effects of CF, however the lungs are always involved in all individuals with CF. Most individuals with CF have fat malabsorption and fat soluble vitamin malabsorption due directly and indirectly to an affected pancreas producing insufficient pancreatic enzymes into the small intestine. Less common complications include gastro-esophageal reflux, CF related asthma, sinusitis, nasal polyps, liver disease, diabetes, arthritis and osteoporosis.

Without aggressive and responsive respiratory management and correction of malabsorption the prognosis for individuals with CF would be poor due to progressive irreversible lung disease and malnutrition. Fortunately in recent times individuals with CF are managed by increasingly knowledgeable specialist CF teams and along with the current treatments available the prognosis for someone with CF has greatly improved.

The right psychosocial support and good adherence to treatments, which includes respiratory and nutritional management is essential. With timely interventions for acute respiratory exacerbations and lots of exercise it is realistic to expect an individual with CF to live well into adulthood.

HOW CF AFFECTS THE LUNGS

In an individual without CF, mucus is produced by mucus glands in the airways. This mucus traps harmful bacteria that have been breathed in. They can then be removed by the small oscillating hairs, called cilia, which line the inside of our airways.

Their rhythmic waft moves the mucus up from our smaller lower airways into our larger upper airways where we eventually swallow it. In an individual with CF, the mechanism for clearing mucus and bacteria out of the lungs is impaired due to presence of thicker & stickier mucus. The presence of abnormal mucus in the lungs causes the cilia to malfunction allowing bacterial organisms to thrive, causing repeated and eventually continuous chronic infection in the airways. Repeated infections in the lungs will result in inflammation which eventually leads to progressive and irreversible lung damage.

THE MANAGEMENT

The paediatric CF team work in partnership with the affected child/young person, their family and other teams such as community health and education to manage the CF respiratory treatments and care from diagnosis through to transition into adult services (typically by 18 years old). The aim of CF respiratory treatments is to maintain a child's/young persons normal lung function by:

- Keeping the airways as clear as possible i.e. mimic normal mucus clearance
- Regularly screen for new or changing pathogens that are known to affect the CF lung
- Responding promptly to the results of respiratory cultures, and/or observed or reported respiratory signs and symptoms which would be indicative of new

infections and/or inflammation in order that the airway is restored and risk of damage reduced.

- Slow the potential damaging effects of chronic infections and inflammation on the airways
- Promoting exercise

Every individual with CF is at risk from chest infections (normally as a consequence of a cold). The frequency is unpredictable, but typically numerous, particularly in the cooler months of the year.

The main treatments to protect the lungs from the potential damaging affects of infection and inflammation are: physiotherapy, antibiotics, exercise and adequate nutrition (discussed later). Vigilance for new or increasing respiratory symptoms is required at all times as this will determine whether treatments or physio management need to be altered.

The typical treatments used in CF are as follows:

i) Physiotherapy

The aim of chest physiotherapy is to loosen the mucus in the lungs so that it can more readily move out of the lungs to be swallowed or spat out. Physiotherapy is tailored to the individual child and family and needs to be performed everyday even if they are well (normally twice a day). When a child/young person starts to develop the signs and symptoms indicative of a chest infection their physiotherapy will normally be increased accordingly.

ii) Antibiotics for acute chest exacerbations

Although many coughs are a response to viral infection, in children with CF we have a low threshold for treating with antibiotics because of the increased risk of suppressed

bacterial infection and the damage it can do. Antibiotic courses are typically 10-14 days. Cough swabs or sputum samples should always be sent to ensure we have a clear idea about possible respiratory pathogens in each patient. However, antibiotics are often started empirically before the results are available.

If a child/young person with CF develops new respiratory symptoms e.g. a new productive cough we would consider that this indicates an infection. Families will have been advised to increase physiotherapy +/- salbutamol and a cough swab or sputum should be sent. In response to these symptoms the family is advised to communicate with the CF team or one of the senior paediatricians based at the Children's Assessment Unit (CAU) & typically oral or, if the child is unwell, IV antibiotics are commenced (often before the cough swab/ sputum result is available).

Contrary to the current community antibiotic guidelines for children not affected by CF, antibiotics are used readily in children with CF. The BNFC and the current CF Trust guidelines for antibiotic treatment are used by Portsmouth Hospitals NHS Trust for prescribing in CF.

iii) Long term antibiotics for prophylaxis and bacterial colonisation

If a childs lungs becomes colonised with a particular pathogen such as Pseudomonas aeruginosa or Staphylococcal aureus they will require continuous oral or inhaled antibiotics. Sometimes they require antifungal treatments.

iv) Exercise

Daily exercise is encouraged in individuals with CF. Exercise helps strengthen respiratory muscles, improves clearance while promoting better posture which in turn helps the lungs work better.

v) Other respiratory treatments

Often individuals with CF require nebulised mucolytics which increase and improve mucociliary clearance in the lungs by reducing the viscosity of the mucus. Some require inhalers, both bronchodilators and anti inflammatory steroids (more commonly used in the management of asthma). Antibiotics that have antiflammatory properties such as azithromycin are also used.

How CF Affects Digestion

Part of our digestive tract is the pancreas; It normally produces digestive enzymes and delivers them via small tubes (ducts) into our gut (small intestine). These enzymes are required to break down the food that we eat so that we can absorb the resulting nutrients into our bloodstream. Once in the bloodstream these nutrients are distributed around the body and are generally used for energy, the body's growth (particularly in children) and repair.

In most people with CF the ducts leading from the pancreas into the small intestine are permanently blocked as a result of damage by thick sticky mucus. The resultant digestive enzyme deficiency means that affected children and young people with CF require pancreatic enzyme replacement; this enables adequate fat and vitamin absorption.

1. Pancreatic enzymes

In affected individuals with CF, pancreatic enzymes (typically Creon) are administered orally before any drinks, meals or snacks containing fat. The dose of these enzymes is adjusted according to the fat content of the drink, meal and snack and varies between individuals.

2. Vitamins

Children and young people with CF need to take extra vitamins every day because of the poor absorption in the intestine. They will commonly include vitamins A,D and E. Some patients also require vitamin K particularly those with CF liver disease.

FOLLOW UP & ANNUAL REVIEW

Children and young people with CF are expected to attend the Children's Outpatient department at Queen Alexandra hospital at least once every 3 months (this may be more frequent in the months after diagnosis or in situations where health is not so good.). A dietician, doctor, specialist nurse and physiotherapist will review them during this outpatient clinic. Once a year they will have an annual review which will include investigations such as a chest x ray, blood tests and often a liver ultrasound, oral glucose tolerance test and bone density scan. A more thorough assessment of their respiratory and nutritional health is made by the CF team and changes made to the management for the following year.

How primary care can support children and young people affected by CF and the CF team at Portsmouth

- Supply repeat prescriptions for at least a months worth of chronic medications including Creon & vitamins and where possible utilise home delivery services especially when oral nutritional supplements are required.
- Where an infant/child requires on oral antibiotic liquid and the family are competent allow them to reconstitute the oral antibiotic at home to prevent unnecessary trips to pharmacy.
- Prescribe plastic ampoules for reconstituting nebulised solutions such as 'BD Plastipak' to avoid the need for needles