Cystic Fibrosis

Diagnosis and Management information leaflet

Cystic Fibrosis services for children in South and West Wales are organised as a shared care network between the local hospital and the central Specialist CF Centre based at Noah's Ark Children's Hospital for Wales in Cardiff. All patients have a named consultant at their local hospital and at the CF centre. For patients in Cardiff, all care is provided by the Specialist CF Centre based at Noah's Ark Children's Hospital for Wales.

The mainstay of treatment is preventative with a structured care plan incorporating optimal nutrition including pancreatic enzyme supplementation, regularly physiotherapy and exercise, prophylactic antibiotics and aggressive early treatment of respiratory infections.
Prognosis is improving and we have an expectation for children with CF to attend school normally, partake in all sporting activities and have the same opportunities in childhood as their friends. We aim to transfer them to adult care with near normal lung function.

Treatment for Cystic Fibrosis is well structured and multidisciplinary. Patients are seen regularly in clinic where they are reviewed by medical staff, physiotherapist, dietician, CF specialist nurse and psychologist.
Newly diagnosed infants and children are reviewed weekly initially by the specialist MDT, at least every month until the age of one year, and then every two months thereafter. You will receive a timely copy of the clinic letter from each appointment.
Every year, all patients have an in depth annual review in Cardiff which includes comprehensive investigations and a thorough assessment of progress.
Treatments are often altered at clinic. We would be grateful if you could continue to prescribe medications as outlined in clinic letters.

If a child with CF develops a respiratory infection for more than 24-48 hours (viral or bacterial) the family is encouraged to contact the CF nurse specialist at their local hospital. If reasonable to do so, the nurses will collect a cough swab or sputum sample for microbiological analysis prior to the child/young person commencing a two weeks' course of antibiotics. The antibiotic may be provided by the hospital. Otherwise a prescription request will be faxed to the GP surgery with the antibiotic and dose required.

Please note that in line with CF Trust (2009), dosages for children/young people with CF tend to be double the usual dose and for at least two weeks. If the child is on continuous prophylactic antibiotics, these should be continued while the additional antibiotic treatment is taken.

Children with Cystic Fibrosis should receive the annual Influenza vaccination at the GP surgery as well as routine childhood immunisations.

Smoking is actively discouraged and we would recommend no exposure to tobacco smoke at home or in homes the child visits. If appropriate, please discuss smoking cessation with families if the opportunity arises.

If you have any questions or would like to discuss one of your patients further, please do not hesitate to contact the local network team or specialist MDT in Cardiff (029 2074 4892). For further information about CF, you can visit our webpage on www.uhwchildren.com/respiratory.