CYSTIC FIBROSIS

This fact sheet has been written by parent carers for parent carers.
Clinical information has been approved by West Sussex practitioners.

What is cystic fibrosis?

Cystic fibrosis (CF) is a genetic condition. It mainly affects the lungs and digestive system, which become clogged with thick mucus. This can make it hard for people with the condition to breathe, and to digest food. There is no cure for cystic fibrosis, but the condition can be managed with physiotherapy, exercise, medication and diet. Many people are carriers of the faulty gene for cystic fibrosis – if two carriers have a baby, there is about a 1 in 4 chance that the baby will have the condition. Around 1 in every 2,500 babies born in the UK has cystic fibrosis.

Characteristics of cystic fibrosis

Signs of cystic fibrosis may be picked up at birth (see 'diagnosis' below for more information). Usually symptoms will develop within the first year of life, but as these can vary in their severity, some people may not be diagnosed with cystic fibrosis until they are older. Symptoms can include shortness of breath, wheezing and recurrent chest infections, along with poor growth and weight gain, prolonged diarrhoea or constipation. There are more than 1,500 mutations of the cystic fibrosis gene, which means that there is a lot of variation in the way in which people are affected by the condition and how severe their symptoms are.

Diagnosis

Some babies may be diagnosed with cystic fibrosis shortly before or after birth if they develop a condition called 'meconium ileus', which is where the bowel becomes blocked with meconium, a thick, dark substance that babies usually pass in the first few bowel movements after birth. Since 2007, the heel prick blood test, or Guthrie test, which is carried out on newborn babies, has included a test for cystic fibrosis. If the blood sample shows abnormalities, doctors will ask for further screening, usually in the form of a sweat test and genetic test to confirm or rule out cystic fibrosis. The sweat test is considered the 'gold standard' for the diagnosis of cystic fibrosis, as children and adults with cystic fibrosis have higher levels of salt in their sweat. Genetic testing is carried out using either a blood sample or a DNA sample which is taken by rubbing a swab on the inside of the cheek.

Types of therapy

When someone is diagnosed with CF, they will be referred to a cystic fibrosis centre, where specialists will be able to advise you on how to manage the condition. This will probably include the following:

- **Diet** – most people with CF will need to take special enzymes to help them digest their food. It is important for them to have a diet that is high in calories and rich in fat and protein, to ensure that they get the nutrients they need and maintain a healthy weight.
- **Insulin** – some people with cystic fibrosis may have diabetes and may need to take insulin and manage their diet.
- **Medication** – this can include antibiotics to prevent and treat infections, inhaled medicines such as pulmozyme (DNase), hypertonic saline, or mannitol powder to thin mucus so it can be coughed up.
- **Physiotherapy** – every person with CF has different needs, so a physiotherapist will create an individual programme of therapy. This can include ways to clear the airway, such as 'Active Cycle of Breathing Technique or (ACBT). A physio can also help with exercise, breathing techniques and posture.

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- **Supplements** – vitamin supplements are usually recommended, and a dietician may recommend that your baby or child takes salt supplements. If your baby has been recently diagnosed with CF, a dietician at your cystic fibrosis centre will be able to advise you on what vitamins and supplements may be needed.
- **Transplants** – in very severe cases of CF, a lung transplant may be needed. Outcomes for people with CF are generally good, as patients are often younger and in better health.
- **Vaccinations** – it is important that people with CF have their vaccinations and also have an annual flu jab and a pneumonia vaccination, as they are vulnerable to infection.

Ask About

- **Child Disability Service** – a social work service for children and young people with severe and/or complex disabilities, it works with families and other agencies to ensure they get the right support. Information can be found via the local offer website: **https://westsussex.local-offer.org/services/265-child-disability-teams-lifelong-services-social-care** or families should call the Integrated Front Door (IFD) for West Sussex Children Services on **01403 229900** or complete the online referral: **www.westsussex.gov.uk/Raiseaconcernaboutachild**.
- **Compass Card West Sussex** – a free leisure discount card for 0 to 25 year olds with SEND, which is run by **Amaze**. Find out more at: **www.compasscard.org.uk** or by calling: **0300 123 9186**.
- **Home start** – support for parents with at least one child under five. For more information see: **www.home-start.org.uk**.
- **Portage** – a home-based educational programme tailored to a child's individual needs. Available to very young children. Chichester: **01243 536182**; Crawley and East Grinstead: **01293 572480**; Horsham: **01243 536182**; Mid-Sussex: **01444 243150**; Worthing: **01903 242558**.
- **Short Breaks for Disabled Children** – commissions holiday clubs, after school clubs, buddy schemes and short breaks at home or in the community. Tel: **0330 222 2562** or email: **SENDCommissioningTeam@westsussex.gov.uk**.
- **West Sussex Children Services** – support from health and social care. If your child is under 18, call the Integrated Front Door (IFD) for West Sussex Children Services, tel: **01403 229888** or email: **WSChildrenservices@westsussex.gov.uk**. For over 18s call the **Adults' CarePoint**: **01243 642121**, or email: **socialcare@westsussex.gov.uk**.
- **Other resources** – the Ashdown club, Worthing: **01903 528607**; Kangaroos, Haywards Heath: **01444 459108**; PACSO, Chichester: **01243 533353**; Springboard Project, Horsham: **01403 218888**.

Further reading and useful links

- **Making Sense of it All** – our handy parent/carer guide contains information and advice on benefits, support for your child at school, getting help from social services and accessing social & leisure activities – **www.reachingfamilies.org.uk**.
- **Children's Cystic Fibrosis Fund Worthing & Chichester** (can be found on facebook) - Supports CF Children up to age 18 yrs and their families that live in Sussex: **www.ccffwc.co.uk**. Contact Jo: **07745 325919**, or Charlee: **07886 888302** or Email: **ccffwc@gmail.com**.
- **Cystic Fibrosis Holiday Fund** – grants towards short breaks for under 18s with cystic fibrosis. See: **www.cfholidayfund.org.uk** for more details (grants for over 18s via the **Cystic Fibrosis Trust**).
- **Cystic Fibrosis Trust** – information and support for people with CF and their families. See: **www.cysticfibrosis.org.uk** or call their helpline: **0300 373 1000**.
- **Disability living allowance (DLA)** – your child may qualify for DLA, a state benefit that will help with their care. For further information visit **www.gov.uk/disability-living-allowance-children**.
- **West Sussex Local Offer** – go to **https://westsussex.local-offer.org** and search for 'cystic fibrosis'.