CYSTIC FIBROSIS

This fact sheet has been written by parent carers for parent carers.

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 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 help break down mucus in the lungs.
- **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, inhalation techniques and posture.

- **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

- **Information, Advice and Support (IAS)** – Amaze supports families of children and young people with SEN and disabilities in Brighton and Hove. We have a helpline, publications and a website. We also run workshops and courses and can offer one-to-one support with EHC planning and making DLA and PIP claims. Call Amaze on: **01273 772289** or visit: **www.amazebrighton.org.uk**.
- **Compass Card** – a free leisure discount card administered by Amaze for 0 to 25 year olds registered on The Compass who live or go to school in Brighton and Hove or West Sussex, or who are looked after by social services in these areas. Call Amaze on: **01273 772289**:
- **Independent Support** – Amaze's Independent Supporters provide extra advice and support to young people and parent carers who are going through the process of getting an EHC Plan in Brighton and Hove and Sussex. For Brighton and Hove Independent Support, call: **01273 772289**. For Sussex Independent Support, call: **0300 123 7782.**
- **Integrated Child Development and Disability Centre (Seaside View)** – where many children are diagnosed and where health, education and social care can work together to support and treat your child. Assessments will usually be via a health or education professional but you can contact Seaside View directly on (01273) 265780
- **Kaleidoscope** – a group for parents and carers of children with physical disabilities. Little Wheels is for children aged 0 to 8 and provides early support and info, regular swimming sessions and meet-ups. Wheels is for 8 to 16 year olds and they organise regular leisure activities. Contact Gillian on: **07788 924940** (Little Wheels) or Sandra on: **07946 344220** (Wheels).
- **Making a claim for DLA** – if your child's care or mobility needs are significantly greater than their peers you may be able to claim Disability Living Allowance for them. Find out more at: **www.gov.uk**. Amaze can give you advice and practical support with making a claim. Call the helpline on: **01273 772289** or email: **helpline@amazebrighton.org.uk**.

Further reading and useful links

- **Cystic fibrosis holiday fund** – funds holidays and short breaks for people under 25 years who have cystic fibrosis. See: **www.cf-holidayfund.org.uk** for more details.
- **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** and search for 'DLA'.