Amaze & Reaching Families FACT SHEET Updated Nov 2019

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 new-born 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, which thins 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

CYSTIC FIBROSIS

the '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

- **Amaze SENDIASS** – the **Special Educational Needs and Disability Information, Advice and Support Service (SENDIASS)** offers impartial, confidential advice on anything to do with special educational needs and disabilities for 0 to 25 year olds, including education, health, and social care. Parents, carers, children and young people under 25 with SEND living in Brighton & Hove or East Sussex can use the service. Contact SENDIASS by email **sendiass@amazesussex.org.uk** or by phone on **01273 772289**. Amaze offers other support for parents and young people including books, fact sheets, workshops, groups and peer support. To find out more visit our website at **amazesussex.org.uk**
- **Compass Card** – a free leisure discount card administered by Amaze for 0 to 25 year olds with SEND who are registered on the compass. Go to: **www.compasscard.org.uk**, email Amaze at: **compass@amazesussex.co.uk** or call: **01273 772289** (Brighton & Hove) or **0300 123 9186** (West Sussex).
- **Disability Living Allowance (DLA) –** if your child's care or mobility needs are significantly greater than the needs of their peers you may be able to claim DLA for them. Visit **www.gov.uk** and search for DLA. Amaze can also give you advice and support with making a claim. Call our helpline: **01273 772289** or visit our website: **amazesussex.org.uk**.
- **Seaside View Child Development Centre** – this is where many children are diagnosed and where health professionals such as paediatricians and occupational therapists 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.**

Further reading and useful links

- **Cystic Fibrosis Holiday Fund** – grants towards short breaks for under 18s with CF. 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**.
- **Local Offer** – the local authority's online listing of all the services and support that are available to families with children with SEND in the area. Visit **new.brighton-hove.gov.uk/special-educational-needs-and-disabilities**