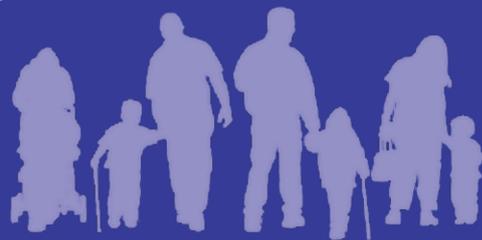


CYSTIC FIBROSIS

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

- **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.
- **Portage** – a home-based educational programme tailored to a child's individual needs. Available to very young children. Crawley and East Grinstead: **01293 572480**, Chichester and Horsham: **01243 536182**, Mid Sussex: **01444 243150**, Worthing: **01903 242558**.
- **Reaching Families** – provides training, information & other fact sheets related to this topic. We also offer benefits advice, peer support, a Facebook group & handbook (*see below*) for parent carers of children & young people with SEND in West Sussex. See: www.reachingfamilies.org.uk.
- **West Sussex Children's Services** – support from health and social care. If your child is under 18, contact the Integrated Front Door (IFD) for West Sussex Children's Services. Tel: **01403 229900** or email: WSChildrenservices@westsussex.gov.uk. The IFD is also an entry point for the Children with Disabilities (social care) service for children & young people with severe and/or complex disabilities. Eligibility criteria apply – See: <https://westsussex.local-offer.org/services/265>. For over 18s call the Adults' CarePoint: **01243 642121**, or email: socialcare@westsussex.gov.uk.

Further reading and useful links

- **Making Sense of it All: From Birth to Adulthood** – Reaching Families' handbook for parent carers of children and young people with SEND in West Sussex. This provides essential information on money matters, including claiming DLA and PIP, as well as social care, health, leisure, travel, education and employment. Go to: www.reachingfamilies.org.uk/guides.html.
- **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**.
- **NHS Health A-Z** has a section devoted to cystic fibrosis. Go to: www.nhs.uk/conditions/cystic-fibrosis.
- **West Sussex Libraries** – For books on this topic available to borrow for free, go to: <https://arena.westsussex.gov.uk/welcome> and search the library catalogue for 'cystic fibrosis'.
- **West Sussex Local Offer** – go to <https://westsussex.local-offer.org> for local services, support and details of Short Breaks providers for children & young people aged 0 - 25 years with SEND.

