

JOURNALIST FACTSHEET

SYMPTOMS OF CYSTIC FIBROSIS

WHAT IS CF?

Cystic Fibrosis (CF) is one of the UK's most common life-threatening inherited diseases affecting 8,500 children and adults. People who have CF produce a thick sticky mucus which clogs their lungs and digestive systems making it difficult to breathe and absorb food properly.

DIAGNOSIS

After long campaigning by the CF Trust, from 2007 all newborn babies in the UK are screened for Cystic Fibrosis at birth using the heel prick blood test.

Around 1 in 10 children with cystic fibrosis are diagnosed at or shortly after birth due to a condition called meconium ileus where the gut becomes blocked with meconium - a thick, dark, sticky substance which is made in all babies' intestines before being born. Urgent surgery may be needed to relieve the blockage.

Some children born earlier than 2007 who were not screened at birth will be diagnosed later after they have become unwell and developed symptoms, often involving many dirty nappies each day, chestiness and failure to thrive.

SYMPTOMS

When a child is born with cystic fibrosis, symptoms usually appear in the first year of life, although occasionally they can develop later. The thick sticky mucus in the body affects a number of organs, particularly the lungs and digestive system.

The symptoms and related problems of cystic fibrosis can vary in severity from person to person.

Lungs

It is common for people with cystic fibrosis to have difficulties such as:

- **Cough and wheeze** - the body tries to shift the thick mucus in the lungs by coughing it up.
- **Recurring chest and lung infections** - Infections are caused by the continual build-up of mucus in the lungs, which provides an ideal breeding ground for bacteria.

Digestive system

Cystic fibrosis can also cause mucus to block the ducts in the pancreas. The pancreas produces essential food-digesting enzymes. Because the flow is blocked, not enough of the enzymes reach the intestines (bowel) to help break down food, which can cause:

- **Large, smelly stools** – If the digestive enzymes are not being produced, food is not adequately digested and excess fat is lost in the stools making them bulky, oily, smelly and difficult to flush away.
- **Malnutrition** – because the body cannot digest essential nutrients in food (particularly fat), it is often difficult to gain weight and infants may struggle to put on weight and grow. Adults with cystic fibrosis often find it difficult to gain and maintain weight. In children with cystic fibrosis, this can result in delayed puberty if they are severely underweight.
- **Diabetes** - in older people with cystic fibrosis, the pancreas can become more damaged. Diabetes can develop if the pancreas fails to produce enough insulin, a hormone that controls the level of sugar in the blood. Diabetes in people with cystic fibrosis is different to diabetes in people without cystic fibrosis. The symptoms of diabetes include feeling constantly thirsty, frequently needing to pass urine, and feeling extremely tired rarely occur in patients with cystic fibrosis. People with cystic fibrosis who develop diabetes may find it difficult to gain weight or may lose weight and may see a decline in their lung function. Cystic fibrosis related diabetes is usually controlled by regular injections of insulin. Diabetes rarely occurs in children with cystic fibrosis.

Ears, nose and sinuses

People with cystic fibrosis may be prone to sinusitis and hay fever, which may need to be treated with nasal sprays or antibiotics. Some older children and adults develop nasal polyps, which are fleshy swellings that grow from the lining of the nose or sinuses. If they become troublesome, they may need to be removed.

Bones and joints

Some older children with cystic fibrosis develop a form of arthritis (swelling and pain of the joints), usually in one or two large joints such as the knee. In most cases, symptoms improve with time and treatment.

Older children and adults may also be prone to osteoporosis (thin brittle bones) for many reasons including repeated infection, poor growth or weight, lack of physical activity and a lack of vitamins and minerals due to the digestive problems. People with cystic fibrosis are more at risk of developing osteoporosis if they are taking steroids to help with lung infections.

Osteoporosis as a result of cystic fibrosis may cause joint pain and bones may fracture (break) more easily in some. Some people need to take drugs called bisphosphonates to help to maintain their bone density.

Infertility

Both men and women with cystic fibrosis can have problems conceiving children. In virtually all men with cystic fibrosis, the tubes that carry sperm do not develop correctly making them infertile. In women who are underweight, their menstrual cycle may become absent or irregular. There is also an increased thickness of cervical mucus in women with CF which may reduce fertility although most women become pregnant without any difficulty.

Liver

In a few people with cystic fibrosis, the tiny bile ducts in the liver can become blocked by mucus. This can be serious as the disease progresses, and in some cases, it may be necessary to have a liver transplant.

Incontinence

People with cystic fibrosis, especially females, are more likely to suffer from incontinence. Urine usually leaks during coughing fits. It is important that people with cystic fibrosis are asked about this and encouraged to talk to a member of their cystic fibrosis team about the problem as soon as possible so they can get help.

Other

Parents often remark that their children with CF have salty-tasting skin. This is because abnormalities in the sweat glands cause a loss of salt, which secretes through the skin.

People with CF often develop clubbing of their fingers and toes due to the effects of chronic illness and low oxygen in their tissues.

MORE INFORMATION

Please visit our website www.cftrust.org.uk or call us on 0208 464 7211

ENDS

For all media inquiries please contact:

Gemma Foy on 0208 290 7912 or email gfoy@cftrust.org.uk

Notes to Editors

- The Cystic Fibrosis Trust is the UK's only national charity dealing with all aspects of Cystic Fibrosis (CF). It funds research to treat and cure CF and aims to ensure appropriate clinical care and support for people with Cystic Fibrosis.
- Cystic Fibrosis (CF) is one of the UK's most common life-threatening inherited diseases. Cystic Fibrosis is caused by a single defective gene. As a result, the internal organs, especially the lungs and digestive system, become clogged with thick sticky mucus resulting in chronic infections and inflammation in the lungs and difficulty digesting food.

- Each week five babies are born with Cystic Fibrosis and three young people die – 90% from lung damage. Around half of the CF population can expect to live over 38 years, although improvements in treatments mean a baby born today is expected to live even longer.
- Further information can be found on our website www.cftrust.org.uk. Help and advice for those affected by Cystic Fibrosis is available through our Helpline on 0300 373 1000. For further information, media should contact Gemma Foy on 0208 290 7912 or email gfoy@cftrust.org.uk

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