



Great Ormond Street Hospital for Children NHS Trust: Information for Families

Cystic fibrosis (CF) is suspected

We suspect that your child has cystic fibrosis (CF) because he or she was born with a bowel blockage called meconium ileus. This information sheet explains the link between CF and meconium ileus and some basic information about what happens next.

What is meconium ileus?

This is the most common type of blockage in cystic fibrosis (CF). All newborn babies have meconium in their bowel. This is the dark faeces passed in the first two days of life. If a baby has CF, the meconium is so thick and sticky that it blocks the bowel. The surgeons will talk to you about the treatment needed to get rid of the blockage. When your baby has had the blockage removed, either in an operation or by washing out the bowel, he or she will need to tests to confirm or rule out CF.



What is CF?

CF is an inherited disease affecting the lungs and digestive system. It happens because the gene that is responsible for making mucus is faulty. Normally, the mucus that lines our internal organs clear, lubricating and protects against infection. In babies with CF, it is thick, congesting and prone to infection.

Human beings have about 30 to 40,000 different genes, each of which has a function in making an individual person. The genes are arranged in pairs (one of the pair from each parent) on 23 chromosomes. Inevitably, some of these genes are faulty; a normal gene can overcome a faulty one, but if both genes in the pair are faulty, the genetic instructions cannot work. About one person in every 25 carries the faulty CF gene.

Most people carry different faulty genes but in CF (and other recessive conditions) parents, though healthy themselves, carry the same faulty genes, and risk passing them on to their children. Each pregnancy carries a 25 per cent chance of the child being affected.



How will having CF affect my baby?

CF affects the lungs and digestive system. More details about the effects follow.

Digestive system

The pancreas is a gland in the abdomen and one of its functions is to produce digestive juices (enzymes) that help digest and absorb the food we eat. In babies with CF, the small channels through which the enzymes flow become blocked with sticky mucus. This means that they cannot digest and absorb food as well as they should. To help your baby digest food and gain weight, we will give him or her pancreatic enzymes before each feed. If your baby needs these enzymes, the dietitian will explain more about this when feeding is established. Taking pancreatic enzymes will not affect breastfeeding.

Lungs

We all have mucus in our lungs, which helps them to function, but in babies with CF, the mucus is abnormally thick. This can block the smaller airways and lead to infection. To help prevent this happening, we start babies on antibiotics and teach parents about chest physiotherapy. As children get older, we encourage them to be very active and do lots of physical activities.

Does CF affect my baby's development?

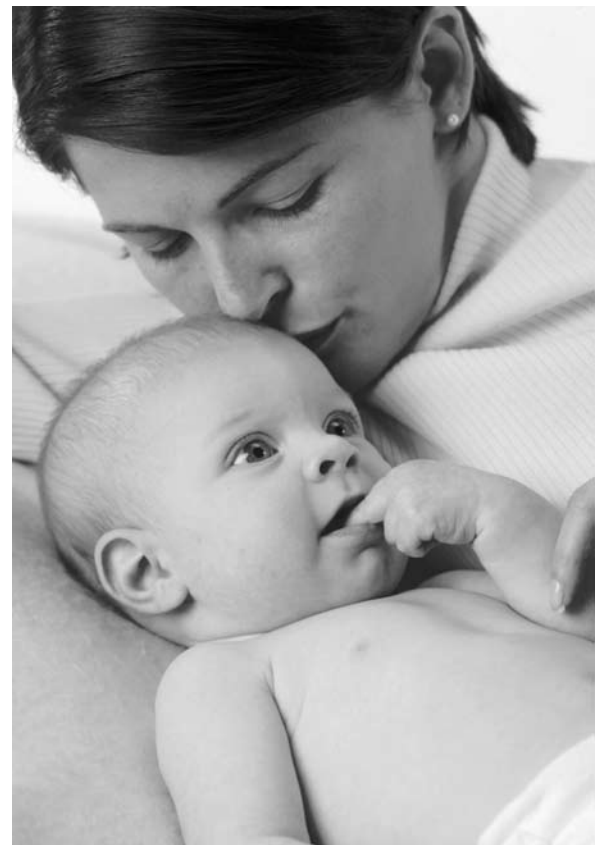
No, it will not affect your baby's development and he or she should go to school and join in all activities with friends. Immunisations are important for all children and these are given to children with CF according to the normal schedule.

How is CF diagnosed?

At Great Ormond Street Hospital (GOSH), a small sample of your baby's blood will be sent to our genetics laboratory. Scientists will look at this sample to find the CF genes. The medical team will let you know as soon as the results are available, usually within a week.

As well as looking at a blood sample, scientists will also look at a small sample of your baby's faeces (poo) to see if the pancreas is working well enough. The results of these tests will show whether your baby will need pancreatic enzymes.

Sometimes, the doctors want to carry out a sweat test when your baby is between four and six weeks old. This test measures the amount of salts and minerals (sodium and chloride) in your baby's sweat. More information about the sweat test is available in another information sheet.





What will happen in the future?

At present, there is no cure for CF but there are many treatments available. If your child has CF, the team looking after him or her will keep you updated on all the latest research.

Until we have the results of the tests, you might want to find out more about CF. Although there is plenty of information on the Internet about CF, some sites are less reliable than others.

We suggest that you contact the CF Trust in the first instance for reliable information that is applicable to people living in the UK.

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Notes

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www.goshfamilies.nhs.uk

www.childrenfirst.nhs.uk