Cystic Fibrosis all about nutrition

Gastrointestinal issues in cystic fibrosis

The Cystic Fibrosis Trust is grateful to the dietitians from the Cystic Fibrosis Dietitian Group UK (CFDGUK) who prepared the information in this leaflet.

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Introduction

Cystic fibrosis (CF) can cause many different gastrointestinal (GI) conditions, which often have varying symptoms for each person and require different treatment. These symptoms can affect your quality of life and should be fully investigated and treated. You may be referred to a gastroenterologist, a doctor who can give further advice about these conditions.

This leaflet talks about GI issues that can happen with CF and how to recognise symptoms and report them to your CF team. The treatment for each issue is individual and you should always follow the advice of your CF team.

The information within this leaflet is for adults and young people over the age of 16. Children under 16 years old can also experience GI problems, but this should be discussed with their CF team.

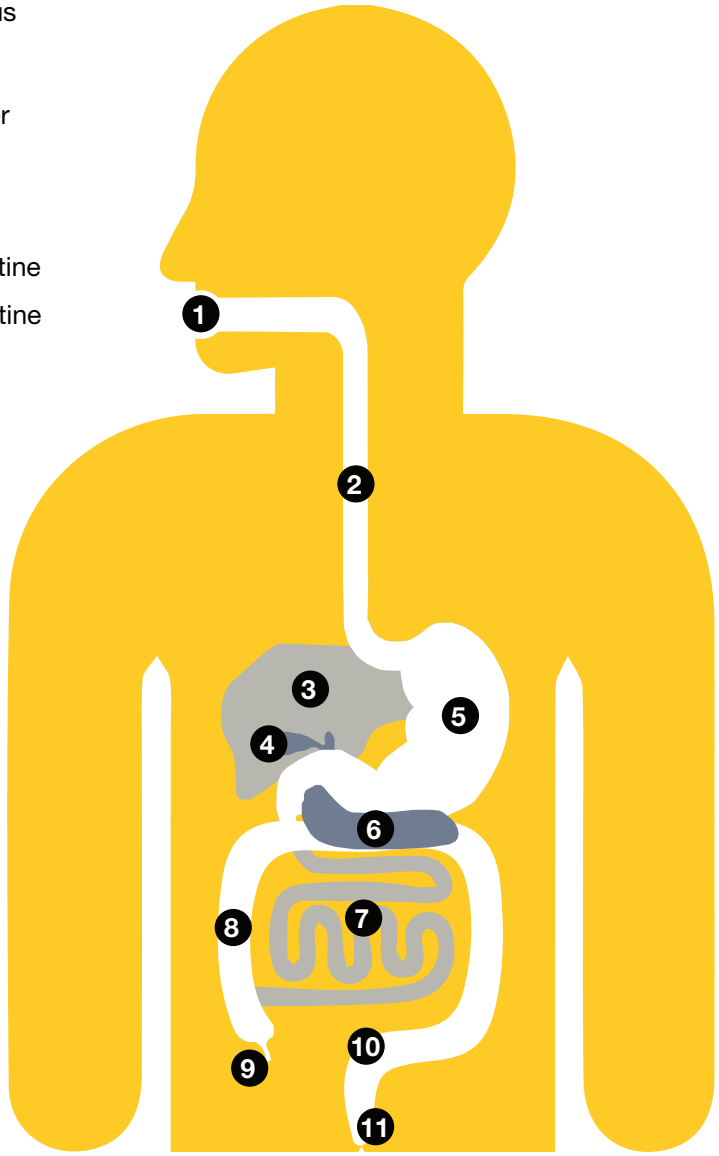
Different parts of the digestive system

Your digestive tract is the route that your food follows after you put it in your mouth. It involves many different organs. Many of these organs can be affected by having cystic fibrosis.

After swallowing, your food goes down your oesophagus and into your stomach, where it is broken down into a pulp. This pulp then moves into your small intestine where digestive enzymes from the pancreas, and bile from the liver help to digest your food further. However, in people with CF the small tubes that transport the digestive enzymes out of the pancreas become blocked with mucus, meaning that the enzymes can build up in the pancreas instead of reaching the digestive system. The pancreas can then become inflamed, and as a result the pancreas is not able to produce enough digestive enzymes. This is called pancreatic insufficiency, and most people with CF have to take supplements such as Creon® or Nutrizym 22® to replace these digestive enzymes.

Once most of the nutrients have been absorbed from your small intestine, undigested parts of your food move into your large intestine and then out into your rectum where your poo (stool) is stored until you need to go to the toilet.

1. Mouth
2. Oesophagus
3. Liver
4. Gall bladder
5. Stomach
6. Pancreas
7. Small intestine
8. Large intestine
9. Appendix
10. Rectum
11. Anus



Bowel habits

'Bowel habits' is the phrase used to mean how often you go to the toilet to poo. Bowel habits can vary from person to person, someone might poo three times a day and another person may poo once every three days. Your pattern will be individual.

Some people have inaccurate or unhelpful beliefs about bowel habits which can lead to stress or negative behaviours. People may believe "I must poo every day", "poos must be a certain shape", or "I must keep straining until I feel everything is out (fully evacuated)". These 'rules' can increase stress levels or lead to someone spending a long time straining on the toilet. Straining is unhelpful because it increases your focus on your bowels and your anxiety about them and can lead to tensing the muscles you actually need to relax, in order to poo.

Recording and reporting gastrointestinal symptoms

A range of GI symptoms can be associated with cystic fibrosis.

- Feeling sick (nausea) and being sick (vomiting)
- Heartburn/acid reflux
- Abdominal pain in any area of your tummy, or sometimes in your back. You may feel different types of pain (eg sharp, stabbing, dull aches).
- Changes in your poo, eg changes to how often you go, how soft or hard it is, its smell, colour, oiliness, difficulty in going, unable to flush, bleeding when you wipe or blood on your poo
- Visible bloating, or a bloated feeling inside your tummy.
- Excessive farting (wind or gas)

Gastrointestinal symptoms can affect your quality of life but taking the correct treatment and keeping your CF team updated with your symptoms can often improve them. If you are experiencing any of the above GI symptoms, it can be very helpful for your CF team if you record them using a diary, example below. You can take this diary to your CF clinic appointment and show it to your team who may then be able to recommend treatment or changes to your diet to help improve things. Always remember to discuss any GI worries that you have at your appointments, even if you are not asked about them.

The example below shows how someone who takes pancreatic enzymes (eg Creon®) can record their GI symptoms.

Time of day	Description of food/drink	Enzyme capsules taken	Description of symptoms eg describing your poo, any pain or noting any sickness or vomiting
9am	Two slices of toast with butter and cup of tea	3	Poo after – smelly, yellow and loose.
11am	Chocolate bar	2	Felt sick after eating.
1pm	Cheese sandwich and crisps with apple juice	4	Felt fine – no symptoms.
6pm	Fish and chips	6	Pain in tummy after eating. Large poo, dark brown and formed.
10pm	Hot chocolate made with full fat milk and two biscuits	4	Acid reflux burning my throat when in bed.

(We suggest you speak to your CF team about the best way for you to record any symptoms. For some people, watching every bowel symptom can make them feel stressed which can sometimes increase their symptoms.)

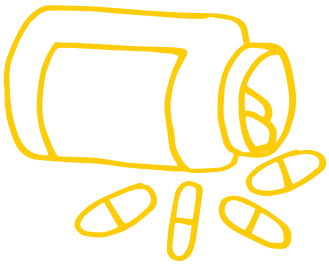
We understand that it may feel embarrassing to discuss your GI symptoms with your CF team, however, the team are very used to discussing these issues and its important that they get accurate information about your symptoms so they can help address any problems.

Gastrointestinal conditions linked to cystic fibrosis

This section briefly covers some of the GI conditions that people with CF can experience. If you are worried you might have any of these conditions, it is important that you discuss them with your CF team.

Distal intestinal obstruction syndrome and constipation

Distal intestinal obstruction syndrome (DIOS) and constipation are very common conditions where the intestine can be blocked with poo. While the symptoms are often the same for both conditions, the treatments may differ, so it is important that you and your CF team work together to make the correct diagnosis.



What is constipation?

This is where thick, bulky poo sits in your intestine for too long. It is possible to poo every day and still have a lot of poo in your large intestine, or you may find that you are only pooing every few days or less. If you have constipation you may also feel stomach pain, bloating or a feeling that you have not completely emptied your bowels after doing a poo.

What is DIOS?

Distal intestinal obstruction syndrome is a condition that is only seen in people with CF, and happens when thick poo sits where the small intestine joins the large intestine. This mass of poo (which is often on the right side of your abdomen) can often be felt when a doctor examines you, or can be seen on an abdominal X-ray. Often this mass is only partly blocking your intestine and you may experience pain, bloating and feeling sick. You may notice changes in what your poo looks like. This can include diarrhoea (very watery poo), which might be overflowing round the blockage.

Sometimes the mass of poo can completely block your intestine, which will usually lead to you vomiting (being sick) bile and having severe pain. Distal intestinal obstruction syndrome usually comes on more quickly than constipation and you may go from having no symptoms to severe pain in just a few hours.

You should contact your CF team if you think you might be experiencing DIOS or constipation symptoms. If you experience severe stomach (abdominal) pain and are being sick then you should seek urgent medical attention by going to A&E.

Risk factors for developing DIOS include:

- your CF genotype (ask you doctor if you are unsure of your genotype);
- being pancreatic insufficient;
- not taking your pancreatic enzymes as recommended by your CF team (as this can lead to poor digestion of fat);
- dehydration, especially in hot weather or when unwell;
- having meconium ileus (bowel blockage) as a newborn baby;
- following a lung or liver transplant; and
- having CF-related diabetes.

While pancreatic insufficiency is a risk factor for DIOS, remember that it is possible to develop DIOS even if you are pancreatic sufficient.

Treatment for DIOS and constipation

There are various medications that are used to treat these conditions and your CF team will advise you which ones are right for you. These include Movicol® or Laxido® and Gastrografin®. It is important that you take these as prescribed and complete the course you are given, ensuring you maintain a good fluid intake throughout your treatment. Discuss with your CF team about what foods you are allowed to eat while being treated for DIOS or severe constipation. Often these conditions can be treated as an outpatient, but more severe cases may require you to be admitted to hospital.

Prevention of DIOS and constipation

Once you have had an episode of DIOS there is a significant risk of you having another episode of DIOS in the following months. You should follow the treatment plan that your CF team give you, which may include preventative laxatives, increased fluid intake and taking your pancreatic enzymes as prescribed. Regular exercise can also help to move the poo through your intestine. All these measures can help to prevent another build up of poo in your intestine.

Acid reflux (heartburn)

Your stomach makes acid to help break down your food, this acid can leak back from the stomach into the oesophagus (see diagram on page 3). Some people with CF experience heartburn or reflux when they eat, but it can also happen at other times of day. This is a very common condition and many people with CF are started on acid-blocking or acid-reducing medication (often Omeprazole®, Lansoprazole® or Ranitidine®) to improve these symptoms. There are a number of tests that are sometimes recommended to investigate reflux further. Discuss with your CF team if you are having reflux symptoms or have questions about your reflux medication.

Delayed gastric emptying (gastroparesis)

Gastroparesis is where the stomach empties slower than normal. This can lead to varying symptoms such as feeling sick, feeling full after eating or being unable to eat much at a meal, abdominal discomfort, or being sick hours after eating. There are tests that can be done to confirm a diagnosis of gastroparesis and there are various treatment options available. If it is not treated, gastroparesis can have an impact on your nutrition. If you are unable to eat enough, you may struggle to maintain or increase your weight. Delayed gastric emptying can also affect your control of CF-related diabetes if you struggle to match your insulin doses to your food absorption.

Small intestinal bacterial overgrowth

Our intestines contain bacteria, which are helpful for our digestion. Having CF can sometimes change the balance of bacteria and lead to there being too many ‘bad’ bacteria which can lead to bloating, excessive wind and changes to your poo. Regular use of antibiotics can make this problem worse. There is some research into using probiotics to put ‘healthy’ bacteria back into the intestine. There are currently limited products available on prescription but there are many over-the-counter probiotics which can be used. Always discuss with your CF team if you are thinking about taking a probiotic supplement, as there may be interactions with your other medication.

CF-related liver disease

Some people with CF have liver disease that is related to their cystic fibrosis. This is commonly diagnosed when you are a child or teenager and you should receive regular scans and blood tests to monitor your condition. You may be seen by a specialist liver doctor (hepatologist) who will help your CF team to manage your condition. Discuss with your CF team if you are concerned about how CF can affect your liver.

Cystic Fibrosis Trust

The information in this leaflet is general, please discuss it with your dietitian for a more personalised look at the topic.

This leaflet is part of a broad series on nutrition. Leaflets are available as online downloads and printed copies and can be found here: cysticfibrosis.org.uk/nutritionleaflets. You can also order the leaflets and our other publications from our helpline or download them here: cysticfibrosis.org.uk/publications.

Our helpline is open Monday to Friday, 9am–5pm, and can be contacted on 0300 373 1000 or by emailing helpline@cysticfibrosis.org.uk. Trained staff can provide a listening ear, practical advice, welfare/benefits information or direct you to other sources of support.

The information in this leaflet is based on clinical best practice, a consensus of opinion by dietitians within the CFDGUK and a consensus document on nutrition management of cystic fibrosis, which you can read here: cysticfibrosis.org.uk/publications.

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