Cystic fibrosis diabetes (CFD)

This factsheet explains what to expect when you are tested for cystic fibrosis diabetes (CFD). We hope it will help to answer some of the questions you may have.

This factsheet does not replace any advice you may receive from your diabetes doctor or cystic fibrosis (CF) team. Please follow their advice.

What is cystic fibrosis diabetes (CFD)?

Cystic fibrosis diabetes (CFD) is common in people who have CF. The average age for being diagnosed with CFD is 18 to 21, and it becomes more common as people get older. Around half of people with CF develop diabetes by the age of 30, and up to 90% of adults with CF have some difficulty with the body's handling of sugar by the age of 40.

Although CFD has some similarities with the other more common types of diabetes, its cause and how it is managed are different.

What are the symptoms of CFD?

Symptoms of CFD may include:

- weight loss
- feeling thirstier than usual
- needing to pass urine more frequently than usual
- weight loss
- tiredness
- an increase in the frequency of chest infections

However, with regular check-ups, we can usually diagnose CFD before symptoms occur.

Why do people with CF develop CFD?

Diabetes is caused by the lack of a hormone called insulin, which is made in the pancreas. CF causes a build-up of thick secretions in the pancreas and these secretions damage the cells that produce insulin. Lack of insulin means that the body is unable to use the glucose (sugar) found in the food we eat normally. As a result, sugar levels rise above normal ranges in the blood.

How is CFD diagnosed and monitored?

Oral glucose tolerance test (OGTT)

We diagnose CFD with an oral glucose tolerance test (OGTT). This test is designed to see how the pancreas copes with a glucose load. The test measures your blood sugars after an overnight fast. We will then ask you to drink a glucose solution (similar to the amount often consumed in the form of regular fizzy drinks). We will measure your blood sugars again two hours afterwards. Being ill or taking certain medicines can affect the OGTT test results, so it's important that we

only do the test when you are well (or at the end of a stay in hospital) and when you haven't recently started taking any new medicines such as steroids.

If your OGTT test results are not normal, we will usually ask you to monitor your blood sugar at home for a period of time. We will show you how to monitor blood sugar levels by finger prick testing and by using a continuous or flash glucose monitoring (CGM) device. Depending on the results, we can decide whether any medication is needed and if so, what type of treatment would help the most.

Continuous or flash device monitoring (CGM)
CGM involves placing a small sensor under the skin that measures the glucose in the tissues of the body. The sensor is then paired with a recording device that displays your blood glucose levels without the need to take finger prick samples. CGM is a very useful tool for monitoring how well the treatment is working and can help the CF team to prescribe the best treatment based on your personal blood sugar readings.

We sometimes use CGM systems (such as I-Pro and Free Style Libre) to help manage your CFD because they provide a more complete picture of your blood sugar trends over several days. In particular, we can identify any abnormally high or very low blood sugar readings.

Haemoglobin A1c (HbA1c) test
This test shows us your average blood sugar levels over the last two to three months. We use this test to help assess how well the treatment is working over a longer period of time.

Why is it important to treat CFD?

High blood glucose levels can increase the risk of chest infections and lead to a fall in lung function.

Treating CFD with good blood glucose control can increase your overall feeling of wellbeing and may also reduce the number of chest infections and hospital admissions.

If CFD is not treated and your blood glucose levels are not well controlled, this may lead to long-term complications and serious effects on eye sight, kidney function and circulation.

How is CFD treated?

The first steps in treating CFD involve making changes to your diet and increasing physical activity.

If lifestyle changes are not enough to keep your glucose levels in the normal range (4 to 7.8mmol/L), we will consider starting treatment with insulin. Insulin is given by an injection under the skin, usually between one to four times per day. Not all people need the same level of treatment and the insulin we prescribe will be tailored to suit you.

Some people may need to have insulin therapy from the start. We will discuss the most suitable insulin regimen with you to make sure that it fits into your daily routine.

Occasionally we may also use tablet treatments to treat CFD.

Managing your diet with CFD

Keeping your body weight healthy is extremely important for people with CF.

Dietary advice for CFD is different to that given to people with other types of diabetes (Type 1 or Type 2 diabetes). People with CFD often (although not always) still need a high-calorie, high-protein and high-fat diet. Fat and protein affect glucose levels less than carbohydrates.

Carbohydrates can be split into sugary carbohydrates and starchy carbohydrates.

Sugary carbohydrates (simple sugars) are absorbed quickly and cause blood glucose levels to rise quickly. This rapid rise in glucose may be followed by a sudden drop to below normal glucose levels, an effect called reactive hypoglycaemia.

Examples of sugary carbohydrates:
- sugary drinks
- syrup
- honey
- jelly sweets

Starchy carbohydrates (complex carbohydrates) are absorbed from the gut more slowly and cause a gentler rise in blood sugar levels.

Examples of starchy carbohydrates:
- rice
- pasta
- potato
- bread
- non-sugary breakfast cereals

People with CFD should have regular starchy carbohydrates with every meal to help keep their blood glucose levels stable. They should limit the amount of sugary carbohydrates, particularly those in the form of sugary drinks, because of the effect on blood glucose levels.

Your CF dietitian will help you identify the types of food and drink that have high sugar levels in your diet and work with you to make small and gradual dietary changes whether you are on insulin treatment or not.

CFD and overnight feeds

Overnight feeds are given over a number of hours and therefore the glucose tends to be absorbed over a prolonged period. There are several insulins that can be given at the start of the feed to help manage blood glucose levels overnight. Your diabetes team will advise on the best insulin for you before starting your overnight feed.

What is a normal blood glucose level?

The normal range for blood glucose levels is between 4 to 7.8mmol/L. One significant difference between CFD and other types of diabetes is that your blood sugar levels may only be high after meals, while the levels before meals and when you haven’t eaten are normal.

When you first measure your blood glucose levels at home, we recommend that you test at

different times of the day to help build a picture of how your blood glucose levels change over the course of the day. You should measure your glucose levels before meals (breakfast, lunch and dinner) and two hours afterwards as well as taking a reading before bed. These can all be recorded on one day or spread throughout the week to give a better picture of what happens on different days (weekdays and weekend days).

This table explains the symptoms and treatment for abnormally low or high blood glucose levels:

	Low: Hypoglycaemia ('hypo') (blood glucose level less than 4.0mmol/L)	**High: Hyperglycaemia** (blood glucose level more than 7.8mmol/L)
Symptoms	• sweating • dizziness • hunger • looking pale • feeling faint • blurred vision • loss of co-ordination • headache • palpitations	• blurred vision • frequent urination • thirst • tiredness • poor weight gain • reduced lung function • increased risk of infection
Treatment	1.Take 15g of fast-acting carbohydrate such as: • five glucose tablets (3g carbohydrate each) • 150mls non-diet drink (1 mini can) • 150 to 200ml fruit juice (1 small carton) • one to two tubes of Glucogel® (10g each) 2. Wait 15 minutes and re-check your blood glucose level: If it is still less than 4mmol/L re-treat (as above). If your blood glucose level has increased, have 15 to 20g of slow acting carbohydrate (see examples below) to prevent sugar levels dropping again: • two digestive biscuits • one cereal bar • one medium slice bread/toast • one medium banana • one small bowl of cereal with milk	If your blood glucose levels are consistently above 8mmol/L, we will need to adjust your diabetes treatment. Please get in touch with your diabetes team. Please note that drinking water and exercise may help lower your blood glucose levels in the short term.

Is CFD for life?

CFD is caused by the body's decreased ability to deal with high glucose loads. Some people notice that when they are exercising regularly and reducing simple sugars in their diet, and their chest is also infection free, their blood glucose levels return to normal. This does not mean that they no longer have CFD but that they are able to keep their glucose levels healthy with lifestyle and dietary changes.

Occasionally, people with CFD may be asked to have another OGTT test after they have gone through puberty, especially if their insulin needs start to decrease. This is because the hormones that are released during puberty can affect how your body reacts to insulin. After puberty there is a drop in the level of these hormones and sugar handling may return to normal. Some people can then stop taking insulin.

If this is the case, we will ask you to have another OGTT test each year for five years. After five negative OGTT results, we would consider that you no longer have diabetes.

Who do I contact if I need help with my CFD?

If you are struggling to manage your CFD, contact your CFD specialist nurse or dietitian. They can either answer your questions over the telephone or book you into a clinic to see your specialist CFD doctor.

Can I drink alcohol?

If you want to drink alcohol, we recommend that you do so in moderation. Alcohol can initially raise your blood glucose levels (particularly if you drink it with sugary mixers) but after a few hours it tends to lower your blood glucose levels, especially if you are drinking on an empty stomach. You should consider eating or having a snack when drinking alcohol or soon after to avoid 'hypos'. Your CF team can give you more information about this.

What happens when I get ill?

Your blood glucose levels can rise or fall when you are unwell.

- If your appetite is reduced, your blood glucose may be lower than normal.
- If you have a chest infection or illness, your body may not respond as well to the effects of insulin, which may lead to raised blood glucose levels.

When you are unwell it is very important to monitor your blood glucose levels regularly (ideally every four to six hours). Continue with your usual doses of insulin unless your blood glucose levels are low, or the CF team advise you otherwise.

If you cannot tolerate solid foods, it is important to have sugary drinks or small snacks every two to three hours to stop your blood sugar levels dropping too low.

CFD and diabetic ketoacidosis (DKA)

Diabetic ketoacidosis (DKA) is a potentially life-threatening condition which can affect people who have diabetes. DKA is extremely rare in CFD and mostly affects people with Type 1 diabetes. However, if you are acutely unwell with very high blood glucose levels, your team will consider testing for ketones to see if you have DKA. A complete lack of insulin in the body results in the body breaking down fat to use as energy, and ketones are formed as part of this process. DKA treatment involves intravenous (IV) insulin and fluids.

CFD and driving

There are strict rules around driving with diabetes. When you apply for a driving licence, you must declare your diabetes if you are being treated with insulin. You will need to confirm that your diabetes is well controlled and that you are testing your blood glucose regularly, especially before driving.

Because driving with low glucose levels is dangerous, the DVLA recommends that you check your blood sugars before driving and every two hours during long journeys. The safe level for driving is a blood glucose of above 5mmol/L ('five to drive'). We recommend that you keep sugary snacks and a blood glucose monitor in the car for safety. Do not drive if your blood glucose level is less than 4mmol/L or you feel hypoglycaemic.

The DVLA now also accepts blood glucose readings produced by CGM systems for driving group 1 vehicles (cars and motorcycles).

If you have diabetes, you must renew your driving licence every three years and inform the DVLA if you are required to take insulin injections for more than three months.

DVLA
Telephone: **0300 790 6806**
Website: **www.gov.uk/diabetes-driving**

You will also need to inform your insurance company that you have CFD. If you don't, this may invalidate your insurance policy in the event of a claim.

Frequently asked questions

1. Can I still drink high-calorie supplements?
Yes. However, we recommend that you choose milk-based rather than juice-based supplements as they are less sugary. These are best taken at the end of meals when they will affect your blood sugar levels less, rather than as a snack between meals. Speak to your dietitian for further advice.

2. What is carbohydrate counting?
Carbohydrate counting consists of matching your insulin dose to the amount of carbohydrate that you eat at each meal. The amount of insulin needed by each individual is different, so the diabetes team working with your CF team will help you work out how much insulin you will need.

3. Do I get free prescriptions?
People with CFD who treat their diabetes with tablets or insulin are entitled to free prescriptions. If you are in England and you are younger than 60 years old, you will need to have a medical exemption certificate to claim your free prescription. This is available from your GP or post office, and will last for up to five years.

4. Do I get free eye tests?
Yes. Everyone with diabetes is entitled to free NHS sight tests.

5. Do I get free dental treatment?
No. Unfortunately having CFD does not automatically qualify you for free dental treatment.

Contact us
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Telephone: **023 8120 6801**

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For help preparing for your visit, arranging an interpreter or accessing the hospital, please visit **www.uhs.nhs.uk/additionalneeds**

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