

# Cystic Fibrosis Related Diabetes (CFRD)

In people with cystic fibrosis (CF), CFRD is caused by damage to the pancreas over a period of time. It is common for people with CF to be diagnosed with CFRD - it occurs in around 20% of adolescents and 40 - 60% of adults with CF. CFRD shares features with other types of diabetes but is a distinctly different condition, with different risks and different ways of managing these risks.

To understand CFRD, you need to understand a bit about the pancreas, glucose and insulin. The pancreas is an organ that has two main functions. The first is to make and secrete a hormone called insulin and the second is to make and secrete digestive enzymes and juices. In CFRD, the pancreas' ability to produce enough effective insulin is affected.

Insulin is a hormone that helps to control blood sugar levels, which can also be referred to as blood glucose levels. Glucose is a type of sugar that is used by cells in our bodies for energy. This is our body's main source of fuel. It is essential for life. Without glucose, our cells can't operate and carry out all their different functions.

We get most of our glucose from foods, particularly carbohydrate containing foods. The carbohydrates we eat are digested in our stomach and broken down into small particles - mostly glucose - and then transported around the body by the blood to the cells where energy is needed.


However, glucose can't get into the cells without insulin. Think of it like a lock and key system. The cells are 'locked' and in order for the glucose to get into the cells insulin is required, to act like a 'key', allowing the glucose in.

## **If glucose cannot enter our cells two things happen:**

1. The cells do not receive the energy they need to work properly
2. Blood sugar levels rise

## **Is CFRD different to other types of diabetes?**

Yes - There are 2 main types of diabetes, which most people have heard of.

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- **Type 1 diabetes** is an autoimmune condition in which the body attacks the cells in the pancreas (beta cells) which produce insulin. Type 1 diabetes accounts for around 9% of the diabetes population. It is usually (but not always) diagnosed in children and young adults and is managed with insulin.
  - **Type 2 diabetes** occurs when the body doesn't produce enough insulin or the insulin that is produced isn't recognised by the cells.
  - For some, but not all people, Type 2 diabetes is preventable. It may be managed with diet and exercise alone in some people and others will need to use medication as well as lifestyle changes to manage it. Type 2 diabetes used to be thought of as a disease only affecting adults, however it is becoming increasingly common in younger people. Type 2 diabetes accounts for over 90% of the diabetes population.
  - CFRD is different to other types of diabetes and the symptoms may vary from person to person. It may be triggered by an exacerbation of CF and treatment for infection or may be a gradual onset. It is often worse during CF exacerbations, as inflammation and infection make the cells of the body less sensitive to insulin, as can pregnancy and some medications.

The major risk from uncontrolled Type 1 and Type 2 diabetes is vascular (blood vessel) damage that occurs as a result of high concentrations of glucose in the blood over time. This is what causes problems like blindness, kidney damage and ulcers, amongst other things, in people with diabetes.

However, this is quite different to the major risks with uncontrolled CFRD. The major risks for uncontrolled CFRD are not vascular. They come from the body not being able to transport enough **glucose** to the cells for them to function optimally. The concerns seen in uncontrolled CFRD include weight loss, loss of protein stores in the body and poor health outcomes such as decline in lung function and increased mortality. Vascular damage can occur but it is not as common in CFRD as in other types of diabetes.

### **What are the symptoms of CFRD and high blood glucose levels (hyperglycaemia)?**

- Lack of energy
- A decrease in lung function
- Weight loss
- Increased hunger
- Excessive thirst
- Increased urine output

### **Does everybody who has CFRD have symptoms like this?**

No. Often people are diagnosed with CFRD during a routine blood test. Some symptoms of CFRD may also be very similar to other CF-related symptoms. Some people may experience only a few symptoms, others may experience no symptoms at all. This is why people with CF are regularly screened for CFRD.

## Why is it important to screen for CFRD?

- If the symptoms of CFRD are left untreated, your CF symptoms can become worse. This can lead to:
  - Increased thickness of mucus which becomes more difficult to clear
  - Reduced ability to fight infection
  - Loss of muscle
  - Loss of weight

Increased blood glucose levels over time can lead to other diabetes-related complications, such as damage to small blood vessels, impaired kidney function and vision impairment.

## Screening and diagnosis of CFRD

- The gold standard for CFRD testing is the Oral Glucose Tolerance Test (OGTT). Ideally this should be done annually. Other tests are used to check for Type 1 and Type 2 diabetes, but can sometimes miss people with CFRD. The OGTT is more sensitive and the best choice for screening in CFRD
- An OGTT is performed in the morning, after a person has fasted for 12 hours. A drink high in sugar (glucose) is given and then a Blood Glucose Level (BGL) is taken at set times following this to monitor insulin function.

## Pregnancy and CFRD

Women with CF are at increased risk of developing diabetes during pregnancy as many women are not able to produce the extra insulin which is needed during this time.

It is very important for the health of the baby to protect it from exposure to elevated blood glucose levels, and to prevent weight loss or inadequate weight gain in the mother during pregnancy.

If you have CF and are planning on having a baby, ideally you should have your OGTT checked prior to conception. If you have become pregnant and have not had your OGTT checked in the past 6 months, you should have it checked right away.

It is best to also repeat an OGTT at the end of the first and second trimesters.

## Managing CFRD

As patients with CFRD are insulin insufficient (don't have enough insulin) taking insulin is the treatment for CFRD. This helps people achieve 'glycaemic control', which means they have the right amount of glucose in their blood. Patients who achieve glycaemic control have better nutritional status, improvements in their weight, better lung function and better survival rates.

In other types of diabetes, losing weight and changes to the diet can play a big part in diabetes management, but a diagnosis of CFRD DOES NOT-



CHANGE THE EXISTING RECOMMENDATIONS for nutrition in CF. Weight loss is not part of the management plan, fats are not restricted and a high energy, high protein diet is still recommended. It is important that carbohydrate containing foods are consumed throughout the day. You should discuss this in more detail with your CF dietitian.

Diabetic Ketoacidosis or DKA is a serious complication of diabetes which can occur in other types of diabetes, but it is extremely uncommon in CFRD.

### What is hypoglycaemia - a “hypo”?


- A hypo is when blood glucose levels drop too low, which can be very serious if not managed properly.
- Mild hypoglycemia is common in CF and also in CFRD. This can occur if you haven't eaten in a while, known as a 'fasting state,' or it can occur after eating as part of a disordered insulin response. This is not what people are usually referring to when they talk about 'hypos.'
- A hypo is a blood glucose level of less than 4.0mmol/L
- Changes in diet and activity levels can mean that your insulin requirements may change. It is important to work closely with your CF team to learn about your individual needs and how to monitor your blood glucose levels.
- In CFRD hypos occur in response to taking too much insulin. They can be severe if not managed immediately. When glucose levels get too low the cells in the brain and muscles are not able to work properly.

### What are the symptoms of hypoglycaemia?

- **Symptoms of hypoglycaemia can vary from person to person and early signs may include:**
  - Feeling shaky and weak
  - Hunger
  - Sweating
  - Lightheadedness, dizziness
  - Headache
  - Pins and needles around the mouth
  - Mood change

### How do you manage a hypo?

- If you think you might be experiencing a hypo you need to eat 15g of fast acting carbohydrates. This is equivalent to 3 big jelly beans, 3 teaspoons of sugar or honey or 50ml of lemonade.
- Wait for 15 minutes and then retest your blood glucose levels.
- If the BGLs are still below 4.0mmol/L have another 15g of fast acting carbohydrate.
- If the levels are over 4.0mmol/L, have 15-20g of slow-acting carbohydrate such as a banana, cereal bar or 250ml of milk.



You could also eat your meal if it is due, to prevent your level from dropping again.

- Talk with your CF team about how to prevent and manage your hypos and always carry fast acting carbohydrates with you.
  - It is very important that you **DO NOT** drive if your BGLs are below 5 mmols.

For more information about CFRD, make an appointment to see a Diabetes Educator.

Contact your local CF office

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