

Cystic Fibrosis Related Diabetes

Cystic Fibrosis Related Diabetes (CFRD) occurs in about 20% of adolescents and 40-60% of adults with cystic fibrosis (CF). CFRD shares features with type 1 and 2 diabetes but is a different condition, with distinct risks and ways to be managed.

One function of the pancreas is to produce insulin; a hormone that helps control the blood sugar, or glucose levels. Glucose is the fuel that the cells need to operate effectively. The insulin is the “key” to allow the glucose to enter the cells. However, when the production of insulin is deficient, the cells do not receive the energy they need and the blood sugar level rises.

CFRD may be triggered by an exacerbation of CF, medication or it could be a gradual onset. With less transport of glucose to the cells, the glucose levels in the blood rise and people will present with:

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

Not everybody with CFRD will have these symptoms; often people are diagnosed during a routine blood test. Some people may experience few symptoms and others have no symptoms at all. For this reason, people with CF are regularly screened for CFRD.



The Oral Glucose Tolerance Test (OGTT) is the most sensitive and the best choice for screening for CFRD and it is recommended annually for people with CF who are 10 years and older.

The Importance of Screening for CFRD

When symptoms are left untreated, CF symptoms can deteriorate and lead to:

- Increased thickness of mucus which is difficult to clear
- Reduced ability to fight infection
- Loss of muscle mass and loss of weight

Over time, long-term complications, such as damage to small blood vessels, impaired kidney function and vision impairment can occur.

Pregnancy and CFRD

Women with CF are more prone to developing CFRD due to an increased need to produce extra insulin during

pregnancy. When planning a baby, an OGTT is recommended for those who have not been tested within the last 6 months.

Managing CFRD

Insulin is the treatment of choice for people with CFRD. Insulin helps control the right amount of glucose in the blood (glycaemic control). When glycaemic control is achieved, people's nutritional status, weight and lung function are improved, and their health outcomes optimised.

Contrary to people with other types of diabetes, people with CFRD need to continue with the recommended CF nutritional guidelines of a healthy fat, high energy diet. They should continue to consume food containing carbohydrates throughout the day.

Low Blood Sugar (Hypoglycemia)

A normal fasting blood glucose level (BGL) is between 4.8 and 8.0 mmol/L. Having a BGL below 4.0mmol/L (hypoglycaemia) can have serious repercussions, because the cells in the body are starved of sugar and not able to function properly. Hypoglycemia could occur when there is a change in diet and activity levels, or in response to taking too much insulin. Eat meals at scheduled times to help avoid hypoglycaemia.

Symptoms of Hypoglycemia

Symptoms vary from person to person; early signs may include:

- Feeling shaky and weak
- Hunger
- Headache and mood change
- Sweating
- Lightheadedness and dizziness
- Pins and needles around the mouth

Actions to Manage Hypoglycemia

Take 15g of fast acting carbohydrates, such as 6-7 jellybeans, 3 teaspoons of sugar or honey, 150ml of soft drink. Then wait for 15 minutes and retest BGLs:

- If your BGL is still < 4.0mmol/L take another 15g of fast acting carbohydrate.
- If over 4.0mmol/L, have 15-20g of slow-acting carbohydrate such as a banana, cereal bar or 250ml of milk.

Important Warning: Please do not drive if your BGLs are below 5.0 mmol/L.

Talk with your diabetes team about how to prevent and manage your hypoglycemia and always carry fast acting carbohydrates with you.

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

Useful Resources

- [CFRD \(Perth Children's Hospital\)](#)
- [CFRD \(NEMO\)](#)
- [CFRD \(CF Foundation\)](#)
- [Diabetes Australia](#)

Thank you to Sandra Wilberforce, Clinical Nurse Diabetes Educator, for input into this factsheet.

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