Hyperglycemia means high blood glucose. High levels of blood glucose are found in people with untreated diabetes. Because hyperglycemia may be harmful, it is important that it be correctly treated. People who have cystic fibrosis (CF) and diabetes, or cystic fibrosis–related diabetes (CFRD), share features of type 1 and type 2 diabetes, but CFRD is a unique and distinct form of diabetes and requires a special treatment approach.

About one third of the people who have CFRD have diabetes with fasting hyperglycemia; that means they have high blood glucose levels all the time, including when they wake up in the morning and before they have anything to eat.

Two thirds of the people who have CFRD have diabetes without fasting hyperglycemia; this means that their blood glucose levels are fine in the morning before they eat but become high after eating.

If you have CFRD with fasting hyperglycemia, you will need intensive treatment with insulin to help keep your blood glucose levels normal and to help you maintain a normal weight. Usually, you can be treated as an outpatient using a team approach.

If you are very ill or are also fighting a lung infection, you may need to be hospitalized. If you have CFRD without fasting hyperglycemia, you may or may not be treated with diabetes medications, depending upon your overall state of health.

What Can I Do?
If you have this form of diabetes, you are at very high risk for progressing to CFRD with fasting hyperglycemia. You should have a home glucose-monitoring device so you can check your own blood glucose levels. We recommend a glucose profile one day a month. A glucose profile is a series of three tests that you perform in one day—in the morning before you eat, before your main meal, and 2 hours after your main meal. Any time you are ill, you should do a
glucose profile daily. This is when you are at greatest risk for higher glucose levels.

**Treatment for People With CFRD With Fasting Hyperglycemia**
Like the team approach to CF lung disease, the team approach to treating diabetes has been shown to be the most effective method of care.

**The Team Approach to Diabetes Management**
You can receive coordinated care from a team of healthcare professionals that includes:

- Physicians with expertise in diabetes (endocrinologists)
- Diabetes nurse educators
- Dietitians
- Mental health professionals

**What Can I Do?**
The most important members of the treatment team are you and your family. The goal is to empower you to become self-sufficient in diabetes management to help keep yourself healthy. You can accomplish this goal by working diabetes care into your daily routines. Although you will have to add some new medical cares, you should not have to significantly change your lifestyle to fit the demands of diabetes management.

**Insulin Therapy**
At present, insulin is the only medical therapy officially recommended for treating people with CFRD and fasting hyperglycemia. At first you might be overwhelmed by the idea of adding insulin injections to your already complex medical routine. But most people become convinced that the injections are worthwhile.

**Benefits of insulin therapy.** Most people find that the additional insulin helps them:

- Gain weight, often much more than you have been able to gain for years
- Feel better
- Have more energy

**Insulin regimens.** People with CFRD usually require only very small doses of insulin to keep their blood glucose level normal. Many different insulin regimens are possible, depending on your needs. The most important time to take insulin is before meals, because it allows the body to fully use the nutrition and calories in the food.

- At the University of Minnesota, the most common insulin regimen chosen by patients who have been on insulin therapy for at least 6 months is rapid-acting insulin three to six times per day with meals plus a small dose of insulin glargine (a 24-hour background insulin) once a day. We use such tiny needles for insulin that most people do not find them to be at all painful. This schedule allows patients to eat whatever they want, whenever they want, as long as they compensate it with insulin.

- Some CF patients use an insulin pump. This pager-sized device can be clipped to a belt or pocket. It delivers insulin constantly through a small IV-like plastic tube inserted into the fat tissue of the abdomen or buttocks. It needs to be replaced every 2 to 3 days. Extra insulin is delivered with the push of a button to cover meals. Many patients appreciate the freedom from injections that a pump allows, whereas others would rather have injections than wear a mechanical device.

**Insulin dose and carbohydrate intake.** The insulin dose is based on carbohydrate (sugar and starch) intake. In other words, if you eat a high-carbohydrate meal of pasta, rice, or ice cream, you will require more insulin than if you had eaten a low-carbohydrate meal of meat or eggs.

If you are ill or experience nausea, you might not know how much food you can tolerate. In this situation, you might be advised to take a rapid-acting insulin after eating. Many people with CF receive nighttime continuous drip feedings. A single injection containing a combination of NPH (longer-acting) and regular (short-acting) insulin just before the feeding usually works well. The regular insulin covers
the first half of the feeding, and the NPH covers the second half. The glucose level 2 hours into the drip can be used to adjust the regular insulin, and the glucose level at the end of the drip can be used to adjust the NPH. The goal is to achieve a glucose level that is less than 150 mg/dL.

**Devices for delivering insulin.** Insulin can be given in a standard 30-unit, 50-unit or 100-unit syringe. Most people with CFRD use a pen device for insulin delivery.

If you are thin, make sure you use ultrafine, short needles for both syringes and pens. Insulin pumps are also effective in treating CFRD, although the rate that the insulin is injected is much lower than the rate in people who have other forms of diabetes. Oral diabetes medications are not recommended except for people who participate in research studies.

**Dietary management while taking insulin.** The diet recommended for people with CFRD is very different from the diet recommended for people with other forms of diabetes.

People with CFRD need a high-calorie diet to maintain a normal body weight, while people with other forms of diabetes usually need a diet that enables them to lose weight.

- People with CF usually have low cholesterol levels, do not appear to develop heart disease, and lose excessive amounts of salt in their sweat. Thus, a high-calorie, high-fat, high-salt diet is recommended for all persons with CF, including those with diabetes. However, people with other forms of diabetes are told to cut back on fat because they are at risk for developing hardening of the arteries (atherosclerosis) and heart disease. They are told to cut back on salt because they are at risk for high blood pressure.
- There is no need to cut back on sweets if you have CFRD or only diabetes, as long as you take the right dose of insulin.

**Monitoring.** While you are on insulin therapy:

- You will be asked to monitor blood glucose levels at least four times a day by sticking your finger to obtain a blood sample. If you have sensitive fingertips, try a lancet device with adjustable depths and a glucose monitoring device that requires minimal amount of blood. The glucose goal is 80 mg/dL to 120 mg/dL (4.4-6.7 mmol/L) fasting and less than 150 mg/dL (8.3 mmol/L) 2 hours after eating.
- You will need to be monitored for the same eye, kidney, and nerve problems as people with other types of diabetes. If these problems are caught early, they can be treated.
- You will also need to have regular blood pressure measurements, foot examinations, annual dilated eye examinations, and annual assessment of urine albumin (protein) concentration.
- You should have your cholesterol and triglyceride levels checked once diabetes has been brought under control. Hyperlipidemia (high levels of fats in the blood) and macrovascular disease (such as heart disease, stroke) do not appear to be of concern for people with CF, but this may change as people with CF live longer. Your cholesterol will probably be low, in which case it does not need to be checked again. If it is found to be high, however, it should be followed more closely by your healthcare provider.

Too much insulin can cause your blood glucose to fall too low, which is called hypoglycemia. Before the relatively recent availability of rapid-acting insulin, hypoglycemia was quite common 3 to 4 hours after meals or during the night. Although your symptoms may not be severe, they can be uncomfortable. However, you might be able to avoid hypoglycemia by using rapid-acting insulin (lispro or aspart) instead of longer-acting insulin. Low blood glucose can be treated by consuming 15 g to 30 g of a quick-acting carbohydrate (such as juice or glucose tablets).
When People With CF With or Without Diabetes Are Hospitalized

During acute illness, you are insulin resistant and blood glucose levels are at their highest. Diabetes may appear even if you have never before had diabetes. If you have fasting hyperglycemia for more than 48 hours, insulin therapy is usually recommended. There is about a fifty-fifty chance that you will be able to stop taking the insulin within a month or two or you will need to continue to take insulin after your health improves.

If you take insulin on a regular basis and then become acutely ill, you will require much more insulin, perhaps even four times your normal amount. An unexpected rise in your insulin needs may be the first sign that you are acutely ill. Insulin doses must be aggressively increased and then lowered as the physical stress waxes and wanes.

Can Diabetes Worsen Lung Function?

Medical researchers have reported that people with CFRD are more underweight and have worse lung function than people with CF who do not have diabetes. The worse the insulin deficiency, the worse the decline in lung function. Insulin deficiency in CF often leads to abnormal protein breakdown and thus negatively influences

- Weight
- Pulmonary function
- Survival

Fortunately, insulin therapy appears to be able to reverse this process. Some researchers have found that insulin therapy helped participants gain weight and lung function that had been declining for several years before diabetes was diagnosed.

Impaired Glucose Tolerance

Impaired glucose tolerance means that your blood glucose levels are abnormal, but not in the diabetic range. If you have CF along with impaired glucose tolerance, you are at greater risk of developing diabetes than people with CF who have normal glucose tolerance. The symptoms of diabetes are often silent. Therefore, be sure to have an annual oral glucose tolerance test to make sure you are not developing a more serious glucose problem.

Summary

Diabetes is common in people with CF. However, diabetes develops gradually, and the beginning stages have no symptoms. Early in the disease process, you won't know you have diabetes unless your healthcare provider tests you for it. Although CFRD shares many features with type 1 and type 2 diabetes, CFRD is a distinct medical condition that requires a unique treatment approach. To prevent setbacks in your nutritional status and perhaps damage to the small blood vessels, be sure to work closely with your healthcare team. Remember, YOU are the most important member of the team.

References


Lanng S, Thorsteinsson B, Nerup J, et al. Influence of the development of diabetes mellitus on clinical status in patients with cystic...


This article, which appears on mycysticfibrosis.com, was written by Antoinette Moran, MD, a professor in the Department of Pediatrics at the University of Minnesota, Minneapolis. Dr. Moran, a pediatric endocrinologist, is also conducting research in cystic fibrosis–related diabetes. She reviewed this article in 2007.