CYSTIC FIBROSIS

CF Fact
cystic fibrosis fact sheet

Nutrition for Adults

Diet is particularly important in the management of cystic fibrosis (CF), as a person with CF who has a good diet may get fewer lung infections, recover more quickly from infections, maintain better lung function and have more energy for everyday activities.

Enzymes

Approximately 85% of adults with CF are pancreatic insufficient. This means that the pancreas is unable to produce or release enough digestive enzymes into the small intestine to digest food properly. To manage this, most people will take pancreatic enzymes capsules with food to aid in digestion.

The main goal of this is to optimise nutrient absorption, improve weight gain, and prevent nutrient deficiencies. Forgetting or taking too few enzymes will result in diarrhoea, abdominal pain, and over time, weight loss or difficulty in gaining weight. It is important to correctly match your enzyme doses with your food and fluid intake to optimise nutrient absorption. See your CF dietitian for further help determining your individual enzyme requirements.

Diet

A balanced diet is important and should include all food groups (unless otherwise advised). Adults with CF can require between 10 to 100% more calories in their daily diet than those without CF because they do not absorb food as easily and the body uses more energy to function.

Specific energy requirements should be individualised and guided by a CF dietitian, as they will differ based on gastrointestinal and pulmonary symptoms, gender, age, weight, height and physical activity levels. Dietary needs may again change with pregnancy, transplant, diagnosis of CF Related Diabetes (CFRD) or changes in certain medications.

Vitamins and Minerals

Vitamins

People with CF, particularly those who are pancreatic insufficient, often have deficiencies in 'fat-soluble' vitamins A, D, E and K due to the body's limited or total inability to absorb these vitamins. VitABDECK is a CF-specific multivitamin that is generally prescribed to people with CF.

Each vitamin is important for different bodily functions:

- Vitamin A for immune health and night

vision.
- Vitamin D for bone health.
- Vitamin E as an antioxidant.
- Vitamin K for normal blood clotting and bone health.

Salt

People with CF lose large amounts of sodium and chloride (minerals that make up salt) in their sweat therefore these minerals need to be supplemented through diet and/or supplements. Ensuring adequate salt intake is important to prevent low salt levels in the body which can lead to dehydration and symptoms such as fatigue, nausea, muscle cramps and thicker mucus. The amount of salt replacement needed for each individual varies according to symptoms, dietary intake, climate and activity levels.

Gaining Weight

Often people with CF can struggle to gain weight despite the use of enzymes and increased caloric intake. It can be difficult to maintain a high calorie diet, particularly during periods of infection when appetite is often poor and other symptoms such as nausea or increased coughing can impact food intake.

Fat 'boosting' is often used to help with weight gain. This is simply adding more fat to a meal so it is higher in calories without having to consume larger quantities of food. For example, adding 2 tablespoons of peanut butter to celery adds an extra 20g of fat and over 200 calories. Fat is the easiest food source to use at it is the most concentrated source of calories.

Other tips include using full-cream milk, avoiding 'low-fat' food options, using nutritional supplement drinks, adding sauces to food, trying to eat more often throughout the day and having ready-to-eat snacks on hand at all times. Supplemental feeding directly into the stomach via a percutaneous endoscopic gastrostomy (PEG) or nasogastric tube (NGT) may also be needed if, despite all other options, weight gain is still an issue.

Overweight and Obesity

Surprisingly, new data from around the world shows that the proportion of people with CF who are overweight or obese is increasing. Part of this can be attributed to advancing CF treatments such as genetic modulator therapies.

There is currently no research into the health risks associated with CF and obesity, however with increasing life expectancy, people with CF may potentially be at risk of the same metabolic risks (heart disease, diabetes, stroke) as the general population. It is important to ensure you are guided by a CF dietitian to maintain a healthy weight.

Dietary Review

At least once a year, people with CF should:
- Meet with the CF dietitian to review diet, weight changes, vitamins and enzymes.
- Make a plan and set clear goals regarding weight and diet.
- Have blood levels of fat-soluble vitamins checked.
- Ask about having an oral glucose tolerance test (OGTT) to check for CFRD.

Useful Resources

- CFFood (CFWA)
- CFCooking (CFWA)
- Pancreatic Enzyme Replacement Therapy for Adults with Cystic Fibrosis (NEMO)
- Cystic Fibrosis and High Energy Diet (NEMO)
- Vitamin Supplementation and CF (NEMO)
- CFWA Nutrition Factsheets

Thank you to Jordan Henderson, Clinical Dietitian, Sir Charles Gairdner Hospital, for input into this factsheet.