

Nutrition & Cystic Fibrosis

A Guide For Adults

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About this guide

This booklet has been developed to provide you with practical, general and factual information about the cystic fibrosis (CF) diet. It is designed to be a general guide only, so it is important to note that some of the information might not be relevant to you.

The dietary recommendations for people with CF have changed a lot over the years and guidelines now suggest that dietary advice should be individualised based on gastrointestinal symptoms, pulmonary symptoms and factors such as gender, age, weight, height and physical activity levels.

Your CF dietitian, in conjunction with the CF care team should advise you on your specific dietary requirements,

and be your first port of call when questions or concerns arise.

To keep on top of your nutritional requirements, at least once a year, you should:

- meet with your dietitian to review your diet, supplemental vitamins and your use of enzymes
- have a plan and set clear goals regarding your weight and/or diet
- have your blood levels of fat-soluble vitamins checked
- ask about having an oral glucose tolerance test (OGTT) to check for CF related diabetes (CFRD)

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1. Nutrition for pancreatic sufficient adults

In Cystic Fibrosis (CF), often the pancreas is affected and this is referred to as pancreatic insufficiency, or being pancreatic insufficient.

If you do not suffer from pancreatic insufficiency, this is termed being 'pancreatic sufficient'.

It means that your body is able to digest food naturally and therefore medication to assist with digestion is not required.

This means that usually you can follow a normal, balanced diet. The diet is the same as what is recommended for the general population and should include a wide variety of nutritious foods from the 5 food groups:

- vegetables
- fruit
- grain (cereal) foods such as breads, cereals, rice, pasta, noodles, polenta, couscous, oats, quinoa and barley

- lean meats and meat alternatives such as poultry, fish, eggs, tofu, nuts, seeds and legumes/beans
- dairy foods such as milk, yoghurt, cheese and their alternatives

You should limit intake of foods containing trans and saturated fats and added sugars.

It is also worth considering that some people who are pancreatic sufficient can be susceptible to pancreatitis and some can become pancreatic insufficient at a later stage.

Some of the hints and tips included in this booklet may still be relevant for pancreatic sufficient people such as salt supplementation, vitamins and bowel issues.

2. Enzymes

Approximately 85% of people with CF suffer from pancreatic insufficiency. This means the pancreas is blocked with mucus, preventing the body from breaking down and digesting food naturally.

If the pancreas cannot work properly then Pancreatic Enzyme Replacement Therapy (PERT) will be required to assist with the digestion of food.

The main goal of treating pancreatic insufficiency is to optimise nutrient absorption, improve weight gain, and prevent nutrient deficiencies.

If you are pancreatic insufficient you will need to take enzymes with most foods. The most commonly used enzyme in Australia is Creon.

You need to take enzymes with any foods that have fats, carbohydrates and proteins in them to help absorb the energy and nutrients contained in food and to prevent malabsorption. It is important to remember to:

- take the right number of enzymes-based on the amount of fat in your food
- take enzymes at the right time - enzymes only work for around 30 minutes, so if you are eating slowly or snacking over a long period of time you will need to take more enzymes after the 30 minutes is up
- take enzymes the right way- they should be swallowed whole as a tablet, not crushed or chewed.

The enzymes tablets will break down fats, carbs and proteins but are dosed based on the amount of fat in food.

Below is a rough guide of how many enzymes are required per gram of fat in food, however dosages change from person to person and your CF team will guide you in what dosage is best for you.

1 Creon 10,000 capsule per 6-8g fat
1 Creon 25,000 per 15-20g fat

3.

The CF diet

Long term incorrect dosage of enzymes can have detrimental effects on your nutrition status, weight and immunity. If you are unsure of how many enzymes you should be taking, contact your CF team and they will help you.

HOW DO I KNOW THE DOSE IS CORRECT?

The effectiveness of your PERT should be routinely monitored by your CF care team. Weight gain is generally a good indicator of how well the enzymes are working and if you are digesting and absorbing your food.

The following symptoms may be signs of malabsorption (inadequate uptake of nutrients from food) and could mean your enzyme dosage may need reviewing:

- constipation, stomach ache, diarrhea or wind
- loose, greasy or floating stools
- weight loss

- very bad smelling stools or wind
- mucousy or oily stools

WHAT IF I FORGET TO TAKE ENZYMES?

Nothing drastic is likely to happen if enzymes are forgotten during a meal, other than possibly an upset stomach.

If enzymes are not taken regularly over a period of time, you will experience weight loss and loss of nutrients from your food, which will ultimately result in poor health outcomes.

If enzymes are forgotten at the start of the meal, you can still take them during a meal or at the end, as long as it's within 5-10 minutes of eating.

Diet is a very important part of the management of CF and plays a significant role in how well you feel.

Good nutrition and a healthy body weight have been directly linked to having better lung function, so making sure you eat enough to maintain a healthy weight is important to your overall health.

A person with CF who has a good diet and is well nourished may:

- get fewer lung infections
- have the ability to fight and recover quicker from infections
- maintain better lung function
- have more energy for everyday activities

It is accepted that most people with CF need to eat more calories than the general population, however individual recommendations should be provided as those with lower body weight or who are unwell may require more daily calories than those who are able to maintain a healthy body weight.

WHAT FOODS SHOULD I EAT?

A balanced diet is important for people with CF. This includes all food groups (unless otherwise advised), including breads and cereals, fruits and vegetables, dairy products and protein foods like meat, chicken, fish, legumes, nuts and eggs.

Specific energy requirements should be individualised and guided by your CF care team as there is not a 'one-size-fits-all' method for determining energy needs; rather they should be based on gastrointestinal and pulmonary symptoms as well as factors such as gender, age, weight, height and physical activity levels.

Our bodies get energy from 3 main nutrients, or 'macronutrients'. These are; Fats, Proteins and Carbohydrates. Each of them is essential and they will be discussed in more detail below.

Diets, 'lifestyles' or eating plans which encourage the removal of any of these three macronutrients are unbalanced

and could present serious risks for a person with CF.

FATS

Guidelines state that adults with CF should get about 35-40% of their daily food intake from fat sources, which is more than the 30% that is recommended for the general population. Your dietitian will be able to recommend your specific daily requirement in grams based on your energy needs.

For the general population, high consumption of ‘bad’ fats can lead to health issues such as cardiovascular disease, obesity, diabetes and more, and many people with CF also worry about the long term health effects of a high fat diet.

Typically, pancreatic insufficient individuals with CF have been shown to have normal cholesterol levels, as well as minimal risk factors for cardiovascular disease despite their high fat intakes.

This is thought to be most likely due to poor absorption of fat. With individuals with CF now living longer, the long term effects on cardiovascular health

are being considered, and concerns may exist particularly for those with a higher body mass index.

For some individuals, increased intake of ‘healthier’ fats rather than ‘bad’ fats may be encouraged.

There are four main types of fats; polyunsaturated, monounsaturated, saturated and unsaturated (trans) fats.

Polyunsaturated and monounsaturated fats are considered ‘healthier’ fats as they come from natural products and contain good cholesterol.

‘Bad’ fats may include saturated and unsaturated (trans) fats. Saturated fats are still often good options for some people with CF as although they are higher in cholesterol, people with CF tend to have low cholesterol levels, so they are not considered such a problem.

Unsaturated (trans) fats are considered the least nutritionally beneficial source of fat, however are a good option for those struggling to maintain a healthy weight.

Here are some example of different sources of dietary fat:

Polunsaturated fats	Monounsaturated fats	Saturated fats	Unsaturated (trans) fats
fatty fish (such as salmon, tuna, herring and sardines)	olive oil	visible fat on meat such as: beef, veal, lamb, pork, poultry fat	commercially baked products (e.g. biscuits and pastries)
vegetable oils (such as sunflower, safflower and soybean oils)	canola oil	butter	deep fried takeaway foods
fish oils	peanut oil	cream	
seeds (such as flaxseed, pepitas, sunflower seeds)	avocado	dairy made from whole milk such as milk and cheeses	
legumes	natural peanut butter	plant fats including coconut oil, palm oil and cocoa butter	
soybeans	nuts (e.g. almonds and peanuts)		
nuts (e.g. walnuts and peanuts)	seeds		



PROTEINS

Generally, protein foods should make up about 10-15% of the diet for people with CF. Your dietitian will be able to recommend your specific daily requirement in grams based on your needs.

Protein plays an important role in helping your body repair tissue. Muscle, skin, hair and fingernails are all made of protein. Here are some suggestions for protein sources and how to add them to your diet:

FOOD	IDEAS
MEAT & MEAT ALTERNATIVES	<ul style="list-style-type: none">› meat, chicken, fish and meat alternatives such as baked beans, lentils, kidney beans, tofu› include at each main meal and as snacks
CHEESE	<ul style="list-style-type: none">› serve on crackers/sandwiches› grate onto vegetables› add to rice/pasta› cut into small blocks as a snack› make cheese sauce to add to meals/vegetables
FULL CREAM MILK	<ul style="list-style-type: none">› try milk based drinks› make soups, puddings, custards, desserts or packet mixes with milk instead of water
EGGS	<ul style="list-style-type: none">› mash egg with mayonnaise as a sandwich topping or stir through potato salad› try an omelette or quiche with chopped meat, vegetables and cheese
NUTS & SEEDS	<ul style="list-style-type: none">› use smooth peanut butter or other nut pastes rather than jam or vegemite› use hummus or tahini as a dip or spread› eat whole roasted nuts as a snack› use in baking (e.g. almond meal)

CARBOHYDRATES

Around 50% of all calories in a CF diet should come from carbohydrates.

Your dietitian will be able to recommend your specific daily requirement in grams based on your energy needs.

Low carbohydrate diets such as ‘the paleo diet’ are popular in media at the moment, however these may be dangerous for people with CF, who have difficulty with digestion and absorption of nutrients and energy.

Carbohydrates are the easiest form of energy for the body to digest. They are naturally found in breads, cereals, grains, legumes, dairy products, fruit, starchy vegetables, and in small amounts in non-starchy vegetables.

They are also found in other foods such as lollies, sweetened drinks, sauces and condiments.

The term carbohydrate refers to a chemical structure and it includes sugars and starches.

Starchy foods such as breads, pastas, crackers, rice, legumes and vegetables like potatoes are great energy sources

and should be included at most meals and snacks.

The energy from starches is released into the body more slowly than the energy from sugar because it takes the body longer to digest it, however it is still digested faster than the energy from fats and protein.

Sugar has had a lot of discussion in the media lately and it has not always been portrayed accurately. Sugar is naturally occurring in many healthy foods (fruit or dairy products) and is a healthy inclusion in the diet.

Sugar that is added to food can be an issue if it is consumed in large quantities, but a small amount is ok.

As people with CF often have increased energy requirements, there can sometimes be more ‘room to move’ with intake of certain nutrients than for the general population.

You are best off speaking to your CF team for individualised advice about sugar consumption.

FIBRE

The term 'fibre' refers to the part of the food which is not digested by the body, and remains intact throughout its passage through the digestive system.

It is a particularly important part of the CF diet as it helps form and move stools through the digestive tract, decreasing the chance of bowel blockages.

A high fibre diet not only helps prevent or minimise risk of Distal Intestinal Obstruction Syndrome (DIOS) and constipation, it helps to maintain a healthy microbiome – in other words it helps to keep the good bacteria (probiotics) that lives in your bowel, healthy. As we learn more about the microbiome we are learning that it may play an important role in supporting our immune system.

Fibre is usually found in foods which contain carbohydrates, and there are some sources of fibre listed below:

FIBRE

brown rice
whole wheat & other whole grains
seeds & nuts
oatmeal
beans & lentils
fruits & vegetables

To maximise your intake of fibre, choose whole grain foods over white, leave edible skins on fruits and veggies, add nuts and seeds to dishes or enjoy them as snacks and choose pulses and legumes at least once a week. Talk to your dietitian regarding how many grams of fibre per day to aim for.



VITAMINS AND MINERALS

Fat-soluble vitamins

People with CF, particularly those with pancreatic insufficiency, 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.

Deficiencies could affect the health of your bones and eyes, as well as

the body's ability to fight infections; therefore most people with CF will need to take vitamin supplements. Vitamin levels should be checked at least once a year at annual review and more frequently if required.

The following table details each of the fat-soluble vitamins, their health benefits and food sources:

VITAMIN	BENEFIT	FOOD SOURCES
VITAMIN A	<ul style="list-style-type: none">› Supports immune function, normal vision, bone and tooth formation, cell function and immunity.› It also works to help fight infections to keep the intestines healthy.	<ul style="list-style-type: none">› liver, egg yolk, whole milk and fortified low-fat milk, fortified cereals, dark coloured fruits and vegetables (e.g. carrot, sweet potato, spinach, broccoli, apricot, rockmelon and peaches).
VITAMIN D	<ul style="list-style-type: none">› Helps build and maintain strong bones and teeth.› Without enough vitamin D, bones can become thin and brittle.	<ul style="list-style-type: none">› The major source of vitamin D in Australia is exposure to sunlight, while small amounts come from foods such as fortified cereals and fortified soy products, fish (e.g. mackerel and canned sardines), fish-liver oils (e.g. cod-liver oil).› Dairy products may not be made with vitamin D, so be sure to read labels.
VITAMIN E	<ul style="list-style-type: none">› Is an anti-oxidant and important for nerve and muscle function.› Helps keep red blood cells healthy and also helps fight infection and maintain the health of the intestines.	<ul style="list-style-type: none">› wheat germ, nuts, eggs, vegetable oils, green leafy vegetables and fortified cereals.
VITAMIN K	<ul style="list-style-type: none">› Important for blood clotting and bone health.	<ul style="list-style-type: none">› green leafy vegetables (e.g. spinach, broccoli), liver, soya beans, and canola oils.

VitABDECK is a CF-specific multivitamin that is generally prescribed to people with CF.

To enhance their effectiveness the vitamins should be taken with enzymes and a fat-containing food or drink. Your dietitian will be able to recommend your specific daily requirement.

SALT

People with CF lose large amounts of sodium and chloride (minerals that make up salt) in their sweat.

As the body cannot make sodium and chloride it must be supplied through diet and/or supplements.

The amount of salt replacement needed varies for each individual according to symptoms, dietary intake, the climate you live in and levels of physical activity.

Further increased amounts of salt may be required when you are unwell, if you are eating less or are having tube feeds, if you are engaging in excessive exercise or are living/holidaying in a hot climate.

Not getting enough salt can reduce appetite and cause stomach pain and dehydration. Your dietitian will be able to recommend your specific daily requirement, which may come from food and/or supplements.

Signs of dehydration are:

- feeling tired
- moody/grumpy
- headaches

- poor concentration
- salt crystals on the skin
- nausea/vomiting
- decreased appetite
- muscle cramps
- dark urine colour
- thicker, harder to clear mucus
- constipation

Tips for avoiding dehydration include:

- add table salt to your food
- always carry a water bottle
- eat salty foods like Vegemite, pretzels, soy sauce, tomato sauce, chips and nuts
- use plenty of sauces/gravies/condiments
- choose the ‘salted’ varieties of foods
- drink sports drinks such as Hydralyte
- take salt tablets

Here are some ways to include salt in your diet:

SALTY FOODS
chips
pretzels
rice crackers
white bread
salted nuts
sauces (e.g. tomato, soy)
stock cubes
bacon
cup a soup
baked beans
Vegemite
salt tablets
sports drinks

CALCIUM

Calcium is needed daily by everyone. It is essential for strong bones and teeth. As the body cannot make calcium it must be supplied through diet and/or supplements. If you don't get enough calcium through your diet, your body can start taking calcium from your bones, putting you at an increased risk of bone breakages and osteoporosis.

Currently the recommended daily intake for people with CF is the same as that for the general population (1200 mg daily), unless you suffer from low bone mineral density, in which case your dietitian may recommend more.

SHOULD/CAN I DRINK ALCOHOL?

Drinking alcohol can be a very enjoyable and sociable part of life, however the effects of alcohol on medications, the liver and your general health should all be taken into consideration.

For people with CF the functioning of the liver is very important. A well-functioning liver may be able to tolerate small amounts of alcohol.

The amount of alcohol to allow in your diet should be a decision made between yourself and your care team.

CALCIUM	APPROX. CALCIUM CONTENT IN MG PER 100G/100ML
Calcium fortified milk (e.g. Lite Start, Shape, Pura tone)	120
Yoghurt	100
Tinned salmon with bones or sardines	277-382
Cheese (cheddar)	721
Calcium fortified soy milk	190
Custard	145
Ice-cream (full cream or low fat)	127
Almonds	248
Tofu	683
Meat (beef)	20
Eggs	25 mg per egg
Baked beans	31
Oranges	38 mg per orange
Bread, whole wheat	138 (approx. 40mg per slice)
Cottage cheese	61
Broccoli	40
Spinach	99 (approx. 3 cups)
Dried Apricot	55 (approx. 4 mg per apricot)

Meal ideas

Here are some ideas for foods that are good to include in your daily diet:

Breakfast

toast with eggs or avocado and sautéed vegetables (mushroom, tomato etc)
buttered toast with Vegemite and a glass of milk
cereal with fruit and full cream milk
muesli with yoghurt
porridge with dates, walnuts and full cream milk
berry smoothie made with full cream milk and pumpkin seeds
vegetable omelette with cheese
pancakes with bananas, chopped almonds and cream
bagel with salmon and cream cheese
beans, cheese and scrambled or fried eggs on toast

Lunch

sandwiches with fillings such as: cheese, ham, chicken or tuna with avocado, chutneys/sauces and salad
veggie and feta frittata and a side salad
tuna pasta bake with a side salad
homemade mini pizzas with cheese, meats and vegetables
chicken, tuna or egg & salad wrap with mayo, avocado, hummus or other dressing
homemade nachos (with mince, kidney beans, cheese, salsa, sour cream)
baked beans, or sardines on toast with a piece of fruit, jacket potato with olive oil/ butter and baked beans or chilli and sour cream

Dinner

quiche with salad and baked potato or bread roll
spaghetti bolognese with grated cheese
fettucini with chicken or bacon , mushrooms, onions, snow peas and creamy sauce
fried rice with egg, vegetables, meat and sesame oil
coconut cream curries with tofu or meat, veggies and rice
grilled chicken and sweet potatoes with sautéed spinach and pine nut pesto
steak with a creamy sauce, steamed veggies and mashed potatoes
fish and roasted vegetables with garlic bread
tacos, enchiladas or burritos with meat, guacamole, cheese, capsicum & onion

Snacks

nuts (almonds, pistachios, cashews) and seeds (pumpkin, sunflower)
fruit with yoghurt
muesli or protein bar
veggie sticks with ricotta or peanut butter
wholegrain crackers with Vegemite or avocado
toasted Lebanese/pita bread with dips
crackers with avocado, cheese, cream cheese, tuna, hummus or cream cheese dips
hard boiled eggs
tuna kit (comes with tuna, crackers and mayo)
dried fruit
cereal and milk

Drinks

smoothie: full cream milk, fruit, ice cream/yoghurt - you can add honey or other flavourings
creamy milkshakes with ice cream, toppings or flavourings such as Milo
Sustagen and other supplement drinks

4.

Gaining weight

Often people with CF will struggle to gain weight. The pressure to eat enough food and maintain or even gain weight can make eating feel like more of a chore than a pleasure.

If you struggle to gain weight it might be difficult at times to deal with other people's reactions, such as the idea that you are 'lucky' to be thin.

These comments can often be frustrating or disheartening. Talking to your CF care team about concerns or anxieties you may have relating to your weight can be beneficial.

If your CF care team want you to gain weight, it can be useful to start by setting small goals, as this can make the prospect less daunting. Agreeing on a target weight might have to be a compromise between you and your team, but it is important you are in agreement with this.

FAT BOOSTING

Eating foods which are higher in fats is often the easiest way to gain weight. Adding fats to your meals will increase the amount of calories without having to increase how much food you eat.

This is known as 'boosting' the calories in your food (for example, if you added 2 tablespoons of peanut butter to some celery, you can get an extra 20g of fat in that snack). The type of fats you choose to eat are up to you.

Here are some tips to boost your daily calorie intake:

USE FULL CREAM MILK	<ul style="list-style-type: none"> › Much higher in calories than skim milk. › Use them with everyday meals like porridge, cereal, or mashed potatoes.
AVOID 'LOW FAT' AND 'FAT FREE' FOODS	<ul style="list-style-type: none"> › You're better off with the normal versions of these foods. › Always choose full fat dairy and salad dressings.
EAT MORE OFTEN IF YOU CAN'T EAT A LOT	<ul style="list-style-type: none"> › Aim to eat six smaller meals rather than three big meals a day, which may be too filling.
PROVIDE HIGH CALORIE DRINKS	<ul style="list-style-type: none"> › Milk, milkshakes, hot chocolate and nutritional supplements.
ADD SAUCES	<ul style="list-style-type: none"> › Sauces, gravies, dips or toppings can add lots of calories to food.
HAVE READY-TO-EAT SNACKS HANDY	<ul style="list-style-type: none"> › Yoghurt, nuts, cheese, dips, and mini muffins.
BOOST FRUIT AND VEG WITH HIGH CALORIE ITEMS	<ul style="list-style-type: none"> › Eat vegetables that are naturally high in fat (avocados, olives, coconut) › Add butter or a cheese sauce to veggies (you can also add salt; this doesn't increase the calories but it does add more flavour). › Make a smoothie by mixing fruit, whole milk (or non-dairy option if dairy isn't tolerated) and ice cream together. Chia seeds can also be a great addition to smoothies. › Add items such as nuts, seeds, croutons, hardboiled eggs and cheese to salads.

SUPPLEMENT DRINKS

Nutritional supplements can be used in conjunction with your normal food to further increase the calories in your diet. They come in both powder and ready to drink forms and are generally available in supermarkets or pharmacies.

NAME	BRAND	FLAVOURS	AVAILABILITY
Powdered Supplements			
Milk powder	any brand		supermarket
Sustagen	Nestle	chocolate/vanilla	supermarket
Sustagen Hospital Formula	Nestle	chocolate/vanilla/neutral	pharmacy
Proform Nutrition & Energy	MG Nutritionals	neutral	gluten free
Ensure	Abbott	chocolate/vanilla	pharmacy
Ready to drink 'milky' supplements			
UP & GO	Sanitarium	variety	supermarket
Protein Revival	Aussie Bodies	variety	supermarket
Sustagen	Nestle	chocolate/vanilla	supermarket
Resource Plus	Nestle	chocolate/vanilla/strawberry	pharmacy
Resource 2.0	Nestle	variety	pharmacy
Novasource 2.0	Nestle	vanilla	pharmacy
Fortisip Multifibre	Nutricia	variety	pharmacy
Ensure Plus	Abbott	vanilla	pharmacy
Two Cal HN	Abbott	vanilla	pharmacy
Ready to drink 'fruity' supplements			
Enlive Plus	Abbott	variety	pharmacy
Fortijuice	Nutricia	apple, forest fruit, tropical	pharmacy
Resource Fruit Flavoured Beverage	Nestle	peach, wild berry, orange	pharmacy



You can also make your own high calorie drinks yourself:

Chocolate Milkshake	Calories (432 total)	Fat (21g total)
1 cup milk 1 tablespoon of milk powder 1 tablespoon of Milo 2 scoops of ice cream	178 37 21 196	8.8g 2g 0.5g 10g
Enriched Milk	Calories (215 total)	Fat (10.8g total)
To every cup of milk add 1 heaped tablespoon of full cream milk powder and whisk until dissolved.	178 (for 1 serve) 37	8.8g 2g
OR To 2 cups of milk add 1 cup of evaporated milk	Calories (244 total) 296 244	Fat (22g total) 18g 4g
Fruit Smoothie	Calories (510 total)	Fat (21g total)
1 cup milk, 1 ripe banana or cup of tinned fruit 1 tablespoon of milk powder 2 scoops ice cream	178 99 37 196	8.8g 0.1g 2g 10g

SUPPLEMENTAL FEEDING

Supplemental feeding may be required if you struggle to gain weight and reach your energy requirements.

Supplemental feeds, otherwise known as enteral feeding can help improve nutritional status by delivering extra calories directly into the stomach via the nose (naso gastric tube) or abdomen (Percutaneous Endoscopic Gastrostomy, or PEG).

They can also provide overnight feeds, allowing you the freedom to enjoy normal meals and activities during the day.

Enzymes are needed with the tube feedings. Some people gain weight with supplemental feeding and the tube can be removed and some people benefit from long term supplemental feeding – every situation is taken case by case and decided between the patient and the CF team together.

5.

Dietary supplements

PROBIOTICS

There are both 'good' and 'harmful' bacteria naturally occurring in the body. The balance of these bacteria may be disturbed by medications such as antibiotics or from infection, stress, poor diet and/or illness.

Probiotics are 'good' bacteria that can assist in balancing bacteria in the gastrointestinal tract, and may also aid digestion, reduce intestinal inflammation and minimise symptoms of reflux.

Probiotics come in various forms including yoghurts, capsules, tablets, beverages and powders. It is highly recommended that you speak to your CF care team before starting any new supplements.

Probiotics have been found to be both safe and beneficial in people with CF, with results showing:

- reduced inflammation
- reduced exacerbations

- increased lung function
- increased gastrointestinal health

Probiotic supplements may be most beneficial during and immediately after antibiotic use.

At times when you're not on antibiotics, daily sources of 'good' bacteria through diet (e.g. yoghurt) should suffice, instead of paying for supplements which become expensive over time. It is advisable to stagger the administration of antibiotics and probiotics with the probiotics being taken at least 3 hours after the antibiotics.

FISH OIL

Fish oil has many great health benefits and may be beneficial to your general health, however there have been no significant biochemical or clinical improvements in CF symptoms.

SUPERFOODS

Superfoods are types of foods which are thought to have a very high








content of certain nutrients, such as antioxidants and nutrients our bodies can't produce. These superfoods are great for our bodies and should be eaten as part of a balanced diet:

- oily fish, such as sardines and salmon – rich in omega 3 fatty acids
- tomatoes – high in lycopene
- olive oil – rich in antioxidants and good fats
- brazil nuts – high in selenium
- berries – high in antioxidants
- broccoli – high in antioxidants and a good source of folate
- beans – high in fibre and antioxidants
- natural yoghurt – contains good bacteria to fight bad bacteria
- soy – rich in protein and high in fibre

Since the term has been introduced, many companies have abused the 'superfood' label in order to promote their products. New products such as spirulina, wheatgrass, maca and so on, may have certain health benefits, however they have no specific CF nutritional benefits, and can be an unnecessary and costly expense.



Bristol Stool Chart

TYPE 1		Separate hard lumps, like nuts (hard to pass)	Very constipated
TYPE 2		Sausage-shaped but lumpy	Slightly constipated
TYPE 3		Like a sausage but with cracks on the surface	Normal
TYPE 4		Like a sausage or snake, smooth and soft	Normal
TYPE 5		Soft blobs with clear-cut edges	Lacking fiber
TYPE 6		Fluffy pieces with ragged edges, a mushy stool	Inflammation
TYPE 7		Watery, no solid pieces. Entirely liquid	Inflammation

HEALTHY BOWEL HABITS

Everyone has different bowel habits. The range for normalcy can be anywhere from 3 bowel movements per day to 3 per week.

Many factors can affect regularity from day to day, such as diet, hydration, travel, medications, hormonal fluctuations, sleep patterns, exercise, illness, surgery and stress.

CF can cause you to have runny, pale, bad smelling stools, however good enzyme management should help to minimise these symptoms.

You should keep an eye out for any changes in your bowel habits, which might signal a problem.

Sometimes it can be difficult to talk about bowel habits, particularly when having to describe them.

A useful tool for this can be the Bristol stool chart, which provides a reference framework for talking about stools (see page 25).

CONSTIPATION AND DIARRHEA

People with CF will often experience constipation or diarrhea at one time or another. This is most likely due to a miscalculation of enzymes over a couple of days.

If you experience diarrhea regularly you can lose a lot of valuable nutrients and calories from your food.

Diarrhea may result from not having enough enzymes, viral or bacterial infections or antibiotic use. Diarrhea can lead to malabsorption, which is the inadequate uptake of nutrients from food. A combination of dietary changes and increased fluid can often help regulate bowel motions.

Constipation is the passing of hard, dry bowel movements. Stools may be difficult to pass and can cause pain, bloating, discomfort and bowel obstruction or blockage.

Constipation may result from having too many enzymes. Treatment of

constipation may include increasing dietary fibre, sodium and fluid intake as well as light exercise. Speak to your dietitian if this occurs as they can help make a treatment plan with you.

DISTAL INTESTINAL OBSTRUCTION SYNDROME

Distal Intestinal Obstruction Syndrome (DIOS) is a complication of CF. It occurs when faecal material and intestinal contents stick to the lining of the intestines and cause a blockage. Symptoms can include:

- cramps
- abdominal pain
- bloating
- hard stools
- reduced bowel movements
- loss of appetite
- vomiting

Symptoms can be similar to constipation, however DIOS is not constipation. In confirmed DIOS there is usually a complete blockage of the bowel.

Sometimes a hard mass can be felt on the right side of the abdomen. DIOS needs to be diagnosed with an abdominal x-ray.

DIOS often occurs with a change in diet, illness, exacerbation and/or dehydration in hot weather.

Having one episode of DIOS increases your risk for another and you may be prescribed laxatives or stool softening medications by your CF care team.

If you have previously experienced DIOS it can be really helpful to learn to recognise and treat symptoms early to prevent another episode. The following tips may be useful:

- make sure your enzymes are correctly matched to your fat intake
- ensure adequate fluid intake to prevent dehydration (8–10 glasses of water a day)
- ensure adequate salt intake, particularly in warm weather
- include adequate fibre in your diet

7.

Maintaining nutrition when unwell

When you are unwell your body needs a lot more energy than normal, even though you might not feel like eating.

You might not feel hungry, food might not taste the same or you could feel constantly full.

However, it is important to remember that food plays a very important role in your recovery, and in fact your body needs *more* food when you are unwell.

Think of food as a fuel for your body or as a medicine which is part of your treatment.

LOSS OF APPETITE

To increase your appetite, you could try the following:

- eat something every couple of hours during the day
- keep food on a small plate as a large full plate may put you off eating
- gentle physical activity can stimulate appetite- try taking a short walk around the block

- eat more meals with family and friends instead of alone

Snack ideas for a poor appetite include:

- milk and drinks containing milk
- yoghurt, custard and ice cream
- Fruche, Yogo and other dairy desserts
- fresh or dried fruit
- nuts, seeds
- cheese and biscuits
- peanut butter on bread
- cream soups
- hard boiled eggs
- crumpets, muffins, pikelets or scones with jam, honey, syrup, butter or cream
- dips made with cream cheese, beans or sour cream
- sandwiches
- cakes and biscuits

NAUSEA

To combat nausea, you can try the following:

- Avoid skipping meals - an empty stomach can make nausea worse. If the smell of food cooking causes nausea, try and stay away from the kitchen - have other family members/roommates cook for you if possible.
- Use mouth rinses and washes or try sucking on sugar free hard boiled lollies or peppermints to reduce bad mouth tastes.
- Try not to lie down or lean your chair back for at least one hour after eating.

Snack ideas for dealing with nausea include:

- Eat cold or room temperature foods as these do not taste or smell as strong e.g. sandwiches, salads, custard, mousse, yoghurts, tinned fruit or jelly.
- Snack on dry foods e.g. biscuits, noodles, cereal, toast or crackers.
- Eat salty foods e.g. clear soup and potato crisps may help.
- Drink cold clear fluids between meals e.g. cordial, lemonade, dry ginger ale or fruit juice.
- Try ginger containing foods e.g. ginger beer, ale, tea or candied ginger.

- Avoid fatty, rich, spicy or very sweet foods if you find they make your nausea worse.
- Try softer foods which require less chewing, so are easier to eat.

MAKING UP FOR LOST CALORIES

Following a period of nausea and/or loss of appetite you may need to make up for lost calories.

Here are some tips to do so:

- Treat yourself to your favourite foods and eat them at any time of the day e.g. breakfast foods at dinner.
- Choose drinks that are high in energy and protein e.g. milk or commercial supplements such as Sustagen or Ensure.
- There might be times of the day when you feel more like eating - make the most of these times by eating well.
- Try to cook with higher energy foods so that your meals are higher in calories.

ADMISSIONS

During admissions, it is important to eat well and maintain your weight (or gain). Here are some tips to manage your weight during an admission:

BREAKFAST IDEAS	› choose a high protein choice everyday e.g. eggs or add cream to porridge and coffee
MAIN COURSE IDEAS	› eat the meat/meat alternatives and/or dessert before your veggies (if your appetite is poor) › add butter or margarine to the veggies › add cream to your soup › ask for a double serve of the protein part of your meal › choose a sandwich as well as your hot meal or salad
DESSERT IDEAS	› order 2 or 3 dessert options › order a side of ice-cream, cream, or yoghurt
SNACKS	› ask visitors to bring favourite snacks › keep your snacks within easy reach › don't rely on your appetite - try to eat something every 2-3 hours
NUTRITIOUS DRINKS	› when your appetite is poor, it may be easier to drink than eat nutritious drinks such plain or flavoured full cream milk › milkshakes, nutrition supplements e.g. Sustagen
MEAL TIMES	› sitting out of bed (if possible) during mealtimes can make it feel more normal › moving around and getting out of bed can help your appetite › encourage visitors to see you at meal times to make meals more enjoyable



Exercise & diet

8.

Exercise is highly recommended for people with CF as it helps clear mucus and increases muscle strength.

Exercise does however also increase energy needs even further so it is important to replace the lost energy, water and salt through your diet.

It is essential that you discuss your training schedule with both your physiotherapist and your dietitian so that a suitable diet can be planned for you.

This will ensure that you maintain your weight and keep well hydrated. It will also help you perform and recover well after training sessions.

HYDRATION

People with CF are more susceptible to dehydration because of the increased amount of salt lost through sweat. Fluid and salt intakes need to be increased when exercising to replace these losses.

You should drink 2-3 litres of fluid a day to remain hydrated, and when you are playing sport or exercising you should drink:

- 500-600ml 2 hours prior to exercise

- 150-350ml right before exercise
- 150-200ml every 15-20 minutes during exercise

Sports drinks can be beneficial for people with CF during exercise, especially in warmer weather. Benefits of use include:

- increased thirst drive and voluntary fluid intake due to the salt content
- improved fluid retention post exercise
- a good source of calories
- improved gastric emptying (the time it takes for food to empty from the stomach and enter the small intestine)

If exercising for more than an hour, sports drinks and oral rehydration solutions may be recommended.

It is important to be aware that although they have benefits, sports drinks are also high in sugar, and many brands are not overly high in salt.

DIFFERENT BRANDS OF SPORTS DRINKS

Hydralyte Sports is considered one of the better brands for rehydration because of the high sodium (salt) content (115mg/100ml).

SPORTS DRINK	SODIUM MMOL/L	SODIUM MG/100ML
Gatorade	22	51
Gatorade G series prime	40	93
Gatorade Endurance	37	84
Powerade isotonic	12	28
Staminade	13	29
High 5 Isotonic	35	80
Musashi Electrolyte Replacement	19	43
Powerade Zero	22	51
Gastrolyte (pre-made)	43	101
Gastrolyte (sachet)	40	92
Gastrolyte (tablets)	20	46
Hydralyte (sachet)	44	103
Hydralyte (tablets)	15	34
Hydralyte Sports	50	115

WHAT SHOULD I EAT BEFORE AND AFTER EXERCISE?

You should always make sure you are well fed before and after exercise. A store of carbohydrates in the body is important prior to beginning exercise (glycogen stores).

Inadequate carbohydrate intake leads to the body breaking down fat and muscle stores for energy, which has detrimental effects on exercise and performance.

You should eat a well-balanced meal 2 to 3 hours before exercising. This will allow enough time for digestion and ensure that your muscle glycogen stores are topped up.

Follow this by a light snack one hour before exercising. The snack should be a familiar food that is well tolerated and won't upset your stomach.

Snack ideas include:

Snack idea	Calories	Fat (g)	Carbs (g)	Protein (g)
mixed nuts (30)	285	26	9	6
Up and Go drink	198	3.8	30.2	8.5
muesli bar - Carman's Original	201	8.6	25.5	4
slice multigrain bread with banana	198	1.2	37	5.2
canned tuna (95g) Greenseas	94	1.8	6.7	12.2
Sustagen Sport chocolate flavour (60g)	225	0.7	39.2	14.7

Post exercise you should aim to replace all losses. The food should ideally contain some carbohydrates and protein. *Try some options:*

Snack idea	Calories	Fat (g)	Carbs (g)	Protien (g)
crumpet x2 with 1 tbsp peanut butter	284	10.8	18.4	10.4
sandwich (wholegrain bread) with 2 tsp jam	260	1.3	34.8	6.8
mu n (apple cinnamon from McCafe)	89	4.3	11	4
banana	76	0.1	16.8	
boiled egg x2	122	8.4	0.6	11
600ml cow's milk	426	21	37.8	21
fruit and nut mix (60g)	300	18	24	6
baked beans (220g)	202	1.1	31.9	11.3
200g Greek yoghurt	268	19.4	14.4	9.4

Aim to eat a full meal within a few hours of finishing exercise. People with CF will generally have a greater energy expenditure after exercising, rather than actually during, therefore energy intake post exercise is particularly important.

SHOULD I TAKE DIETARY SUPPLEMENTS?

SPORTS SUPPLEMENTS

These are not normally required if you are eating a balanced diet. Special sports foods, such as energy bars and carbohydrate gels are available and can be of benefit when participating in endurance events.

PROTEIN SUPPLEMENTS

If you are eating enough food to meet your energy needs for exercise, it is likely you will be eating enough protein too.

Drinking plenty of milk/milkshakes with added milk powder will have a similar effect to drinking protein supplements and will cost you less.

Always talk with your dietitian before taking any protein or sports supplements. They may contain substances that interact with medications.

9.

Managing my diet out of home

It is important to make sure that you maintain your diet even when you aren't at home. This can be tricky, so it is important to be prepared and plan ahead.

If you are pancreatic insufficient it is important to take enzymes with you everywhere as you never know when you might get hungry or be offered food.

Keep spares in other places such as at work, in your car and at family and friends houses you visit frequently (while ensuring they are kept in suitable conditions). This will help to ensure you aren't caught without them.

TRAVEL

Maintaining adequate nutrition during travel is important and can take some consideration and prior planning. Below are some things to consider before you travel:

GET ADVICE FROM YOUR CF CARE TEAM	They will give you advice on what you need to discuss with your travel agent. They may also be able to give information on CF centres near your travel destinations.
HYDRATION	<p>Hydration is very important, especially if you are taking a long flight, visiting a hot and humid place, taking part in outdoor activities, are drinking alcohol, or if you have travellers' diarrhoea.</p> <p>During your trip ensure you:</p> <ul style="list-style-type: none"> • Drink a minimum 2 litres of fluids a day to prevent constipation and dehydration. • Drink every hour (carry a water bottle or camel pack). • Only drink safe water (bottled if needed).

SALT	Adequate salt intake is very important to avoid dehydration. Increase your salt intake by putting extra salt on food, taking salt tablets and sports drinks. Talk to your CF team about how much you will need.
FOOD HYGIENE	<p>Risk of dehydration and salt loss is very high with travellers' diarrhoea. If you get diarrhoea, maintain fluids and seek medical help.</p> <p>To avoid diarrhea:</p> <ul style="list-style-type: none"> • Consume only safe food and water (pay attention to advice given in travellers' guides). • Avoid dirty premises and street vendors. • Avoid raw or poorly cooked foods and peel all fruit. • Hot foods should be served steaming hot and cold foods served cold. • Drink bottled or treated water even for brushing teeth.
KEEP UP THE CALORIES	<ul style="list-style-type: none"> • Order special meals for your flights, beforehand. • Follow recommendations from guidebooks on good value cafes and restaurants. • Make use of the well-known fast food outlets. • Buy extra foods and drinks at supermarkets, and carry snacks with you when out and about.
SUPPLEMENTS	<p>To take or not to take? This depends on excess luggage and customs.</p> <ul style="list-style-type: none"> • Alternatively you can use powdered versions such as sachets of Scandishake. • You can make your own milkshake or smoothie drinks using full cream UHT milk/ milk powder, ice cream, yoghurt, and toppings from local stores. Take a "shaker" to make your own high calorie drinks.
ENZYMES	Carry enzymes in your hand luggage for use on the plane and in case your bags get lost. Take enough enzymes (and even extra in case of delays in returning home) for the whole trip or check to make sure you can buy the correct supply at your travel destination.



DINING OUT

Eating out can be difficult because it can be hard to work out how many enzymes you should take when you haven't prepared the food yourself.

Fast food restaurants often have nutritional information on the packaging of the food or online, or if you are at a restaurant don't be afraid to ask the waiter about the ingredients or how the food is cooked.

Further information like this will help you determine the fat content.

Also, when ordering from a menu, descriptive words can tell you a lot about how the food is cooked.

Words such as sautéed, fried, crispy, butter sauce, pan fried, cream sauce, in its own gravy or hollandaise are likely to be higher in fat, than words such as grilled or steamed.

BEING ORGANISED

If you know you are going to be out most of the day, plan ahead the night before and think about what you are going to eat, how you will store your food (does it need refrigeration?) and remember to pack your enzymes.

If you know you won't be home until late, such as if you're going straight to the gym from work, you should also make sure you pack plenty of snacks.

Here are some other ideas to make packing meals easier:

- When cooking, make enough to pack a meal for tomorrow's lunch-use plastic containers to freeze meals that you can easily 'grab 'n go'
- Work out the fat content of recipes, such as heavy soups, chilli or curries before you make them and then divide into portions to freeze.
- Write how many grams of fat/how many enzymes are needed on the container/freezer bag. This way on a busy day you will have a meal ready to go, with the enzymes already worked out for yourself.
- Cook extra rice or pasta at dinner the night before as these can be used to make quick and easy cold salads - simply add some meat, chickpeas or beans, vegetables and salad dressing, or add pesto.
- Buy individually wrapped snacks so you can easily carry them in your bag.
- Convenience foods such as freezer meals or canned soups are easy and

inexpensive, although you want to make sure they still have good nutritional content.

- Pack some leftovers in a thermos for a tasty warm meal that you can eat on the go.
- Make some pizzas or savoury muffins that you can freeze so you always have a meal or snack ready to grab as you run out the door.
- Quick and simple snacks such as avocado dip with crackers or vegetables, are nutritious and easy.
- Cooking daily meals may be a challenge for many people. With the demands of work, family activities and daily treatment, it can seem like there is no time for planning meals, shopping, and cooking.

Here are some tips to make meal preparation easier:

- Have a list of quick and easy 'go to' meals that you know won't take you long to prepare and cook e.g. meat and vegetables, pasta dishes, omelette etc. (meals should be 'boosted' as required).
- Plan your meals for the week using a meal planner and shopping list to ensure your shopping trip is quick and easy.

- Try 'batch cooking' in a slow cooker – meals like curries, stews and heavy soups can be made easily and portioned into single serve meals for times when you are too busy or unwell to cook.
- Purchase some instant foods or frozen meals that can be quickly prepared-not all frozen meals are healthy, so look for those that have ingredients from at least two food groups.
- Create a shelf in your kitchen or refrigerator just for your 'grab 'n go' favourites.

10.

Pregnancy & nutrition

Most women with CF can have healthy pregnancies. If you are planning to become pregnant or if you become pregnant it is essential that you discuss this with your CF care team right away.

They will be able to advise you about screening tests that you may want to have carried out, any changes to your medication that may be required, the possible effects on your health and to answer any questions you might have.

WEIGHT

Pre-pregnancy weight and weight gain during pregnancy are important determinants of the baby's birth weight. A healthy weight prior to conception is ideal; also conception can be more difficult when you are underweight.

Additional calories will be required to support pregnancy for someone with CF. The increase in energy requirements required to grow a baby can make gaining weight a challenge, however those who gain adequate

weight during pregnancy tend to have better outcomes and bigger, healthier babies.

If a healthy weight cannot be achieved through a high energy diet, then supplements or more invasive nutritional support may be considered.

Daily calorie needs will vary from person to person so you should be advised by your dietitian. They will be able to help you reach your target weight pre and post pregnancy.

FOOD

It is recommended you eat a range of foods from the 5 food groups in three meals and many snacks daily to meet the nutritional needs for yourself and your baby.

Speak to your CF care team about your energy, vitamin and mineral requirements throughout your pregnancy. Fat-soluble vitamins, calcium intake and folate should all be considered..

NAUSEA

Nausea, vomiting and 'morning sickness' are commonly experienced in the first 12 weeks of pregnancy due to hormonal changes.

These problems often (but not always) go away after the first trimester, however some women really struggle and need to work closely with their health care team.

Tips for dealing with nausea are detailed on page 27. You should seek advice from your CF care team if you can't eat enough because you feel sick and/or are throwing up.

CONSTIPATION AND GASTRO-OESOPHAGEAL REFLUX DISEASE

Constipation is common during pregnancy. See page 24 for more information on constipation.

Gastro-oesophageal reflux disease (GORD) may be worse in the last 12 weeks of pregnancy due to slowing movement of the gut and pressure on the stomach from the large uterus.

Discuss your symptoms with your CF care team as you may benefit from medication. For more information on GORD, see page 48.

CYSTIC FIBROSIS RELATED DIABETES

If you have Cystic Fibrosis Related Diabetes (CFRD) and are considering pregnancy you will need to do an Oral Glucose Tolerance Test (OGTT).

Untreated diabetes and high Blood Glucose Levels (BGLs) may lead to a loss in energy, weight, and increase your risk for illnesses. It can also have harmful effects on your baby.

If you have CFRD, monitoring your BGLs is necessary so that you can achieve optimal control prior to pregnancy. The OGTT will need to be repeated once pregnancy is confirmed and then again in the 2nd and 3rd trimesters.

Pregnant women with CFRD should consult with their CF team regarding best management of BGLs. If you have poor weight gain during pregnancy, you may be advised to use nutritional supplements, or supplemental feeding at an earlier stage than for non-diabetics, which in turn will require modification of the insulin regimen.

There should be close liaison between the CF dietitian and the diabetes team, preferably with access to a physician or nurse with particular expertise in diabetic pregnancy.

BREASTFEEDING

Women with CF are encouraged to breastfeed and many have done so successfully. Information regarding the transfer of medications from mother to infant via breastmilk is often lacking and patients should review their entire medication list with the CF team when considering breastfeeding.

Breastfeeding also increases the caloric needs of women with CF so they will need to further increase their calorie intake

11.

Post transplant health & nutrition

Nutritional requirements before and after a transplant can be quite different. Pre-transplant, you may have required extra food to provide your body with enough energy.

Now, after your surgery, your body is not using all that extra energy to breathe so your daily calorie requirements may change. Also, the side effects of steroids are excellent for appetite and weight gain, so if you continue to eat the way you did before your transplant you may gain excess weight.

Gaining weight after your surgery, so that you are in a healthy weight range, is very important to help you to heal and stay healthy, however gaining too much weight can put you at risk of other health complications.

To stay healthy, focus on a balanced diet which includes fruits, vegetables, whole grains, milk and milk products and meat and meat alternatives. Talk with your dietitian if you have questions about healthy eating, your

healthy weight range and/or your daily nutritional needs.

FOOD SAFETY

Due to your immunosuppressive therapy you are at an increased risk of food poisoning. Symptoms of food poisoning may include fever, headache, tiredness and aches and pains.

Less common symptoms are diarrhoea, nausea and abdominal cramps. Symptoms may progress to more serious forms of the illness, such as meningitis and septicaemia.

Symptoms such as diarrhoea and vomiting can cause poor absorption of the anti-rejection medications and increase your risk of rejection.

Here are some tips to ensure your food is safe to eat:

- check packaging is intact and observe 'use-by' dates
- once food is opened use within the recommended time frame, usually 2 to 3 days

- refrigerate or freeze foods immediately after a meal and eat leftover foods within 24 hours
- keep your fridge at 5.0 C, freezer not above -18.0 C
- store raw and cooked foods separately
- wash knives, cutting boards and food preparation surfaces with hot water and soap after contact with raw meat
- use separate chopping boards and knives for raw meats, vegetables, fruits etc.
- wash hands well before preparing food, after touching raw meat, after touching animals, and after going to the toilet
- cook all food thoroughly
- only reheat foods once and ensure it is piping hot all the way through
- keep your kitchen clean
- wash dishcloths daily
- wash all fruit and vegetables before eating


High risk foods include:

- unpasteurized milk
- cheeses (Brie, Camembert, Stilton), 'blue'/mouldy cheese
- raw or lightly cooked eggs and foods containing them (Caesar dressing, cookie dough, other batters)
- raw fish/shellfish/cold smoked fish
- rare meats
- cooked chicken
- deli meat
- soft ice cream

CYSTIC FIBROSIS RELATED DIABETES

The chance of developing Cystic Fibrosis Related Diabetes (CFRD) after transplant is high. This is because people with CF have a higher chance of developing diabetes in general, and steroid use can cause diabetes.

If you already have diabetes, continue to monitor your Blood Glucose Levels

A background image showing a glass of orange juice on the left and a bowl of cereal with blueberries on the right. The cereal is topped with several fresh blueberries.

(BGLs) and adjust your insulin doses with the help of your medical team.

If you did not have diabetes before transplant, your BGLs will be checked as part of your transplant bloodwork.

It is important to watch for the following symptoms and let your transplant team know if you experience them as it may indicate you have developed CFRD:

- unexplained weight loss
- feeling more tired
- feeling very thirsty
- change in appetite
- urinating more often
- blurry vision
- skin infections

BONE HEALTH

After transplant, bone health continues to be very important in CF. Steroids can thin your bones further and increase your risk for fractures.

In order to maintain a healthy bone status it is important to:

- maintain a healthy weight
- take recommended amounts of calcium and vitamin D
- include weight-bearing exercise in your daily activity (as prescribed)

KIDNEY HEALTH

High BGLs and some of the transplant medications can cause kidney damage. *To help prevent this:*

- drink plenty of fluid to help your kidneys remove the remainders of drugs from your body (unless advised otherwise by your doctor)
- keep your BGLs under good control

BOWEL HEALTH

It is important to remember that your bowels still have CF, which means if you are pancreatic insufficient you will still need to take enzymes and watch for signs of a bowel obstruction.

The chance of a bowel obstruction

could be higher after transplant because of pain medications (which are constipating) and from not getting enough fibre and fluids.

With drinking adequate fluids, taking your enzymes regularly, and monitoring your bowel movements, you can minimise chances of a bowel obstruction.

12.

Cystic Fibrosis-Related Diabetes

CFRD is a type of diabetes unique to people with CF. It is different to other types of diabetes and the symptoms may vary from person to person. It is caused by damage to the pancreas over a period of time.

CFRD can also be triggered by an exacerbation and treatment of an infection and/or be a gradual onset.

It is not related to diet.

Individuals with CF should be tested annually for the development of CFRD and following diagnosis, should have a six monthly dietary review.

One of the pancreas' main jobs is to make and secrete a hormone called insulin. Insulin helps glucose to be used by cells in our bodies for energy.

If glucose cannot get into our cells they cannot work properly.

A reduced amount of insulin production results in high blood glucose levels (BGLs), known as hyperglycaemia - or 'hyper'.

In a person with CF, symptoms of a hyper may include:

- a lack of energy
- a decrease in lung function
- weight loss
- increased hunger
- excessive thirst
- increased urine output

Hypoglycaemia or a 'hypo' is when the opposite occurs and BGLs drop too low.

Signs of a hypo can include hunger and feeling shaky followed by sweating and possibly fainting.

Insulin and other diabetes medications are used to help the body use glucose effectively and keep BGLs at near-normal levels.

This will help to maintain your weight and ensure good health. Everyone is different and so individualised dietary advice from a specialist diabetes educator experienced in the management of CFRD is essential.

FOOD

People with type 1 or type 2 diabetes are often advised to eat a low-fat, low-salt, and sometimes low-calorie diet.

People with CF have different nutritional needs. In general, if you are pancreatic insufficient and have CFRD you still need to eat your normal CF diet that has been recommended by your dietitian to help you maintain a healthy body weight.

A healthy diet means eating a variety of foods from all food groups. The only change is that, now that you have CFRD, you need to learn how to measure or count the foods that affect your BGLs.

Foods that contain carbohydrates affect BGL the most because the body turns them into sugar. Foods that are made up mostly of protein and fat have much less of an effect on BGLs.

CARBOHYDRATES


Carbohydrates are the best energy source for your body. When digested in the gut they breakdown into glucose which is absorbed in your blood stream.

It is transported around the body to be used as energy in cells, in muscles, the brain and all other organs. Glucose is also stored in the liver for slow release energy.

Try to eat sufficient amounts of carbohydrates each day. A very low carbohydrate diet is not recommended for people with CFRD.

Your dietitian or CFRD educator will also be able to help you learn how to carbohydrate count and manage CFRD.

Carbohydrates produce glucose energy at different rates; some fast, some slow. The Glycaemic Index (GI) indicates how a carbohydrate food affects BGLs. The lower the GI, the slower the rise in BGLs will be when the food is consumed.



It is recommended to eat more low and intermediate GI foods and less of the high GI foods, but not to exclude them.

This helps to optimise blood glucose control and maintain caloric intake.

- Low GI foods have a GI less than 55 and are broken down slowly by the body
- Intermediate GI foods have a GI between 55 and 70.
- High GI foods have a GI greater than 70.

Low GI foods include:

- legumes (e.g. kidney beans, baked beans, chickpeas)
- sweet potato
- low-fat dairy products (e.g. milk, yoghurt)
- many whole fruits (e.g. apples, oranges)
- pasta (except for 'quick cook' varieties)
- wholegrain breads
- wholegrain cereals (e.g. oats)

FATS

People with type 1 or type 2 diabetes are often advised to eat a low-fat diet to help prevent other health issues such as obesity and heart disease.

This advice does not usually apply to people with CF.

Most people with CF will be advised to continue to eat a high fat diet, however with the advice to select more of the good fats, such as monounsaturated and polyunsaturated fats (see page 8 for more information on fats).

Fat does not have a big effect on BGLs. It can slow carbohydrates from being released by the stomach when you eat a lot of fat at a meal, though, so it has an indirect effect on BGLs.

The more fat in your food, the later your BGLs may peak. Protein does not have a big impact on BGLs but can slow down carbohydrate absorption if eaten together. Still, you need to eat enough protein for good health.

ALCOHOL

If you plan to drink alcohol, you should be aware of the effect it has on your BGLs. When you are drinking, your liver is busy breaking down the alcohol so it will release less glucose into the blood.

This puts you at risk for low BGLs. Signs of being drunk are a lot like signs of low BGLs. If alcohol has clouded your thinking, you may not treat your low BGLs the right way.

Those around you may not know that you have low BGLs and this puts you in grave danger.

If you are going to drink:

- talk to your doctor or diabetes educator about how alcohol may affect you and how to best manage your CFRD
- only drink alcohol when BGLs are well-controlled
- do not drink on an empty stomach and continue to eat carbohydrate snacks while drinking e.g. chips (nuts are protein and don't increase glucose levels)
- check your BGLs after drinking to learn your response to alcohol (you may also need to check your BGLs during the night especially if you drank too much or you have been physically active while drinking)
- have a bedtime snack after drinking to prevent a hypo
- never drink alone; be sure to tell your friends that you have diabetes when you drink alcohol
- carry identification that states you have diabetes (diabetes ID bracelet or necklace)

- be aware that alcohol may impair your awareness of hypos
- do not count the carbohydrates in alcohol towards your insulin dose

EXERCISE

Routine exercise can help control your BGLs by making your body respond better to insulin.

Be mindful that your body will use up more carbohydrate stores with exercise and you will need to monitor your BGLs during exercise.

People with CFRD can work out safely however should be aware of the following:

- check your BGLs before, during and after your workout so you can watch your blood sugar patterns
- carry a source of carbohydrates with you e.g. glucose tablets
- you may need an extra 15 to 30 grams or more of carbohydrates for each hour of intense or lengthy exercise
- the blood-sugar-lowering effect of a workout can last as long as 12 to 24 hours, so you may need to eat an extra bedtime snack with carbohydrates on the days you've exercised really hard

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Other cystic fibrosis-related conditions

CYSTIC FIBROSIS RELATED BONE DISEASE

People with CF may develop brittle bones, a condition known as osteoporosis, which can result in broken bones, bone pain and changes in the way bones are shaped.

You may not be aware that you have problems with your bones. In CF, malabsorption of vitamin D impairs bone formation. Inactivity, malnutrition and chronic lung disease can all lead to bone breakdown.

Following a lung transplant, bone density loss is increased due to immunosuppressive and steroid medications. Diabetes, alcohol and smoking may result in a more rapid onset of osteoporosis.

To reduce your risk of CF related bone disease:

- eat a healthy diet including vitamin D and calcium

- exercise regularly to strengthen bones
- get at least 1 hour of sunlight daily to help your body make vitamin D
- take vitamin D supplements

GASTROESOPHAGEAL REFLUX DISEASE

Gastroesophageal reflux disease (GORD), more commonly known as reflux, is the backward flow of the stomach contents into the oesophagus.

GORD results from improper functioning of the lower end of the oesophagus and can cause symptoms of heartburn, chest or abdomen discomfort and chronic cough.

Those with CF tend to experience GORD more than those without CF. Untreated, GORD can also cause pain or difficulty swallowing, damage to the oesophagus and oesophageal narrowing.

You may find certain foods trigger your symptoms. These foods may irritate the oesophagus.

Discovering which foods make your symptoms worse can help you deal with your reflux.

Common triggers of reflux include spicy foods, coffee, chocolate, alcohol, soft drinks, citrus fruits, tomatoes and high fat foods.

Triggers are different for different people. In order to figure out your triggers, it helps to keep a food diary. This diary should track:

- what food you eat
- what time of day you eat
- any symptoms that you experience

Keep the diary for at least a week. You may want to go longer if you eat a varied diet. You can then use the diary to identify the specific foods and drinks that affect you.

Talking to your CF dietician about your reflux is important as everyone has different needs and dietary requirements. Some people will require medications to treat the reflux.

Avoiding alcohol, carbonated drinks and spicy foods before airway clearance may be useful in preventing episodes.



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Other resources

APPS	<p>Calorie King®</p> <p>A quick and easy way to check calories, carbohydrates and fats. This app contains over 22,000 Australia foods. Provides information including protein, fibre, fats, cholesterol and salt</p>
FACT SHEETS	<p>Bone Health Cystic Fibrosis Related Diabetes Dehydration Healthy Fats Nutrition for Adults PEG</p> <p>All factsheets available from: www.cfwa.org.au/what-we-offer/resources</p>

CFfood

cystic fibrosis nutrition program

CF Food:

CF Bites: High Energy Recipes for Cystic Fibrosis

A recipe book for people with CF but with great dishes and snacks which can be enjoyed by all the family.

Available in hard copy from CFWA or online at www.cfwa.org.au/what-we-offer/resources

CFfit

cystic fibrosis exercise program

CF Fit:

A series of booklets for personal trainers and people who have CF, which provide information about CF and exercise. Available in hard copy from CFWA or online at www.cfwa.org.au/what-we-offer/resources/

CFcooking

cystic fibrosis healthy cooking

CF Cooking:

Healthy cooking demonstrations for people with CF, including how to boost energy levels, how to make healthy CF meals when tired, how to fuel up for exercise and more. Available online at www.cfcooking.org



www.cfwa.org.au

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