

Cystic **Fibrosis** *our focus*

Nutrition: A guide for adults with cystic fibrosis

Introduction

Achieving and maintaining a good weight is essential for maintaining good lung function and helping withstand infection, and is associated with improved outcomes for people with cystic fibrosis (CF). This factsheet provides you with practical ideas to maximise your intake of food, gain and maintain a healthy weight. It is not intended to replace any advice you receive from your dietitian.

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Does diet matter?

Diet definitely does matter. Achieving and maintaining a good weight is essential for maintaining good lung function and helping you to withstand infection, and is associated with improved outcome.

Dietary requirements vary considerably and whilst some people may have normal energy requirements, most people with cystic fibrosis have higher energy needs than people without CF, because they do not absorb their food as easily. In addition, chest infections can increase energy needs. As a general principle, people with cystic fibrosis often require 20 to 50% more calories each day than people without CF, however some may need considerably more than this. For this reason we usually encourage you to have a high calorie diet.

This high calorie intake is often hard to achieve as many people with cystic fibrosis experience reduced appetite, especially during episodes of infection – the very time when your body's energy requirements are at their highest. Many people with cystic fibrosis need to take enzymes to digest and absorb fat-containing foods. Forgetting or taking too few enzymes will cause you to lose fat (energy) in your stools and this results in some of the fat you have eaten being wasted. This can also lead to weight loss or difficulty in gaining weight despite eating well. Even if you are taking your enzymes appropriately you will continue to lose some fat in your stools and this contributes to your increased energy requirements.

More recently we have begun to see some patients who have gained too much weight. These people may need to reduce their fat and energy intake and should be assessed and advised on an individual basis. The information within this factsheet may not be appropriate to these patients.

How often should I see the dietitian?

Your dietitian will give you practical help and advice on achieving and maintaining your ideal weight. This may involve using supplements and/or tube feeding. If you require pancreatic enzymes a dietitian will advise you regarding dose adjustment and timing with foods. It is therefore important to see your dietitian regularly, ideally at each clinic visit.

What is a high energy diet?

This is a diet that contains an increased amount of calories. Calories are a measure of how much energy a food contains. Dietary fat provides the most energy (calories) in the smallest volume and is the reason that we encourage you to use high fat foods and snacks whenever possible, together with the appropriate number of enzymes. This should be in combination with the right balance of foods from other food groups – starchy foods (bread, pasta, rice and cereals), proteins (meat, fish, cheese and meat substitutes), dairy foods (milk, cheese, and yoghurts) and fruit and vegetables.

Although a diet high in energy can be hard to achieve, there are simple guidelines that you can follow to help increase your calorie intake:

Fatty foods

- Fat is the richest source of energy (calories). One gram of fat contains nine calories, which is more than twice the amount found in protein or carbohydrate. Fat is also a good source of essential fatty acids and fat-soluble vitamins

- Where possible you should choose unsaturated fats and oils as they give the same amount of calories but in a healthier form. Monounsaturated fats include olive oil and rapeseed oil. Polyunsaturated fats include sunflower, soya, sesame and corn oil. Soft spreads made from olive oil, rapeseed oil etc can be used to replace butter and lard
- The essential omega 3 fats are also important and are found mainly in oily fish. You should try to include these in your diet (see below)
- Try to eat plenty of fat, making sure you take enough enzymes with your food, as fatty foods require more enzymes
- Fry your food in olive oil or rapeseed oil, add olive oil, butter or margarine to vegetables and potatoes and spread butter or margarine generously on bread
- Olive oil can be drizzled on food to increase the flavour and energy content
- Toss pasta in olive oil before serving or drizzle olive oil over pasta dishes
- Add mayonnaise, olive oil, dips or dressings to sandwiches and salads
- Choose higher fat foods such as pastries, crisps, nuts, seeds, chocolate, cakes, biscuits and chips
- Single, double or whipped cream can be added to puddings, sauces and soups
- Add extra olive oil/butter/margarine to vegetables, potatoes, pasta, bread and toast
- Try to have more fried foods
- When grilling or roasting foods, drizzle them with olive oil or vegetable oil and baste with oil regularly
- Add olive oil/mayonnaise/salad cream to sandwiches and salads
- Always add a dressing (oil or mayonnaise) to your salads
- Avoid using low fat spread or eating reduced fat foods

Starchy (carbohydrate) foods

- Starchy foods are another good energy provider and should be a part of every meal
- Starchy foods include breakfast cereals, pasta, potatoes, rice and bread
- Make them higher in calories by brushing with olive oil, garnishing with a drizzle of oil, frying them or adding lots of milk, butter, olive oil, mayonnaise etc
- Mash potatoes with butter, olive oil spread or margarine and mix in some cream, crème fraiche or cheese
- Toss pasta in olive oil before serving or drizzle olive oil over pasta dishes
- Add chopped nuts or dried fruit to cereal, yoghurt, milk pudding, fruit and ice cream

Sugary foods

- Sugary foods such as jam, honey, marmalade, syrup, fizzy drinks, tinned fruit in syrup, cakes, biscuits, sweets and chocolates are also energy rich
- Put plenty of sugar into hot drinks, on cereals and in desserts
- Spread jam, honey, marmalade and chocolate spread thickly on bread and toast
- Add sugar, honey, jam or syrup to cereal, yoghurt, milk pudding, fruit and ice cream

- Your teeth are very important to your health. Remember to clean your teeth after eating sugary food or taking sweetened drinks and to visit your dentist regularly
- ***Avoid foods which are labelled as being 'low sugar' or sweetened with artificial sweetener, such as diet yogurts, diet squashes and diet fizzy drinks***

Milk and dairy products

- Milk and dairy products are an important source of energy and calcium
- Use full cream or Channel Island milk and try to have between one and two pints daily in drinks, desserts such as custard, instant whips or milk pudding, savoury sauces such as cheese or parsley sauce and on cereals
- Add double cream to ordinary milk when making any milk-based savoury and sweet dishes or milky drinks
- Use milk instead of water when making up condensed or packet soup. Stir in double cream or olive oil just before serving
- Always add milk and olive oil/butter/margarine to mashed potato
- Cheese is an ideal snack; serve as cheese on toast, cheese and crackers etc. In addition cheese can be used to add extra calories to your food, for example it can be added to mashed potatoes, baked beans or pasta dishes
- Sprinkle grated cheese on top of vegetables, potatoes, soups and sauces
- Yogurts and ice cream make a quick dessert or snack. Try to choose whole milk, thick and creamy, Greek- or custard- style yoghurts and real dairy ice creams
- ***Avoid low fat dairy products such as low fat yoghurts, semi-skimmed and skimmed milk and low fat cheeses such as cottage cheese. If these are the only types of dairy products you like they are still a good source of calcium***

Protein foods

- Protein foods come in two varieties:
 - Animal proteins such as meat, fish, eggs, milk and dairy products
 - Vegetable proteins such as beans, peas, lentils and nuts
- Try to keep your protein intake high by having a good helping at each meal, including breakfast. It will also help if your snacks include some protein
- Fatty or oil-rich fish are good source of protein as well as a good source of the omega 3 fatty acids. Try eating oil-rich fish such as salmon, herring, sardines, mackerel, pilchards, trout, kippers and fresh tuna regularly
- If you are vegetarian make sure you replace meat or fish with a variety of protein sources such as soya mince or other meat substitutes, beans, seeds, lentils, tofu, nuts, cheese or eggs
- ***Avoid eating raw eggs and do not add them to high energy drinks. They do not add many calories and may be a Salmonella risk***

Fruit and vegetables

- Fruit and vegetables are generally low in energy, but do provide vitamins and some minerals, so they are an important part of a balanced diet

- Try to eat fruit and vegetables everyday – fresh, frozen or tinned – but if your appetite is poor do not fill yourself up with fruit and vegetables at the expense of higher calorie foods
- Some fruit and vegetables e.g. bananas, beans, pulses and root vegetables such as potato, turnip and parsnip are higher in calories
- Try to have a glass of fruit juice or squash with added vitamin C daily
- Use fruit in pies and crumbles or serve with double cream or dairy ice cream
- Add double cream, ice cream or custard to fruit
- Add sauces, olive oil or butter to vegetables or fry them

Calcium

- Calcium is essential to the body for maintaining strong and healthy bones. Therefore it is important that you have plenty of calcium rich foods each day
- The best sources of calcium are dairy products such as milky drinks, cheese, yogurts and dairy ice cream
- If you don't like these types of foods, non-dairy calcium sources include baked beans, tinned fish with bones (salmon, sardines and pilchards) and white bread
- Also look out for foods that may have calcium added to them such as soya milk, drinks and cereal bars
- Generally non-dairy foods contain less calcium and therefore you will need to eat more of them to ensure an adequate calcium intake. If you only like low fat varieties of milk and milk products continue to have these as they contain similar amounts of calcium as full cream versions

If you are concerned about your calcium intake talk to your CF dietitian

Alcohol

Drinking alcohol can be a very enjoyable and sociable part of life and a moderate amount will do you no harm. However, drinking too much regularly or drinking a large amount all at once can be harmful to your body. Alcohol is broken down and processed by the liver. Taking large amounts of alcohol, especially over a short period of time, will put a strain on the liver and cause damage if it occurs repeatedly. Check with your CF team that it is safe for you to drink alcohol.

It is recommended that men should aim for no more than three to four units of alcohol per day and women two to three units per day*. Exceeding these limits and binge drinking at the weekend should be avoided.

To assess your alcohol intake, use the following as a guide:

***1 unit = ½ pint beer or lager (3.5–4%ABV), a single measure of spirits (40% ABV), a small glass (100ml) of wine (10%ABV).**

Remember that home measures are more generous and many alcopops, wines and beers contain nearer to two units per glass or bottle due to their high alcohol content.

Suggested meal plan

You should aim to have at least three main meals and three snacks daily. Here are some ideas for these meals and snacks:

Breakfast

- Cereal with full cream milk and sugar and orange juice
- Toast/crumpets/croissants with butter/margarine and jam/marmalade/honey/cheese/peanut butter
- Beans/cheese/scrambled or fried eggs on toast
- Fried sausage, bacon and eggs with hash browns or fried bread

Lunch

- Ready meal/pizza/sausage roll/fish fingers/pasty and chips
- Baked beans/cheese/scrambled or fried eggs on toast
- Sandwiches with meat/tinned fish/cheese with bag of crisps
- Jacket potato with olive oil/ butter and baked beans/cheese or curry/chilli con carne
- Whole milk yogurt/chocolate bar/cake

Evening meal

- Fried, grilled or roast meat/fish/eggs/chicken
- Or vegetarian meat substitute or lentils with chips/potatoes/rice/pasta
- Fresh/frozen/tinned vegetables with olive oil, butter/margarine or a sauce or salad with mayonnaise/olive oil-based dressing
- Sponge pudding with custard or fresh or tinned fruit with double cream or ice cream
- Milk pudding with jam or syrup/thick and creamy yogurt/individual trifle or dessert/cheesecake/gateaux
- Cheese and biscuits

Morning/afternoon/bedtime snack

- Milkshake/fizzy drink/fruit juice/glass of whole milk/coffee, hot chocolate or malted drink made with milk
- Biscuits/cake/crisps/dried fruit and nuts/chocolate bar/cheese and crackers/ sandwiches/ individual fruit pies/crisps and dips/peanuts/toast/yogurt

Is this really a healthy diet?

For most people a high fat and high calorie diet would not be considered a healthy diet. However for many people with cystic fibrosis this is the most appropriate diet to help achieve and maintain a good healthy weight. This in turn helps to fight infections and stay healthy for longer. It is almost the opposite of the diet recommended for most other adults which is low in fat, sugar and salt with an increased intake of dietary fibre. This type of diet is not suitable for most people with cystic fibrosis because it is bulky and filling and is unlikely to provide enough energy.

Not everyone is the same and some people with cystic fibrosis may gain too much weight. These patients may need to reduce their fat and energy intake and should be assessed and advised on an individual basis by a specialist CF dietitian.

To give your high fat diet a more healthy emphasis where possible you should try to use monounsaturated cooking oils or spreads such as olive oil, rapeseed oil, olive oil spread instead of lard or butter and to eat fruit and vegetables daily. These can be incorporated as high fibre, high energy snacks including dried fruit, fruit pies and crumbles served with cream or ice cream. Additional fat e.g. olive oil, butter, cream, cheese can be added to dishes after the rest of the family has been served to ensure the whole family get the type of diet they need.

What about pancreatic enzymes?

Approximately 85% or more of adults with cystic fibrosis are pancreatic insufficient. This means the pancreas is unable to produce or release enough digestive enzymes into the small intestine and so food is not digested and can not be absorbed in the gut. To help to compensate for the lack of digestive enzymes most people with cystic fibrosis have to take replacement pancreatic enzymes to digest their food properly. The enzyme dose needed varies from one adult to another. Your CF team will advise on appropriate enzyme doses and it is important to discuss any changes to your enzyme intake with the team before changing your dose. There is a wide range of strengths of pancreatic enzymes available. The most commonly used preparations in the UK are Creon® and Nutrizym®. When taking pancreatic enzymes it is also important to drink plenty of fluids.

Making your enzymes work for you

To get the most benefit from your enzymes, follow these guidelines:

- Take enzymes with every meal, fat-containing snack or milky drink
- Spread your enzymes throughout the meal
- High fat meals will need more enzymes than low fat meals
- Most snacks will require fewer enzymes than a meal but be cautious as some snacks are very fatty and can require as many or even more enzymes than a meal
- Be flexible about the dosage and timing of enzymes
- Enzymes do not need to be taken with fruit, jelly, sorbet, boiled, chewy or jelly sweets, squash, fizzy drinks or fruit juices

Remember there is no standard dose of enzyme. You should take the amount required to control your bowels. Sometimes people with cystic fibrosis produce too much stomach acid and this can make pancreatic enzymes less effective. If you are experiencing problems with heartburn, reflux or abdominal cramps then please discuss with your CF team and they can prescribe appropriate medications to help.

Distal intestinal obstruction syndrome, fluid and fibre

Distal intestinal obstruction syndrome (a blockage in the bowel) – usually referred to as DIOS – may be caused by not taking enough enzymes and if you are dehydrated this can make things worse. Sometimes it can occur for no known reason. You should make sure you drink enough fluid and aim to have 30 ml per kg body weight of fluid each day. For example if you weigh 50 kg this is 1500 ml per day, the equivalent of about six to eight glasses/mugs of liquid each day.

- Some drinks such as sweet, fizzy caffeine-containing drinks or strong coffee may dehydrate you further. Water or diluted squash are best

- When the weather is hot or you are in a hot room you will need more fluid
- If you are doing a lot of exercise the fluid you lose by sweating will need to be replaced by drinking more
- Your body will lose fluid if your blood sugars are too high. Make sure that if you have diabetes it is well controlled (see section Cystic fibrosis-related diabetes on page 21)
- Taking more fibre in your diet may help prevent DIOS. Try including a high fibre cereal at breakfast, some wholemeal bread, and increasing the amount of fruit and vegetables you eat. If you increase your fibre intake you will need to increase the amount of fluid you drink.

Are vitamin supplements necessary?

Malabsorption of the fat-soluble vitamins A, D, E and K is likely in most people with cystic fibrosis, especially those who are pancreatic insufficient. Without supplements, blood levels of these vitamins may become low and occasionally deficiency symptoms can occur. Fat-soluble vitamins should be taken at a mealtime when enzymes will also be taken.

Recommended daily supplements (starting doses):
Doses from:

- Vitamin A – 4000-8000 iu (International Units)
- Vitamin D – 400-800 iu
- Vitamin E – 50-200 iu

How can I gain weight?

It can be difficult to maintain your weight at the right level, especially if you are getting a lot of chest infections. Trying to gain weight requires just as much effort and planning as a slimming diet. The following ideas might help:

- The best time to actively try to gain weight is when you are well. You will use a lot of extra energy during episodes of infection and at these times you may have to concentrate on simply maintaining your weight
- Eat regularly; an erratic meal pattern could mean that your average energy intake is actually quite low, even though you might eat well on some days. Organise a meal plan and make sure that you stick to it
- Eat six times a day (three meals and three snacks). Remember that meals do not have to be cooked to be high in energy. Snacks can often provide the same if not more energy than meals or cooked foods if you choose appropriately
- High energy drinks and supplements can help to increase your intake further. Take them last thing at night when they won't affect your appetite. If you take them during the daytime, have them at the end of a meal and not in place of food. Remember that over-reliance on these in place of food can mean that you tire of the flavours more quickly and have nothing to fall back on when your appetite is poor
- Increase the energy content of the food you eat, so that you have more energy in the same quantity/portion of food. Easy ways to do this are by adding extra olive oil, butter, margarine, cream, mayonnaise, dressings or cheese to dishes; frying your food, basting or drizzling food with olive oil
- Try to concentrate on buying high energy foods and avoid low fat, low sugar varieties. Check the label on products and be aware of what foods contain

- Remember that if you require enzymes these should be taken with all fat containing foods and drinks. You will waste the energy in meals and snacks if you have no means of digesting them properly
- If you are having difficulty gaining or maintaining your weight, even though you are trying to eat as much as you can, discuss this with your dietitian/doctor. They may have more ideas or alternatively there may be other reasons why you are not gaining weight which can be easily corrected

Appetite

A poor appetite and weight loss are often the first signs of a chest infection and are very common. Appetite can be slow to return and it is very hard to eat when you have little or no appetite.

It is important to try to keep to a high energy diet even if you do not feel like eating.

The suggestions below might help:

- It may be a good idea to avoid set mealtimes. However, schedule regular eating times for a snack or high energy drink every two hours. This helps to form a habit of eating; you will expect to eat at these times and your body will begin to prepare for food
- Liquid calories are often easier to take when you are unwell; you may find it easier to take high energy drinks/nutritional supplements
- Abandon main meals if you find them too filling – snacks can be just as high in energy, less filling and can be eaten any time
- Alcohol can act as an appetite stimulant – sometimes having a small drink before meals can stimulate your taste buds
- If you feel unwell in the mornings don't force yourself to eat, but do make sure you then eat throughout the day and during the evening
- Limit mealtimes to about 30 minutes and space mealtimes three to four hours apart. This will give you the chance to get hungry and look forward to the next time
- Make the most of any times that you might feel hungry
- A trip to the supermarket may spark your interest in new types of foods
- Looking at magazines with pictures of food or watching food programmes on TV may help stimulate your appetite
- Light exercise or walking before meals in fresh air, if possible, can increase the appetite
- Try not to eat alone; eat with other people whenever possible
- Use small plates and serve smaller portions, this may seem more manageable
- Drink lemonade or juice with the meal. Juices contain acids that can stimulate the appetite
- Try spicy foods. Spicy foods make our mouths water and change the taste of foods
- Eat food slowly and chew food well
- Eat food cold. A cold temperature downplays the smell and taste of food
- Sprinkle more sugar and salt on food to try to heighten its taste
- Drink beverages between meals instead of with meals. Liquids at mealtimes can make the stomach feel full. Drinking less while eating allows more room for food

Nutritional supplements

A wide range of special products are available if your weight, weight gain or appetite is poor. They may also be useful during episodes of infection. Nutritional supplements should always be used in addition to the meals and snacks that you eat rather than as a replacement. Your dietitian or doctor will advise on the most appropriate type, the quantity you should take and whether enzymes are required.

Nutritional supplements come in a variety of forms and most are available on prescription.

Nutritionally complete supplements e.g. Ensure Plus, Fortisip, Fresubin, Entera, Resource. These contain a balance of all nutrients including energy, protein, vitamins and minerals. They are available in a variety of flavours (including savoury) and can be taken as a sip feed or added to recipes.

Energy and protein rich supplements e.g. Scandishake, Calshake, Enshake, Enlive, Fortijuice.

Glucose polymers e.g. Maxijul, Polycose, Polycal. These are available in powder and liquid form. They can be added to drinks and soft foods to increase the energy content without increasing bulk.

Fat emulsions e.g. Calogen, Liquigen. These are available in a liquid form that can be added to milk based drinks or can be taken alone in small quantities as a medicine. They are a very rich source of calories but require careful supervision due to their high fat content.

Energy and protein supplements e.g. Pro-cal. This is available in powder and liquid form. It can be added to drinks and soft foods to increase the protein and energy content without increasing bulk. The liquid can be taken alone in small quantities as a medicine.

Other supplements are also available such as protein supplements and fat and carbohydrate mixtures. Discuss this with your dietitian and CF team.

Commercial milkshake supplements such as Build Up or Complan are also available in chemists and supermarkets. They are available in sweet and savoury flavours and can be made up with milk or water. However these products are not available on prescription.

Tube feeding

Despite concerted efforts eating and/or taking nutritional supplements, there are times when it becomes increasingly difficult to gain weight without some help. This can be particularly difficult if you suffer from repeated chest infections. Although you may feel you are eating well and your enzyme dose is correct, you may still fail to gain weight because your energy requirements are so high.

Tube feeding can supplement daytime food intake, make gaining weight easier, and relieve the pressure of constantly having to eat.

Tube feeding is usually administered overnight to allow maximum daytime food intake. It involves either a nasogastric (down the nose) tube, which can be passed nightly and removed in the morning, or a gastrostomy tube, which is a tube placed directly into the stomach. The advantages of a gastrostomy tube are that it is hidden and cannot usually be dislodged or coughed out. For longer term feeding the gastrostomy may be the more suitable option for many people with cystic fibrosis.

The dietitian and doctor will discuss with you the type of feed to use, the

timing and rate of the feeding, and the correct dose of enzymes with the feed.

The timing of the feed is very important to ensure you feel hungry the next day and can eat normally.

Many people with cystic fibrosis have commented that tube feeding helps relieve the pressure of having to keep eating to gain weight. Following the experience of having a tube in place and gaining weight, they look upon tube feeding as a positive intervention.

Tips on managing your diet

You may be starting work or going on to further education and living away from home for the first time. It can be a busy time with lots to do. Eating well and maintaining a healthy weight will boost your energy levels, helping you to cope with new challenges. The advice below may help you.

Shopping

- Shopping at a supermarket is often cheaper than local shops. If you are on a tight budget look out for supermarket value labels
- Bulk-buy a selection of snacks such as nuts, dried fruit, crisps, chocolate bars, peanuts, seeds etc at the supermarket to take to work or college and keep in your desk or locker. This will be less expensive than buying them on site and you will always have a snack to hand
- If you find shopping a physical challenge, look into home delivery services; many stores offer internet or phone shopping services

Preparing meals

- If you are planning to live away from home for the first time, try to prepare yourself. Get in some cooking practice before you leave. Collect some easy recipes of meals you like
- Convenience food and ready meals can be useful if you do not have very much time or don't enjoy cooking. They are nutritious and require little time or effort to prepare (see page 14 for ideas).
- Think about having your main meal at work or college, especially if there is a subsidised canteen

General

- Try not to skip breakfast – at least have some cereal or a Danish pastry with juice or milk, which is quick and nutritious
- Be prepared; if you cannot buy food at work, either take a packed lunch or stop at a bakery or sandwich shop on your way to work
- If you have to make long journeys or spend a long time in the car keep a supply of drinks and snacks with you
- If you find it hard to maintain your intake, you may find prescribable high energy drinks useful to take to work. Speak to your dietitian or doctor if you have not used these before
- Keep your cupboards, fridge and freezer well stocked with food in case you cannot get to the supermarket (use the list on the following page as a guide)

Foods to keep in stock

Cupboard

- Tinned meat – ham, corned beef, stew, curry
- Tinned fish – tuna, sardines, mackerel, pilchards
- Tinned vegetables – baked beans, tomatoes, sweetcorn
- Tinned soups – creamed or condensed varieties
- Jars of pasta, curry or stir-fry sauces
- Olive oil, rapeseed oil, vegetable oil, gravy and instant sauce mixes
- Salt, pepper, herbs and spices
- Sugar
- Pasta, rice, noodles
- Breakfast cereals, biscuits, crackers
- Chocolate bars, sweets
- Savoury snacks – crisps, nuts, seeds
- Dried fruit
- Dips
- Jam, honey, peanut butter, chocolate spread
- Tinned sponge and milk puddings, fruit in syrup
- Ready to serve custard, evaporated milk
- Instant whip, milkshake powder or syrup
- Long-life milk and juice

Fridge

- Margarine/butter
- Cheese, cheese spread
- Milk, cream, yogurt
- Mayonnaise, dressings, dips and bottled sauces, including ketchup

Freezer

- Ready meals – lasagne, curry, nut roasts
- Breaded fish or chicken portions, sausages, beef burgers, vegetable or bean burgers
- Frozen fresh meat
- Pizza, garlic bread, naan bread, pitta bread
- Oven chips, croquette potatoes, potato waffles, hash browns, potato rosti
- Frozen vegetables including peas
- Dairy ice cream

Quick and easy meals

If you don't feel like cooking, try one of these quick and easy meals. Most are made from convenience or frozen foods that are readily available and easy to store.

- Baked beans or tinned spaghetti on toast with grated cheese
- Omelette with filling e.g. ham, cheese, tomato, mushrooms. Serve with

bread/toast and margarine or butter

- Tinned mini sausages and baked beans or ravioli with toast and margarine or butter
- Pizza with extra grated cheese
- Ready prepared quiche, pork pie, pasty or sausage roll with salad and mayonnaise or olive oil dressing
- Cold meat/ready cooked chicken with oven or microwave chips
- Cooked pasta tossed in olive oil with ready prepared sauces (always try to buy cheese or cream based varieties)
- Puddings – microwavable sponge puddings, crumbles served with tinned or packet custard, tinned rice pudding, whole milk yogurts, or tinned fruit in syrup served with ice cream

Make sure that you always have some frozen ready-made meals that can be used as needed, such as:

- Shepherd's pie, hot-pot or casserole with tinned or frozen vegetables
- Microwave curry or chilli served with naan or pitta bread
- Lasagne, pasta bake, macaroni cheese
- Ready made Yorkshire pudding with minced beef, chicken casserole or sausage and gravy filling

Snacks

Use the following in plain or toasted sandwiches or on crackers/savoury biscuits. Try bagels, ciabatta bread, French sticks and wraps as a change.

- Grated cheese with pickle/chutney or onion/mayonnaise
- Ham, cheese and tomato
- Salmon, tuna or prawns with salad and mayonnaise
- Cream cheese with pineapple, raisins or chopped dates
- Cream cheese with chives, chopped onion and salad
- Avocado and bacon
- Egg mayonnaise
- Cheese and coleslaw
- Hummus and salad
- Chicken, bacon and mayonnaise
- Peanut butter and jam/mashed banana and honey

Jacket potatoes

- Tinned or ready prepared curry/chilli
- Baked beans (try curry/barbecue flavour or baked beans with chopped streaky bacon/sausage)
- Coronation chicken
- Cheese and coleslaw
- Tuna and mayonnaise
- Grated cheese and olive oil, margarine or butter
- Grated cheese and mayonnaise
- Creamed mushrooms

Other snacks

- Soups (try to have the creamed varieties)
- Packets of crisps, tortilla chips, peanuts, nuts and raisins
- Chocolate
- Teacake, malt loaf, scone, crumpets or croissants with butter/margarine and jam
- Doughnut, cake, biscuits
- Full fat yoghurt, creamy yoghurt or Greek yogurt (with fruit and honey)
- Cereal with sugar and full cream milk
- Individual trifle, cheesecake, ice cream

Exercise and diet: getting the right balance

Regular exercise is good for everyone. For people with cystic fibrosis, exercise also helps clear mucus and increases lung muscle strength. It can help build muscles and strong bones and generally increases the sense of wellbeing. Whether you take part in recreational sport or exercise as part of a healthy lifestyle, what you eat and drink is important.

We already know that most people with cystic fibrosis have higher energy needs than people who do not have cystic fibrosis. Regular exercise can increase your energy needs even further so it is important to increase the amount you eat to avoid losing weight.

If you have cystic fibrosis-related diabetes (CFRD) you should discuss your exercise programme with your dietitian and physiotherapist. They will help you to understand the effects of exercise on your diabetes and will be able to give you more specific advice.

Food for energy

Whenever you exercise you need energy or fuel. The harder you exercise the more energy you use. This energy comes from the carbohydrate and fat in our diet.

Carbohydrate is the most important fuel for an active individual. It is stored in the liver and muscle as glycogen. These stores are used during exercise when energy is needed quickly. However during exercise, especially high intensity exercise such as sprinting and team sports, these stores are used up rapidly. When you run out of glycogen you will start to feel tired and your performance will be affected.

Starchy foods are excellent sources of carbohydrate and should be included at all mealtimes. Eat one large helping at each mealtime e.g. breakfast cereals, pasta, bread, potatoes, rice.

Fruit and fruit juices will also provide carbohydrate e.g. bananas, apples, pears, grapes and dried fruit.

Sugary foods are an important part of the cystic fibrosis diet and can provide additional calories to meet the needs of exercise. These should be taken in addition to your usual high energy starchy foods e.g. soft drinks, sweets, jam, honey, cakes, cereal bars, muffins, pancakes, biscuits and puddings.

How much carbohydrate do I need?

This will depend on the intensity and duration of exercise. The longer you exercise the greater your energy requirements. If you are exercising more than one hour a day you may need to increase the amount of carbohydrate at mealtimes and as snacks.

Suitable high carbohydrate snacks

These are ideal to have before and after exercise and include: milkshakes, smoothies, yogurts, cereal bars, jam sandwiches, fruit buns, bagels, crumpets, fig rolls.

What about fat?

Fat is converted into energy much more slowly than carbohydrate. A low fat diet is not generally recommended for people with cystic fibrosis, as it is unlikely to provide enough energy. Exercise increases your energy requirements even further; therefore your aim is to achieve a balance between sugar and fatty foods for energy, whilst maintaining a healthy body weight.

If you are exercising regularly for an hour or more on most days, it is important to make up the energy deficit by eating plenty of both carbohydrate-rich foods and fatty foods. Some people with cystic fibrosis have successfully completed endurance events such as half-marathons or marathons. It is however essential that you discuss your training schedule with your dietitian so that a suitable diet can be planned for you to ensure that you maintain your weight, keep well hydrated, perform well and recover well after training sessions and enjoy the run!

What about protein?

Protein rich foods are not the main source of energy for exercise. In fact most people have enough protein in their normal diet to meet the demands of training and exercise provided they are eating a balanced diet with sufficient carbohydrate and energy to meet their needs.

Muscle is gained through a combination of strength/resistance exercise and diet, which contains adequate energy and carbohydrate to fuel the exercise. It is the exercise training, which brings about the adaptations in the muscle, not the amount of protein consumed.

However it is still important to eat good sources of protein daily such as meat, poultry, fish, cheese, eggs, pulses, nuts, Quorn, tofu, soya mince.

Are protein supplements necessary?

If you are eating enough food to meet your energy requirements and the extra energy costs of exercise you will be eating enough protein too. Protein supplements are unlikely to be necessary. A pint of milk will provide a similar amount of protein as a serving of protein supplement at a much lower price. Drinking plenty of milk and milkshakes fortified with milk powder will have the same effect.

You may also benefit from taking a prescribable supplement drink, which will provide you with additional protein and energy to meet the demands of exercise. Ask your dietitian/doctor for more details.

How soon before exercise can I eat?

Ideally you should eat a well balanced meal two to three hours before exercising. This will allow enough time for digestion and ensures that your muscle glycogen stores are topped up. It is also a good idea to have a light carbohydrate snack one hour before exercising.

Refuelling after exercise

Start refuelling your energy stores with high carbohydrate snacks as soon as you have finished exercising. Follow with a full meal within two hours after exercising.

Fluids and hydration in sport

Most exercise results in the body becoming warmer. As the body can only operate efficiently at a narrow temperature range it takes steps to cool you down by sweating. The sweat then evaporates and cools the body. You cannot stop your body losing fluid by sweating but you can prevent it from becoming dehydrated.

People with cystic fibrosis tend to lose more salt in their sweat than those who do not have cystic fibrosis. It is important to ensure an adequate fluid intake to prevent dehydration. It may also be necessary for you to take additional salt supplements if you sweat a lot. This is especially important in warm weather and for prolonged periods of exercise. Thirst is a very poor indicator of hydration, especially in CF: by the time you are thirsty you are already dehydrated.

Signs of dehydration

Signs of dehydration include fatigue, dry mouth, thirst, headache, feeling irritable, flushed skin, dark, scant urine, weakness, dizziness, cramp and profuse sweating. Your sputum may become thicker, making airways clearance more difficult. You may feel too tired to eat, which means that you do not adequately refuel your body's energy stores and this could lead to unintentional weight loss.

Later symptoms include impaired mental performance, stumbling, dizziness, shrivelled skin, sunken eyes, muscular spasm and nausea.

Consequences of dehydration

Even mild dehydration can result in reduced sporting performance. Severe dehydration may lead to heat stroke and sweating will stop. By this stage, you will need medical treatment.

How much fluid do I need?

In general, most people need about two to three litres of fluid per day to remain fully hydrated. You can lose 0.5-1.5 litres per hour of sweat during exercise. If you are exercising during hot and humid conditions the amount of sweat you lose may be even greater than this.

It is important to start exercise fully hydrated. You should aim to drink 500-600ml of fluid, such as water or a sports drink, about two hours before exercising, followed by an additional 150-350ml, depending on tolerance, before exercise commences.

For low intensity exercise such as walking or gently cycling or for exercise lasting no more than 30 minutes, the risk of becoming dehydrated is probably very small, unless it is very warm. However drinking a sports

drink or sugary drink will provide you with energy to help maintain a healthy weight and provide fluid to replace any sweat loss.

During exercise, aim to drink smaller amounts more frequently, ideally 150ml-200ml every 15-20 minutes.

What should I drink?

During the day, you should aim to enjoy a range of drinks including water, fruit juices, squash and soft drinks. Tea and coffee contain caffeine, which can act as a diuretic and stimulate urine production. It is therefore best to include a variety of drinks each day.

During low intensity exercise of less than 30 minutes, water is useful to replace your fluid losses. However, it will not provide any energy.

Hypotonic drinks

These are absorbed into the body more quickly than plain water. They usually contain low levels of carbohydrate (less than 3g/100ml) and may have a little salt added. They quench thirst and provide fluids but do not provide a significant amount of energy. They may be used during low intensity exercise, lasting less than an hour.

Isotonic drinks

These are absorbed as fast as or faster than plain water. They contain a little salt and 5-8g carbohydrate/100ml, which will provide energy to fuel the muscles. Most commercial sports drinks fall into this category e.g. Powerade, Lucozade Sport, Isostar.

In cystic fibrosis there is evidence that individuals who exercise lose more salt and water as sweat than people who don't have cystic fibrosis. Drinking a flavoured sports drink has been shown to encourage drinking and reduce the effects of dehydration. Therefore it may be beneficial to consume sports drinks even if you are exercising for less than an hour, particularly in warm conditions. It may also be necessary to take additional salt supplements.

Hypertonic drinks

These drinks contain more than 10g carbohydrate/100ml. Hypertonic drinks such as pure fruit juice, many canned drinks and energy drinks, are absorbed more slowly than plain water. They replace lost energy rather than fluids and are therefore not an effective way to re-hydrate. They can provide extra energy in the CF diet and may be used to top up your daily carbohydrate intake.

Stimulant drinks

These usually have a high carbohydrate content plus other additives which are claimed to give you more energy, such as herbs, caffeine, taurine, guarana, ginseng, carnitine etc. There is no evidence that these will improve sporting performance and they should therefore be avoided during exercise.

Key points

- Fluid intake is essential during exercise
- Start exercise fully hydrated
- Drink before you feel thirsty

- Water will replace fluid loss during low intensity exercise lasting less than 30 minutes
- Isotonic sports drinks are useful to replace salt, fluid and energy lost during exercise
- Salt supplements may be needed especially during prolonged exercise or in warm weather
- Avoid carbonated drinks which can cause gastric disturbances
- Start rehydration immediately after exercise

Vitamins and minerals

Most people with cystic fibrosis are prescribed additional fat-soluble vitamins. It is unlikely that you will need to take any other additional vitamins if you are exercising regularly. In fact, an excessive intake may be harmful. There are however, some situations in which your doctor or dietitian may recommend additional specific vitamins or minerals for certain individuals e.g. if iron stores are low. It may also be necessary for you to take additional salt supplements if you sweat a lot, especially during periods of warm weather. These should only be taken as directed or prescribed.

Calcium

Calcium is essential for bone health. Adolescents and adults with cystic fibrosis may be at risk of low bone density – see page 21 for more details.

Although physical activity generally improves bone mass, an inadequate calcium intake can lead to an increased risk of stress fractures. It is therefore important to ensure an adequate intake of calcium. The best sources of calcium in the diet are milk and milk products, tinned fish with bones, white bread and fortified breakfast cereals.

Iron

Iron is an important trace mineral found in haemoglobin in red blood cells, and in myoglobin in the muscle cells. Iron deficiency can impair oxygen transport in the body and can lead to such symptoms as extreme fatigue and increased risk of infections. It is therefore important to include foods which are rich in iron. Iron can come from animal (haem iron) and vegetable (non-haem iron) sources. Good sources include meat and meat products, cereals, green vegetables, peas, beans, lentils and dried fruit.

Special sports foods and supplements

Special sports foods, such as energy bars, sports drinks and carbohydrate gels can be of benefit when participating in an endurance event such as distance running.

Creatine

This is a naturally occurring compound found in meat and fish. Most of the body's stores of creatine are found in muscle. Research in athletes has shown creatine to have short term benefits in repeated bouts of maximal exercise such as sprinting. Current research in cystic fibrosis is looking at the potential benefits of creatine supplementation to improve muscle strength.

Many sports supplements claim to improve muscle strength and performance but lack any scientific evidence to back up these claims. As a result of poor manufacturing claims, some products may contain

impurities, which could potentially interfere with other medications and be harmful.

Most people do not need to take sports supplements if they are eating a balanced diet. Always consult with the dietitian or doctor before taking any sports supplements.

Why else is diet important?

Low bone density

As more people with cystic fibrosis move through adulthood, new complications are emerging. Cystic fibrosis-related low bone mineral density (which means thin bones/low bone density and is sometimes termed osteopenia and osteoporosis) is one such area. This means that bones slowly become more fragile and in time more likely to break. The causes of reduced bone mineral density in CF which can lead to osteopenia and osteoporosis (thin bones) are very complex. There are several causes of low bone density in people with cystic fibrosis. Dietary risk factors include poor body weight and poor stores of vitamin D and calcium.

Achieving and maintaining good bone health is important. Bone density increases yearly until around the age of 20 and then gradually decreases over the following decades. For some people with cystic fibrosis the process is speeded up. Although specific treatments for osteoporosis are available, good nutrition, a good body weight, a high calcium intake and weight-bearing exercise are essential for good bone health.

Vitamin D is also essential for bone health. It comes mainly from sunlight (just a few minutes each day is sufficient), but it is also important to take any additional vitamin D supplements that are prescribed for you.

Cystic fibrosis-related diabetes (CFRD)

Diabetes is common in adults and adolescents with cystic fibrosis. It occurs in approximately 30% of people with CF by the age of 25 years. CFRD is less common in children.

What is diabetes?

Diabetes is a condition where the amount of glucose (sugar) in the blood is too high. Normally a hormone called insulin controls the amount of glucose (sugar) in our blood. Insulin is needed to move glucose from the blood stream into the cells. In diabetes the pancreas does not produce enough insulin and the insulin produced does not work properly. When people talk about blood glucose or blood sugars they mean the same thing and often use the terms interchangeably.

Why is cystic fibrosis associated with diabetes?

It is well known that in most people with cystic fibrosis the pancreas does not work properly. This results in two important medical problems:

- Firstly, the pancreas is unable to produce enough digestive enzymes or juices, which break down foods such as carbohydrate, proteins and fats so that they can be absorbed in the gut. This is why most people with cystic fibrosis have to take additional enzymes with food and snacks containing fat.

- Secondly, the pancreas can stop producing enough insulin and this can result in CF-related diabetes.

Cystic fibrosis-related diabetes is different from the two main types of diabetes (Type 1 and Type 2 diabetes) and has features of both.

In addition, people with cystic fibrosis may develop high blood sugar levels during periods of lung infection or while taking oral steroids. This may be temporary and may resolve when the acute infection is treated or when the steroids are reduced or stopped.

Some people may be concerned that the sugary diet that they have eaten may contribute to them developing CF-related diabetes. There are many other risk factors that cause or contribute to people with cystic fibrosis developing CF-related diabetes. Any concerns should be discussed with your doctor or dietitian.

Treatment

While some people can control their blood sugar levels by taking tablets, in CFRD most people are best treated with injections of insulin. Insulin cannot be taken by mouth because it is destroyed in the stomach. It is usually given as an injection two to four times a day.

Maintaining a healthy body weight is one of the most important steps you can take to ensure good health. People with CFRD still need to eat their usual high calorie, high protein and high fat diet to help achieve and maintain a healthy body weight. This is the opposite of the usual advice for diabetics and it can become confusing.

In cystic fibrosis, more energy (calories) is needed in the diet and the dose of insulin can usually be tailored to individual requirements and people can be taught to adjust their insulin dose to their dietary intake. Keeping your blood sugars at a near normal level will also help to maintain/improve your weight.

The Cystic Fibrosis Trust produce a factsheet called Cystic fibrosis-related diabetes, which contains a lot of detailed information (see 'Further information' for details of how to access CF Trust literature).

Pregnancy

If you are planning to become pregnant it is essential that you discuss your plans with your CF consultant. 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 may have.

Anyone planning a pregnancy should spend time preparing. This is especially important for women with cystic fibrosis. People who are underweight may have an irregular menstrual cycle. Achieving a normal, healthy body weight helps to correct this and may increase the likelihood of conceiving a baby. In the preconception or planning phase it is important to make sure you are in the best possible health and that you optimise your weight and nutritional status. An oral glucose tolerance test should also be carried out at this time to screen for diabetes. It is also usual to repeat the oral glucose tolerance test during the pregnancy to screen for gestational diabetes.

The healthy high energy diet you have been recommended should continue. In addition, all women who are planning a pregnancy should take a daily supplement of folic acid (a B vitamin) from the time they stop

contraception and for the first twelve weeks of pregnancy. Additional folic acid is recommended as it has been found that increasing folic acid intakes before and during the first three months of pregnancy may prevent neural tube defects such as spina bifida. The recommended dose is 400micrograms (0.4mg) and this is available either on prescription or you can buy it from your local chemist or many supermarkets.

There is often confusion surrounding other vitamin supplements in women with cystic fibrosis who become pregnant. Most people take a combined supplement containing both vitamin A and D. Vitamin D is essential in pregnancy when requirements for this vitamin increase. In contrast, very high intakes of vitamin A in women who do not have cystic fibrosis have been reported to increase the risk of birth defects. However, for patients with cystic fibrosis supplements of fat soluble vitamins (vitamins A, D, E and possibly K) are given to correct deficiencies that occur because of malabsorption. Deficiencies of vitamins in pregnancy can also be detrimental. It may be appropriate to reduce the dose of vitamin A to a "safe limit" but most women with cystic fibrosis should continue to take their routine vitamin supplements. You should discuss this with your CF doctor or dietitian.

Adequate weight gain during pregnancy is very important and in some women tube feeding may be necessary to help achieve this.

Should you have any concerns you should discuss them with your CF team.

Further information

The Cystic Fibrosis Trust provides information about cystic fibrosis through our factsheets, leaflets and other publications.

Most of our publications are available through our helpline and can be downloaded from our website or ordered using our online publications order form. Visit cysticfibrosis.org.uk/publications.

The Cystic Fibrosis Trust helpline can help you with a range of issues, no matter how big or small. Our trained staff can provide a listening ear, practical advice, welfare/benefits information or direct you to other sources of support. The helpline can be contacted on 0300 373 1000 or helpline@cysticfibrosis.org.uk and is open Monday to Friday, 9am – 5pm.

Calls to 0300 numbers cost no more than 5p per minute from a standard BT residential landline. Charges from other landlines and mobile networks may vary, but will be no more than a standard geographic call and are included in all inclusive minutes and discount schemes. If you are worried about the cost of the call please let us know and we'll call you back.

You can also find more information at our website cysticfibrosis.org.uk.

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More factsheets available at:
cysticfibrosis.org.uk/publications

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The information included in this publication is not intended to replace any advice you may receive from your doctor or CF multidisciplinary team and it is important that you seek medical advice whenever considering a change of treatment.

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