

The Cystic Fibrosis
Association of Ireland

**Nutrition for
Adolescents
& Adults
with
Cystic
Fibrosis**



What is Cystic Fibrosis?

Cystic Fibrosis (CF) is an inherited disease, affecting mainly the lungs and digestive system. It is Ireland's most common life threatening genetically inherited illness.

Approximately 1 in 19 people are carriers of the CF gene.

CF affects the secretory glands, damaging many organs including the lungs, the pancreas, the digestive tract, the liver and the reproductive system. It causes a build up of mucus blocking the bronchial tubes and preventing the body's natural enzymes from digesting food.

CF affects primarily the lungs and digestive system. A build up of mucus can make it difficult to clear bacteria which leads to cycles of lung infection and inflammation. Mucus can also block the ducts of the pancreas making it difficult to digest and absorb adequate nutrients from food. The result is that people with CF can be prone to constant chest infections and malnutrition.

Some people with CF must consume pancreatic enzymes with food to absorb nutrients and must also perform daily chest physiotherapy to ensure they stay fit and healthy.

Not all people are affected in the same way or to exactly the same degree – some are affected more or less than others.

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This booklet has been written to assist you.

It is not intended to replace any advice you may receive from your CF doctor or Clinic.

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Introduction

Eating well and maintaining a good nutritional status is a very important part of the treatment of CF. There is evidence that improving and maintaining a good nutritional status has a positive impact on prognosis and general well-being in CF. This booklet is designed to provide general nutrition advice for adolescents and adults with CF, however all persons with CF should see their dietitian for individualised nutrition advice.

Malnutrition in CF results from a combination of three main factors

1. Having repeated chest infections causes your body to work harder, this means you burn more calories and these calories need to be replaced. People with CF have higher daily energy requirements than the non-CF population.
2. Pancreatic Insufficiency, which is present most people with CF can result in poor digestion and absorption of particular nutrients especially fat. Loss of these fat-calories puts the person with CF at further risk of poor weight gain and malnutrition.
3. Poor appetite in CF is common. Many people with CF will lose their appetite especially during periods of infection.

If you are well nourished, you will:

- Look and feel better
- Be better able to fight infections
- Have a store of energy to help you through periods of infection

HOW MUCH FOOD DO YOU NEED?

It is important to eat enough calories to gain weight, grow and be strong enough to fight infections.

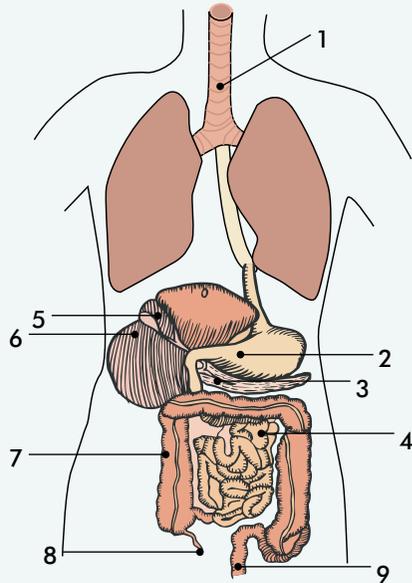
All people with Cystic Fibrosis are encouraged to eat a diet high in calories and fat. Many years ago the dietary advice given to people with CF was very different. Restriction of dietary fat was common due to the inadequacy of pancreatic enzymes available. Since the development of effective pancreatic enzymes there is now, no reason to restrict dietary fat.

THE NEED FOR ENZYMES

The majority of people with Cystic Fibrosis have difficulty digesting the food that they eat. This is because in CF an organ called the pancreas does not supply enough of the chemicals (enzymes) that are needed to digest food. This is often referred to as Pancreatic Insufficiency (PI). When you do not produce these enzymes in your body you have to take them in the form of capsules when you eat.

The Gastro-Intestinal Tract

The Gastro Intestinal Tract (GIT) or "gut" is a tube that connects your mouth to your anus. The function of this tube is to take in food, digest it and then absorb it into your bloodstream where it is used as a source of energy and for other functions. When food is eaten, it passes into your stomach where it is mixed with stomach acid. It then passes along into your small intestine (small bowel). Your gut releases enzymes that digest the food and allow it to be absorbed. Food that is not digested passes into the large intestine (bowel) and from here it is excreted as waste, i.e. a bowel motion/stool.



- | | |
|--------------------|--------------------|
| 1. Oesophagus | 6. Liver |
| 2. Stomach | 7. Large intestine |
| 3. Pancreas | 8. Appendix |
| 4. Small intestine | 9. Anus |
| 5. Gall bladder | |

FOOD IS MADE UP OF 3 MAIN NUTRIENTS:

- FAT
- CARBOHYDRATE (CHO) sugars and starches
- PROTEIN

Nutrients are found in different amounts in different foods. In food, these nutrients are made up of long chains of smaller units, like beads on a chain, for example:

Fats O-O-O-O-O-O

Carbohydrates □-□-□-□-□-□

Proteins Δ-Δ-Δ-Δ-Δ-Δ

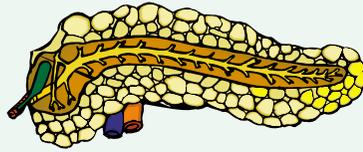
ENZYMES

The function of enzymes is to break these chains into single units small enough to be absorbed into your blood system. There are different enzymes to break down each type of nutrient chain.

LIPASE IS THE ENZYME WHICH BREAKS DOWN FAT

These enzymes are produced all along the GIT. However, the lipase enzyme that breaks down fat comes mainly from an organ called the pancreas.

Pancreatic Enzymes



In CF the pancreas may not produce enough lipase to digest the fat in your food.

If the enzymes are not available to break the chains of nutrients, the food will pass through you without being properly digested. When undigested food ends up in your bowel it can make your stomach bloat causing cramps and wind which can be very uncomfortable. The undigested food passing through will also cause you to have more frequent bowel motions and so lose valuable nutrients, making it difficult to gain weight.

To help prevent this most people with CF need to take Pancreatic Enzymes. This is often referred to as “Pancreatic Enzyme Replacement Therapy” – PERT.

ENZYME PREPARATIONS

There are many different types of enzymes available. Most enzymes come in capsule form. The active enzyme is contained in granules. The capsule dissolves in the stomach and then enzymes are released. The enzymes mix with the food you eat and digest it as it moves down through your intestine. There are a variety of enzymes preparations available. Generally they come in two forms:

Enzyme Capsules: Creon, Pancrease, Nutrizyme

- available in a standard or high strength preparation

Loose Enzyme Granules:

- primarily prescribed for infants. A standard scoop is used to measure out recommended doses.

Your dietitian /doctor will prescribe the most suitable enzyme preparation for you.

A GUIDE TO TAKING ENZYMES

You need to take your enzymes with almost everything you eat. Taking your enzymes with all your food enables you to digest the food you eat and get the beneficial nutrients from it.

REMEMBER

It is very important to take enzymes as advised by your dietitian

Unfortunately, there is no standard dose of enzyme and doses can vary greatly from person to person. The correct dose is that which prevents symptoms, e.g. steatorrhoea (loss of fat in stools), abdominal distension, cramps etc. and promotes weight gain.

The following general points should be remembered when taking enzymes:

- The higher the fat content of the meal, the higher the dose of enzyme needed to digest it.
- Bigger portions of foods generally need more enzymes.
- All foods except pure sugar foods need enzymes.

METHOD OF TAKING ENZYMES

SWALLOW THE CAPSULES AND ENZYME GRANULES WHOLE

- Enzymes capsules should be swallowed whole. For people who have difficulty swallowing, loose enzyme granules may be used or the capsules can be opened and the contents taken from a spoon with water, juice, jam or fruit puree.

Enzymes will not work effectively if:

- They are chewed
- They are added directly into foods other than those mentioned previously
- They are taken too far in advance of a meal/snack or taken too long after the food has been eaten.



ROUGH GUIDE TO THE FAT CONTENT OF FOOD

NO FAT FOODS (You do not need to take enzymes with these foods)	Sugar, jam, syrup, boiled sweets, jellies, mints Minerals, squash, glucose, polymers, fruit juice, black tea or coffee Ice lollies, jelly,
LOW FAT FOODS	Cereal, bread, plain biscuits, boiled potatoes. Steamed/poached/grilled fish, chicken
MEDIUM FAT FOODS	Yoghurt, cheese, mousse, milk & milk based drinks Mashed potato, oven chips, Meat, tinned fish in oil.
HIGH FAT FOODS	Quiche, pizza, breaded chicken or fish, processed meats - sausage rolls, pies, chips, roast potatoes, fried foods, crisps, nuts, cream cakes, doughnuts, chocolate bars

REMEMBER ..MORE FAT = MORE ENZYMES

Divide the enzyme dose

- If your meal is eaten over a period of time ($> 1/2$ hour), it is best to divide the enzyme dose over the meal. This ensures that all of the food has some enzyme mixed with it. It is also useful to divide the dose if you are unsure how much food will be eaten, i.e. give a small amount of enzymes at the start of the meal, and give the remainder of the dose if the meal is finished.

ARE YOU HAVING PROBLEMS WITH YOUR ENZYMES?

If you are having frequent bowel motions daily you are losing a lot of the nourishment and calories from the food that you eat. Therefore you may need more enzymes, especially if your bowel motions are bulky, foul smelling or difficult to flush. If this is the case, discuss it with your dietitian or CF team.

OTHER THINGS THAT MAY NEED REVIEWING ARE:

- Are you taking enough enzymes with all of your meals and snacks even when you are eating away from home?
- Are you taking enough enzymes with higher fat foods?

- Are you dividing your dose of enzymes over your meals?
- It is best to take some enzymes before and some during the meal. This ensures that there are enough enzymes mixed with all that you eat.
- Are you having heartburn or indigestion?
- If your stomach is too acidic it can reduce how well the enzymes work. You may need to be put on a medicine to correct this.
- It is also important not to chew the microspheres (granules). This breaks the protective shell and exposes the enzymes to stomach acid, which can destroy them.
- Are your enzymes in date and being stored correctly?
- Always check the expiry date on each batch and store them in a container in a cool dry place out of direct sunlight.
- If you are adjusting the dosage of enzymes, do so gradually and wait 24-48 hours before adjusting further. You should discuss enzyme changes with your dietitian or doctor

Dietary Advice

WHAT SHOULD YOU EAT?

- It is important for everyone with CF to eat a variety of foods, including dairy foods, meat and fish, cereals and breads, fruit and vegetables.
- It is especially important to include lots of energy and protein rich foods. You should try to include as many of these as possible in your diet every day.
- Even if you do not feel very hungry, try not to skip meals, but eat something small more frequently instead.
- Try to include foods containing protein with each meal and snack.
- Remember that you are likely to need pancreatic enzymes with everything you eat (except foods that only contain sugar). Foods containing a lot of fat will need extra enzymes.
- If you find it difficult to gain weight. You might need to boost your diet with extra energy supplements, nourishing drinks or tube feeds. This can be discussed with your dietitian.

PROTEIN RICH FOODS

You need protein to build muscle mass. Therefore, it is important to eat lots of protein-containing foods every day. For example milk, yogurt, cheese, eggs, meat,

chicken, fish and pulses. All these foods are a rich source of protein. Remember that many of these foods will require pancreatic enzymes for digestion.

ENERGY RICH FOODS

Foods that contain a lot of fat or sugar provide lots of calories. For this reason fats and sugars are especially useful because they can add extra calories to foods without making it too bulky and filling you up.

HIGH FAT FOODS

These include pure fats such as butter, margarine, oil and cream.

Suggestions to increase the fat content of food include:

- Fry foods whenever possible.
- Add lots of butter or margarine to potatoes or vegetables and spread thickly on bread, hot toast, crackers etc.
- Add mayonnaise to salads and sandwiches with meat, cheese or eggs.
- Add cream to soups, sauces, puddings, desserts, fruit and milky drinks.



- Choose other high fat foods such as crisps, ice-cream, chocolate, pastries, ready to eat individual pots of desserts etc. as snacks between meals.
- Include other high fat protein rich foods, e.g. pate, cheese, nuts and processed meat as much as possible.
- Use plenty of sugar in drinks, cereals and desserts etc.
- Spread jam, honey, marmalade, etc. thickly on bread, biscuits or crackers.
- It is best to use full sugar varieties of foods for maximum calories (i.e. not "diet" products).

SUGAR AND SWEETS

Sugar is very useful, as a source of calories. Suggestions to boost the energy content of your foods with sugar include:

- Chocolate, sweets, cakes and biscuits contain lots of energy and may be given as snacks and after meals, but not instead of meals.

HOW TO INCREASE THE ENERGY CONTENT OF SOME FOODS

Instead of: Plain Potatoes:

- add cream/butter/margarine
- fry with bacon and onion
- make chips or sauté potatoes in butter or oil
- add fillings to baked potatoes, such as sour cream or greek yogurt cheese and bacon coleslaw baked beans

Vegetables:

- stir fry vegetables using butter or oil
- add cheese and melt under grill
- serve with cheese sauce
- add melted butter

Plain fruit:

- stew fruit with sugar
- serve with cream, ice-cream, yogurt or custard
- try dried fruit as a snack or added to cereal
- choose tinned fruit in syrup rather than in juice

Milk

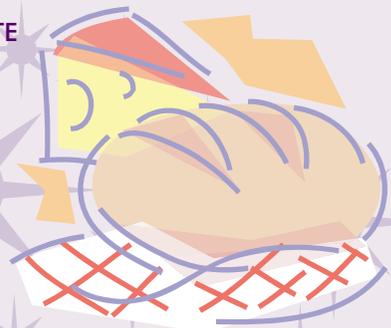
- add cream and/or skimmed milk powder to fortify milk
- use this fortified milk in cereals and to make up milky drinks, soup etc.

SAMPLE ENERGY & PROTEIN RICH MEAL PLAN

Breakfast: + Enzymes	Juice Cereal + milk + sugar/ Toast + butter + jam Coffee made on milk/ Milky tea/ Juice
Mid morning: +Enzymes	Scone/ bun + spread + jam Milky drink or high protein high energy drink
Lunch: + Enzymes	Chips/ Rice/ Pasta/ Potato + butter Vegetable + butter/ Salad + dressing Meat/ Fish/Eggs/ Cheese/ Beans + sauce/ gravy Dessert + cream/ ice-cream/ custard Milk/ High protein energy drink
Mid Afternoon: + Enzymes	Snack (see suggestions) + milky drink or a high protein high energy drink
Dinner: + Enzymes	Chips/ Potato/ Rice / Pasta/ Bread + butter Vegetables + butter/ Salad + dressing Meat/ Fish/ Eggs/ Cheese/ Beans/ Pizza Bun/ Cake/ Biscuit/ Pasty/ Yogurt Milk/ High Protein High Energy Drink
Supper: + Enzymes	Snack (see suggestions) + Milky drink or a High Protein High Energy Drink

WHAT TO DO IF YOU HAVE A POOR APPETITE

When you have an infection you need a lot more energy than normal, even though this might be the very time when you may not feel like eating. Although you might have a small appetite, it is essential to keep up your energy intake. Try snacking frequently with small snacks or nutritional supplements during the day.



Nutritional Supplements

NUTRITIONAL SUPPLEMENTS

There is a wide range of special nutritional products that can be used to supplement your dietary intake if your weight or appetite is poor. They should not be used as substitutes for meals and snacks, but rather as an extra source of calories. For this reason they are generally best taken between meals or before bed. However they can also be useful if your appetite is poor during an infection as you might find it easier to take a drink instead of solid food during this time.

REMEMBER

Supplements should not take the place of your meals (unless you are unwell)

ORAL SUPPLEMENTS

Oral nutritional supplements come in a variety of forms. The main types are:

- Nutritionally complete supplements
- Energy and protein rich supplements
- Protein supplements
- Glucose polymers
- Fat emulsions
- Fat and carbohydrate mixture

You should discuss what supplements might be suitable for you with your dietitian.



Tube Feeding in Cystic Fibrosis

Some people with CF may find it especially difficult to gain or maintain their weight despite eating and taking supplements. Supplementary tube feeding can be a useful option. This involves using a special tube to deliver feed into your stomach at night time. This is called "Gastrostomy Feeding".

Getting the tube placed involves a minor procedure in hospital. The tube is very discreet and can remain hidden under your clothes. There are various types of tubes available (e.g. PEG tubes, or low profile "button" tubes) – your dietitian will discuss the different types of tubes with you.

The tube can then be used at night by attaching it to a pump that delivers the feed into your stomach while you sleep. You will not feel anything and by the morning you will have had the equivalent

of two large meals overnight! During the day you should continue to eat an energy and protein rich diet as normal.

If you have a feeding tube, it can also be used if you are unwell or have a poor appetite, to supplement your intake during the day.

Depending on what feed is used you may need to take enzymes with your night feed.

Some people may prefer to use a nasogastric tube for overnight feeds. This involves having a tube passed from your nose into your stomach. This feeding method is not suitable for everyone.

If you would like more information about tube feeding please ask your dietitian or CF team.

Vitamin Supplementation

Because most CF patients have pancreatic insufficiency they will have some degree of fat mal-absorption which can also lead to some vitamin deficiencies.

There are two types of vitamins:

FAT-SOLUBLE – vitamins A, D, E and K

WATER-SOLUBLE – the B group of vitamins and vitamin C

In CF replacing fat-soluble vitamins is very important to prevent deficiencies.

Almost all patients with cystic fibrosis will be prescribed vitamin supplements – you should discuss what to take with your Dietitian.

It is important to take only vitamins prescribed for you by your CF dietitian or doctor.

Cystic Fibrosis Related Diabetes

WHAT IS DIABETES?

Diabetes is an inability to control the level of sugar in your blood.

A normal blood sugar is between 4-6mmol/litre. Insulin is the hormone that controls the level of sugar in your blood. In diabetes the blood sugar level remains high because you do not have enough insulin to keep it normal. This may result in weight loss due to sugar being lost in the urine. The main symptoms of untreated high sugar levels are:

- Increased thirst
- Passing lots of urine
- Tiredness
- Blurred vision
- Weight loss

Many teenagers and adults with Cystic Fibrosis (CF) develop diabetes, this is known as CF Related Diabetes (CFRD). It is not treated in the same way as other types of diabetes. For this reason people with CF and diabetes should **not** follow a strict low sugar, low fat diet. You may require insulin or tablets to help control your diabetes.



WHAT CAN CAUSE DIABETES?

The main cause of diabetes in people with Cystic Fibrosis is insulin deficiency. During a chest infection or course of steroids you may develop glucose intolerance. This is when your body cannot control your blood sugar level for a short period of time. It may return to normal when the infection resolves or the steroids are stopped. You may only require medication temporarily. Repeated chest infections and steroids may lead to diabetes over time

HOW IS DIABETES DIAGNOSED?

An oral glucose tolerance test is the best way to diagnose diabetes. This involves measuring fasting blood sugar levels, then measuring blood sugar levels at regular intervals for up to 2 hours following a specific amount of glucose (sugar) given as a food or drink.

HOW CAN I CONTROL MY DIABETES?

1. Insulin

Most people with CF and diabetes require insulin. If you do need insulin, it has many benefits. It can improve your blood sugar control and help you to gain weight. Using insulin may also make you feel better as improving your blood sugar levels, you may find your chest also improves. There are many different types of insulin which when used correctly can allow for dietary flexibility.

Insulin is injected under the skin using a very small needle. Most people with CFRD will take an injection before each meal.

2. Tablets

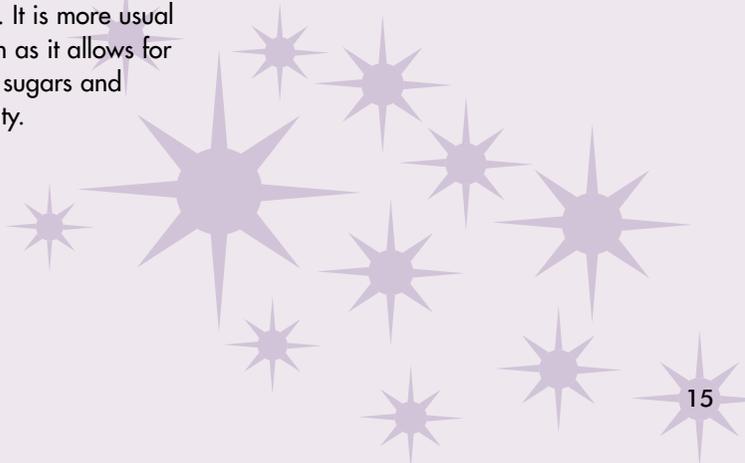
Tablets are occasionally prescribed. These need to be taken with foods once or twice daily to help control your blood sugar level. It is more usual to be prescribed insulin as it allows for better control of blood sugars and greater dietary flexibility.

DIETARY ADVICE

The recommended diet for someone with diabetes who does not have CF is a diet low in fat and sugar to help control blood sugar and prevent too much weight gain as well as reducing cholesterol level.

With CF, more calories are needed in the diet to help growth and weight gain. Because fatty foods are twice as rich in calories as sugars, a high fat diet is encouraged with very little sugar restriction and you are certainly not encouraged to lose weight. In CF related diabetes there is much more flexibility in the diet to allow for weight gain and maintenance of a good nutritional status.

It is very important that you get specific advice from your dietitian in the event that you develop diabetes.



Osteoporosis

Osteoporosis is a reduction in bone mass which may lead to brittle bones which are prone to fractures. People with CF can have increased risk of developing osteoporosis due to various factors including:

- Poor nutritional status i.e. low body weight.
- Poor intake of certain nutrients such as calcium and vitamin D.
- Low hormone levels.
- Certain medications.
- Low levels of physical activity.
- Malabsorption

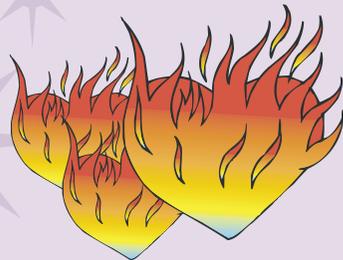
The following will help to reduce the risk of osteoporosis:

- Ensure you eat a high protein and energy diet.
- Ensure you take enough enzymes to maximise absorption of nutrients.
- Ensure adequate Calcium intake. Dairy products such as milk, cheese and yoghurt are major sources of calcium. If you do not include dairy products regularly, consult your dietitian regarding suitable alternatives.

- Ensure adequate vitamin D intake. It is essential to take your prescribed vitamins to prevent vitamin D deficiency.
- Include physical activity in your daily routine.

GASTRO-OESOPHAGEAL REFLUX OR HEARTBURN

Heartburn or indigestion is a burning sensation in the chest, which may extend up to the throat. It is also known as gastro-oesophageal reflux. The main cause of heartburn is reflux of acidic stomach contents up into the oesophagus. It is important to inform your doctor if you suffer from heartburn as medication can be prescribed to help you. Uncontrolled heartburn can lead to malabsorption as pancreatic enzymes do not function in an acidic environment. Fizzy drinks, alcohol, coffee and spicy foods, such a curry or chilli can worsen symptoms. It may be useful to avoid these foods if they cause a problem.



Constipation

Constipation can be a problem for some people with Cystic Fibrosis. A combination of dietary changes and fluid can help regulate your bowel motions. Therefore if you have problems with constipation you should contact your dietitian for further advice. It is important to note that constipation is different to Distal Intestinal Obstruction Syndrome (DIOS). DIOS is discussed below.

Many people with CF occasionally complain of stomach cramps. This may be for several reasons so it is important to tell your doctor if you suffer from any abdominal upsets. Things that may help to alleviate this problem include:

- Eating small amounts frequently rather than large meals.
- Ensure you are taking enough enzymes. If you are not, your food may not be properly digested and so end up in your large bowel. This can lead to the production of excess gas, which can make your stomach bloat, cause cramps and wind that can be very uncomfortable.



Distal Intestinal Obstruction Syndrome

Distal Intestinal Obstruction Syndrome (DIOS) is a common complication of CF. It was formerly known as Meconium Ileus Equivalent (MIE). It occurs when thick sticky mucus and semi digested food collect in the intestine and cause a blockage. This can result in abdominal bloating and discomfort and pain that can in turn cause a poor appetite and weight loss.

POSSIBLE CAUSES OF DIOS

DIET AND ENZYMES

- Poor compliance with enzymes
- Inadequate enzymes to ensure nutrient absorption. Changes in eating habits or dietary patterns.
- Dehydration or lack of fluid can also contribute to DIOS. This is of special concern in hot weather or in sunny climates

MEDICATION

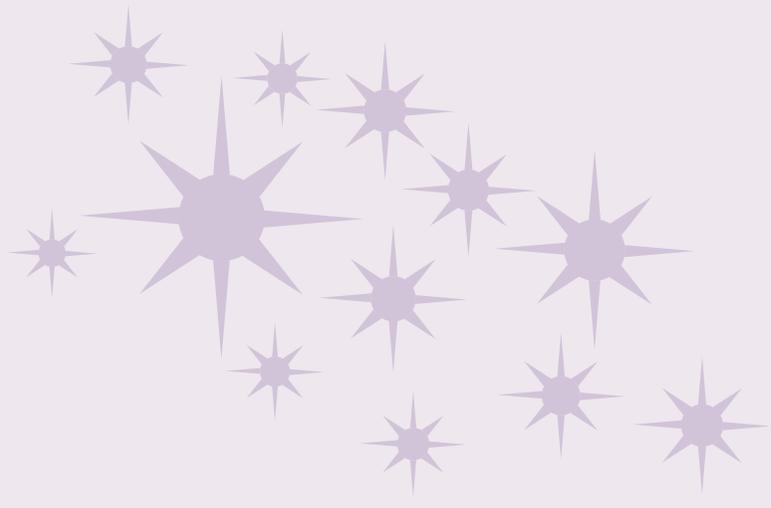
- Some drugs (e.g. some pain killers or cough mixtures containing codeine) can affect your bowel and be a factor in DIOS. If you are on these medications and have bowel problems you should discuss it with your dietitian or doctor.

TREATMENT

Treatment of DIOS will vary between patients depending on the severity of the obstruction. You will need to have an x-ray of your abdomen and a laxative regime will be prescribed which should clear the blockage.

Once the blockage has cleared you will be advised on a treatment regime to prevent further episodes of DIOS.

Nutrition is a vital part of the overall management of all people with Cystic Fibrosis. Regular review by your dietitian in your CF Unit is important to monitor and optimise your nutritional status



BOOKLETS

THE FACTS & FINDING OUT

An introduction to the causes and effects of Cystic Fibrosis. A guide for parents of newly diagnosed children with Cystic Fibrosis.

NUTRITION

Eating well with Cystic Fibrosis –
A guide for children and parents.

TREATMENT

Physiotherapy for Cystic Fibrosis. Illustrates methods of physiotherapy in CF.

RIGHTS & ENTITLEMENTS

A guide to rights and entitlements for people with CF and their families.

LEAFLETS

CFAI GENERAL LEAFLET

Outlines the structures and help available from the Cystic Fibrosis Association.

CYBER CAMPUS LEAFLET

Description of online computer course for young adults with CF.

THE FACTS

General introduction to causes and effects of CF.

MAGAZINES

ANNUAL NEWSLETTER

Yearly magazine which looks at the work and achievements of the Association.

FUTURE FORCE

Magazine designed and written by CF adults covering large range of topics and issues affecting CF adults.



Further enquiries about literature, including booklets produced by the Association can be obtained from

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