

**The Cystic Fibrosis
Association of Ireland**

**Paediatric
Nutrition
and
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.

Written by and with thanks to Ann-Marie Brennan, Paediatric Dietitian, Cork University Hospital.

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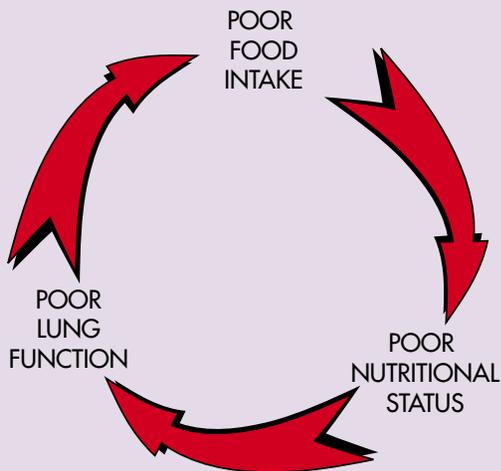
Nutritional Status

DOES NUTRITIONAL STATUS MATTER IN CF?

Nutrition plays an important role in your child's overall wellbeing. Your child can achieve normal growth, fight off infections and improve lung function when nutritional needs are met.

WHY DO CHILDREN WITH CF GET MALNOURISHED?

- **Malabsorption**
Malabsorption is common in children with cystic fibrosis. Calories and other nutrients pass out of the body in the stools, therefore eating extra food is necessary to replace what is lost.
- **Frequent Infections**
Cystic fibrosis causes the body to work harder, for example when your child coughs or has an infection. This means you use more calories and this energy needs to be replaced.
- **Reduced Food Intake**
Due to lack of appetite, chronic chest infections, abdominal pain and vomiting.



Introduction to Enzymes

DOES YOUR CHILD NEED ENZYMES?

Most infants and children with CF do not have the ability to digest their food properly and therefore need enzymes. There is no standard amount of enzymes for children and every child should be treated individually. The correct dose is that which controls stomach and bowel symptoms i.e. amount of stools, consistency of stools, abdominal pain. You should discuss your child's enzyme requirement with a dietitian or member of the CF team.



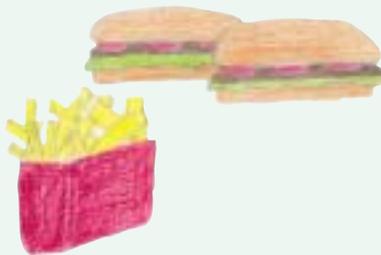
WHAT ENZYME PREPARATIONS ARE AVAILABLE?

Pancreatic enzymes come in several forms. The most commonly used pancreatic enzyme preparations in Ireland are Creon and come in capsule form. The capsules are made up of microspheres or granules, which contain enzymes. The capsule dissolves in the stomach and the enzyme granules are released. From there the enzymes pass into the small intestine where they digest food.

The two main types of enzyme preparations are:

- Loose Enzyme Granules, available for infants.
- Enzyme Capsules, available in a standard or high strength preparation.

Use of Enzymes



HOW SHOULD ENZYMES BE TAKEN?

- Swallow capsules whole at as early an age as possible (greater than 2yrs of age). Discuss this with the your dietitian or member of the CF team.
- Avoid mixing enzymes directly with food. In infants or for children who have difficulty swallowing, the capsule may be opened and mixed with fruit puree / water / jam and taken from a spoon. Alternatively, loose enzyme granules may be prescribed for infants e.g. Kinder Creon.
- Avoid crushing /chewing capsules or leaving enzymes beads in the mouth after meals.
- Give with a meal rather than after.
- Give some at the beginning and some during the meal if mealtimes last greater than 30 minutes.
- Give extra enzymes with fatty or large meals and snacks.
- Give enzymes with snacks e.g. milk, biscuits, yoghurts.
- The dose required mainly depends on the fat content of the food and the portion size eaten. Increase the dose gradually if stools are pale, loose, fatty or difficult to flush. Wait 1-2 days before adjusting further between each increase.

- There is no need to take enzymes with sugar, jam, fizzy drinks, fruit, dry cereal or jellies.
- Store enzymes as directed and avoid keeping them in the glove box of the car (they can get too hot).
- If enzymes are forgotten at a mealtime, they should be taken within 30 minutes after the meal.

HOW DO I KNOW IF MY CHILD IS TAKING THE CORRECT AMOUNT OF ENZYMES?

Children who are gaining weight and growing well are probably getting enough enzymes.

TOO LITTLE:

- Number of stools may increase.
- Stools may change in appearance (large, greasy, orange spots).
- Stools may float in the toilet or smell worse than usual.

TOO MANY:

- You might notice your infant's bottom becoming red or your child complaining of an itchy bottom or constipation.

Feeding Infants & Toddlers

WHICH MILK?

- Breast milk or standard infant formula is a suitable milk for most babies.
- Babies who have undergone bowel surgery or who need milk that is easier to absorb may be prescribed special milks.
- High calorie ready-made baby milks or calorie supplements added to milk may be necessary for babies who have poor growth and weight gain.
- It is recommended that breast milk and/or formula be continued until 1yr of age and sometimes beyond it if the quantities of spoonfeeds are small. Cow's milk should not be given as a drink before 1 year of age.
- Meat, fish, eggs contain protein and require extra enzymes.
- Mixing dry baby foods with formula instead of water is a good idea, as this will increase nutritional value considerably and will require extra enzymes.

The volume of milk taken in the early stages of introducing solids should remain the same.



WHEN SHOULD I INTRODUCE SPOONFEEDS?

Spoon feeds are recommended from 4-6 months of age. Some solids will require enzymes. Follow the guide:

- Fruit, vegetables, baby rice contain carbohydrate and do not need enzymes.
- Milk based foods e.g. yoghurts, cheese, fromage frais, butter contain protein and fat and require extra enzymes.

>> TIP:

Try to include solids before large milk feeds, so your child has a good appetite and is not too full.

Feeding Older Children

HIGH ENERGY / HIGH PROTEIN DIET

The diet should be based on a good routine and include foods from all food groups but must also include foods which are high in fat and sugar.

- **Favour energy & protein rich foods**
 - ✓ dairy products e.g. butter / oil / cream / ice-cream / cheese / eggs.
 - ✓ meat / fish / poultry / nuts* / pate / pizza.
- **Fatty Foods**
 - ✓ sausages / chips / crisps / chocolate / biscuits / croissants / pastries / cakes.
 - ✓ fry foods / baste with oil instead of grilling, steaming, boiling.
- **Fortify Foods**
 - ✓ add cream / sugar / milk / mayonnaise / jam / marmalade / gravy to foods.



>> TIP:

Don't forget to encourage fruit and vegetables i.e. stirfry vegetables, choose tinned fruit in syrup instead of natural juice.

* **NOTE:** Do not offer nuts to children under five years of age.

Planning Your Child's Diet

PLANNING A HIGH ENERGY / HIGH PROTEIN DIET

Fitting extra calories into your child's diet may take some planning.

- You need to plan ahead what to buy at the shops.
- You need to find high calorie snacks that are easily available (or snacks your child can prepare alone).
- You need to plan for the extra time and preparation these meals will require.

THE FOLLOWING ARE USEFUL SUGGESTIONS FOR MEALS:

Breakfast:	Cereal with Milk, Cream, Sugar Bread and Butter, Jam, Marmalade, Peanut-butter Bacon / Sausage / Egg Milk / Juice	
Lunch:	Bread / Bread Roll / Pitta Bread / Bap / Crackers etc with butter, mayonnaise Meat / Fish / Egg / Cheese Vegetables or Salad Yogurt / Chocolate / Crisps / Popcorn / Biscuits / Fruit Milk / Juice	
Dinner:	Potato with butter, cream / Chips / Rice / Pasta / Waffles / Pizza Meat / Fish / Egg / Cheese Vegetables with butter or salad with dressing Dessert: Cake / Biscuits / Custard / Rice Pudding / Yogurt / Ice-cream Milk / Juice	
Snacks:	Cheese cubes/triangles Yoghurt Chocolate bar Custard / Rice pudding Nutritional Drinks	Muffin Ham slices Crisps Fruit Scones
		Crackers and cheese Cocktail sausages Cereal with milk Mini pizzas Biscuits

DIETARY SUPPLEMENTS

If weight gain or appetite is poor your child may need a supplement to give them a boost. Supplements should not replace food at mealtimes. Ideally they should be prescribed on an individual basis. Discuss this with your dietitian or member of the CF team.

Guidelines on their usage:

- Offer 2-3 times a day after meals or at bedtime.
- Avoid overuse.
- Extra enzymes are needed with milk / yoghurt based supplements.
- Check appetite for meals does not decrease.
- Supplements come in a variety of styles e.g. juice / milk / yoghurt based and a variety of flavours, it is important to find one that your child enjoys.

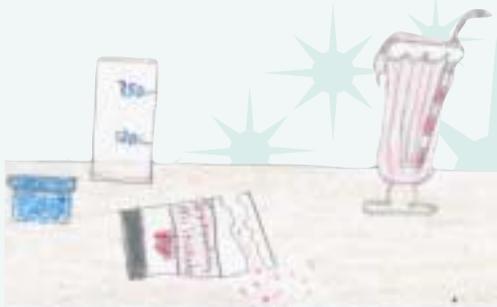
TUBE FEEDING

Some children who are not gaining weight or are not growing well may need a tube feed to help them gain weight and grow better. The thin tube may be passed down a child's nose into their stomach i.e. nasogastric tube or a tube may be passed directly into the stomach i.e. gastrostomy tube, which involves a minor operation.

A high energy liquid feed is delivered through the tube and this usually happens at night time when the child is sleeping. Children are encouraged to eat normally during the day and take their usual nutritional supplements. Your CF team will advise if tube feeding is necessary for your child.

VITAMINS

In CF, it is likely that the Fat Soluble Vitamins which are A,D,E,K will not be absorbed well by the body. These vitamins may be lost with fat in the stools. Therefore it is important that a Fat Soluble Vitamin supplement is taken daily to prevent deficiency. Your dietitian or health professional will advise which vitamin supplement is suitable for your child in order to meet the recommendations. The amount of vitamins required will vary as your child grows and develops.



TODDLERS & BEHAVIOURAL FEEDING PROBLEMS

Food Refusal may be associated with:

- parental anxiety about food and excessive focus on food and weight gain.
- force feeding.
- vomiting/gagging associated with coughing.
- acute infections.
- frequent hospital admissions.

MANAGEMENT PLAN

- Do not force feed.
- Praise food eaten & ignore food refusal.
- Encourage regular family meals, so children learn correct feeding behaviour.
- Set the child up for success at mealtimes e.g. use small portions.
- 'Likes' & 'Dislikes' change daily.
- Limit feeding time to 20-30 mins.
- Be consistent in your approach.
- Do not discuss your child's feeding behaviour in front of them .

DENTAL HEALTH

Children with CF have normal teeth, however the sugary drinks and sweet foods that are recommended as part of the diet may cause tooth decay.

The following guidelines will help keep teeth and gums healthy:

- Brush teeth first thing in the morning and last thing at night.
- If less than 2yrs of age use soft toothbrush and water only.
- If between 2-7yrs of age, use a pea-sized amount of fluoride toothpaste and ensure toothpaste is not swallowed.
- Encourage taking sugary foods and drinks at main meals. Dairy products are one of the most tooth friendly snacks e.g. glass milk, yoghurt, cheese.
- Infants and toddlers should not sleep with a bottle in their mouths.
- Visit a dentist regularly.



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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