

CF Health Matters

Enzyme Guidelines for Children with Cystic Fibrosis



Good nutrition is important for normal growth and development in children, and for body maintenance in adults. Good nutrition includes a healthy diet with all the needed nutrients, as well as proper digestion and absorption of these nutrients.

GOOD NUTRITION =
healthy diet + digestion + absorption

Digestion

When food is eaten it goes into the stomach where it is broken down into small pieces. These small pieces pass into the small intestine. In the healthy body, digestion takes place with the help of bile from the liver and enzymes from the pancreas. The nutrients from the food are then taken in by the body (absorbed) and used for energy and growth. Food not broken down (not digested) passes out of the body in the stools.

In most people with cystic fibrosis, the tubes in the pancreas are blocked with the same type of sticky mucus found in the lungs. Pancreatic enzymes cannot get into the small intestines so food is not properly digested. This leads to poor nutrition and/ or poor growth. Most

people with cystic fibrosis need to take pancreatic enzyme capsules to help with digestion of food.

Enzymes

Pancreatic enzymes contain the enzymes lipase, protease, and amylase which digest fat, protein and larger sugars such as starch. Enzyme capsules are available in two forms, powder and enteric-coated, and are made by different companies in different strengths. The “number” gives the amount of lipase in the product.

“ **Most children can learn to swallow enzyme pills between three and seven years of age.** ”

Powdered enzymes are available in a loose powder and tablet form. Since these enzymes do not have a special coating some of the enzyme is broken down (therefore it does not work anymore) by acid in the stomach. The enzymes that do reach the upper small intestine help to break down and digest food.

Enteric coated enzymes are used by

most children and adults with cystic fibrosis. The coating protects the enzymes from acid in the stomach so that the enzymes reach the small intestines. The coating may take some time to dissolve, so the enzymes are released further down in the intestine than is normal.

The amount of enzymes needed by a person with CF varies with age, body size, and amount and type of food eaten. Your CF physician or dietitian will decide the type and dose of enzymes to use. This may change over time, especially in children.

Enzymes and Different Types of Food

Foods are made up of protein, fat and carbohydrate. Examples:

Protein foods: meat, fish, poultry, dairy products

Fat: butter, margarine, oils and salad dressings; many dairy products, including milk; many meat and meat alternatives; snack foods such as chips, cookies, crackers, chocolate bars; pastries and desserts

Carbohydrate: starches-breads and cereals, rice, pasta, vegetables; sugars- fruit and fruit juice, sugary foods such as candy.

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In general, foods that are high in fat and protein need more enzymes than foods high in carbohydrate. Enzymes are not needed for foods that contain only sugar because someone with CF is able to digest these foods with the enzymes found in saliva. Examples of these are fruit, fruit juice and fruit drinks, soft drinks, hard candy, popsicles, Jell-O.

Giving Enzymes

Enzyme capsules may be swallowed whole. Most children can learn to swallow pills between three and seven years. For younger children and infants, the capsules can be opened and given by spoon. Enzymes should be given either at the beginning of the meal, or given throughout the meal (i.e. at the beginning and the middle). Enzymes mix with the food eaten, and pass into the stomach and small intestines. It is recommended to take additional enzymes if more food is eaten ~ 1 hour after the last enzyme is taken.

Enzymes in Powder Form

- Open the capsule and mix with a small amount of applesauce (e.g. 2.5-5 ml) or a small amount of water. Feed applesauce mixture by spoon or water mixture with a small syringe. Once the enzyme is mixed with food, it must be eaten right away.
- Try to mix enzymes with the same food each time. This will mean that your child will only associate that food with enzymes

- Place Vaseline around the outside of the baby's mouth to protect the skin in case the baby spits some enzyme out. The Vaseline should be wiped away before feeding. This is especially important for mothers who are breastfeeding their infant with CF. Otherwise, latching may be difficult.

“ **Never increase enzyme dosage without consulting your CF team. Too many enzymes can be harmful.** ”

- Clean the inside of the mouth out with a large cotton tip or small towel mixed with sterile water to be sure that there is no enzyme left in the gums or under the tongue. Enzymes can cause the mouth tissue to become sore and bleed if left in the mouth.

Enteric Coated Enzymes - Infants

- Open the capsule and mix the beads with applesauce or other strained fruit. Give with a spoon. Another way to give enteric coated enzymes to young babies is to place the beads on a spoon or in a small flexible medicine cup. The beads can then be placed or poured into the back of the baby's mouth and then given the breast or bottle to feed.
- Check baby's mouth with either a large cotton tip or with your clean finger to see that no beads are left in the

mouth at the end of the feeding.

- Do not crush beads; the coating will be destroyed.
- Do not add beads to your baby's bottle. Beads will block the nipple.
- When your baby begins solids, they usually take only a small amount of food. At this point, they do not need any more enzymes than they are already taking with their breast milk or formula. However, when your baby is eating 50–125 ml ($\frac{1}{4}$ - $\frac{1}{2}$ c) of strained food or solids or more, the amount of enzyme will need to be increased.

Enteric Coated Enzymes - Older Children

- Enzyme capsules should not be chewed, crushed, or sprinkled on food.
- Make sure your child has a drink after taking enzymes.

Tips

- Once enzymes are started, weight gain is a good way to see if someone with CF is digesting and absorbing their food.
- Remember that it is important to adjust your enzymes to the amount of fat you are eating in a given meal. Ask your dietitian to help you with this.

Other signs to watch for:

- Diaper rash, mouth sores and irritation around the mouth may be a problem during the first month that your baby takes enzymes. Let your CF Team know

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if this happens. Applying zinc oxide or Vaseline in generous amounts to the buttocks can help.

- Stools usually decrease in amount and/or size, and should be less foul smelling or oily looking once enzymes are started. The amount of enzyme given may not be right if you notice the following: loose stools; stooling more often; greasy or floating stools; stomach pain or cramps; gas. Remember that there are other reasons for loose stools such as viral or bacterial infections, antibiotics, or too much juice. Let your CF Team know if you notice these signs of poor absorption. Even after your older child or teen has been taking enzymes for a long time it is helpful to check the stools every so often.

It may help you to know how many stools a day are “average” at different ages:

- Toddlers: 1-3 bowel movements/day
- Children or adults: 1-2 bowel movements/day
- If your child completely forgets his or her enzymes at a meal, the enzyme can be taken within 30 minutes after a meal (although it works best when given with the meal).
- Enzymes are very sensitive to heat and may degrade sooner than the expiry date indicated on the medication bottle. Do not store enzymes in a cupboard that is in direct sunshine, in the bathroom, in the glove compartment of a vehicle, or any place you think may be too warm in

your house. If you carry enzymes loose in a pocket or small container, discard the enzymes once weekly and exchange for a fresh supply from your stock bottle.

Do not increase enzymes without checking with the CF Team. Too many enzymes can be harmful.



Getting Your Child to Take Enzymes

Getting children to take enzymes can be a challenge. For example, toddlers may simply resist taking enzymes especially if they can tell that you are upset. School age children may be embarrassed to take enzymes at school.

Your child is most likely to cooperate with taking enzymes if, from the beginning, you are consistent and matter-of-fact about it. Starting in the preschool years you can talk with your child about the need for enzymes with food. You

can explain that taking enzymes will help them grow taller and gain weight. They also prevent gas, tummy aches, and loose stools. By taking the enzymes your child will have energy to play and keep up with friends.

School age children may want to hide taking enzymes in front of friends. This can lead to problems of not taking the enzymes at school. A better approach is to be open about the enzymes and to make enzymes a routine at home and at school.

Your Questions Answered

Q. How will I know if the enzyme dose is correct?

A. Healthy weight gains, having stools less often or in smaller amounts are all signs that your child is getting the right amount of enzyme.

Q. Which foods do not require enzymes for digestion?

A. Fruit (including dried fruit), juice, soft drinks, hard candy, popsicles and Jell-O are examples of foods that do not need enzymes.

Q. Will I be able to breastfeed my baby?

A. In most cases, you will be able to breastfeed your baby. Enzymes are still needed with each feeding. If your baby was very underweight at the time of CF diagnosis, a combination of breastfeeding and bottle feeding with fortified breast milk may be required for at least

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a few weeks. Once your child's weight is normal for their height, exclusive breast-feeding can resume.

Q. What should I watch for when my baby starts taking enzymes?

A. Watch for diaper rash, mouth sores or irritation, and let your CF Team know. Keep track of your child's stool pattern, and call the clinic if your child is having loose, greasy or floating stools, stomach cramps or gas.

Q. Does my child need enzymes with liquids, such as milk, if that is all that he is having?

A. Yes. Milk and other drinks such as milk-shakes and yogurt drinks contain protein and fat that require enzymes for digestion. The only liquids that do not require enzymes include juice, pop and sports drinks.

Q. How do I get my 5 year old child to swallow her enzymes?

A. Your child can try swallowing smaller capsules such as TicTac candy. Try dipping the capsules in margarine or butter to make the coating less sticky. Offer fun rewards such as stickers or crayons to help encourage her. Remember, though, that some children are ready to swallow pills early, at 2 years of age, while others are not ready until after they are over 5 years. Do not force your child to swallow the pills if they are not ready or if they are very resistant.

Enzyme Dosing Guidelines* (using enzymes with 8000 U Lipase/capsule)

AGE	TYPE OF FOOD	ENZYME DOSE	
		POWDER	ENTERIC COATED
0-12 months	Breast milk or formula whole milk	1 per feed or 1 per 125 mL	1 per 250 mL
	Solids: cereal, meat or vegetables	1 per 1/2 jar strained or 60 mL pureed or mashed food	1 per jar strained or 120 mL pureed or mashed food
1-4 years	Meal	4-5	2-3
	Snack	2-3	1-2
5-12 years	Meal	n/a	3-5
	Snack	n/a	1-3
over 12 years	Meal	n/a	5-8
	Snack	n/a	2-3

NOTE: The enzyme dose for snacks may be increased or decreased depending on size of the snack.

* Your CF Clinic may use different enzyme strengths. Follow the dose recommended by your CF Clinic.