The pancreas is a gland in the body connected to the small intestine. The small intestine is where most of the digestion and absorption of food occurs. One of the functions of the pancreas is to make enzymes that digest food. Digestive enzymes from the pancreas enter the small intestine through a small duct, or passageway. In cystic fibrosis (CF), the ducts in the pancreas become clogged with thick, sticky mucus. This mucus blocks the enzymes from reaching food in the small intestine. This can lead to poor digestion and absorption of food, as well as problems with weight gain. Fortunately, pancreatic enzyme replacements or “enzymes” are available to help people with CF digest and absorb their food.

What Are Enzymes And How Do They Work?
Pancreatic enzyme replacements come in capsule form. Inside each capsule are many small “beads” that contain digestive enzymes. Each bead is covered with a special “enteric coating.” This coating allows the beads to dissolve in the small intestine. The digestive enzymes are then released in the small intestine to help digest food. The main functions of enzymes are:
• to digest carbohydrate, protein and fat (the three parts of food that supply calories);
• to help with weight gain; and
• to promote nutrient absorption.

Because there appears to be a connection between better lung function and higher body weights, it is very important to take enzymes with all meals and snacks. More than 90% of people who have CF take pancreatic enzyme replacements.

How Are Enzymes Given?
Enzymes should be taken just before meals and snacks. For older children and adults, the capsules should be taken with liquid and swallowed whole. For infants and small children, the capsules may be opened up. The beads inside each capsule may be mixed in a small amount of soft food, such as applesauce, and given from a spoon. For babies, it may be helpful to offer breast milk or formula just after the beads are given.

Troubleshooting Guidelines
Some very young babies may “spit” out the beads. If this happens, gently scoop the bead mixture back into the baby’s mouth until the entire dose of enzymes has been given. It may take time for your child to learn how to swallow beads due to the new texture. Enzymes do not have a taste. Be patient, calm, and reassure your child. Call your CF dietitian or care center if your baby is having trouble taking enzymes.

How Are Enzymes Given?

Toddlers may refuse to take enzymes as they become more independent. If this occurs, try giving your toddler acceptable choices, such as, “Would you like to take your enzymes in applesauce or pears today?”

For many reasons, some children, teens, and even adults refuse to take their enzymes. Enzymes are essential in the care of CF. Talk with your CF dietitian or care provider if taking enzymes has become a problem.

Signs Of Maldigestion and Malabsorption
• Poor weight gain despite a good (sometimes ravenous) appetite;
• Frequent, loose and/or large bowel movements;
• Foul-smelling bowel movements;
• Mucus or oil in the bowel movement;
• Excessive gas and/or stomach pain;
• Distention or bloating.

Persons with CF who have not yet started taking enzymes may have any or all of the symptoms listed above. Improvement is generally noted once enzymes are started. Sometimes, people with CF who already take enzymes experience these symptoms. This may suggest that the dose or type of enzymes may need to be adjusted. Do not increase or decrease the dose of enzymes without talking to your CF dietitian or care provider. Always give the prescribed dose of enzymes.

General Information About Enzymes
• Enzymes should be taken just before eating.
• Enzymes are needed with all meals and snacks, milk, breast milk, formula and nutritional supplements (see “Foods That Do Not Need Enzymes” listed at the end for exceptions).
Nutrition  •  Pancreatic Enzyme Replacement

- Avoid “skipping” enzymes. Keep a supply of enzymes with you if meals or snacks may be eaten away from home.
- Enzymes work for about an hour after taking them.
- Always take the right dose. Do not change the dose without talking with your CF dietitian or care provider.
- Slightly higher doses of enzymes may be needed with foods that are high in fat (like “fast foods,” fried foods, and pizza). Tell your CF dietitian or care provider if higher fat foods cause problems.
- If the enzyme capsules are opened, don’t crush or chew the enzyme beads. They won’t work as well.
- For infants and small children who need the capsules opened up, mix the beads with a soft, acidic food, such as applesauce. Avoid mixing enzymes with milk-based foods (e.g., yogurt or pudding).
- Do not mix enzymes into foods ahead of time.
- Keep enzymes at room temperature (59˚ to 86˚ F). Keep enzymes away from heat. Don’t store them in places such as on top of a toaster oven or inside a hot car.
- Do not refrigerate enzymes.
- Enzymes have an expiration date. Check the expiration date on each bottle to make sure they are “fresh.”
- Always keep the lid tightly sealed on the bottle of enzymes.
- Ask the pharmacist to give you the enzymes in the original, sealed, unopened bottle from the manufacturer.
- For persons who supplement their diet with tube feedings, enzymes may be taken by mouth before starting the tube feeding.

Generic Enzymes
DO NOT ACCEPT GENERIC PANCREATIC ENZYMES. Generic enzymes have less activity than name brand enzymes and don’t work as well. If you notice that you are receiving generic enzymes, contact your CF dietitian or care provider and they will help you get name brand enzymes.

Foods That Do Not Need Enzymes
Pancreatic enzyme replacements contain enzymes that digest fat, protein and complex carbohydrates. Some foods and drinks do not require enzymes because they contain only simple carbohydrates that digest easily. Examples of foods and drinks that do not require enzymes are:

- Fruits
- Juice, juice drinks
- Soft drinks or sports drinks
- Infant rehydrating formulas (like Pedialyte®)
- Tea, coffee (without cream)
- Hard candy (like lollipops)
- Fruit snacks
- Jelly beans
- Gum
- Popsicles, freezer pops, flavored ice

Except for fruits and some fruit juices, there is little nutritional value in most of the foods and drinks listed above.

Thus, it is not recommended to offer these regularly or in large amounts. Ask your CF dietitian or care provider if you are unsure if enzymes need to be taken with a certain food.

If you have questions about “Pancreatic Enzyme Replacement In People With Cystic Fibrosis,” or any aspect of CF care, call your CF dietitian or care provider.

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