Pancreatic enzyme replacement therapy (PERT)
for children and adolescents with Cystic Fibrosis

Who needs Pancreatic Enzyme Replacement Therapy?
Pancreatic enzyme replacement therapy (PERT) is essential for children and young people with cystic fibrosis (CF) who are pancreatic insufficient. PERT helps the body digest and absorb nutrients from foods. Without PERT, children are at risk of poor nutrient absorption leading to poor growth and weight gain.

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 (to supply the body with energy)
- Help with weight gain
- Promote nutrient absorption.

Because there appears to be a link between better lung function and higher body weights, it is very important to take enzymes with all meals and snacks.

Dosing PERT
People with CF vary in their degree of pancreatic insufficiency. This means one person may require different amounts of PERT to another. Your dietitian will help you determine your dose. It is important to discuss any changes to your enzyme intake with the CF team before changing your dose.

A good guide for enzyme dosing is:
- 1 scoop of Creon Micro (5000IU) per 3-5g fat
- 1 Creon 10 000 capsule per 6-8g fat
- 1 Creon 25 000 per 15-20g fat

It is better to give a little bit more enzyme than too little. For example, if a food has 10g of fat, give 2 Creon 10 000 rather than 1. The maximum enzyme dosage is 10 000IU of lipase/kg body weight.

Enzyme doses need to be monitored regularly and adjusted based on amounts of food and fluid consumed. Doses also be changed if there are signs...
malabsorption. These signs include stomach pain, excessive gas, bloating, frequent, loose and / or oily stools, poor weight gain and poor growth.

**When should I take the enzymes?**

- Take pancreatic enzymes immediately before meals/snacks. Enzymes last in the small intestine (where they work) for around 30 minutes.
- Capsules and beads should not be crushed or chewed. Beads should be mixed with an acidic food (e.g. fruit puree, fruit gel, jam or tomato sauce) until capsules can be swallowed.
- If the child is regularly taking longer than 30 minutes to eat, give half the dose at the start of the feed and the rest half-way through.
- High fat meals will need more enzymes than low fat meals.
- Snacks will generally require fewer enzymes than a meal. Be cautious with high fat snacks, as these can require as many (or even more!) enzymes than a meal.
- Be flexible about the dosage and timing of enzymes. You need to adjust the amount of enzyme you take depending on how much fat is in your meal or snack.

**How do I know if I am taking enough?**

- Stools should return to a more normal appearance and colour, and flush away easily.
- You should not experience stomach pains on eating and you should have less wind.
- If you are gaining weight, it is a good sign that you are taking enough enzymes to digest and absorb your food.

Note: Sometimes people with CF produce too much stomach acid and this can make pancreatic enzymes less effective. If you have heartburn, reflux or abdominal cramps, discuss this with your CF team.

**Are there any foods and fluids that don’t need PERT?**

Enzymes need to be taken with ALL food and fluids. The exception is foods that contain mainly simple sugars. Some examples include:

- Fresh, dried and canned fruit
- Non - starchy salad vegetables
- Lollies, jellies, sorbet, Roll-ups
- Fruit juice, cordial, soft drink, electrolyte replacement drinks, water

Remember that while enzymes are dosed according to how much fat is in food, enzymes work on fat, carbohydrate and protein. Therefore, they are still needed with foods that are lower in fat such as...
rice cakes, lean meat, baked beans, plain spaghetti and rice and egg whites.

**How should the enzymes be stored?**
- Store enzymes in the fridge.
- Enzymes will deteriorate if kept in a hot place for example; above your fridge or in the car.
- Top up enzyme carriers daily to avoid them deteriorating in hot weather.
- Check the expiry date of enzymes; use them while they still have a good shelf life (e.g. 6 months minimum).
- Always keep the lid tightly sealed on the bottle of enzymes.
- Discard enzymes that are out of date, or if they have changed colour or texture or have been exposed to hot temperatures.

**Do not** put enzyme beads in bottles or cups, or down nasogastric tubes or gastrostomies.

My usual enzyme dose is ____________ for every_________ g dietary fat

**Things I can do to improve managing my PERT:**
1. 
2. 
3. 
4. 

*For further information contact your Dietitian or Cystic Fibrosis Centre*