Overview of Pancreatic Enzyme Therapy for Cystic Fibrosis (CF)/Pancreatic Insufficiency (PI)

Determining the need for enzymes is done with:
- Fecal Elastase (< 100 indicates PI)
- 72 Hour Fecal Fat with Diet Diary
- CF Gene mutations consistent with pancreatic insufficiency (PI)
  - ΔF 508 – most common CF gene associated with pancreatic insufficiency
  - R117H – most common gene associated with pancreatic sufficiency
- Pancreatic Stimulation Testing

Dosing enzymes for infants
- Enzymes need to be given with breast milk and all types of infant formula
- Open the capsule and sprinkle the “beads” on a small (1/2-1) teaspoon of applesauce
- Give the applesauce and “beads” by spoon at the beginning of the feeding
- Wipe away any remaining “beads” that are noticed at the end of the feeding on the child’s mouth, skin or mother’s breast
- Do not crush or try to dissolve enzymes – the coating on the enzymes protects the enzyme from stomach acid
- Repeat dose of enzymes if next feeding is greater than 45 minutes from last enzyme administration
- Use barrier ointments (Bag Balm, Triple Paste, Desitin) when enzyme therapy is started to prevent diaper rash
- NICU babies with pancreatic insufficiency who have been on TPN usually start enzymes when the oral intake is ~ 20-30 ml Q 3 hrs

Enzyme dosing for children and adults
- Give enzymes with each meal and snack that contains fat and/or protein
- Give at the beginning of meal or snack
- If large numbers of capsules, divide between beginning, middle and end of meal
- Enzymes are effective for 45 minutes
- Repeat dose if meal or snack longer than 45 minutes

How much enzyme is needed?
- Starting dose for CF is usually 1000-2000 units lipase/kg/meal
- CF Foundation recommends a dose no higher than 2,500 units lipase/kg/meal or no greater than 10,000 units lipase/kg/day

Calculating Enzyme Dosage
- Each capsule of enzyme contains the amount of lipase listed in the enzyme name:
  For Example: Creon 6,000 contains 6,000 units of lipase per capsule

Increasing Enzymes
- Enzymes need periodic adjustment
  - During times of rapid growth
• Infancy
  • Adolescence
    – Development of malabsorption symptoms
    – Poor weight gain
    – Lower levels of absorption based on 72 hour fecal fat

Symptoms of Fat Malabsorption
• Large, bulky BMs
• Loose BMs
• Foul-smelling BMs
• Mucous or oily in BMS
• Excessive Gas and/or stomach pain
• Distention or bloating
• Poor weight gain despite good (sometimes ravenous) appetite

Note: Diarrhea is NOT on this list

Enzyme Therapy for Tube Feedings
• The “beads/microspheres inside a Creon or Zenpep may be crushed.
  • Families should purchase a mortar and pestle to grind the beads.
    Generally, a pill crusher will not work, especially with Creon.
    • For inpatient use, the mortar and pestle is accessed through the
      Med Select system (on W11) or by contacting an inpatient
      pharmacist. At discharge, the mortar and pestle will be sent home
      with the patient.
    • The beads are crushed using a circular, rotating movement of the pestle.
      All beads must be flattened or pulverized into a fine powder. Avoid
      breathing in the dust from the crushing process.
    • If a family has difficulty crushing Creon, replace tube feeding enzymes
      with Zenpep in a similar dose. Zenpep crushes much easier.
  • This dosing of crushed Creon or Zenpep may be used as an approximate starting
    point for tube feedings:

<table>
<thead>
<tr>
<th>Caloric Density of Formula</th>
<th>Amount of Crushed Enzyme per 240 mL</th>
</tr>
</thead>
<tbody>
<tr>
<td>Infant formula 20-27 Cal/oz</td>
<td>1 Creon 6,000 or 1 Zenpep 5,000</td>
</tr>
<tr>
<td>1.0 Cal/mL pediatric or adult formula</td>
<td>1 Creon 12,000 or 1 Zenpep 10,000</td>
</tr>
<tr>
<td>1.5 Cal/mL pediatric or adult formula</td>
<td>1 ½ Creon 12,000 or 3 Creon 6,000 or 1 Zenpep 15,000</td>
</tr>
<tr>
<td>2.0 Cal/mL adult formula</td>
<td>1 Creon 24,000 or 1 Zenpep 20,000 or 25,000</td>
</tr>
</tbody>
</table>

  • Mix the crushed enzymes into the entire amount of formula used.
  • Allow the enzyme/formula mixture to sit for 15-20 minutes before starting the
    feeding to allow the enzymes to digest the formula.
  • Hang only enough enzyme/formula mixture for a 4 hour infusion in the hospital
    environment. Refrigerate the remainder, and refill as needed to complete the
feeding. Formula and crushed enzymes can be premixed and stored in the refrigerator for up to 24 hours.

- Partially or predigested formulas may require less crushed enzymes
- If malabsorption is present, a partial dose of enteric coated enzymes may be given at the beginning of the tube feeding to optimize digestion of the first 30-60 minutes while the crushed enzymes are “digesting” the formula
- It is important that ALL formula be infused for maximum Calorie/nutrient intake

Testing to Determine Efficacy

- 72 Hour fecal fat analysis with a corresponding diet diary is the gold standard for assessing fat absorption
- To Calculate:
  \[
  \text{Fat grams in (diet) - Fat grams out (stool)} = \text{Fat absorbed}
  \]

  \[
  \text{Fat grams in (diet)}
  \]

  \[
  \text{Goal for absorption} = > 90\% \text{ in CF/malabsorption}
  \]

  \[
  \text{Healthy normal is 93-100\% absorption}
  \]

Foods that don’t require Pancreatic Enzymes (however, these food/drinks are generally not going to promote wt. gain.)

- Fruits
- Juice, juice drinks
- Plain vegetables
- Soda or sport drinks
- Pedialyte
- Tea, coffee (without cream)
- Candy without chocolate or nuts
- Fruit snacks
- Popsicles, freezer pops, flavored ice

Nutrition Therapy for Pancreatic Insufficiency

- High Calorie, high protein
- >40\% of Calories from fat
- Supplement with fat soluble vitamins
- General Calorie goals are DRI x active / very active
- Protein needs are generally 2 gm/kg

Vitamin Therapy for Fat Malabsorption

- Fat soluble vitamin deficiency associated with fat malabsorption
- CF specific vitamin supplements designed to provide higher A,D, E, & K levels
  - Drops (AquADEK drops used at CHW)
  - Chewable tablets (Complete Formulation bubblegum chewable used at CHW)
  - Softgels (Complete Formulation D3000 softgels used at CHW)