

## CF Specific Patient Handouts

Rich Carroll, Pharm.D.  
Clinical Pharmacist  
Children's Memorial Hospital  
Chicago, Illinois 60614



## Patient information comes from several sources

- ✘ Leaflet (handout) from hospital pharmacy
- ✘ Leaflet from mail-order pharmacy
- ✘ Leaflet from local pharmacy
- ✘ Other sources, i.e. internet



## Information to help patient understand medication

- ✘ How to administer
- ✘ What adverse effects to watch for
- ✘ What to do if a dose is missed
- ✘ When to call your physician
- ✘ Other information to be helpful



## Leaflets are vague

- ✘ Most handouts are designed for one handout per drug
- ✘ Not related to age of patient
- ✘ Not related to disease specific issues
- ✘ Sometimes seem to be too bland, perhaps for legal concerns



## We felt the need to develop CF specific handouts

- ✘ Allows patients and caregivers to feel like the information is more carefully tailored to their situation
- ✘ Avoid the impression that these are routine medications for a routine purpose
- ✘ Give the patient much more appropriate information



## We need to pass some specific, needed information

- ✘ Use the CF leaflet to "add to" the standard leaflet
- ✘ Let the standard leaflet be the basic information
- ✘ The CF specific handout should supersede the standard information.



## Some handouts are misleading

- ❖ Calcium is used as a source of the mineral calcium, not primarily as antacid
- ❖ Antibiotics are routinely labelled as take until finished, which may not be appropriate in CF, certainly not if the antibiotic is used for prophylaxis
- ❖ Many drugs may have handouts that state "this drug is not recommended for use in pregnancy."

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## Some handouts make illegal or improper suggestions

- ❖ FIRST DATA BANK is the largest provider on online, integrated patient handouts
- ❖ As of 10/13/04, First Data Bank was suggesting that the pharmacist may be able to offer a generic substitute for pancreatic enzymes.
- ❖ The field is an option that would have to be manually deleted, and would be deleted for all handouts; most pharmacies are not aware of this option.

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## Some handouts do not specify storage conditions as completely as needed

- ❖ Pulmozyme® will routinely say "store in a refrigerator"
- ❖ It will not likely say that the foil pouches may be stored at room temperature up to 77 degrees F for up to 24 hours.
- ❖ If it is unrefrigerated for more than 24 hours (such as during shipping), it should be discarded.

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## Differences in Storage Conditions

- ❖ Tobii® nebs will come with instructions that it should be refrigerated
- ❖ Tobii® can be stored in or out of their foil pouches at room temperature (77 degrees F) for up to 28 days.

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## Dose schedules

- ❖ PI leaflets often do not mention or stress the correct importance of what to do about missed doses, late doses.
- ❖ PI leaflets seldom have the correct information for a patient who is taking prophylactic antibiotics only on some days of the week.

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## Drug Use

- ❖ Seldom will the PI leaflet describe the use of the drug for a CF patient.
- ❖ Usually, only common uses for drugs are mentioned, and in very general terms.

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## Enzymes - dosing

- ❖ No current PI leaflets discuss the multiple needs of CF patients
- ❖ No allowance is made for varying dose regimens, such as "3 with meals, 2 with snacks, one at bedtime"
- ❖ No allowance is made for dietary variations

## Enzymes - Substitution

- ❖ We need to make the CF patient and caregiver aware of the problem
- ❖ We need to arm the patient with information about what to do if a pharmacist wants to substitute a different product

## Tobi® Substitution

- ❖ Patient may need to be aware that there are three separate products
- ❖ Should know which ones contain preservatives, which have longer stability
- ❖ Should know how each is packaged, and which they prefer and why

## Legal Issues

- ❖ Involve everyone, CF director, CF physicians, Nurses
- ❖ Make it become your standard of practice
- ❖ Some national momentum for this practice
- ❖ National acceptance

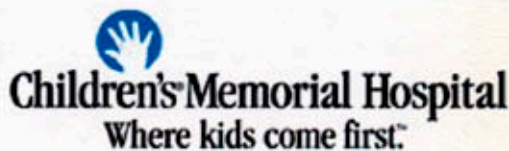
## Disclaimer

With your prescription for pancreatic enzymes, your pharmacy may provide you with an information sheet, which may not be optimized for use in Cystic Fibrosis. Cystic Fibrosis patients will need some additional information. This handout is intended as a supplement to the standard handout.

## Suggested – Two Columns

- ❖ **Generic Substitution**
  - Your handout now tell you that a generic drug is available at a cost saving.
  - Generic enzymes are not made to the same standards as most generic drugs. Tell your pharmacist that you do not want a generic form of enzymes. You may refer to: <http://www.cfdp.org/pancrelipan.htm> and <http://www.cfdp.org/veloxin.htm>
  - The CF patient and their caregiver need to be an advocate for obtaining non-generic enzymes. Wide variations in product composition, formulation, and manufacturing have been identified between current suppliers. Variations from product to product may affect the safety and efficacy of these medications. As of April 20, 2014, all manufacturers of enzyme products must transmit a new drug application to the FDA.
  - Please do not simply tell your CF physician that you are changing brands or using a generic. Please discuss this matter with your CF physician before changing sources.

# Pulmozyme<sup>®</sup> Nebs (dnase, dornase alpha)



With your prescription for dnase nebs, your pharmacy may provide you with an information sheet, which may not be optimized for use in Cystic Fibrosis. Cystic Fibrosis patients will need some additional information. This handout is intended as a supplement to the standard handout.

The standard handout may say:	CF specific information
<p><b>Use:</b> Your patient handout may say that these inhalations are used to reduce the thick mucous in the lungs.</p>	<p>In your case, these nebulizations are very important to break up the thick, sticky mucous in the CF lung. It is imperative that this be nebulized as directed by your physician or respiratory therapist. Do not mix with Tobi (Tobramycin) in the same inhaler</p>
<p><b>Precautions:</b> Your medication sheet may say that you should only take if clearly needed during pregnancy. It may also say that this drug is not recommended for use in pregnancy.</p> <p>It may say that breast feeding your infant while using Tobi is not recommended</p>	<p>Obviously, CF patients should make their CF physician aware of their pregnancy, but most will continue the use of dnase inhalations</p> <p>Discuss with your CF physician if you plan to breast feed your infant.</p>
<p><b>Missed Doses</b> Take any missed dose as soon as possible, but not if it is almost time for the next dose. If it is time for the next dose, skip the missed dose and resume your regular schedule</p>	<p>If you missed a dose, take the dose as soon as possible. If you miss more than one dose in a row, or miss several doses in a week, please consult with your CF physician. Do not "double-up" on doses to catch up.</p>
<p><b>Storage:</b> Store in the refrigerator</p>	<p>Store in the refrigerator between 36 and 46 degrees F. away from light and moisture. However, if necessary (e.g. transporting this drug) you may store the foil pouches (opened or unopened) at room temperature up to 77 degrees F for up to 24 hours. Do not freeze.</p>

## **You and Your CF Meds: What your retail pharmacist may not be telling you?**

### **NUTRITION RELATED MEDICATIONS**

Emily Griffith RD, LD

Children's Memorial Hospital

#### **A. Pancreatic Enzymes**

1. Why do many CF patients need pancreatic enzymes?
  - a. Background-the pancreas functions to release "enzymes" during digestion to help the body to break down and absorb food.
  - b. Pancreatic insufficiency affects 85-90% of patients with CF. With pancreatic insufficiency, the ducts of the pancreas become blocked by thick/viscous mucus.
  - c. If the pancreas is unable to release sufficient digestive enzymes, the body is unable to absorb food adequately. Thus causing GI discomfort resulting in nutrient deficiencies and poor weight gain and growth.
2. How do they work?
  - a. Non-enteric coated-still available, however rarely used.
  - b. Enteric-coated enzymes-are protected by a special coating that prevents activation of the enzymes until they reach a nonacidic environment. Therefore they can bypass the stomach and not be activated until they reach the small bowel.
3. When should they be taken?
  - a. Enzymes should be taken before all meals, snacks, and tubefeedings that contain fats or proteins. (including beverages)  
They work for 45min to 1hr.
4. How should they be taken?
  - a. Infants and small children-if unable to swallow pills, the enzyme capsules should be opened and the "beads" or "tablets" put in a small amount of an acidic food such as applesauce or pears (not with nonacidic foods such as yogurt or pudding) and feed it to the child by spoon.
    - Enzymes can be inactivated with crushing or chewing.
    - It is important to make sure there are no residual beads in the mouth because they can start to "break-down" the child's mouth tissue.
    - Enzymes should not be "pre-loaded."
  - b. Children and adults-should swallow capsules with a liquid prior to eating.
  - c. Split dosing
  - d. Tube feedings-taken prior to feeds.

4. How should pancreatic enzymes be stored?
  - a. Enzymes should be stored at room temperature. (not stored in refrigerator or left in a hot area)
  - b. Should be used within their expiration date.
5. Why are brand name enzymes important?
  - a. Products will be required to have formal FDA approval by 2008, but until then, it is recommended to use the brand name enzymes such as Creon, Pancrease, Pancreacarb, and Ultrase.

## B. Vitamins

1. Why are they needed?
  - a. Fat-soluble vitamins (vitamins A, D, E, and K) are dissolved in fat and absorbed; CF patients with pancreatic insufficiency often need additional fat-soluble vitamins to meet their needs.
2. How are vitamins administered?
  - a. Available in liquid (for infants and small children), chewable tablets, and capsules.
  - b. For best absorption, vitamins should be taken with a meal and enzymes.
3. How should they be stored?
  - a. Vitamins should be stored at room temperature

## C. Histamine 2 blockers and proton pump inhibitors

1. What are they?
  - a. Medications that help to suppress gastric acid production in the stomach.
2. Why are they needed?
  - a. To help prevent reflux
  - b. To suppress acid production in the stomach to help active enzymes.
3. How are they administered?
  - a. Available in oral suspensions, granules, capsules, tablets (dissolve)
  - b. Labels stated they should be taken on an empty stomach, however not imperative.
4. How should they be stored?
  - a. Room temperature; in a dry, cool place.