



Exciting New Vest Therapy Adherence Research

Airway clearance techniques, including high-frequency chest wall oscillation devices (vest therapy), serve as a central treatment approach for patients with CF. However, providers have had to rely on patient and parent self-reporting to determine adherence to this important therapy.

Currently there is not a reliable objective method for measuring a patient's adherence to vest therapy. This is limiting to both clinicians seeking to provide optimal patient care and researchers seeking to establish evidence to support specific therapeutic approaches.

This project aims to leverage new technology developed by Hill-Rom, Inc. to remotely collect usage data directly from Bluetooth-enabled vest systems. We are attempting to enroll 100 participants with CF aged 2-22 years and collect vest usage data over a 12-month study period. Participants will not be asked to come to the clinic for extra visits or to change their care or behavior in any way. Families will trade in their current Hill-Rom vest systems for a new vest system provided by Hill-Rom.

We hope all eligible families will consider participating in the study whether they currently use their vest therapy routinely or not. Our goal is to get a realistic snapshot of how families are using vest therapy and begin to examine the impact it may have on our patients' health.

Hill-Rom vest therapy adherence research is taking place at Children's Minnesota.

If you would like to learn more about the Vest Adherence Study, please contact Christine Benoit at 651-220-6254 or christine.benoit@childrensmn.org

The CF Breeze will be distributed electronically for patients and families of Children's Hospital and CRCCS.

To be added to the distribution list or to update your e-mail address, please complete a listserv form available in the lobby of CRCCS or by contacting Mary Sachs or Sandy Landvik.

Upcoming Events

CF Community Parent Support Group meets from 7 p.m. to 8:30 p.m. at Children's Minneapolis.

- Monday, May 23
- Monday, July 25
- Monday, September 26

For CF Foundation activities, please visit:

www.cff.org/Chapters/minnesota



Having Siblings with CF

by Alexis (16)

I have two siblings with cystic fibrosis. Their names are Easton(3) and Edith(18 months). Since I've been helping with them for almost four years now I've gotten used to it but in the beginning it wasn't so easy. It was hard to wrap my head around it at first. There were a lot of questions but once we were educated on what it was and the treatments we had to do, it got easier. Their lives may not be as long but they will be just as amazing. I can't wait to see what comes next.



Hospitalizations

Will my child have to be hospitalized for CF?

Anytime that your child with CF is hospitalized it can be a stressful time for you and your family. Many families do not know what to expect the first time or even reasons why a hospitalization may occur. Here are a few of the more common reasons:



| Common Reasons | Symptoms |
|-------------------------------|--|
| Pulmonary exacerbation | Hospitalization is often indicated when outpatient treatment with an inhaled or oral antibiotic has not been effective in improving their pulmonary function or respiratory symptoms. Other symptoms your child may have include a decreased appetite or weight loss, a decline in lung function of 10% or more, cough that is increased from baseline, an increase or new onset of sputum production, or fatigue. |
| Bowel obstruction | This a rare but not uncommon complication of CF typically treated with Miralax, stool softeners, or enemas. The obstruction is often diagnosed by an abdominal x-ray. |
| Sinus infection | May require hospitalization and surgery if symptoms not relieved by an antibiotic treatment course at home. Nasal polyps can also occur and may require surgery. |
| Inadequate weight gain | On rare occasions, an admission may be required to evaluate ability to gain weight if all outpatient strategies have been unsuccessful. |

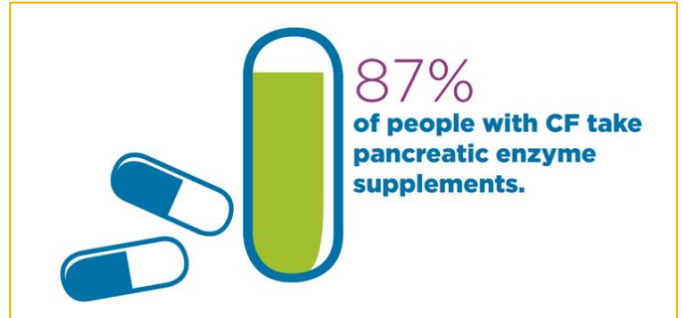


Medication 101

Pancreatic Enzyme Replacement Therapy (PERT)

What is pancreatic insufficiency?

Pancreatic insufficiency that requires PERT occurs in between 80-90% of all patients with Cystic Fibrosis. It results from CF transmembrane conductance regulator (CFTR) dysfunction in the pancreas that leads to inadequate secretion of pancreatic enzymes into the small intestine. Pancreatic enzymes are substances released by the pancreas into the small intestine that help to break down protein, carbohydrate and fat into smaller more easily absorbable forms. Without enough pancreatic enzymes in the small intestine, malabsorption of nutrients will occur and can lead to gastrointestinal discomfort, frequent stooling, large oily or foul smelling stools and nutrient deficiencies.



How does pancreatic enzyme replacement therapy work?

PERT is prescribed for people with CF who have pancreatic insufficiency in order to make up for the enzymes that are not released by the pancreas. Taking enzyme capsules supplies the small intestine with the pancreatic enzymes needed to break down food and nutrients into small enough forms that they are able to be absorbed.

When and how are pancreatic enzymes taken?

It is recommended that enzymes be taken before eating all meals and snacks. For infants and children who are not yet swallowing enzyme capsules, enzymes should be given in a small amount of acidic pureed baby foods (applesauce) prior to eating. Patients who receive supplemental nutrition via feeding tube should work with the CF team to determine the best enzyme plan to cover the nutrients provided by their tube feedings. Taking enzymes before eating and drinking helps to ensure that the enzymes needed to help digest food are present in the small intestine when the food gets there rather than trailing behind the food if enzymes were taken after eating.

How is the brand of enzyme prescribed for me or my child determined?

All pancreatic enzymes must be approved by the Food and Drug Administration prior to being used by patients with CF. There are six formulations that have been approved to date. They are: Creon, Pancreaze, Pertzye, Ultresa, Viokace and Zenpep. Your pulmonology provider will work with you to determine which formulation is best for you/your child based on individual circumstances.



Amped Up Nachos

As winter is drawing to a close and we are starting to get small glimpses of spring, we are getting excited for foods that remind us of warm breezy days and sunshine. A quick and easy meal for spring and summer is nachos. Nachos provide an excellent canvas for including fun ingredients that offer extra bang for their buck in the calorie and protein department.



- Try using **tortilla chips** with a hint of lime or other citrus flavoring as this creates a mouth-watering sensation that will get kids craving more. For fun in the kitchen, consider making your own tortilla chips by first cutting fresh corn tortillas into 6 equal sized wedges. Next, pour canola oil into frying pan to a depth of 1/8-1/4". Heat to 350° F. Fry tortillas for about 2 minutes until crisp and slightly darkened in color. Remove from pan with metal tongs and place on a plate covered with paper towel, add salt as desired and allow the chips to dry. Squeeze lemon or lime juice over chips if preferred.
- **Shredded cheese** – 110 calories and 6 grams protein per ¼ cup. Try melting it on for an extra gooey delight. Choose bold varieties such as pepper jack or sharp cheddar to make the flavor more exciting.
- **Guacamole** – 60 calories and 1 gram protein per 2 Tbsp. Using guacamole is an easy way to introduce kids to avocado which provides heart healthy, anti-inflammatory, monounsaturated fat.
- **Sour Cream** – 60 calories and 1 gram protein per 2 Tbsp. Try adding taco seasoning to sour cream for an extra punch of flavor and sodium.
- **Plain Greek Yogurt** – 15 calories and 2.8 grams protein per 2 Tbsp. Greek yogurt offers approximately twice the protein as regular yogurt and can be used in many recipes to increase protein content of foods.
- **Refried/black beans** – 120 calories and 7 grams protein per ½ cup. Adds interesting texture and is rich in fiber and protein.
- **Chorizo sausage** – 250 calories and 14 grams protein per 2 ounces. Cut sausage into disks or remove meat from casing to make sausage crumbles for interesting flavor and a new take on ground meat for nachos.
- **Barbecue pulled pork or chicken** – 70-90 calories and 6-9 grams protein per 2 ounces. Spring and summer are a great time to start breaking out the barbecue flavor and the addition of barbecue sauce provides extra sodium to meet increased sodium needs for CF.

No matter how you make them we hope you enjoy this easy and delicious meal!
Buen Provecho