COOK CHILDREN'S
CYSTIC FIBROSIS CENTER

FORT WORTH, TX.

COOK CHILDREN'S CYSTIC FIBROSIS CENTER NEWS

VOLUME VI

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The Cystic Fibrosis Center at Cook Children's Medical Center is one of more than 115 accredited CF centers throughout the nation. These centers are accredited by the Cystic Fibrosis Foundation, a non-profit organization founded in 1955 dedicated to funding research to find a cure for CF and improving the quality of life for people with the disease.

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Stay Healthy This Flu Season

Here are some helpful ways you can stay "BUG" free.

1) Wash your hands-this is the single most important method of reducing your risk for the FLU.

2) Get the Flu Vaccine

3) Be aware of your surroundings including the people that are near you who may be ill with the flu at home or in the office or anywhere around you

4) Stay on top of doing all your nebulizer breathing treatments and airway clearance—the better you breathe with less mucous build up the better off you will be this flu season

5) Avoid time spent in crowded areas (malls, movie theaters, etc) during peak flu season (October-March)

6) If you start feeling like your symptoms have increased (sputum production, cough, fever, shortness of breath, chest congestion) please call the center for further evaluation.

This is a reminder that all CF patients are highly recommended to receive the FLU VACCINE. Please be sure contact us if you have already received this vaccine via phone. If you have NOT received the flu vaccine please schedule an appointment any day of the week.

** It is important that CF patients as well as family members also receive their flu vaccinations.

The **CF**Chef program is an online nutrition resource that allows those touched by cystic fibrosis to share and receive support, recipes and meal tips.

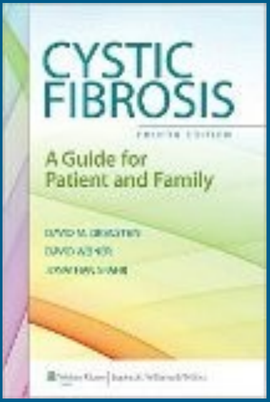
People who have cystic fibrosis (CF) usually require calorie intake that is greater than what is recommended for the general population. This is because CF makes it hard for people to properly digest food, causing malabsorption of the calories and nutrients (vitamins and minerals) in food. Malabsorption can cause problems such as stomach pain, frequent bowel movements and poor growth.

People who have CF are advised to work with their Registered Dietitian (RD) and CF Center team member to develop a suitable nutrition and meal plan. The CFChef program offers a CF nutrition guide, online cookbook, Q&A with CF Nutrition Expert, and opportunity to share your recipes.

Do you have the new version?

Cystic Fibrosis: A Guide for Patient and Family

David Orenstein, MD has released the newest version of his best selling book, Cystic Fibrosis: A Guide for Patient and Family. This new version was just released on 8-10-11. The book is a guide to informed and compassionate care– designed specifically for patients with CF and their families,. Check it out!



Therapy YOUR Way

Personalize your Vest and Airway Clearance System with these fun and unique designs!

The Vest Skins are designed to perfectly fit the unique contour of your device. The high quality prints are easy to put on and remove.

Choose from hundreds of designs or download your own photo to make The Vest® Airway Clearance System uniquely your own!

Transform your ordinary device and **Make it Yours!**

It you would like some more information, visit the official website:

www.thevest.com

The MOBILAIRE 50psi

Cook Children's Home Health Care now carries this heavy duty compressor for your nebulizer treatments. The MOBILAIRE 50psi is the ULTIMATE compressor is designed to provide you with durability (5 year warranty), effective air flow, and efficient time delivery. In fact, when studies were made looking at the delivery of TOBI and Pulmozyme they found that good drug delivery into the small airways of your lungs were best achieved at a psi of 30. This new Mobil air will allow for this effective air flow of at least 30 psi to get the medication where it needs to go and allow for these meds to work better for you.

Many of you have asked for the wonder machine that would deliver your meds in a timely manner. This is it. Medication delivery time with the Mobil air is as follows (Psi should be dialed between 30-40):

Albuterol/Xopenex ---8 minutes

Pulmozyme---8 minutes

Hypersal---8 minutes

Tobi (made by Novaritis—NOT the premade tobramycin)---11 minutes

Colistin---11 minutes

But note that MobilAire is not easily portable, as it weighs about 11 pounds. Your Pari or even Pari Trek are much easier to travel with.

The eflow device was not created to be utilized with albuterol, xopenex, pulmozyme, hypersal and TOBI. The altera device CAN only be used with Cayston. Though there is a temptation to use these devices for other meds because it saves so much time, it does not guarantee proper delivery of these medications to the airways.

Cost: The total cost for this device is $240. It is covered by most private insurances (PPO/HMO) and under Medi-care. It is not covered by GHPP or MediCAL. However, patients may purchase this device with a credit card in full or in a payment plan if you are not financially able to pay it all at once. Please feel free to ask your pulmonologist or respiratory therapist about this device at your next clinic visit.

Be sure to replace filters and clean as needed.

MOBIL AIR COMPRESSOR SETTINGS

Use As Directed:

Medication Name Mobil Air PSI Setting TOBI Less than 30 PSI

Colistin 30 PSI

Pulmozyme 30 PSI

Albuterol/Xopenex 35-37 PSI

Hypersal 35-37PSI

Pill Swallowing Tips

It is common for children to have difficulty swallowing tablets and capsules, but children over 5 years old can usually master this skill with a little practice. Teaching your child the technique of pill swallowing requires patience, so set aside a time when you won't be disturbed and when your child is calm and receptive. Work in short intervals. Sit down at a table with your child and explain that you are going to help him learn a new skill. First, check your child's swallowing reflex by asking him to take a mouthful of water and swallow it. If no water dribbles out of his mouth, your child is ready to start learning to swallow pills. (If your child has trouble swallowing water consult his pediatrician or speech therapist.) If your child has nasal congestion, have him blow his nose or use saline drops before attempting to swallow the medication.

The simplest way to teach your child to swallow pills is to **practice swallowing** candy cake decorations as pill substitutes. These decorations are available in the baking department of most grocery stores. Buy about 5 types, from tiny round sprinkles to large silver spheres so that you have " pills" of gradually increasing size. Also purchase some small candies such as tic- tacs or mini m&ms.

Once your child has swallowed water successfully, you can move on to swallowing candy sprinkles. Demonstrate for your child before he tries. (If you find it difficult to swallow pills ask someone else to teach your child!)

- Place the smallest candy sprinkle on the middle of the tongue.
- Take a good sip of water.
- Keep the head level (don't tip the head back).
- Swallow the water (and the pill).
- Have another sip of water to keep the " pill" moving.

If the pill doesn't go down with the first swallow, just say, "keep drinking" and it will probably wash down with the next gulp. Let your child try as many times as he needs to until he can swallow this tiny sprinkle every time he tries. If he struggles, go back to just swallowing water, praise him for this, and calmly suggest that you will try again another time.

When your child has mastered swallowing the first size, move on to the next (don't say bigger) size and so on. If your child is unsuccessful twice with the next size, let him return to the previous size " pill" before ending the session. This ensures that he ends the practice session with success. Limit each practice session to a few minutes or less as tolerated.

At the next session, start with the smallest sprinkle size and ask your child to swallow each size 5 times before moving to the next. When your child can reliably swallow the tic-tacs or m&ms, ask him to try swallowing an actual pill. Children need regular practice in order to maintain this new skill, so daily practice is important. Some children will need 6 or more sessions in order to master swallowing pills.

If the above method doesn't work for your child there are other techniques that you can try:

- Put the pill **under** the tongue and take big gulps of water. This will usually wash the pill out from under the tongue and down the throat.

- Place the pill on the **middle** of the tongue and fill the mouth with water until the cheeks are full, then swallow the water. The pill should slip down too .

- Put the pill right at the **back** of the tongue rather than in the middle.

- Have a **few sips of water before trying to swallow** the pill, this should help the pill to slip down more easily.

- Put the pill on the tongue then ask your child to take **3 gulps of water using a straw**. When he swallows the water he will probably swallow the pill too.

- Have your child try swallowing pills **standing up** rather than sitting down.

- Try the **pop-bottle method** (This method reduces the tendency to gag on the pill.) Place the tablet anywhere in the mouth. Take a drink from a soda-pop bottle, keeping contact between the bottle and the lips by pursing the lips and using a sucking motion. Swallow the water and the pill.

- Try the **two-gulp method** (This method helps to fold down the epiglottis (the flap of cartilage at the back of the throat that folds down and protects the airway during swallowing.) Place the pill on the tongue. Take one gulp of water and swallow it, but not the pill. Immediately take a second gulp of water and swallow the pill and the water together.

- If your child's medication is in capsule form, try the **lean-forward technique**. Capsules are lighter than tablets and have the tendency to float forwards in the mouth during swallowing. Leaning the head slightly forward while swallowing causes the capsule to move towards the back of the mouth where it more easily swallowed.

- You could give your child **different liquids** such as milkshake or yogurt drinks to take the pills with. Thicker drinks slow down swallowing and make the pill less likely to separate from the liquid. Some children can swallow pills in spoonfuls of peanut butter, applesauce, pudding or jello. Pills can also be tucked inside mandarin orange segments, and the segments can then be swallowed whole. Chewing a cookie or some crackers and popping the pill in the mouth just before swallowing can also be effective. **Always check with your physician or pharmacist before your child takes his medication with anything other than water in order to avoid a medication interaction with food.**

- If your child isn't ready to learn how to swallow pills explore alternative forms of the medication. Many medications come in liquid, sprinkle or chewable forms and some can be crushed or dissolved. **Never crush, break or dissolve tablets or capsules unless your doctor or pharmacist has advised you to.** Some specialized pharmacies can make up an elixir that contains a palatable tasting liquid containing the required medication if your child cannot swallow pills or capsules.

If swallowing pills becomes essential, e.g. a condition for entering a research study or if the pill only comes in pill form and cannot be cut or crushed, ask for a referral to a **therapist** who has experience teaching children how to swallow medication. Your child may learn this new skill more easily from a neutral figure than from a parent.

Be sure to reward your child's efforts with praise even if he is not successful at each try The goal is to help your child succeed with a variety of techniques that will make taking daily routine medication less of a challenge for you both.

Pancreatic Insufficiency Patient Care Programs:

CFCareForward and ZPoints

CREON and ZENPEP are two types of digestive enzymes used to treat the pancreatic insufficiency in CF patients. Did you know the manufacturing companies offer patient care programs to provide you with free nutritional support for people with cystic fibrosis.

CFCareForward

Abbott Laboratories, the marketer of FDA-approved CREON®, has offered a variety of resources to support the CF community. **CF**CareForward provides nutritional support with free multivitamins in softgel, chewable, and liquid formulations plus a variety of nutritional drink choices (some restrictions apply*).

Ex: Vitamax Chewable Tablets or Liquid Drops
SourceCF softgels
Pediasure/Ensure/Ensure Enlive!/Ensure Plus

www.cfcareforward.com

ZPoints

The ZPoints Program is designed for patients with exocrine pancreatic insufficiency due to cystic fibrosis who have been prescribed ZENPEP. With each prescription for a 1 month supply, eligible patients will earn points that can be applied toward products they use daily.

Ex: Source CF chewable multivitamins/liquid drops/softgels
NUTRA/Balance Protein Fortified Cookies
NUTRA/Balance NUTRA/SHAKE
SCANDISHAKE Calorie Rich Shake Mix
AquADEKs Vitamins

ZPoints

www.zenpep.com

Please be sure to speak with your dietician to learn about how you are eligible to sign up for these special benefits.

You will have choices in mileage varying from 15 miles to 40 miles. Cycle at your own pace.

The Ride for 65 Roses Cycle for Life is fully supported with well-stocked rest stops, bike mechanics, ride marshals, lunch, and so much more.

The Aptalis cf Cycle FOR LIFE®

Wheels in Motion to Cure Cystic Fibrosis

Join us for our inaugural Ride for 65 Roses Cycle for Life bike tour

November 12, 2011

Walnut Creek Country Club

Mansfield, Texas

Register now at: http://FortWorth.CFF.org/CycleforLife

Living with cystic fibrosis means making time to fit your treatments into your busy schedule. Now on [CFLiving](www.cfliving.com), you can watch a video short in which CF patients, experts and caregivers discuss the importance of establishing a routine and understanding how your daily treatments work to help keep you healthy

www.cfliving.com

Click on Videos/Webcasts

How To Encourage Kids To Stick To Their CF Treatments

Videos and Webcasts

Cystic Fibrosis Video Short: Motivating Adherence

How to Encourage Kids to Stick to Their CF Treatments

We know that living with CF means fitting time-consuming treatments into your busy schedule. In this second video in this series, parents of CF kids discuss how they motivate their children with the power of choice. Plus, CF patients share how they manage to stay on top of treatments. Before the video begins, please answer two quick questions that will appear on your screen — and we would love your feedback afterwards with a brief survey.

Cystic Fibrosis Related Diabetes (CFRD)

People with cystic fibrosis (CF) are at risk for developing cystic fibrosis-related diabetes (CFRD). CFRD can be confusing and frightening when you don't have all the facts. We hope this article helps answer your questions and ease your fears.

What is CFRD and how is it different from Type 1 and Type 2 diabetes?

CFRD is a type of diabetes that shares features of both Type 1 and Type 2 diabetes, but is unique to people living with CF. As in Type 1 diabetes, the body does not make enough insulin to keep blood sugars at a healthy level. Thus, people with CFRD, like those with Type 1 diabetes, typically require insulin to manage blood sugars. As in Type 2 diabetes, the body may not respond normally to the insulin that the body does produce so many people with CFRD are also insulin-resistant. Unlike Type 2 diabetes, diet and exercise alone are not recommended to manage blood sugars.

Fact:
CFRD is one of the most common health issues in people with CF. It occurs in 20% of adolescents and 40-50% of adults.

What causes CFRD?

CFRD may be caused by any *one* or a *combination* of the following:

The body does not make enough insulin because the pancreas (the organ that makes insulin) becomes scarred over time by thick mucus within the body.

The body does not respond to insulin normally and insulin resistance develops.

The body produces too much cortisol, a steroid made when the body is under stress that can interfere with insulin's action. Steroid containing drugs (corticosteroids) can also have this effect.

Fact: CFRD cannot be prevented or avoided. Limiting the amount of sugars or carbohydrates in your diet is NOT recommended.

What are the symptoms of CFRD?

1. Weight loss or struggles to gain weight
2. Loss of lung function (FEV_1%)
3. Being very tired
4. Increase thirst

How is CFRD diagnosed?

CFRD is best diagnosed by an Oral Glucose Tolerance Test (OGTT). OGTTs are typically scheduled in the morning because it is important that the test be obtained fasting – nothing to eat or drink other than water for at least 8 hours prior. The test takes approximately 2 hours. During this time, your/your child's blood sugar will be measured two times – once before drinking a glucose solution (fasting) and again 2 hours after drinking the sweet drink. A diagnosis of CFRD is made if:

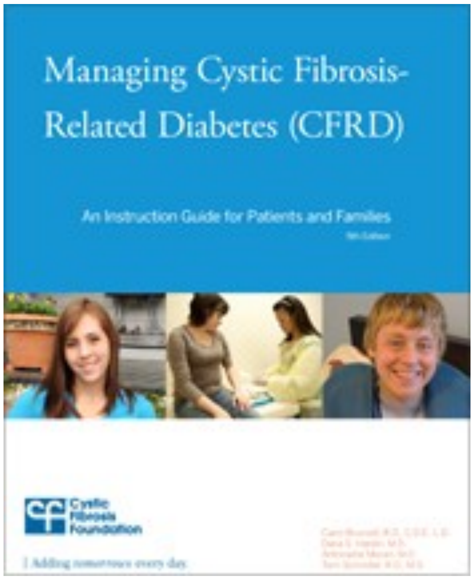
Fasting blood sugar is 126 mg/dL or greater
OR
2-hour blood sugar is 200 mg/dL or greater

An OGTT may reveal that although you/your child does not have CFRD, the body is not managing blood sugars correctly and further testing may be recommended. If you/your child is diagnosed with CFRD, a referral will be made to an Endocrinologist for further management.

Fact: Many people with blood sugar problems will have no clear symptoms for a long time. Cook Children's is recommending that people 10 years and older have an OGTT annually to test for CFRD. This test may be recommended outside of a yearly appointment if CFRD is suspected.

If you have any additional questions or concerns, please feel free to contact the CF clinic.

Cristina Puga RD, CSP, LD
Staci Brummett RD, LD



Managing Cystic Fibrosis-Related Diabetes (CFRD)

An Instruction Guide for Patients and Families

Cystic Fibrosis Foundation

| Adding tomorrows every day.

Download the Managing Cystic Fibrosis Related Diabetes Manual, An Instruction Guide for Patients and Families, 5th edition (80 pgs).

www.cff.org/LivingWithCF/StayingHealthy/Diet/Diabetes

CookChildren's

Cook Children's Medical Center
Cystic Fibrosis Center

Pulmonary Services
901 7th Avenue, Suite 420
Ft. Worth, Texas 76104-2724

Phone: 682-885-6299
Fax: 682-885-1090
Email: www.cookchildrens.org

Making CF stand for "Cure Found"

Pediatric CF Center Staff:

Center Physicians: **James Cunningham, MD– Center Director** (Nurse: Stacy), **Nancy Dambro, MD– Center Director** (Nurse: Charity), **Maynard Dyson, MD** (Nurse: Stacy), **Sami Hadeed, MD** (Nurse: Sharon), **John Pfaff, MD** (Nurse: Tiffany), **Karen Schultz, MD** (Nurse: Paulette)

Adult CF Center Staff:

Center Physicians: **John Burk, MD– Adult Center Director, Steve Davis, MD, Stuart McDonald, MD**

CF Coordinators:	
Janet Garbarz, Carrie Stradley	682-885-6299 (#6)
Dietitian:	
Staci Brummett, Cristina Puga	682-885-7496
Respiratory Therapists:	
Deanna Pinckney, Alex Rasmussen, Crystal Thompson, Cindy Corn	682-885-4189
Child Life Specialist:	
Leah Fox	682-885-4892
Social Services	
Jamie Cole	682-885-2863

After hours and on weekends/holidays, call 682-885-4000 and ask the operator to page the pulmonologist on call.

Cook's Corner: Recipe for Success

Peanut Butter and Jelly French Toast
For ages 4 and above

Ingredients

4 slices whole wheat bread
¼ cup peanut butter
2 tablespoons jelly
2 eggs
¼ cup heavy cream
3 tablespoons butter

Directions

1. Make two peanut butter and jelly sandwiches using the bread, peanut butter and jelly.
2. In a mixing bowl, beat eggs and cream together.
3. Melt butter in a frying pan.
4. Dip the sandwiches in egg mixture, coating both sides of the sandwich with the mixture.
5. Place in frying pan and brown on both sides. Serve with jelly or syrup.

2 servings
700 calories per serving
Fat: 53 g
Sodium: 722 mg
Protein: 20 g
Calcium: 99 mg