

Cook Children's Cystic Fibrosis Center

NEWS

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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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Fun in the Sun: CF and Exercise

Summer is here and it's time to get outside and participate in some good old fashioned exercise!

Exercise used to be discouraged for people with cystic fibrosis (CF) because it was thought that overexertion would increase breathing problems. Now, we know that the opposite is actually true. Studies have shown that regular physical activity provides many benefits to people with cystic fibrosis.

Why Exercise?

The health benefits of an active lifestyle are widely recognized for all people, but exercise is especially important for people with cystic fibrosis because it can:

- Increase lung capacity
- Increase strength and endurance
- Increase energy
- Increase life expectancy
- Improve airway clearance, which helps prevent respiratory infections
- Increase bone density and prevent bone loss

What Type of Exercise Should People with CF Do?

While most people with cystic fibrosis can tolerate some form of physical activity, the amount and type of exercise that a per-

son can tolerate will vary based on the severity of his or her illness. People with CF should work with their health care providers to develop an exercise routine that is right for them. For people that can tolerate it, aerobic exercise provides the most benefit. Aerobic exercise includes things like running, swimming, cycling, or any other vigorous activity that raises your heart rate and makes you breathe harder.

How Much and How Often Should People with CF Exercise?

The general rule of thumb is that to receive the most benefit, exercise routines should include 20-30 minutes of aerobic activity three times per week.

However, any amount of exercise is better than no exercise at all and exercise routines should be adjusted according to each person's level of tolerance.

Tips for Success: In order to get the most out of their workouts and prevent potential setbacks, people with cystic fibrosis should take a few extra precautions when participating in exercise.

Avoid Dehydration: Sweat and salt loss during exercise can cause anybody to become dehydrated, especially in hot weather.

Dehydration can be avoided by replacing fluids and salt lost by drinking sports drinks or water and eating salty snacks.

Avoid Weight Loss: Exercise increases the body's energy requirements, which are already quite high in people with cystic fibrosis. In order to prevent



weight loss, it is important for people with CF to eat enough calories to replace the calories burned in exercise. Exercise plans should be discussed with the CF

dietitian who will recommend appropriate dietary additions.

Airway Clearance: Many people with cystic fibrosis find that doing airway clearance treatments prior to physical activity helps to increase their exercise tolerance. As an added bonus, the exercise itself could loosen more secretions so it might be necessary to do another airway clearance treatment after the workout.

Consult with your CF Physical Therapist (PT) to advise you on the right exercise and activities.

Therapeutics Development Network Centers

- Baylor College of Medicine, Houston
- Children's Hospital & Regional Medical Center, Seattle
- Children's Hospital Boston
- Cincinnati Children's Hospital Medical Center
- Johns Hopkins University, Baltimore
- Massachusetts General Hospital, Boston
- Nationwide Children's Hospital, Columbus, Ohio
- Rady Children's Hospital, San Diego
- Rainbow Babies & Children's Hospital, Cleveland
- St. Louis Children's Hospital
- Stanford University, Palo Alto, Calif.
- The Children's Hospital, Denver
- University of Alabama at Birmingham
- University of Iowa, Iowa City
- University of Minnesota, Minneapolis
- University of North Carolina at Chapel Hill
- University of Utah, Salt Lake City

High Calorie SALTY Snacks

- Pretzels with cheese dip
- Cottage cheese with pears
- Salted bagel with cream cheese
- Dry roasted peanuts
- Potato chips and spinach dip
- Nachos
- Salami and cheese sandwich with pickles
- Smoked ham and cheese sandwich
- Hot dog and bun
- McDonald's Happy Meal
- Spaghettios

Cook Children's Honored as a Therapeutics Development Network Center

January 2009, Cook Children's Medical Center has been honored as a TDN coordinating center by the CF Foundation to participate in Phase 3 clinical trial studies for the CF patient.

The Therapeutics Development Network (TDN) is a nationwide network of 18 CF clinical research centers. These centers specialize in conducting clinical trials to evaluate the safety and effectiveness of new CF therapies. The TDN centers work together to promote quality, safety, and efficiency in CF clinical trials by centralizing and standardizing the research process.

Please join us in welcoming Sara Scott, RN, MA, CCRC, who will be joining our department to assist in development of the clinical trials for our CF patients at CCMC

CF 101:

Getting the Inside on Fluids and Salt

During the sweltering summer months and while participating in outdoor activities and exercise, maintaining good hydration is imperative. With cystic fibrosis, this can be a challenge. In CF, there are increased sodium and chloride losses through sweating. Dehydration can occur more rapidly.

Be aware of the signs and symptoms of dehydration because severe dehydration can be dangerous. Increased thirst, chills, clammy skin, headaches, poor concentration, fatigue, irritability, small amounts of dark urine, nausea, cramps, dizziness, weakness, and dryness of the mouth are all signs of dehydration. If you feel you are suffering from any of the above, you should replenish with fluids immediately. It is best to be proactive with your fluid intake and avoid these symptoms of dehydration-often this is harder than it sounds.

Choose fluids that work for you or your child. Some considerations include:

- Water: Because it's calorie free, it shouldn't curb your appetite
- Sports Drinks (Gatorade, Powerade, etc): Not nutrient dense, but a good choice in prolonged exercise or heat exposure; can be made into popsicles too
- Fruit Juices and Slurpees: High in sugar and can suppress the appetite; perhaps good for those that don't drink well
- Popsicles and gelatin desserts: Small volumes of fluid, but good for the challenged drinker
- **Avoid caffeinated beverages because caffeine is dehydrating. Limit carbonated drinks because they can be filling and may decrease appetite**

Increasing salt (sodium) intake will help replenish the sodium lost in sweat. How much salt is needed? Although there is no single recommendation for CF, the average American adult consumes 2,300-7,000 mg of sodium/day. Individuals with CF should liberally increase salt shaker use and choose more salty foods during warm weather.

2008 North American CF Conference (NACFC) Highlights

More than 6,000 basic scientists and health care professionals from the United States, United Kingdom, Canada, Argentina, New Zealand, Australia, France and many other countries attended sessions, workshops, symposiums, short courses, roundtables, and luncheons where they discussed the latest and greatest in CF research and care.

Some of the main highlights from NACFC 2008 include:

- Preventing CF Lung Disease**

This presentation discussed aggressive treatments based on earlier pioneering studies in Europe, and suggested that this approach may be a way to preserve lung function.

Preston W. Campbell III, MD, executive vice president of medical affairs with the CF Foundation said, "CF is the most amazing story of medicine today."

- The CFF Pipeline: The Amazing Story of Progress, Hope, and Challenge**

The presentation described how the various potential therapies in the Foundation's pipeline are developed through collaborations with both industry and academia. The exciting results from the trials of the CFTR modulator were also discussed with much enthusiasm. The trials involve small compounds designed to improve the function of the defective CFTR protein in the cells of individuals with CF.

- Taking the CF Battle to Extremes: Healthy Starts**

with Newborn Screening; Healthy Aging with Improved Adult Care

As CF newborn screening becomes a nationwide practice, it is vital to focus on enhancing the quality of the screening process to make it as efficient and error-free as possible. Currently 47 states and District of Columbia are actively screening newborns for CF. Infants who are diagnosed with CF through newborn screening

often lead to healthier adolescents and adults. Texas is one of the remaining states not participating in the screening process.

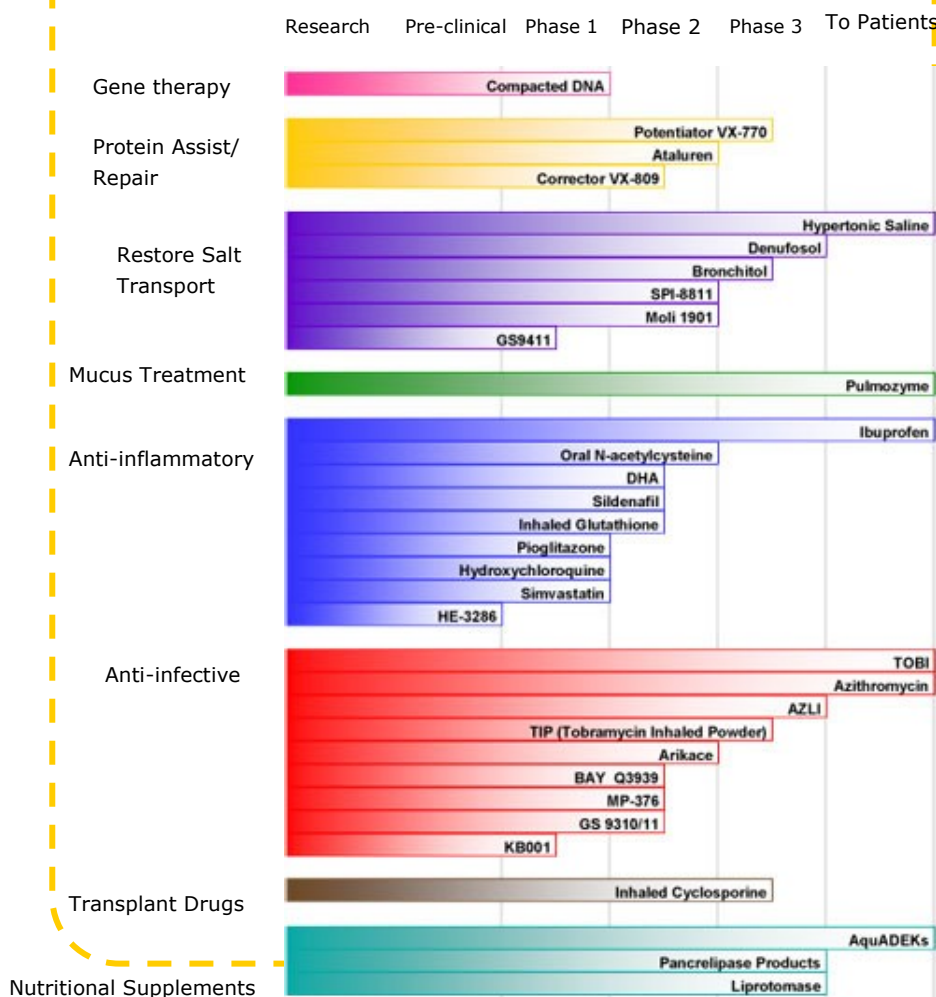
The session continued to discuss the striking improvements in

lung function with adults with CF over the past 20 years. Finding ways to help adults adhere to time consuming treatments and strengthening adult CF care centers is important for continued improvement. The CF community should also anticipate the needs of an elderly patient population with CF.



For more information on the 2008 NACFC please visit www.cff.org. Online web presentations are available for viewing

Below is a "snapshot" of those potential CF therapies that are currently in development as of June 1, 2009





Parent members include: Paula Ramsey, Kim Otten, Shelly Hale, Paul Romer, Jennifer Jaetzold. Not pictured: Carol Shepherd, Lisa Dykes

Cook Children's Cystic Fibrosis Family Advisory Council (CCCFFAC)

Who we are

We are parents and primary caregivers of people with CF 17 years or younger that get their care from Cook Children's Medical Center (CCMC) in Fort Worth, TX. We are dedicated to working with and getting to know the CF Care team and each other in an atmosphere of trust and compassion with the purpose of improving care and quality of life for those with CF.

The CCCFFAC is a recognized volunteer project at CCMC

that began in August 2008. We adhere to the standards of conduct that apply to the hospital volunteers including the obligation to maintain the confidentiality of patient information.

Our Mission

The CCCFFAC exists to enhance the medical care and quality of life for those with CF at CCMC. Through collaborative efforts with the medical community, we seek to promote an open learning environment that results in

personal empowerment and individualized care driven by proven best practices. In all we do, we seek to support CF patients and families with compassion by standing together as one in our common struggle against CF.

Please feel free to contact us with any questions, comments, suggestions at:

cook_childrens_cf_fac@googlegroups.com

(Please remember to contact your doctor with any medical questions)

Call for New Committee Members!

Join the CCCFFAC Team

The Cook Children's Cystic Fibrosis Family Advisory Council (CCCFFAC) is now accepting applications for new committee members. Each committee member will be committed to accepting a two year term on the council as well as

attend monthly meetings on the first Thursday of the month.

Lauren Morrow, CF social worker, will be receiving the completed applications. Applications are available at the CF office or can be requested by

email:

laurenmorrow@cookchildrens.org.

Please refer any questions to

Lauren Morrow.

The Cook's Corner: Recipe for Success

Cheesy Potatoes

Ingredients

- 32 oz Ore-Ida Hash Brown Southern Style Potatoes, frozen
- 2 cups condensed cream of chicken soup
- 2 cups sour cream
- 1 stick butter melted with 1/2 cup diced green onions
- 4 cups grated cheddar

cheese

- 1 cup slightly crushed corn flakes

Directions

Mix all ingredients in a large bowl. Place in a 13x9 inch pan. Sprinkle cornflakes on top. Bake at 375 degrees for 1 to 1.5 hours.

Yield: 15 servings

Nutrient analysis,
per serving:

406 calories, 13 gm protein, 28 gm fat, 378 gm calcium, 21% Vitamin A requirement

Note: Boost the nutrient density by adding any of the following: chicken, turkey, carrots, peas or broccoli. Ample enzymes recommended.



Fuel Efficiency: Increase your diet's mileage



done in the healthiest way?

People with Cystic Fibrosis are encouraged to eat a high calorie, high fat diet, rich in protein and salt. Have you ever considered why? Or how this can be

Why do CF patients need so much?

The human body is made up of so many complex systems constantly at work, even while sleeping. Therefore, energy (or calories) is continuously used in at least four ways. Calories burned can be drastically increased in certain conditions, including CF. It is not uncommon for a person with CF to require 50% more calories than a healthy person of the same age and sex.

The most obvious use of energy is physical activity. During exercise, muscles need energy to move and to support the increased work of the heart and lungs.

But surprisingly, most of our energy is expended while our bodies are at rest. This quiet metabolic activity accounts for 60-65% of our total energy used. While resting, the body still burns calories to support all the basic processes (ex. Maintain body temperature, keep heart beating, keep lungs breathing, etc.)

A third use of energy is for digesting and absorbing food. Although energy demands for processing food aren't higher for people with CF, most don't efficiently absorb foods— even when enzymes/medications work at their best. Thus, some calories eaten are lost and never available to

the body.

A fourth use of energy occurs when the body is under stress. In the process, a significant amount of calories can be used. This accounts for extra energy spent when a person has to adjust to circumstances (ex. Illness, lung infections, fevers, etc.) Therefore, a person with CF burns more energy when not in the best of health.

With all this going on in the body, it is not surprising the calorie demands can be so high in individuals with CF.

How do I achieve high calorie goals?

Make every bite count! Create nutritionally rich meals and snacks high in fat, calories, vitamins, and minerals. Fat is important for a variety of reasons. Gram for gram, it has twice the number of calories as protein or carbohydrate. Because it is more calorie dense, fat is twice as efficient as carbohydrate or protein in achieving calorie goals. At the same time, it is important for our overall health to choose nutrient dense foods. That means getting more in every mouthful. For example, a snack size bag of chips vs.

trail mix are both high in calories. However, trail mix made with favorite snack foods: tiny pretzels, cereal (corn, rice, wheat squares), nuts (peanuts, almonds, soy nuts, pine nuts), dried fruit (raisins, dried berries, banana chips, etc— with or without yogurt, or chocolate coating), small chocolates (candy coated or chips), and/or seeds (sunflower, sesame, pumpkin) has significantly more fiber, protein, vitamins, and other nutrients. Choosing this snack will give the body more than just calories. Plus, it is easy to buy or make and can be tailored to suit your taste (sweet, salty, spicy).

So, while meeting energy needs is a challenge for people with CF, it can be accomplished in a healthy way. It simply takes the right tools and a little planning to make the most of what is eaten.

Make every bite count!

Create nutrient dense meals and snacks that are high in fat, calories, and nutrition!

Nutrient Dense Meal Planning: Some Beginning Strategies

What works for me might not work for you. Lifestyles, food preferences, and the nutrition goals for other family members are just a few of the reasons why. There are several ways to boost nutrition:

- **Buy high-calorie versions** just for your CF consumer (rich ice cream, whole milk, or half and half)
- **Prepare high-calorie, nutrient dense dishes.** See the recipe for *Cheesy Potatoes* in *The Cook's Corner: Recipe for Success*.
- Is someone else on a healthy diet or watching their weight? Then **boost calories** in a single serving using gravy, butter, salad dressing, Alfredo sauce, or shredded cheese.
- **Use convenience items** for the busy lifestyle. It's better than running off without eating.
- **Need help with school lunch?** Time too short to eat? Your CF dietitian has some ideas.
- **Drink milk with most meals** and snacks; stock up on dairy foods because they are packed with nutrition.
- Fruits and vegetables are low in calories but high in nutrients. Use them in small portions often throughout the day to increase the nutrient density without filling you up. **Always add a dip, salad, dressing, or peanut butter.**

Questions? For questions about this or any other nutrition issue, please speak with your CF dietitian: **Staci Brummett at 682-885-7496**

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"

Pulmonary Exacerbation Score (PES)

Cook Children's was selected in January 2008 to work with the Cystic Fibrosis Foundation and other centers nationwide through the Learning and Leadership Collaborative (LLC) to improve the care of people with cystic fibrosis (CF). Since that time, clinic staff, patient/family representatives, and the Cook Children's Cystic Fibrosis Family Advisory Council have been working side by side on numerous projects to improve lung health and nutritional status as well as the quality of life of people with CF.

We are currently working on a new project we are excited to tell you about! We are adding a new tool to our toolbox that will help us optimize your lung health. **As of June 1st,**

your physician will begin calculating your pulmonary exacerbation score (PES) at each clinic visit.

Your PES is based on:

- Information you give us about your lung and nutritional health (such as changes in sputum, appetite, and energy level).
- Results of your lung function tests (FEV1), chest x-ray, and oxygen saturation.

A PES of ≥ 5 is defined as a pulmonary exacerbation.

Your physician will decide how and when to treat your exacerbation. Please feel free to ask your physician if you have any additional questions.

*****NOTE*****

Future CF newsletters will only be available via email or at the clinic.

If you would like to continue receiving the CF quarterly newsletter via email, please contact Carrie Stradley at

carrie.stradley@cookchildrens.org

This newsletter is published by the Cook Children's Cystic Fibrosis Family Advisory Council. Physicians listed herein are members of the medical staff of Cook Children's Medical Center and are neither employees nor agents of the hospital.