

# Cystic Fibrosis Center News



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## CF-RELATED DIABETES MELLITUS

Cystic Fibrosis Related Diabetes Mellitus (CFRDM) is a common and serious complication of CF. Frequency increases with age, with as many as 40% of adults over age 19 and 25% of children with CF developing CFRDM. It is also distinctly different from the more common non-CF types of diabetes, which go by the names of Type I or Type II or juvenile or adult onset diabetes. Although "classic" symptoms of diabetes include increased hunger while losing weight, increased thirst, and increased urination, few if any patients with CFRDM will have these.

In CFRDM, there is too little insulin to let glucose, the "energy molecule," into the cells. Insulin is a product of the pancreas, one of the main organs affected by CF that enables sugar to be moved from the blood to the cells where it can be used or stored. Thus in diabetes, even if there is plenty of glucose in the blood, the body cannot use it for energy and must turn to other sources, like protein that has been stored in the body. This creates a situation in which weight loss is common and weight gain is almost impossible, issues that already challenge those with CF. In addition, a high blood glucose level, known as hyperglycemia, has a detrimental effect on other organs of the body, including the eyes, kidney, and heart. White blood cells that are critical for fighting infection, also do not work well when blood sugar levels are too high. As you can guess, in persons with cystic fibrosis, undiagnosed or untreated CFRDM can contribute to weight gain problems, more frequent pulmonary exacerbations or other serious complications.





# Frequently Asked Questions

In each issue we will be addressing a few frequently asked questions from our Center. Please feel free to submit questions for future issues to our nurse coordinators or Judy Kirby at 650-724-3474.

## Should I be on azithromycin?

The answer to this frequently asked question is, "We'll let you know!" Azithromycin, or Zithromax,<sup>™</sup> is a commonly prescribed antibiotic in non-CF-related sinus and lung infections. Researchers found that it is helpful in another severe lung disease, diffuse panbronchiolitis (DPB). While DPB has many similarities to CF, it is a different disease. Researchers are encouraged by azithromycin's effect on inflammation and lung destruction in DPB and its possible application to CF. Although it does not appear to kill *Pseudomonas aeruginosa* or *Staphylococcus aureus*, the two most common infections in CF, it may inhibit their growth. In addition to its antibiotic properties, azithromycin may have an anti-inflammatory effect in the small airways of the CF lung. There may be benefits to persons with CF, and there have been some initial positive research findings. Currently the CF Foundation is sponsoring a very large scale Phase III clinical trial to test the efficacy, safety and best dose of azithromycin in persons with CF. Although we are interested in this as a potential new weapon against the ravages of CF, we cautiously await the results of this study before routinely prescribing it to our patients. (Our CF Center is not participating in this study.) Feel free to discuss the benefits and risks of this or other therapies with your physician.

## What is the Best Form of Airway Clearance?

The one that you use! If you are like most patients, you don't use therapies that you do not think help you or your child. Airway clearance *has* been shown to be an important treatment for preventing the scarring that occurs in CF lung disease, however different methods appear to be equally effective when used properly and routinely. It is important to be educated about the potential benefits and limitations of each method and then select the method that works best for you and your family. Our team, and in particular, Kristin Shelton, our respiratory therapist, is here to help train, retrain and coach you in both selection and technique.

Chest physical therapy (CPT) has been the gold standard, however not everyone has a healthy adult available to perform CPT whenever needed. Other forms of airway clearance that do not require assistance include the Flutter

Valve and Acapella, hand-held devices that you blow into to vibrate the lungs, IPV and PEP valves that are used in combination with inhaled medications and air compressors, and the ThAIRapy Vest. Autogenic drainage and the active cycle of breathing, both of which require training, are other forms of clearance that can be performed without assistance. Any form of aerobic exercise is also great for airway clearance as long as you push yourself and are not afraid to cough and spit while you are exercising. Exercise has the added benefits of building muscle strength and mass, protecting the bones from osteoporosis, and increasing bowel motility. We recognize that airway clearance can get boring, so mix it up! We don't care what you do, just do it!

## My pharmacy substituted generic enzymes and they're not working as well. Help!

Substitution of trade name enzymes with generic enzymes is occurring at pharmacies with alarming frequency, even though we know that different enzymes work better for different people. Although the generic preparations are meant to be exact substitutions, in many cases there are subtle differences that may affect an individual. *You do not have to accept a generic substitution.* Most pharmacists are aware that the generic preparations are not exactly the same as the prescribed forms and will not substitute without permission. However, if you are having trouble with your pharmacy, please call your CF nurse coordinator (Monica for pediatrics, Mary for adults) for assistance.

## What is Stanford's infection control policy?

Protecting our CF patients from infection with resistant organisms is a great concern. We work to schedule patients to minimize cross-infection, and persons with resistant organisms are requested to wear masks in the common areas. Wearing a mask in the clinic common areas not only protects other patients, but it also protects you. If you would like to wear a mask, you can request one from Chandra McDuffie when you check in. In May, the CFF held a consensus conference on the topic of infection control. When the results are published, our clinic will be quick to adopt the recommendations.

## Making the Diagnosis

It is important for those with CF over the age of 16 years to be screened annually for the condition since the external symptoms may be subtle and are so closely associated with more typical CF symptoms. Younger children should be tested if weight gain or frequent infections are problems. Prednisone, a medication used in some patients, can cause glucose intolerance (high blood sugar) while you are taking it, but does not promote the development of CFRDM.

The diagnosis of CFRDM is made by doing a 2-hour glucose tolerance test that requires three blood tests immediately following a period of fasting. Patients are instructed not to eat or drink after midnight so that the first blood test may be taken first thing in the morning. Following the first blood test, the person drinks an orange-flavored sugar solution. One hour later the second blood test is taken. The final blood draw occurs one hour after the second one. Oral glucose tolerance tests should be done when one is well, as being sick may alter the results.

## Stages of CFRDM

There are three stages of CFRDM, all of which can occur without symptoms. The first stage is impaired glucose tolerance. In this stage, blood sugar is usually normal except at times of stress, as with a pulmonary exacerbation. The second stage is called CFRDM without fasting hyperglycemia (high blood sugar). In this stage, blood sugar goes too high within two hours after eating, but returns to normal between meals. The third stage is called CFRDM with fasting hyperglycemia. With this stage, blood sugar levels are always high. It is believed that patients start with Stage 1 and then progress through Stage 2 before reaching Stage 3. Although we cannot prevent someone's diabetes from progressing, early identification and intervention helps prevent significant weight loss and more severe complications. In times of stress, the body may become more glucose intolerant. Thus, someone with Stage I can have features of Stage II or someone with Stage II can have high between-meal blood sugar. However, when the stress is removed, such as following successful treatment for a pulmonary exacerbation, glucose tolerance will return to normal. Studies on the impact of CFRDM on the long-term health of people with CF have been inconclusive, however we do know that maintaining weight and nutritional status are strong contributors to maintaining good health.

## Treatment Options

Treatment options vary by stage and symptoms, and are different from treatment of non-CF diabetes. The Stanford CF Center is working closely with Judy Kalinyak, M.D., a diabetes specialist in the Stanford University Department of Medicine who has a significant research and clinical interest in CFRDM. Anna Simos, a diabetes educator, and Julie Matel, the CF Center nutritionist, work closely with Dr. Kalinyak. Anna is often available to come to clinic to help evaluate patients with evidence of CFRDM. One of the key points in treating CFRDM is *NOT TO CUT CALORIE INTAKE*. So, unlike other forms of diabetes, restricting one's diet to limit total calorie intake is not healthy and special dietary needs must be recognized and incorporated to maintain weight. Treatment varies for each individual and may include different eating strategies, pills or insulin shots.

## CFRDM Research

The CF Center is participating in a study funded by the National Institutes of Health to identify optimum therapy for patients with CFRDM without fasting high blood sugar (Stage 2). If you are diagnosed with this stage of CFRDM, you may be approached to participate in the study. If you wish to learn more about CFRDM, ask your doctor or Julie Matel, the CF Center nutritionist on your next visit.



*It is important for those with CF over the age of 16 years to be screened annually...*



New team members Monica Smith, R.N. (above) and Lauren Witcoff, M.D. (below, left)

## Our New Team Members

Please join us in welcoming new team members, Lauren Witcoff, M.D. and Monica Smith, R.N. Dr. Witcoff, a pediatric pulmonologist, joins our team as a staff physician. She will be seeing both inpatients and outpatients as part of the CF and general pediatric pulmonary service. Dr. Witcoff has treated CF patients for more than 20 years, most recently as CF Center Director & Director of Pediatric Pulmonary Services at Lutheran General Children's Hospital in Park Ridge, Illinois. She previously served as co-director of the CF Center at Cedars-Sinai Medical Center in Los Angeles. Dr. Witcoff is pleased to return to Stanford just in time to celebrate her 25th undergraduate reunion.

Monica Smith, R.N. is acting Pediatric Nurse Coordinator. Monica joins the Center after working with inpatients on 3West at LPCH. Prior to that she spent ten years at UCSF on the pediatric unit. She can be reached at 650-736-1359 if you need advice or assistance with your care.

We also hope you'll join us in welcoming Sophia Pegg who arrived on July 7, 2001 to Holly and Phil Pegg. We are sad to report that Holly has decided not to return to Packard. We will be starting the process of hiring a replacement in the next few weeks.

## TRANSITION TO ADULT PROGRAM

One of our Center's goals in developing a Center of Excellence program is to create a model program for persons of all ages with CF. Over the next year you will be hearing more about this as we develop programs and services to facilitate a successful transition from adolescence to adult independence. Although we operate as a team of professional staff, inpatient care occurs in two different facilities. Our hospitals are physically connected, however Stanford University Hospital and Lucile Packard Children's Hospital are separately licensed and each has staff specializing in the care of adults or children. We are fortunate to have units specializing in the care of CF patients in both facilities. We believe familiarizing our older adolescent patients with Stanford Hospital before the need for a hospitalization will result in better care and a better understanding of the different admitting and rooming systems, and provide us with an opportunity to facilitate the transition. In September we began a Tour/Orientation Program for persons with CF who are turning 17 or older during the coming year. It is our goal to minimize the stress and concerns over this part of the transitioning process. For more information, please contact Mary Helmers, Adult CF Nurse Coordinator at 650-736-1358.

## EXPANDING CF EDUCATION

We are pleased to announce another very important addition to our program. The Department of Medicine is beginning a formal CF service rotation for its pulmonary fellowship training program. Each month, one of the adult pulmonary fellows will be the "CF Fellow" and participate in the adult CF clinic and adult CF inpatient care at Stanford Hospital as they work closely with the CF attending physicians. The fellows are an energetic group of young physicians who are training to specialize in pulmonary medicine. We are particularly excited about this opportunity to advance the academic mission of Stanford, and to be a leader in educating the next generation of adult pulmonologists in the care of adults with CF. Also, their round-the-clock availability will enhance access to physicians and provide greater continuity of care, especially for adult CF inpatients.

## CF CENTER IN THE NEWS

Over the past few months, members of our CF team have been involved in these efforts to advance the knowledge of CF and clinical care:

**Center Director Richard Moss, M.D.** chaired the first Cystic Fibrosis Foundation Consensus Conference on Diagnosis and Treatment of Allergic Bronchopulmonary Aspergillosis (ABPA) in Persons with CF in June. This conference continues the CFF commitment to bringing focus on advancing the standards of care for clinical treatment while sponsoring research to find a cure for CF and its many complications. In October, Dr. Moss presented two invited lectures on aerosolized antibiotics to the largest event of Pediatric Pneumology that has ever taken place in South American, the combined Congresses of five Brazilian and Latin American pediatric pneumology and cystic fibrosis organizations.

**Adult Center Director Noreen Henig, M.D.** addressed the Cystic Fibrosis Research Inc.'s Teen and Adult Retreat on the topic of CF and Pregnancy, Sexuality, Fertility, and Parenting in August.

This summer **Terry Robinson, M.D.** addressed the Northwest Consortium of the Epidemiologic Study of CF (ESCF) and the CFRI annual conference on "Advances in CT Scanning: Preliminary Data from the mild CF Lung Disease Pulmozyme Intervention Study."



Medical Student Michele Lai's study on pediatric patient perceptions of physicians provided valuable opportunities for CF children to express themselves. Children were asked to create a mandala of the emotions

they felt when they were with their physician. They were given a choice of eight positive and eight negative emotions, and were asked to use colors and shapes to represent their feelings.

Gianna Altano created this mandala with the legend above. She says that the swirl in the middle represents her port, the vascular access device implanted in her chest to ease the administration of antibiotic therapies.

## ACTIVE RESEARCH RECRUITMENTS

We are actively recruiting subjects for these trials:

- **Cystic Fibrosis Once Daily Aminoglycoside Collaborative Trial (CFODACT).** Please consider participating in this study if you are being admitted for an exacerbation.
- **A Multicenter, Double-Blind, Placebo-Controlled Phase II Study of Aerosolized tgAAVCF in Cystic Fibrosis Patients with Mild Lung Disease**
- **Standardization of the Measurement of the Nasal Membrane Transepithelial Potential Difference**
- **Diabetes Therapy to Improve Body Mass Index and Pulmonary Function**
- **Validation of Sputum Induction as an Outcome Measure for Lower Airway Sampling in Patients with CF**
- **A Phase I/II Study of Interferon Gamma-1b by Inhalation for the Treatment of Patients with Cystic Fibrosis**
- **Sweat Rate Procedure Standardization for CF Research Centers for patients who have had genotype testing**
- **Health Buddy telephonic/web-based home monitoring system for persons with CF**
- **Hi-D FACS with CF Blood & Lung Leukocytes study of chronic oxidative stress in CF (cell samples needed from blood/sputum/bronchoalveolar lavage)**

Please consider participating in our research since it is through the efforts of all of us that better treatments will become available. Ask your physician or call our research staff if you are interested in learning more about participation.

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THANKFUL
SAFE
HOPEFUL

## HELP US TO HELP YOU!

Our team is actively working to develop programs to meet your needs as we define our Center of Excellence. Your input is important to us!

- **Adults with CF** survey was mailed a couple of months ago, and we would like more responses. If you would like another copy, please call Mary Itelmars, and either fax or send it in!

- **Parents of Children with CF** questionnaire was sent in June—please return them at your earliest convenience or call Joanne Asano if you need another copy!
- Our **CF Cookbook** is under development by Julie Matel and needs your recipes and contributions. More information and recipe forms are in Clinic E and on our Web site.

All of these may be mailed to the CF Center or faxed to the office at 650-723-5201. Please call us if you have questions or need more copies.

## EDUCATION DAY HIGHLIGHTS

March 31, 2001, the LPCH CF Center Family Education Day was attended by more than 110 persons with CF, families and others. Twelve topics were covered, including how to use the CF Center, how to interpret diagnostic tests, clinical issues such as CF-related diabetes and liver disease, infection control, and advances in CF research. All participants received notebooks with information on the presentations. We still have a few notebooks left, and a video of the conference is available from Cystic Fibrosis Research Inc. (CFRI) in Palo Alto. The response was overwhelmingly positive and we look forward to hosting another one next spring. Please forward any ideas on topics for this or future issues of the newsletter to Joanne Asano at the CF Center or [joanne.asano@medcenter.stanford.edu](mailto:joanne.asano@medcenter.stanford.edu).



Lucile Packard Children's Hospital

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## CYSTIC FIBROSIS CENTER AT STANFORD

Center Physicians: **Richard Moss**, Director;  
**Noreen Henig**, Adult Center Director;  
**Carol Conrad**, **Terry Robinson**,  
**Lauren Witcoff**

### IMPORTANT PHONE NUMBERS

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for more information about our center, CF  
and current topics.

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