

**SAVE THE DATE: 21ST ANNUAL ROSS MOSIER CLASSIC GOLF TOURNAMENT, JULY 29, 2002**

This event was created in memory of Ross who died of CF in 1980 at the age of 12 at Children's Hospital at Stanford. Since its inception, more than \$1,250,000 has been raised to support our CF research program. For more information and to register, download a brochure at [www.rossmosierclassic.org](http://www.rossmosierclassic.org) or call Clyde at 1-800-500-5598.

Caption for this picture?



**CYSTIC FIBROSIS CENTER AT STANFORD**

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

Clinic E Scheduling (Chandra McDuffie)	650-497-8841
Clinic E Fax	650-497-8837
Nicole Eden, Pediatric Nurse Coordinator	650-736-1359
Mary Helmers, Adult Nurse Coordinator	650-736-1358
Kristin Shelton, Respiratory Coordinator	650-724-0206
Julie Matel, Nutritionist, Dietitian	650-736-2128
Joanne Asano, Social Work	650-736-1905
Zoe Davies, Research Coordinator	650-498-5315
Colleen Dunn, Research Coordinator	650-736-0388
Janie Perez, Research Coordinator	650-723-5193
Judy Kirby, Webmaster	650-724-3474

**For Urgent Issues**

Monday-Friday 8:30-5:00 pm	contact RN Coordinator
All Other Times (ask for Pulmonary Physician On-Call)	650-497-8000
Medication Refills: Call pharmacy where medication was last filled	
LPGH Pharmacy Refill Line	650-497-8289

See our website at <http://cfcenter.stanford.edu> for more information about our center, CF and current topics. To subscribe to this newsletter please email or call Judy Kirby at the number listed above.

Non-Profit Org.  
U.S. Postage  
**PAID**  
Palo Alto, CA  
Permit No. 29

Summer 2002

# Cystic Fibrosis Center News

**EAT, DRINK AND BE HEALTHY!**

Nutrition is among the most important strategies for maintaining good health for people with CF. Studies confirm a clear link between poor nutrition and decreased lung function and survival rates. It is important for both children and adults to achieve optimal growth, prevent bone loss, fight infection, and improve quality of life. Nutritionists, nurses and physicians working with the CF Foundation recognize the importance of nutrition: a 2001 CFF Nutrition Consensus Conference adopted more aggressive clinical care guidelines for nutrition monitoring and earlier intervention to prevent or minimize the adverse effects of poor nutrition.



Nutritionist Julie Matel measures the growth of a CF patient.

In 95% of persons with CF, the pancreas does not secrete the digestive enzymes needed to digest and absorb sufficient vitamins, minerals and calories. This is known as "pancreatic insufficiency." Enzymes are taken orally with food to aid in digestion and absorption. However, the oral enzymes require monitoring and adjustment to ensure the right amount is available at the right time. People who take enzymes know they don't automatically "solve" the equation—it still takes work to get enough calories and it takes vigilance to adjust amounts with food and fat intake. Even for the 5-10% of the CF population who do not require enzymes because they are "pancreatic sufficient," maintaining a healthy weight and good nutrition are important for staying healthy and fighting infection.



CF Center at Stanford  
701 Welch Road Suite 3328  
Palo Alto, CA 94304

**Cystic Fibrosis Center News**  
Summer 2002



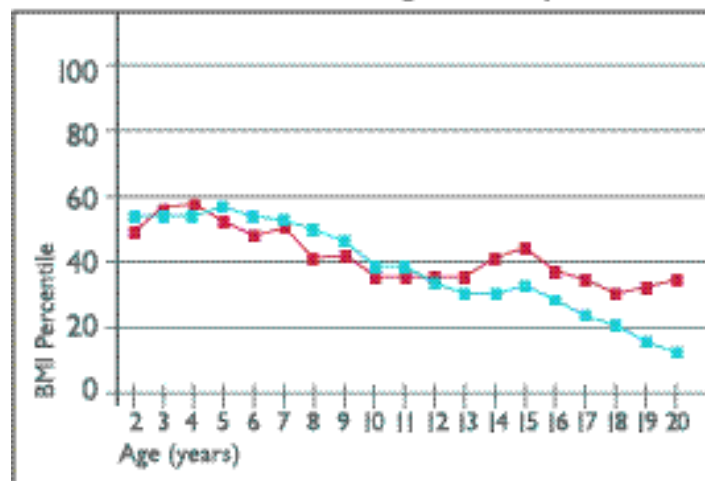
## The Importance of Nutrition

Study after study has shown that good nutrition, which is indicated by weight appropriate for one's height, normal growth rates and normal levels of fat soluble vitamins, is associated with overall maintenance and even improvement in lung function. In addition to good lung function, the immune system requires a certain amount of protein, vitamins A and B complex, iron, zinc, and selenium to function optimally. Good nutrition is also important to optimize bone growth and density. People with CF are prone to decreased bone mass for a number of reasons, including inadequate calcium intake, poor Vitamin D absorption, low body weight and in some, long-term prednisone therapy. Without strong bones, osteoporosis and debilitating fractures can occur which can compromise lung function and the ability to enjoy life to the fullest!

## Improvements in Nutritional Status

The past decade brought significant gains in the nutritional status of persons with CF, a result of better enzymes, earlier intervention, improved respiratory therapy, the promotion of high fat diets and improved understanding of nutritional health. Nutritional status is measured by comparing persons with CF to growth and body mass index charts for the normal population, since our goal is to achieve normal growth and development. These data are collected as part of the CFF national registry, which provides valuable data on clinical outcomes that guide the development of clinical care protocols for CFF Centers and identifies trends and issues that need to be addressed. Between 1989 and 1999 registry data showed improvements in weight percentiles in all age groups: the percent of persons with CF that fell below the 10th percentile for weight has decreased from 36% to 28%, a 22% decline! Less improvement was seen when comparing height to the general

Median CDC Body Mass Index (BMI) Percentiles by Gender for Persons with CF, Ages 2 to 20 years.



population. In 1989, 33% of persons with CF fell below the 10th percentile for height, versus 30% in 1999. Thus, despite significant improvements in average weight and modest gains in growth and stature, the CF population still lags behind the general population. The CFF Consensus Conference set new standards that focus on nutritional measures and early intervention with a goal of building on the advances of the past decade.

## Risk Factors and Detection of Poor Nutrition

We use a number of measures to monitor and detect poor nutrition in its early stages. National growth charts, established by the Center for Disease Control, are used to compare growth and weight gain of children and adolescents since these are proven markers of nutritional status and health. Weight for length and body mass index values allow us to determine the appropriateness of a person's weight for their height. Body mass index (BMI) is one's weight (in kilograms) divided by height (in meters-squared). A child or still-growing adolescent is considered at risk for poor nutritional status when they have no weight gain for more than 3 to 6 months, a weight for length less than the 25th percentile, or a BMI less than the 25th percentile. Adults are considered at risk when they fall below 90% of their ideal body weight.

Julie Matel, our CF Center dietitian and nutritionist, charts these measurements at each clinic visit to monitor changes and identify when more aggressive strategies may make sense. Please ask her to share them with you if you're interested at your next clinic visit.

## Strategies to Improve Nutrition

First and foremost, optimizing medical treatment to prevent and promptly treat lung infections is important. Your body requires more calories when it is fighting infection, and weight loss due to loss of appetite and increased caloric needs can contribute to more severe infections. Good respiratory care and early intervention in the treatment of infections often can prevent the weight loss associated with pulmonary exacerbations.

Optimal enzyme therapy is another important factor. Frequent, foul-smelling or oily stools, may indicate malabsorption of valuable



## Studies confirm a clear link between poor nutrition and decreased lung function and survival rates.

calories and vitamins that can be corrected with a simple adjustment in the number, type or timing of enzymes. Since too many enzymes can cause problems like stomachaches that take away your appetite, make sure you talk to your dietitian about how to adjust your dosage.

For persons with CF, maximizing calories, fat, and protein is important to achieve adequate growth, maintain a healthy immune system and prevent weight loss. One trick is to add as many calories as possible to the foods you enjoy and routinely eat—add two slices of cheese instead of one and use generous portions of mayonnaise on sandwiches. Use plenty of margarine or butter on vegetables and pasta. Add dry milk powder, half-and-half or even cream to soups, potatoes, and milk-based beverages. Choose high calorie snacks such as peanuts (any nuts will do!), dried fruit, granola bars, and smoothies. Getting enough salt and fluids is also important. Commercial supplements are convenient for adding calories. Look for the "Plus" varieties for extra calories and protein.

For some people, nutritional needs for growth, weight gain or maintenance cannot be met by eating alone and your doctor may recommend a gastric or "G" tube that can be used to deliver extra calories while you sleep. A high-calorie liquid formula is usually run overnight through a tube that has been endoscopically or surgically implanted with direct access to the stomach. When not in use, the device lies flat to the skin and looks like a small button. The goal is to supplement, not interfere or replace food eaten normally during the day. Sometimes it makes sense to consider a G-tube before a nutritional crisis hits. Our nutritionist monitors nutritional status for each patient individually and can discuss when and if this is something you should consider.

Appetite stimulants, such as Megace, a form of the hormone progesterone, or Marinol, a derivative of marijuana, are occasionally prescribed to increase appetite and weight gain. However, there can be side effects which you should discuss with your physician or nutritionist before use.

## The bottom line...

Early detection and intervention for both children and adults with poor nutrition and growth failure is paramount to maintaining good health and a strong immune system. Our CF Center recog-

## MEET NEW PEDIATRIC NURSE COORDINATOR NICOLE EDEN

Nicole Eden, R.N., B.S.N. joined our team in February as pediatric nurse coordinator. Nicole is a native of Canada, who worked on the multi-organ transplant unit at the Toronto Hospital for Sick Children and in general pediatrics at UCSF before coming to LPCH in 2000. Working with children with CF was a highlight of her first job as a staff nurse on 3West. She said the teens were her favorite patients and always made her laugh, so when she saw the CF coordinator job ad, it seemed ideal. Nicole and her husband love the outdoors and she is an avid fan of white water rafting, mountain biking, hiking and SCUBA diving. Before starting as coordinator, she spent several weeks in Hawaii and Mexico. Nicole describes herself as an optimist who believes in living life to the fullest, a philosophy she sees in many of her patients. She also likes hugs!



Nicole looks forward to the program development opportunities that the LPCH CF Center of Excellence initiative promises. Her immediate goals are to improve clinic organization, patient flow and patient satisfaction as she gets to know the patients, systems and staff. She wants to be accessible to patients and their families. She will also be assisting Dr. Henig and Mary Helmers with adult patients on days when Mary is unavailable. Feel free to call or email her at (650)-736-1359 or [neden@stanfordmed.org](mailto:neden@stanfordmed.org) if you have questions or need assistance.

nizes the importance of nutrition and as part of our Center of Excellence plan provides a full-time clinical nutritionist to help our patients. Julie Matel is a registered dietitian and nutritionist with more than ten years of experience working with CF patients. She and other members of our CF center team are available to discuss what therapies are best for you or your child. Our goal is to work together to promote good nutrition and keep you in the best possible health!

# 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.

## How do I get a disabled parking placard?

Individuals with CF may be eligible for a disabled parking placard or license plate. A disabled person does not have to own or drive a vehicle to use the placard, however, only the person for whom it was issued may legally use it. Disabled license plates are only issued to persons with permanent disabilities, whereas placards may be obtained for temporary conditions. For persons with CF, a physician must certify that the individual's spirometry, specifically the forced expiratory volume for one second or FEV1, is less than one liter, or arterial oxygen tension (PaO2) is less than 60 mmHg on room air at rest.

To obtain a placard or plate, first complete an "Application for Disabled Person Placard or Plates" which includes a certification that must be signed by your physician. The certification and application are mailed with a check or money order for \$6.00 to: DMV PLACARD, P.O. Box 942869, and Sacramento, CA 94269-0001 (There are no fees for permanent parking placards/plates). Forms and more information may be downloaded at <http://www.dmv.ca.gov>, or requested by calling 1-800-777-0133. Bring the form to your clinic visit to be filled out by your doctor.

## What insurance options are available for persons over 18 who have CF?

If a parent provides insurance coverage for a child through an employer's health plan, coverage usually continues while the child is enrolled as a full-time student. Coverage often lasts at least until the age of 21, and in some cases even longer. Age and time limits differ by insurer so it's important to check your policy or contact your employee benefits department. CCS (California Children's Services) coverage is available to qualifying persons up to the age of 21.

Another insurance program to consider when a person turns 21 is GHPP (Genetically Handicapped Person's Program). GHPP is a state-funded program that helps pay medical costs for adults with CF and other defined medical conditions. If you have CF and are a California resident, you may apply. You must complete an application, and you may be asked to apply for Medi-Cal as part of the process. Call GHPP directly for an application at (800) 639-0597 or (916) 327-0470.

## Are there any programs for people with private insurance who are having difficulty meeting out-of-pocket expenses for doctor's visits, medications, etc.?

CCS is available to qualifying persons up to age 21, if your insurance and income do not cover your out of pocket costs. Also, if your annual family income is over \$40,000 yet under \$300,000 and you do not qualify for CCS, you can apply for GHPP as a secondary insurance. Even if a child is not yet 21, GHPP may cover some services not paid by primary insurance. Financial screening is required, and you may be required to pay an enrollment fee. You may have to provide proof that you applied and were denied by CCS (including denial of an appeal). Bypassing this process is sometimes possible by providing copies of income tax returns. It's always best to call GHPP for more specific advice on your particular circumstances.

Rhiannon submitted the following poem in her scholarship application:

Who I am  
*I am stronger than I thought I was  
 I can make my own choices  
 I can do what everyone else does  
 I can embrace my inner voices*

*I am braver than I thought I was  
 I can face my fears  
 I can live like everyone else does  
 I can accept when I have tears*

*I am smarter than I thought I was  
 I can learn from my mistakes  
 I don't have to do what everyone else does  
 I am ready to raise the stakes*

*I am more determined than I thought I was  
 I am committed to my goals  
 I have dreams like everyone else does  
 And I won't be raked over the coals*

## picture being sent from Rhiannon to Jen

## 2002 CREON FAMILY SCHOLARSHIP PROGRAM & AWARD

For the 10th consecutive year, Solvay Pharmaceuticals, Inc. will award 20 scholarships nationally to outstanding students with CF who have demonstrated academic excellence and the ability to act as role models for other teens and young adults. Every winning recipient will receive a \$2000 scholarship for up to four years of matriculated study in addition to one-year supply of Creon Minimicrospheres. Scholarships are available to all high school, vocational school, college and graduate students with CF who will begin or continue higher education in the Fall of 2002. See your CF Center nurse coordinator or social worker for an application. Application deadline is June 28, 2002.

This year Rhiannon Perez became the first for "this" year Stanford CF patient to win this generous scholarship. A resident of Carmichael, Rhiannon is majoring in philosophy at California State University in Sacramento. She hopes to pursue a Ph.D. in philosophy, and possibly a career in bioethics. Rhiannon loves school, and has found college to be a great place to learn how to deal with the real world. She credits CF Research Coordinators Zoe Davies and Colleen Dunn with encouraging her to apply for the scholarship and continue her educational goals.

*I am more unique than I thought I was  
 I am special in my own way  
 I can do what nobody else does  
 I can say what I need to say*

*I have learned that I have strength  
 That I can conquer my fears  
 I have learned that I am smart  
 And that my goals have been made clear*

*But most of all my life has taught me  
 That we are all unique to each other  
 And there is nothing I would ever change  
 Because I am proud to be like no other*

## ADULTS WITH CF MENTOR STANFORD CF TEENS

A year ago our Center's CF social worker Joanne Asano began a mentoring program that matches adults and teens with CF. The program now has three pairs who regularly communicate through phone and e-mail. The program goal is to provide teens support and the perspective of an adult with CF so they can learn about making the transition to independent adulthood. It gives adolescents a chance to talk to someone who shares similar life experiences, such as coughing in public or deciding when to tell people that they have CF. Joanne believes the program will provide opportunities to build bonds and relationships similar to those that used to develop when adults and children were hospitalized on the same units.

One shining example of the program is the dynamic duo of Kathy Schaal, age 38, and Sandra Aguirre, a high school sophomore who loves to read, dance and hang out with friends. Kathy calls and e-mails Sandra regularly. They both have diabetes as well as CF. Sandra says it's been great having someone who can share experiences and understand what coping with CF means since it's often hard to believe that a



Sandra Aguirre (left) and mentor Kathy Schaal (right)

parent or friend really understands what life with CF is like. They talk about what it takes to stay healthy, and how to balance daily routines and exacerbations with other priorities. Sandra says her relationship with Kathy has given her hope that if she takes care of herself, there is a full life ahead. Kathy enjoys serving as a mentor and being available for support. She thinks it is important for young people to see someone living a full and independent life that is not dominated by CF, despite having to deal with occasional hospitalizations and daily routines.

Another enthusiastic report comes from Debbie Contreras, parent of 18-year-old high school senior Desiree. Debbie reports that connecting her daughter with 31-year old Liz Nash could not have

been timed more perfectly. Liz has offered friendship, support and an empathy that someone without CF will never have, plus she has helped Desiree to see a different side of having CF. Desiree has found a friend who can relate to her own experiences—such as what to tell her boss now that she has her first part-time job. Debbie perceives that Liz's caring phone calls, positive outlook, and ready availability to talk, have really opened a new outlook on the future, which is invaluable as Desiree looks toward college and an independent life.

You may contact Joanne Asano if you have an interest in participating in the program.



Student Desiree Contreras

## 2002 FAMILY EDUCATION DAY HIGHLIGHTS

Our second family education day was held March 2 at LPCH with ten speakers presenting a broad range of topics. Keynote speaker Jane Burns, M.D., Associate Professor of Pediatrics and Infectious Disease at the University of Washington discussed CF lung infections, how they are spread, and how to prevent cross-infection. She outlined why droplets, direct touch, and contaminated surfaces are the primary ways germs are spread. Maintaining a 3-foot distance between persons with CF, thorough hand washing and not sharing utensils or equipment are among the most effective ways to prevent cross-infection. Dr. Burns also noted that when cleaning nebulizers and other equipment, allowing the parts to dry is more important than what soaps you use. She stressed the importance of knowing your bugs, but also being aware that you, and others, may not know when you get a new bug. Therefore precautions are recommended to protect yourself and others at all times. Dr. Burns participated in the 2002 CFF Consensus Conference that established new infection control guidelines for CF Centers.

Dr. Laura Bachrach, Professor of Pediatrics and Endocrinology at Stanford spoke about bone health and CF, noting that bodies build bone up to the age of 29, with bone loss generally occurring after that age. Persons with CF are at higher risk for osteoporosis, however studies have found that if body mass is maintained their risk is no higher than the general population. She stressed the importance of weight-bearing exercise such as

jumping, dancing and running (walking & swimming are not as effective) to maintain bone strength. At least 800 international units (IU) of Vitamin D are needed daily, with as much as 300-400 IU more needed by persons with CF on high salt diets since this causes loss of Vitamin D. 1300 mg per day of calcium are also needed (one glass of milk=300 mg).

Dr. Winston Vaughn, Assistant Professor of Surgery at Stanford discussed CF sinus infections and options for treatment. He showed a video of the endoscopic nasal surgery techniques that he has pioneered at the Stanford Sinus Center.

From our CF Center, Dr. Henig discussed reproductive issues, Dr. Moss spoke on new research, Anna Simos described the new CF Diabetes Center, Mary Helmers presented a survey on adult CF issues, and Julie Matel spoke on the basics of nutrition. We hope you will join us next year at this full day on the latest issues in CF research and clinical care.

Another picture: Dr. Ross or group?

## STANFORD CF IN THE NEWS

Dr. Noreen Henig presented "Cystic Fibrosis: No Longer just a Childhood Disease" at Stanford Internal Medicine Grand Rounds on May 16.

The presentation focused on what physicians of all specialties need to know about caring for the aging CF patient, including common complications, increases in late diagnoses and new treatment regimens.

Dr. Rick Moss gave Grand Rounds lectures on CF at Los Gatos Community Hospital in March and Santa Clara Valley Medical Center in May. His chapter on allergic bronchopulmonary aspergillosis (ABPA) will appear in a new book on CF to be published by Humana Press in August. Dr. Moss co-chaired the CF

Foundation's Consensus Conference recommendations on diagnosis and treatment of ABPA and he will present the report at the North American CF Conference in New Orleans in October.

Zoe Davies, MS, PNP, RN, Research Coordinator published a continuing education article for nurses entitled "Cystic Fibrosis: No Cure Yet, But Treatment Advances Continue" in the May 6, 2002 journal Nurseweek.

## Research News

### INFANT PULMONARY FUNCTION PROGRAM OPENS

Stanford's infant and toddler pulmonary function program opens this summer, with initial tests to be dedicated to the CF research program. The new equipment, one of only eight sites in the country, allows physicians to measure lung function in children previously too young for standard pulmonary function testing. The program builds upon the generous support of The Hedco Foundation. Infants and children up to about 40 pounds and 30 inches in height can be tested by this new technique. The equipment will also be available for routine diagnostic testing. The test takes about 20 minutes once the child is asleep. A sedative is given to induce sleep and a state of deep relaxation so that lung capacity and other measurements can be made accurately without the child waking up. Dr. Carol Conrad directs the program and is the principal investigator on the research. If you have an interest in enrolling your young child, or in obtaining a baseline measurement of lung function, please contact research coordinator Colleen Dunn or discuss it with your physician.



### INFORMED CONSENT FORMS NEEDED FOR DATA COLLECTION

New research and reporting rules require us to obtain patient and minor's parents' signed permission to include clinical information in two ongoing epidemiological studies, the national Cystic Fibrosis Foundation Registry and the Epidemiological Study of Cystic Fibrosis. These registries, which have collected data on the national CF population for many years, have led to almost all of the important advancements in understanding and treatment of CF. You only need to sign a consent form to be included in these studies. The registry is also one way the CFF determines the number of patients at a CF Center and establishes funding and program participation levels. We will be asking all patients to sign the consent form as soon as possible, and are happy to answer any questions you may have about use of the data.

### ACTIVE RESEARCH RECRUITMENTS

We are actively recruiting subjects for the following trials:

#### New Trials:

- CF.Doc internet pilot project, an internet model of clinical care for pediatric & adult patients of Stanford's CF Center
- BILL study to determine the safety and tolerance of repeated doses of a previously tested once daily medication when given every day for 15 days. The medication is thought to reduce the lung inflammatory response to infection. Open to children over age 6 and adults
- Infant and toddler pulmonary function testing (newborns to 3 years of age)
- Duration of Pseudomonas Aeruginosa eradication by TOBI in children under age 6

#### Ongoing Trials:

- Cystic Fibrosis Once Daily Aminoglycoside Collaborative Trial (CFODACT)
- Multicenter, Double-Blind, Placebo-Controlled Phase II Study of Aerosolized tgAAVCF (gene therapy) in patients with Mild CF (enrollment closed)
- Standardization of Measurement of Nasal Membrane Transepithelial Potential Difference
- Diabetes Therapy to Improve Body Mass Index and Pulmonary Function
- A Phase I/II Study of Interferon Gamma-1b by Inhalation for the Treatment of Patients with Cystic Fibrosis
- Hi-D FACS with CF blood & lung leukocytes of chronic oxidative stress in CF (cell samples needed)
- Health Buddy telephonic monitoring and health education program.

Please consider participating in our research studies since it is through your efforts that better treatments will become available. Ask your physician or call our research staff if you, or someone you know, is interested in learning more about participation. Trial participants do not have to be patients of Stanford CF physicians.