

**CENTER HOSTS
NEW PARENT
GET-TOGETHER**

In June we held our first luncheon for parents of newly diagnosed children. Joanne Asano, Center Social Worker, organized the event as a joint effort of the CF Parent Advisory Group and the LPCH Family-Centered Care Program. Veteran parents Kathleen Flynn, Siri Vaeth and Sandy Schumacher shared strategies for managing the ups, downs and daily routines of CF. New parent Christina Paul benefited from the warmth and information provided by the group. She said it helped alleviate her sense of isolation to find parents who were willing to share tips on working with schools, helping a child with social issues, and learning to deal with complex medical issues. If you wish to participate in a future new parent get-together, please call Joanne Asano at 736-1905.



Kathleen Flynn, parent mentor in the Family-Centered Care Program, with son Devon Wakefield.

**CYSTIC FIBROSIS
CENTER AT STANFORD**

Center Physicians

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

Important Phone Numbers

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
LPCH 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.

Fall 2002

Cystic Fibrosis Center News

Teen Mat Vitousek wears a mask in the clinic common areas to minimize the risk of cross infection among persons with CF.



Bacterial Resistance and Infection Control

One of our CF Center's primary concerns is reducing the risk of transmission of antibiotic resistant bacteria among people with CF. We strive to foster a sense of support and community. However, the importance of infection control in the clinic and hospital setting cannot be overstated. Both our Center and the CF Foundation (CFF) have adopted new measures to address these concerns. The following information will explain some of the factors that lead to our new policies and procedures.

WHAT DOES BACTERIAL RESISTANCE MEAN?

It is essential to understand what is meant by bacterial resistance to antibiotics. When a sputum or other specimen is analyzed by a laboratory, a culture is grown to identify the type of bacteria present and what classes of antibiotics will effectively treat it. Sensitivity and resistance are the terms used to describe whether a specific antibiotic will work against an organism. There are 4 categories of bacterial sensitivity to antibiotics that are reported by the lab:

- **Pan sensitiv e** means the identified bacteria can be killed or its growth can be inhibited by all the antibiotics tested.
- **Sensitiv e** means the bacteria is sensitive to **several** of the tested antibiotics, but it may be resistant others.
- **Multiresistant** means the bacteria is resistant to all antibiotics in **two or more classes of drugs**. Currently 3 classes of antibiotics are used to treat *Pseudomonas aeruginosa*.
- **Pan resistant** bacteria are resistant to **all** tested antibiotics.

Fall 2002
 Cystic
 Fibrosis
 Center News

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



Lucile Packard Children's Hospital

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CASE STUDY

The following is an antibiotic sensitivity report for fictional patient Jane Smith whose expectorated sputum culture grew three strains of *Pseudomonas aeruginosa* (PA). It illustrates the antibiotic sensitivity pattern for only 1 of the 3 strains of PA obtained from Jane's culture report:

3+ mucoid *Pseudomonas aeruginosa*

Tested Drugs	Kirby Bauer	Results
Ticarcillin	Resistant	
Piperacillin	Resistant	
Ceftazidime	Intermediate	
Ciprofloxacin	Resistant	
Imipenem	Resistant	
Gentamicin	Resistant	
Tobramycin	Resistant	
Amikacin	Resistant	
Aztreonam	Intermediate	
Cefepime	Sensitive	
Meropenem	Resistant	

Tobramycin	E test (not yet FDA approved)
MIC 32	No interpretation

*Kirby Bauer is the name of the testing system used to determine antibiotic sensitivity.

Masks...



Nurse coordinator Nicole Eden reviews test results with Renee Guzman.

All contacts by caregivers should be preceded by proper hand washing and proper cleaning of instruments.

How is this bug characterized, how should we treat Jane, and what is the appropriate infection control protocol for Jane on her next clinic visit?

The PA in question is characterized as follows: First, it is described as "mucoid". This defines the way it grows in culture, and distinguishes it from "smooth" and "rough" strains. Mucoid PA is thought to be naturally more resistant to antibiotics. Next, the PA is characterized by how many organisms grew: the "3+" describes the **quantity** of bacteria that grew in the culture. The quantification system is crude, and the scale goes from 1-4. Finally, the PA is characterized by its pattern of sensitivity and resistance. In this case, the organism would be classified as multidrug resistant because it is resistant to two categories of antibiotics, the quinolones (ciprofloxacin) and the aminoglycosides (amikacin, gentamicin, tobramycin). It is sensitive to one beta-lactam (cefepime) and intermediately sensitive to two others (aztreonam and ceftazidime). The fact that it is sensitive to some beta-lactams, even if not all beta-lactams, keeps this organism from being classified as pan resistant.

The next question is how to treat Jane. First, it is very important to recognize that multidrug resistant bacteria are almost always treatable! The decision on what to use will be made based on her symptoms and other data, such as her pulmonary function tests. If she needs aggressive therapy, her physician would choose one of the drugs that she is sensitive to and likely add a second agent to help the first antibiotic do a better job, a phenomena called **synergy**. If Jane has only a few symptoms, her physician may choose TOBI®, an inhaled form of tobramycin. Although the conventional lab test shows resistance to tobramycin, the "E test" done at Stanford (not com-

monly done elsewhere) shows the bug can be killed by tobramycin if a concentration of 32 micrograms per milliliter [mcg/ml] can be achieved at the point of infection (her lungs). It is difficult to do this by IV tobramycin without risking toxicity to kidneys or the inner ear, but it is easy with twice daily inhalation of 300 milligrams of tobramycin solution—a standard TOBI® dose. TOBI® produces a sputum level close to 1,000 mcg/ml, far above that needed to kill the bug. The downside of using inhaled TOBI® is that it is not necessarily distributed evenly throughout the lungs and may not get to the sites of worst infection.

The final question is what infection control measures are needed to protect other visitors to our CF Center. Since Jane's culture shows the presence of a multidrug resistant bacteria, our infection control policy requires her to wear a mask in the common areas for clinic visits, and if hospitalized, she will have respiratory isolation procedures in place.

THE IMPORTANCE OF CENTER-BASED CARE

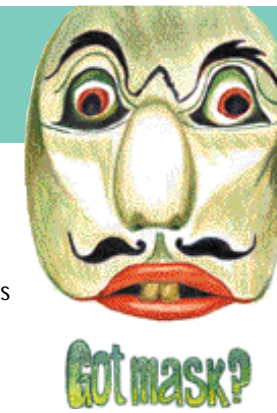
This example illustrates the value of receiving care and bacterial testing at a qualified and accredited CFF Center since many community hospitals do not routinely identify and test for the varieties of bacteria that are found in persons with CF. Accurate antimicrobial testing is critical to ensure effective treatment and accurate assessment of resistance. The CF Foundation accreditation process



involves extensive review of laboratory procedures and testing equipment to facilitate identification of problematic bacteria, and to identify the optimal drugs for treatment.

THE CONCEPT OF RESISTANCE: A RED HERRING IN CF?

Scientists are challenged to define what resistance means in the world of CF. The word resistance refers to lab tests done in a liquid suspension with a set "dose" of bacteria. Defined cutoffs between "sensitive" and "resistant" refer to levels of that antibiotic traditionally associated with curing systemic (i.e. blood-borne) infections. From a treatment perspective, we know in CF that the conventional term "resistance" is questionable. Even if the lab classifies a bacterium as "resistant," it doesn't always mean a drug will not work. Many conditions cannot be analyzed in the lab. Also, the labs test individual organisms rather than groups of organisms, and we know that PA likes to grow as large groups in the lung. It is like testing the strength of a country by fighting only one citizen! Finally, airway clearance, which rids the body of the infected sputum, assists the drug treatments so an antibiotic may not need to completely kill a growing organism for it to alleviate acute symptoms if it is combined with airway clearance.



that are each tested for antibiotic sensitivities, plus cultures obtained for molds, mycobacteria, etc. and the many other tests we monitor.

Our staff focuses priority on prompt identification and informing people who convert from a negative to a positive culture for PA so that we can try to eradicate it. (This is sometimes possible in infants and young children but rarely in adolescents or adults.) In older people, it often is not possible to fully eradicate infections, thus treatment is aimed at controlling the quantity of bacteria, and we expect culture results to be similar over time. We do not routinely inform you of each culture result in these cases, although we are always willing to share results at clinic visits. This is because, besides our clinic volume, we know that a typical bacterial strain that has chronically infected an individual for many years may go from "resistant" to "sensitive" and back again over time, confusing many people and raising needless anxiety. Instead, we focus on dealing with the health of each individual at each clinic



INFECTION CONTROL PROTOCOLS

Infection control procedures are employed to minimize the risk of transmission of bacteria. Our clinic has adopted what are known as universal direct and indirect contact precautions for ALL persons with CF to minimize risks. The simplest way to describe this is to imagine a three foot zone around your body that should not be entered by another person with CF (unless you live with them). Also, there should be no shared object used without proper cleaning in between uses. All contacts by caregivers should be preceded by proper hand washing and proper cleaning of instruments such as stethoscopes.

We are concerned about acquisition and spread of PA and other bacteria, and these new measures are designed to reduce the chance of spread. For multidrug resistant and panresistant bugs, we have added precautions that include use of a mask when in the clinic and hospital common areas, and isolation when hospitalized. This is done to reduce the chance of transmission, not because the person is more ill! The mask is an additional level of precaution, but contact precautions are the key element for ALL persons with CF. We encourage our patients to protect themselves as well as others in the clinic and hospital, since there is no way of knowing when someone has acquired new bugs or new resistances.

REPORTING CULTURE RESULTS

We track levels of resistance in all cultures and strive to inform you promptly of results. In practice, this is very difficult. LPCH/Stanford follows over 230 persons with CF, on average 4 times annually, yielding 880 "surveillance" bacterial cultures alone. Add to this cultures obtained when patients are sick and under treatment. Each specimen often contains multiple bacterial strains

visit, and more particularly when aggressive treatment is indicated.

The infection control guidelines currently being prepared for publication by the CFF do not distinguish between persons with sensitive or resistant PA outside health care settings, but rather recommend uniform infection control behaviors for all persons with CF. Our Center urges adherence to universal precautions (the three foot zone) both inside our facility and in your everyday life. Cultures from your last report are checked before the next clinic visit so we can institute appropriate infection control for that visit. The Center will be sending a letter that informs people with resistant organisms that they are required to wear a mask in the common areas at all clinic visits. Resistant organisms are considered multidrug resistant PA, *Stenotrophomonas maltophilia*, *Achromobacter xylosoxidans*, *methicillin-resistant Staphylococcus aureus*, and *Burkholderia cepacia*. We hope this system minimizes surprises about our infection control measures. It is critical to remember that a "resistant" bug report does not and should not be taken as a bad thing in and of itself, and that our main interest is focused on protecting everyone who seeks care in our Center. The new policy will provide notification in advance of a clinic visit of any carriers of resistant bacteria that will need to adhere to the mask policy. As always, we encourage everyone, not just those who have cultured resistant bugs, to be pro-active in the use of masks and the universal direct and indirect contact precautions that have been shown to reduce the transmission of bacteria among people with CF. Masks are available at the Clinic E check-in desk.

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.

With health plan open enrollment approaching, we have some pointers on selecting a health plan that may facilitate access to your preferred caregivers.

What is the best health plan for someone with CF?

Many factors go into selection of a plan: cost, participating physicians and hospitals, quality of service and prior authorizations to name a few. If you want access to our CF Center, you should look at which plans are accepted by *both* the hospital and the physicians, and what authorizations you need to see us. Most HMOs require prior authorization to see a specialist, and many consider our physicians “out-of-network” and subject to a higher co-payment. Point of Service and traditional insurance plans often let you seek care from any doctor or hospital without prior approval, though often with higher prices for non-network affiliates. Also consider:

- Drug co-payments and formularies (covered drugs), especially since many CF drugs such as TOBI® and Pulmozyme have no generic equivalents.
- Prior authorization process (frequency, comprehensiveness)
- Referral contracts with other specialty centers for CF care and transplants.
- Covered services: diabetic supplies or enzymes? Home care or medical equipment?
- Frequency of visits with a CF specialist?
- Deductibles and co-payments?

In some cases, California Children’s Services and GHPP will cover services such as equipment and home care if your plan does not. Joanne Asano, our social worker can help you navigate the process.

SUMMARY OF YOUR RIGHTS FROM THE CALIFORNIA PATIENT’S GUIDE

- You have the right to receive uninterrupted care from your doctor and HMO and to be referred to other health care providers when necessary.
- You have the right to receive a second opinion when you or your doctor request one.
- You have the right to receive an authorization from your health plan for referral to a specialist within 3 days.
- You have the right to have your doctor freely discuss your medical treatment options and care with you, without interference or restrictions by your health plan.

See www.calpatientguide.org for more detailed explanations of your legal rights.

Do I have the right to continue health care with my current doctor if I change plans?

If you have a serious condition like CF, a plan is required to provide you with continuity of care and appropriate providers. This does not mean they must refer you to the same physician. If a contract with your physician is discontinued, the plan must give you 30 days notice and provide you with services for up to 90 days or the amount of time needed to transfer your care to another doctor. Plans are required to file with the state a written plan for continuity of care and referral to other providers for acute conditions that explains how requests are reviewed and that takes into account the effects a change of doctor could have on treatment for an acute condition. Many HMOs have contracts with specific physicians or other CF or transplant centers which make it difficult to receive care at our Center. It is best to review the list of plans accepted by Stanford/LPCH physicians and hospitals before selecting a plan. This is particularly important if you are on, or anticipate being on, a transplant waiting list.

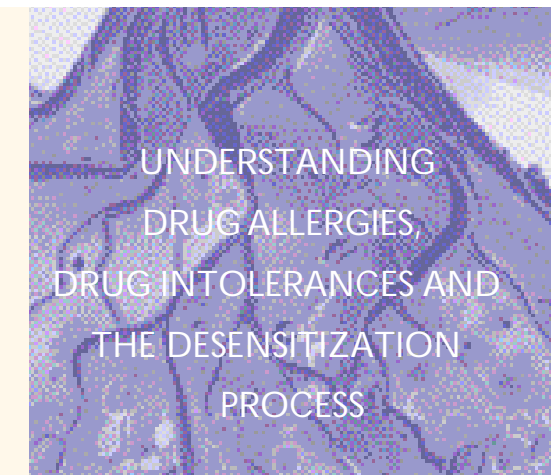
What health plan will ensure access to Stanford/LPCH physicians and hospitals?

Stanford and LPCH hospitals hold separate contracts from Stanford physicians, thus it is very important to check which plans are accepted by each organization. 2003 contract renewal negotiations are underway, however we don’t anticipate major changes. Our hospitals have contracts with most major health plans, HMOs and insurers. Our physicians have contracts with many major insurers, however we have only a limited number of HMO contracts. We have referral contracts with some HMOs and medical groups that allow access with prior authorization. It is best to check the following websites, call the hospital, and check with your primary care physician since contracts can change:

For Stanford Hospital: <http://www.stanfordhospital.com/forPatients/financial/healthPlansSUH.html>

For Lucile Packard Children’s Hospital:
<http://www.lpch.org/ForPatientsVisitors/FinancialInsurance/healthPlansLPCH.html>

For Stanford and LPCH Physicians:
<http://www.stanfordhospital.com/forPatients/financial/healthPlansSFP.html>



Drug allergies are among the most confusing issues in medical treatment. CF often requires intensive and prolonged use of many drugs, thus it is important to understand the difference between drug allergies and drug intolerances, and what can be done to minimize adverse effects.

Drug allergies result when the immune system of the body reacts against a specific drug or class of drugs. There are two basic categories: immediate hypersensitivity and delayed hypersensitivity. Immediate hypersensitivity, also called anaphylaxis, is a severe reaction that results in swelling of the lips and throat, low blood pressure, and difficulty breathing. It can happen the first time a drug is given, or more commonly, upon subsequent treatments (often the first dose of the next course). This is a serious but rare form of allergy that can be life-threatening if untreated.

The second category of drug allergy, delayed hypersensitivity, is less severe and more common. These manifest as rash, fever, malaise, and/or joint pain. The rash can be quite itchy and red, and continued administration of the drug can make it worse. Drug rashes often appear after a few days though they can occur after a few weeks of treatment, and even after finishing a drug course. Once someone has developed a rash to a specific drug, the next time she receives that drug, the rash is likely to appear sooner and may be more severe.

Drug Intolerances

Drug intolerances are defined as reactions that make someone feel poorly, however they are not usually mediated by the immune system and are not considered a drug allergy. Some common drug intolerances include vomiting with codeine, wheezing with TOBI®, or joint pain with ciprofloxacin. Drug intolerances are unpleasant but not life threatening. They are also much more common than true drug allergies.

What Treatments are Available

With most drug allergies, the safest course of action is to avoid the drug. Sometimes this isn’t possible if that drug is the most effective choice to control the bacteria causing symptoms. Various treatments can often minimize adverse effects while allowing the best treatment for the infection to proceed.

Desensitization is recommended for patients with a documented history of true anaphylaxis or immediate hypersensitivity. Desensitization involves giving increasing amounts of the drug up to the full dose over a 6 hour period. This treatment must take place in the hospital with proper monitoring for signs of serious reaction, and to provide a strict dosing schedule. If a reaction is going to occur, it is usually less severe than full anaphylaxis and can be treated with Benadryl™ or a corticosteroid. Patients who tolerate desensitization can receive the drug in its full amount, although 25-30% may develop a rash during the treatment. Missing a dose during the treatment is like starting over, and requires desensitization again. Desensitization does NOT prevent the second category of drug allergies, delayed hypersensitivity, and it must be undertaken each time a new course of that drug is initiated. Delayed hypersensitivity reactions do not require desensitization for each new course of the drug that caused the reaction. Often, an antihistamine (e.g. Benadryl™) or a corticosteroid prevents or minimizes this type of allergy.

Drug intolerances are treated by methods specific to the intolerance, such as anti-nausea medicines for drug-induced nausea. There are persons for whom certain drugs should absolutely be avoided, even when the reaction is considered intolerance. *Desensitization never prevents drug intolerances.*

Cross-reactivity, allergies to all medicines in one class of antibiotics, is another confusing

topic. Many antibiotics such as penicillin are in the beta-lactam class. It is very rare for a person to be allergic to all beta-lactams. A person allergic to penicillin is likely to be allergic to amoxicillin and ampicillin, however he usually can tolerate the beta-lactams used to treat persons with CF, such as piperacillin, ticarcillin, and ceftazidime. Also there is very little cross-reactivity found in the cephalosporin group of drugs. Some drug intolerances are more likely to be seen within a group of drugs, for example ringing in the ears from tobramycin is likely to be experienced with other aminoglycosides such as gentamicin and amikacin.

It is often challenging to determine if a person has experienced an immediate or delayed hypersensitivity reaction or a drug intolerance. Physician or hospital-documented reactions are the most reliable source of information since vital signs and symptoms have been observed and recorded in a person’s medical record. Patient-reported descriptions are also helpful, though it is often difficult to identify or remember all of the specific symptoms. It is common for a person to recall a “bad” experience as a youngster with a specific drug without remembering the details.

A New Treatment Approach

We are adopting a new approach to identification and treatment of drug allergies and intolerances. In the past, we performed a large number of desensitizations when the history was questionable or there was cause for concern of cross-reactivity. We think this has resulted in over treatment for many patients who only experienced delayed hypersensitivity or drug intolerances rather than the more serious immediate hypersensitivity, which can and must be treated with desensitization procedures. This over treatment caused unnecessary hospitalizations and desensitization treatments, and precluded treatment that could be effectively managed without desensitization.

For patients with a history of multiple allergies, we will perform drug challenges in the hospital under observation. Patients will receive a test dose of the drug in question. The test dose is a partial dose of the medicine that will allow us to observe any form of reaction—itchy throat, redness, tingling, fever, etc. If any of these occur, a full dose of the medication will not be given without a desensitization procedure. By definition, the test dose is looking for immediate hypersensitivity reactions, the type of reactions that desensitization can prevent. If no immediate reaction occurs with the test dose, then we will proceed to give the medication at the therapeutic dose and schedule. The test dose will not identify patients who have delayed hypersensitivity; but remember, desensitization would not prevent this type of reaction anyway. A list of test doses and reactions will be documented separately, so that a test dose does not need to be done each time the drug is given if there has been no immediate reaction. If you have questions about the new procedure or questions about your own history of drug allergies, intolerances or side effects, please discuss it with your doctor at your next visit.

HEDCO FOUNDATION GIFT TO FUND INTEGRATED PULMONARY ASSESSMENT PROGRAM FOR THE PRESCHOOL CHILD

We are pleased to announce a generous gift from the Hedco Foundation to the Lucile Packard Foundation for Children's Health to develop an integrated pulmonary assessment program for preschool children. Young children diagnosed with CF and other lung complications such as recurrent pneumonia or gastroesophageal reflux will benefit from the new program. Adult-style pulmonary assessments require patients to cooperate fully and repeat test procedures in a consistent pattern, and to date there have been limited diagnostic tools for children in the 2 to 6 age range.

The new program will have state-of-the-art pediatric bronchoscopy, pulmonary function testing and imaging analysis tools to enable us to diagnose, monitor disease progression, and assess the benefits of new treatments. For example, clinicians may be able to directly assess the efficacy of gene therapy in preschool children. We believe the new program will offer a unique integration of techniques not previously available for preschool children: bronchoscopy combined with autofluorescence provides a view of the inside lining of the airways and access to fluids, cells, and tissues. Pulmonary function tests provide a global assessment of lung function and disease process. Imaging analysis allows detailed viewing of lung structures without invasive procedures. Synthesizing the results of each of these assessments will facilitate more precise and effective care for children in this previously difficult-to-assess age group. The Hedco Foundation gift will be matched by the Campaign for Lucile Packard Children's Hospital to provide the equipment, space, technical staff and support that will allow continued development of the LPCH Pediatric Pulmonary and Cystic Fibrosis Center of Excellence.



Assistant Professor of Pediatric Pulmonary Medicine Terry Robinson will continue development of diagnostic imaging tools as part of the Hedco gift.

NEW ENDOCRINOLOGIST JOINS STANFORD TEAM

Tracy McLaughlin, M.D. has joined Stanford as an expert in glucose intolerance and osteoporosis. Dr. McLaughlin will work with our CF and transplant physicians and diabetes educator Anna Simos in the clinical management of endocrine-related CF issues. Dr. McLaughlin attended UCSF School of Medicine, with post graduate training at Santa Clara Valley Medical Center and Stanford. She has performed clinical research on insulin resistance, diabetes, and heart disease at Stanford. She joins Stanford as a clinical instructor, senior clinical research associate, and practicing diabetologist. She has an interest in osteoporosis prevention among persons with CF, complementing the efforts of pediatric endocrinologist Laura Bachrach. Dr. McLaughlin holds weekly clinics at Stanford and is available for inpatient consultations. She has received research awards from the American College of Physicians, the Endocrine Fellows Foundation and most recently, a Career Development Award from the National Institutes of Health. She looks forward to working closely with the CF team in providing comprehensive expertise in the management of all aspects of CF.

STANFORD CF IN THE NEWS

Stanford participants at the October 2002 North American Cystic Fibrosis Foundation Conference included Terry Robinson, M.D. who presented to the Insights into CF Lung Disease Symposium on "Composite CT/PFT Score: An outcome measure which markedly improves sensitivity to change in early cystic fibrosis lung disease"; the first scoring system that combines High Resolution Computerized Tomography (HRCT) component scores with pulmonary function measurements. Dr. Robinson also presented a poster on Quantitative HRCT air

trapping analysis in CF subjects with mild lung disease during the Pulmozyme® early intervention study, a research project in which many of our Center patients participated. Noreen Henig, M.D. and Penny Stroud presented a poster on "Influences on Access to CF Center-Based Care in California"; an analysis of hospitalization and treatment trends. Richard Moss, M.D. chaired the NACFC symposium on Diagnosis and Treatment of Allergic Bronchopulmonary Aspergillosis in CF and Asthma. He also presented the results of the Multicenter, Double-Blind, Placebo-

Controlled Phase II Study of Aerosolized tgAAVCF™ (gene therapy) in patients with Mild CF, the results of which will be discussed in an upcoming issue. Jeff Wine, Ph.D. co-chaired the Symposium on Proteomics: CF and the Proteomics Approach and also served on the program committee for this year's conference. This year most of the Stanford CF clinical and research teams attended the conference to learn about new directions in research and treatment.

We are actively recruiting subjects for the following trials:

NEW TRIALS:

- Infant and toddler pulmonary function testing (children ages 6 months up to 40 pounds & 30 inches height).
- Concentrated TOBI®: Comparison of Safety and Delivery Time of a Concentrated Tobramycin Solution for Inhalation in Infants and Children with CF, ages 6 months to 12 years.
- TheraCLEC® Total: An Open-Label Safety and Tolerability Study of Oral TheraCLEC® Total Enzymes (requiring two 5 day in-patient admissions) in CF Patients Ages 13 to 45 with Pancreatic Insufficiency Secondary to CF.
- Topical Aminoglycosides: Study of Topical Aminoglycosides to Activate CF Genes in Persons with CF with Premature Stop Codon Mutations in the CFTR Gene. (Also seeking control subjects with CF who have two identified CFTR mutations, neither of which is a premature stop mutation). Open to children over age 6 and adults.
- CF.Doc Internet Pilot Project, an Internet Model of Clinical Care for Pediatric & Adult Patients of Stanford's CF Center.

ONGOING TRIALS:

- Duration of *Pseudomonas aeruginosa* eradication by TOBI® in children under age 6.
- BIIL Study to Determine the Safety and Tolerance of Repeated Doses of a Previously Tested Once Daily Medication when Given Daily for 15 Days. The medication is thought to reduce the lung inflammatory response to infection. Open to children over age 6 and adults.
- Cystic Fibrosis Once Daily Aminoglycoside Collaborative Trial (CFODACT).
- Standardization of the Measurement of the 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 and contributing to the search for better treatments. Ask your physician or call our research staff if you are interested in learning more about participation. You do not have to be followed by our CF Center physicians to participate in most research studies.

Active Research Recruitments

DR. HENIG MATERNITY LEAVE

Adult CF Center Director Dr. Noreen Henig begins a four month maternity leave November 1, 2002, ending February 28, 2003. Adult patients are encouraged to schedule quarterly visits and GHPP annual visits prior to her leave or soon after her scheduled return in March 2003. Drs. Conrad, Robinson, Moss and Witcoff will be seeing adult patients for urgent needs in addition to hospitalized patients. Nurse coordinators Mary Helmers and Nicole Eden will be available as usual.