

# CENTER FOR CYSTIC FIBROSIS USC UNIVERSITY

## FALL 2009 NEWSLETTER

### Welcome to the New Center for CF USC University Newsletter

**The purpose of the newsletter** is to help keep you up to date with what is happening at our clinic at USC. The most recent news is that the clinic has developed a Patient Advisory Committee (PAC) in the Fall of 2008. The PAC was developed to provide patients with the opportunity to collaborate with the CF team in order to improve the quality of care provided by the center. Increased involvement of patients can facilitate the enhancement of the care center's ability to provide responsive care. Overall, more responsive care results in greater patient satisfaction with their care center. It was also developed with the purpose of making life a bit easier for all the patients represented here.

The CF PAC team includes several clinic patients who have been meeting monthly on conference calls with Debbie, Lorelie and sometimes Dr. Rao and Paul.

We have been working on a Patient Handbook that is full of information about the clinic, transitioning to the clinic if you are a new patient from a children's CF center or another doctor or center, information about being an inpatient here at the hospital as well as a wealth of other things.

We have developed a Wallet Card to list medications you are using, a Yearly Test Form which you can keep with you and hopefully has all that medical info at your fingertips. We have developed our own clinic web site which will be a one stop location for clinic and hospital information, but also will include updates on research opportunities, a FAQ section, upcoming events and links to other sites, among other topics.



### Success Stories

**Everyday those living with Cystic Fibrosis** achieve something they never deemed possible or realize a goal they set out for themselves. Here at the Center for Cystic Fibrosis at USC University Hospital we want you to share those very stories of inspiration, accomplishment, and success that you are proud of. We will highlight these stories in the hopes that our patients can find encouragement and inspiration in the success of their peers, to open the doors of possibility and achievement within their own lives.

### Your Cystic Fibrosis Team

A. Purush Rao, M.D.— Director: 323-442-5100

Kamyar Afshar, D.O.—Ass. Director: 323-442-5100

Debbie Benitez, RN, MSN,ACNP—Nurse Coordinator: 323-442-8522, email: [debbie.benitez@health.usc.edu](mailto:debbie.benitez@health.usc.edu)

Paul Beringer, Pharm.D—Clinical Pharmacist: 323-441-1401, email: [beringer@usc.edu](mailto:beringer@usc.edu)

Heather Owens, Pharm.D.—Fellow: 323-442-2631, email: [howens@usc.edu](mailto:howens@usc.edu)

Lynn Fukushima, RN, MSN, CCRC—Clinic Nurse, Clinical Nurse Research Coordinator: 323-442-8522, email: [LynnK.Fukushima@health.usc.edu](mailto:LynnK.Fukushima@health.usc.edu)

Stephanie Low, RD—Dietitian: 323-442-5308, email: [stephanie.low@health.usc.edu](mailto:stephanie.low@health.usc.edu)

Christina Estrada—Medical Assistant: 323-442-8522, email: [christina.estrada@health.usc.edu](mailto:christina.estrada@health.usc.edu)

Lorelie Evangelista, MSW—Social Worker: 323-442-8358, email: [Lorelie.evangelista@health.usc.edu](mailto:Lorelie.evangelista@health.usc.edu)

Autumn Clark – Research Coordinator: 323-442-9580, email: [autumncl@usc.edu](mailto:autumncl@usc.edu)

## Medication & Equipment



### *Which nebulizers are you suppose to use for which meds?*

Answer: You should have a nebulizer for each inhaled medication prescribed to you. For example, if you are prescribed 4 inhaled medications, then you should have 4 nebulizers. Nebulizers must be sterilized after each treatment to reduce bacteria.

### *What do all of the inhaled medications do? In what order are they supposed to be taken?*

Answer: Inhaled medication should be done in the following order:

1. Bronchodilators (Albuterol) – open the airways to help clear mucus and make breathing easier. They are given **before** performing ACTs.

2. Hypertonic Saline – helps to stimulate cough and to moisten thick mucus in the airways.

3. Pulmozyme – helps to thin the mucus so it is easier to cough up. Some patients switch the order of Pulmozyme and Hypertonic Saline.

4. TOBI or Colistin – antibiotic that helps to fight infection, this medication should be taken **after** performing ACTs

5. Advair or other inhaled steroid before or after Tobi or Colistin.

The Vest or other Airway Clearance techniques (ACTs) should not be done after or during inhaled antibiotics or steroids.

## Mentorship Program

The Peer Mentor Program was established in July 2008. The purpose of the program is to improve the quality of life of other patients with cystic fibrosis by providing peer support and encouragement. Specifically, the goal is to enable patients with CF who would like to receive more information or support regarding specific issues to be able to contact another CF patient who has experience in those areas. The mentors are identified by the CF Team and there may be one or more mentors for certain categories such as those listed below:

### Mentor Categories

- |                            |                      |                       |
|----------------------------|----------------------|-----------------------|
| 1. Travel                  | 6. G-Tube            | 11. Bipap             |
| 2. Disability              | 7. Portacath         | 12. CFRD              |
| 3. Family Planning         | 8. Center Transition | 13. Medications       |
| 4. Career & CF             | 9. VEST              | 14. Exercise          |
| 5. Transplant (Pre & Post) | 10. Oxygen           | 15. Daily Maintenance |

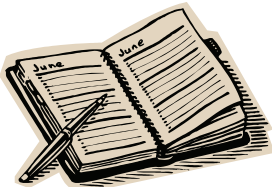
Any patient interested in speaking to a Mentor should contact Debbie or Lorelie. They will then contact the Mentor. Only first names and contact information will be disclosed to the Mentee and Mentor. Communication will be by email or phone only.

## Clinical Trials and Research

### **Bayer/ Ciprofloxacin Study**

The intent of this study is to evaluate the use of ciprofloxacin in an inhaled, powder form, making it more easily accessible to the lungs. There are 4 weeks of active treatment with study visits once a week, and one brief visit conducted by phone. If necessary, there can be a 30 day washout period, (prior to active study drug) for subjects who are taking exclusionary medications. After the active drug period, there are 2 follow-up visits. There are 8 In-Clinic visits total. You will be randomly assigned to receive either cipro as 50 or 75 mg or matching placebo taken twice daily. Three in four patients will be assigned to active study drug.

There is no charge to participate in the study, and you will be compensated for your participation.



### **Tiger 2/ Denufosal Tetrasodium Study**

The objective of this study is to evaluate the use of denufosal in a nebulized form to aid in the clearing of mucus from the lungs. There are 48 weeks (about one year) of active treatment which include 9 clinic visits and 7 brief telephone contacts. There is one follow-up visit. There are 10 In-Clinic visits total. You will be randomly assigned to either Denufosal or placebo, administered three times daily. You will have a 50% chance of receiving study drug. There is no charge to participate in the study, and you will be compensated for your participation.

## Survey Monkey is Coming

The USC PAC has put together a Wallet Card and a Yearly Test Form. You should have received them in clinic and have had an opportunity to try them out. You should be receiving via email or snail mail a survey in the near future. Please take the time to fill that out so that the PAC can know how useful these items are and if you have suggestions to make them more useful. Our aim is to help you with some of these tools and your feedback will be helpful to us.



Please take time to fill out the Survey Monkey

## Questions and Answers

### ***What happens when you're sick and need to be admitted for IV's?***

Answer: If you are having symptoms of an exacerbation (e.g. increased cough, increased sputum, shortness of breath, recent loss of weight, poor appetite, etc) call the Nurse Coordinator at (323) 442-8522. The Nurse Coordinator will contact Dr. Rao and will submit your admitting paperwork to the Admitting Case Manager. The Admitting Case Manager will contact you as soon as your paperwork is processed and a bed is available for you.

### ***How do you contact the doctor after hours?***

Answer: You can contact the CF Center Monday through Friday from 8:00 to 4:30pm at (323) 442-8522. For after hour emergencies, you can call (323) 442-5100 and ask to speak with the Pulmonary Attending on call.



### ***What is the protocol if you have to visit the ER due to a sudden emergency? Who do you call to notify and how do you get in touch with your CF doctor once you're there?***

Answer: If you go to the Emergency Room, inform the doctor that you have CF and you are a patient at the CF Center at USC University Hospital. Ask the ER doctor to call the CF Center or the after hours phone number to speak to Dr. Rao. If your ER visit results in an admission, you can request for your care to be transferred to USC University Hospital. The doctor will contact Dr. Rao to initiate the transfer to our hospital.

## Recipes

### Date Bars

This is great with a cup of coffee or tea. And it makes a nice Christmas treat or gift.

**Preparation Time:** 20 minutes,  
**Cooking Time:** 35 minutes.  
Makes 16 –20 squares.

#### **Crumb Mixture:**

1 1/2 cups flour, all purpose  
1/2 teaspoon baking soda  
1 teaspoon baking powder  
1/4 teaspoon salt  
1 cup butter  
1 cup brown sugar  
1 1/2 cups rolled oats

#### **Date Filling:**

2 cups chopped dates  
3/4 cup cold water  
2 Tablespoons brown sugar  
Grated rind of 1/2 orange (optional)  
1 tablespoon orange juice (optional)  
1 teaspoon lemon juice

#### **Directions:**

- 1) Preheat oven to 325 F
- 2) Combine flour, baking powder, baking soda and salt. Rub or cut in butter until mixture is crumbly. Add sugar and oats. Mix well.
- 3) Spread crumbs in greased, shallow, 9-inch square pan. Pat mixture until firm and smooth.

4) Cook dates, water, orange rind and sugar in small saucepan over moderate heat until thick and smooth. Remove from heat and add fruit juices. Mix well.

5) Bake at 325 F for 35-40 minutes until lightly browned. Cut squares while hot and allow to cool in pan.

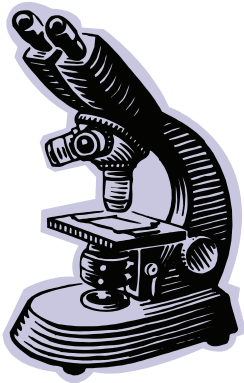
#### **Nutrition Information:**

If cut into 1 1/2 in squares;  
115 Calories, 1.1 grams protein each.  
Total calories 4,163, Protein 39 gms.



## More Research Opportunities

Our clinic will be spearheading two investigator-initiated CF research studies.



### FLU SEASON IS COMING

Influenza can worsen lung disease in CF leading to a decline in lung function or respiratory infection requiring hospitalization.

**SWINE FLU VACCINE** once available is also **RECOMMENDED** for **ANYONE** who has a chronic pulmonary disease which of course includes CF.

Be sure to get your Flu Shots as soon as they are available.

### #1 The effect of pioglitazone on sputum bio-markers of lung inflammation and remodeling in patients with cystic fibrosis.

The purpose is to determine the short-term effects of pioglitazone on the bio-markers of airway inflammation and remodeling in patients with cystic fibrosis. The pharmacokinetics and pharmacodynamics will also be assessed. The study will take place within the Clinical Trials Unit (CTU) at USC Academic Medical Center and will last for 84 days. The number of participants needed is 24. They must have CF and be at least 18 years old, have *P. aeruginosa* in the sputum and an FEV<sub>1</sub> of greater than 40% predicted.

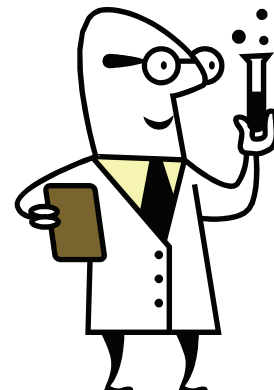
### #2 The effect of doxycycline on sputum bio-markers of lung inflammation and remodeling in patients with cystic fibrosis.

The purpose is to determine the short-term effects of doxycycline on bio-markers of airway inflammation and remodeling in patients with cystic fibrosis. The pharmacokinetics and pharmacodynamics will also be assessed. The study will take place within the Clinical Trials Unit (CTU) at USC Academic Medical Center and will last for 42 days. The number of participants needed is 24. They must have CF and be at least 18 years old, have *P. aeruginosa* in the sputum and have an FEV<sub>1</sub> greater than 40% predicted.

### The purpose of these two studies

is to determine the short term effect on airway inflammation in patients with CF and to measure the amount drug in the blood and sputum with different doses of the study medications. Sputum induction with hypertonic saline is performed in both studies periodically during the study period and the samples are then analyzed to determine the amount of inflammation. If the results with either or both of these drugs are positive the plan is to submit a grant to the CFF to perform a multi-center clinical trial.

*Continued on next page.*



## More Research Opportunities (continued)

**The investigators for both studies** are Paul Beringer, Pharm.D., Heather Owens, Pharm.D., Ayana Boyd-King, M.D., Adupa Rao, M.D., Helen Baron, M.D., Kamyar Afshar, M.D., Debbie Benitez, RN, MSN.

**The sponsor for both studies** is Webb CF Research Award.

**Some definitions for the more detail oriented**

**Pioglitazone:** a drug typically used for treatment of diabetes.

**Doxycycline:** a tetracycline antibiotic.

Both drugs are also known to have anti-inflammatory properties.

**Biomarker:** a distinctive biological or biologically derived indicator (as a biochemical metabolite in the body) of a process, event, or condition (as aging, disease, or exposure to a toxic substance), age-related *bio-markers* of disease and degenerative change. i.e. PSA in prostate cancer, high white blood

count in infection, high enzymes in MI or liver disease.

**Remodeling:** Process of structural changes or deterioration in the disease process.

## Why do I have to wear a mask and gloves at the clinic and hospital?

This is because of the risk of cross-infection.

Cross-infection is an issue of considerable concern for the CF community. Those with CF attract different bacteria or 'bugs' that grow in their lungs. These 'bugs' are rarely harmful to those who do not have CF but may be harmful to others who have CF but who do not have the same 'bugs'.

This became a real problem in the 1990s in relation to Burkholderia cepacia complex, some strains of which can cause a very serious health problem in those with Cystic Fibrosis. It was agreed that those with B. cepacia complex should be segregated from other people with Cystic Fibrosis. It subsequently emerged that there are several strains of B. cepacia complex and some are more serious than others so people with B. cepacia complex were then further segregated according to the strain they had.

Pseudomonas aeruginosa is another 'bug' people with CF regularly acquire, mostly from the environment. This can usually be eradicated or kept at bay with early antibiotic treatment. It is expected that CF patients will pick up Pseudomonas from time to time. There are thousands of different strains of Pseudomonas. If this 'bug' gets well established and starts to become resistant to antibiotics, it can cause problems.

There is some concern that CF patients are more likely to pick up strains of Pseudomonas from each other that are more difficult to treat than strains picked up from the environment. For this reason, CF Centers and Clinics are now encouraged to offer separate clinics for CF patients according to whether or not they have Pseudomonas and if so, whether the particular strain is known to be transmissible, that is, has been shown to have the ability under some circumstances to pass from patient to patient.

'Bugs' such as B. cepacia complex and Pseudomonas aeruginosa can be transmitted from person to person by close personal contact, such as sharing rooms, sharing medical equipment, sharing cutlery or crockery, and by kissing or coughing.

There is little risk of transmission of 'bugs' in an outdoor environment, but traveling with other people with CF in a car, or meeting them socially would introduce a higher level of risk.



## COME JOIN THE PAC!



## WE WANT YOU!

### WE ARE ALWAYS LOOKING FOR SUB-COMMITTEE MEMBERS FOR THE PATIENT ADVISORY COMMITTEE

The PAC or Patient Advisory Committee consists of adults with CF who receive their care at the *extraordinary* Center for CF at USC University Hospital. Our PAC for 2009/2010 is in place, but there are openings for people who might be interested in one of the sub-committees. This past year was the first year of the PAC and so far the subcommittees have developed the Wallet Medication Card, the Annual Testing Sheet, have made huge headway in developing the Patient Handbook. And the Website is almost ready to go. These and other areas could use the input of those who would like to be a part of developing new ideas to improve our quality of life by enhancing the clinic and hospital experiences. **For more information, please contact Lorelie at 323-442-8358 or email: [Lorelie.evangelista@health.usc.edu](mailto:Lorelie.evangelista@health.usc.edu).**

### Clinic Updates

- 1) New team Member Lynn Fukushima -- Clinic Nurse and Clinical Nurse Research Coordinator has joined our team!
- 2) Inpatient Hospital Rounds now on Fridays at 1:30pm.
- 3) Starting in September Portfluses to be scheduled on MONDAYS ONLY.

### Center for CF @ USC

#### CLINIC

Dohene Eye Institute, 5th Floor  
1450 San Pablo St  
CF Center: 323-442-8522

#### MAILING ADDRESS

USC University Hospital  
1500 San Pablo St.  
Los Angeles, CA 90033  
Attn: Center for Cystic Fibrosis

Though Summer is almost gone, the days can still be HOT!

Be sure to drink plenty of water. Maybe have some Gatorade or other sports drink on hand for those really hot days.

PAC Email:  
Coming Soon

CF Website

Coming Soon