7011 Thurston Bowles Building
CB#7248
Chapel Hill, NC 27599

Seasonal Tips — Flu Season

The flu season is upon us. It is important that patients with CF receive a flu vaccine each year. Our goal is to have all patients seen in the Adult CF clinic receive their flu vaccination by December 31.

The Adult CF clinic has flu shots in stock.

However, we also encourage you to contact your local primary care provider, health department and pharmacy to inquire about flu shot availability near you. The Adult CF group will be happy to provide you with a letter verifying your need to receive a flu shot. Patients with CF should not receive the nasal flu vaccine preparation. We ask that you contact Kathy Hohneker, our nurse-coordinator, when you receive your flu shot. Contact her at (919) 966-7049 or by email at khohneker@med.unc.edu.

During flu season, please remember these infection control guidelines:

- Avoid contact with people who are sick with flu or viral illness. Droplets from cough and sneezing spread the influenza virus.
- Wash hands frequently with soap and water, or with alcohol-based hand sanitizers.

Contact us immediately (919-966-4131 for the pulmonary fellow on-call) if you experience fever greater than 100.5°, chills, dry cough, body aches, fatigue and/or headache. Medications are available to help alleviate flu symptoms, but they must be started within 48 hours of the onset of symptoms.

Please contact us at (919) 966-7049 if you have any questions. These websites may be helpful:
http://www.cdc.gov/flu/keyfacts.htm
http://www/nfid.org/influenza

UNC CYSTIC FIBROSIS CENTER

Winter 2008

Quality Improvement at UNC

We hope that this newsletter finds everyone well! This newsletter was created as a way to improve communication with our patients. It includes updates on new staff and changes in CF clinic, information on new research studies, and health information pertinent to you. This is being done as part of our Quality Improvement initiative here at the UNC Cystic Fibrosis Center. You may be wondering who exactly "we" are, what this initiative is, and how it might affect you.

"We" are the Adult CF Quality Improvement team, which is comprised of CF clinic physicians and nurses, your social worker and dietician, hospital-based caregivers, research coordinators, and very importantly, CF patients. This team has grown since its inception in 2004, and our QI efforts continue to expand. Our goal is to evaluate all aspects of CF patient care at UNC in order to pinpoint areas on which we can improve in order to optimize your health and your care experience.

Most recently, our team has set out to change the way we take care of pulmonary exacerbations. Importantly, having an exacerbation is not only unpleasant to go through, but if it happens too often can cause significant and permanent worsening of your lung disease. Therefore, one of our goals is to better detect the onset of pulmonary exacerbations, so that we can initiate treatment sooner and lessen the severity and/or duration of these episodes. We also want to change how care is delivered in the hospital, when that is required to treat the exacerbation. During the upcoming months in CF clinic, your CF caregivers will give you more information on the changes we would like to make.

Since we are aiming to improve your health care, the most important input is YOURS! If there are aspects of your CF care experience with UNC which you feel need improvement or if there are topics you would like to see addressed in this newsletter, please let us know. You may e-mail these comments to pulmonary@med.unc.edu. Please also send us an email if you would like to be involved in the QI team, or would like to receive future correspondence with us about your CF Clinic and our combined efforts to improve it. Please know that your email won't be shared with anyone else, and will only be used to facilitate your care and to seek your input on our QI projects.

Best Wishes,

The Adult CF QI Team

Inside this issue:

Introducing: Jennifer Hendrix

Hello, I would like to introduce myself to you. My name is Jennifer Hendrix and I am your new CF social worker. I have been working at UNC Hospitals as an inpatient social worker for the past two years and have recently accepted the full time CF social work position. CF is new to me so I have lots to learn and am hoping you will share your knowledge with me. Most importantly, I want to know what your needs are and how I can best help you. I am excited to begin working with you.

<u>What does a social worker actually do?</u> I am here to advocate for you and address any barriers that prevent you from receiving the health care you need. I can identify and provide needed resources, make referrals, and offer emotional support and counseling as needed. I can also help with financial and insurance issues, obtaining medications, transportation problems, emotional well being, substance use, questions regarding SSI (Supplemental Security Income), SSDI (Social Security Disability Insurance), Medicare info, Medicaid info, assistance with setting up home services, end of life counseling, life changes (e.g., marriage, having children, disability, etc.), relationship issues, school/work challenges, spirituality.

I will also be available to visit you while you are in the hospital to provide support and address any concerns that you may have. Please feel free to contact me at 919-966-7873 with any questions. If there are any issues related to getting to your clinic appointment, please contact me beforehand and I will explore resources with you. I look forward to seeing you!

Respectfully,

Jennifer Hendrix, MSW, P-LCSW
JHendrix@unch.unc.edu

Need Help During the Holidays? ***Members of the CF /Pulmonary Research and Treatment Center have collected donations to help patients from our clinic who are having a difficult time affording essential items such as food, utilities, clothing and gasoline during the holiday season. Small, one-time awards may be available to assist patients with specific needs. Please contact Jennifer Hendrix (JHendrix@unch.unc.edu, 919-966-7873) or Kathy Hohneker (khohnekr@med.unc.edu, 919-966-7049) by* DECEMBER 15 *if you would like to request assistance from this fund.***

Introducing: Sarah Jones

Hi there! I am your CF dietitian specializing in adult and post-transplant care. I am excited to start working with you and your family. I completed my Master's in Nutrition at UNC and first became interested in CF while doing my final work in the pediatric and adult CF clinics. I joined the UNC Healthcare team shortly thereafter and have spent the last year working in the hospital, where I have met many of you.

My goal is to help you manage nutrition-related aspects of your CF. We often focus on your weight, which is extremely important, but there is much more: bone health, diabetes management, GI issues, enzyme use, antioxidants, vitamins, and anything else that is important to you.

Please feel free to give me feedback and/or share your ideas. I consider you my best teachers. I am always ready to learn more and find new ways to do things. I look forward to meeting you!

Sincerely,

Sarah Jones, MPH, RD
(919) 966-1294
sjones7@unch.unc.edu

North American Cystic Fibrosis Conference — October 23-25, 2008

In October, the North American CF Conference was attended by 3,800 researchers and caregivers who are committed to curing this disease and providing healthy lives for our patients. This year, results from 3 clinical trials with drugs that target the underlying problem in CF were presented. This is a tremendous advance, and makes us optimistic that we will fundamentally change the course of disease in the near future.

VX-770 (Vertex Pharmaceuticals): This drug is a pill designed to make certain mutant CF proteins function more normally. Because it is a pill, it has the potential to correct many aspects of CF, and not just the lung disease. In the study, it was taken by CF patients with a specific CFTR mutation (called "G551D") for 2-4 weeks. Patients taking the drug showed dramatic reductions in sweat chloride values, improved CFTR function in the cells that line the nose, and a trend toward improved lung function tests. Longer term studies should be initiated within the year. If you would like to learn more, go to http://www.youtube.com/watch?v=_KpTnKPi8FE to see Drs. Bob Beale and Frank Accurso talk about this exciting study.

Denufosol (Inspire Pharmaceuticals): Denufosol is an inhaled medication that is aimed at restoring normal salt and water transport in CF lungs. A large study involving 352 patients was recently completed, in which patients with mild lung disease (FEV_1 >75%) inhaled the drug or a placebo 3 times a day for 6 months. Importantly, lung function was found to be significantly improved during this time. All patients were then allowed to use Denufosol for an additional six months. During this later phase, lung function continued to show even greater improvements. Another large trial is underway to see if these encouraging results can be repeated prior to seeking FDA approval.

PTC-124 (PTC Pharmaceuticals): PTC-124 is another oral drug that is targeted against CFTR "stop mutations", which lead to an incomplete CFTR protein. About 10% of CF patients in the US have these mutations. In Israel, a 3-month long study showed evidence of normal CFTR protein production and improved functioning the cells that line the nose. Patients also were noted to have less cough, and a trend towards better lung function. The company plans a rigorous, long-term study to more definitively test their product in the coming year. Stay tuned!

Get Involved: Current Clinical Research at UNC

The most important ways to improve the lives of people with CF are to learn more about the disease and develop new therapies to treat it. Around the country and the world, physicians and researchers are doing just that! However, none of these efforts would be successful without CF patients' help. Your participation in clinical studies is one very effective way to help the fight against CF. If you'd like to learn more, go to www.cff.org or http://www.youtube.com/watch?v=FxyxWJcw4rA&feature=related to see one patient's experience in CF research. Check out the information below to see how you can be "the key" at UNC!

Mucus Clearance During Acute Exacerbations of Cystic Fibrosis (GCRC 2483)
This study is designed to understand why CF patients develop pulmonary exacerbations, by testing the idea that the lungs' ability to clear mucus deteriorates after a trigger (like a virus infection), which then leads to an exacerbation. Patients who are at least 18 years old, have an FEV_1 of at least 30%, and who have required IV antibiotics at least twice in the last 2 years are eligible. The study involves 8 visits to UNC.

Evaluating Regulators of Mucus Hydration Changes in Adult Lung Diseases (IRB 07-1778)
This study examines how the CF lung produces such a thick mucus, and how this changes across the course of disease from childhood to adulthood. Adult patients with CF who have an FEV_1 between 30 and 60% of predicted are eligible. The study can be completed in one visit (about 1.5 hours), and can be done after a routine clinic visit while you are doing well.

A Double Blind, Cross-Over Study Comparing Aerosolized Lucinactant and Vehicle on Mucus Clearance for CF Lung Disease (GCRC 2710)
Lucinactant (surfactant) is a compound that has detergent properties that reduce mucus stickiness in the lungs. The compound has been used in other forms to treat respiratory distress in infants, and this study will test its effectiveness as a CF therapy in aerosolized form. CF patients who are at least 14 years old and have an FEV1 of at least 40% are eligible. The study involves 8 visits to UNC.

Scott Donaldson, MD is the primary investigator for these studies. If you are interested in participating, contact Nadia Bendahmane at ***nadiab@med.unc.edu*** *or* ***919-966-9198****. Compensation for study visits/travel is provided.*

This information has been approved by the UNC IRB.