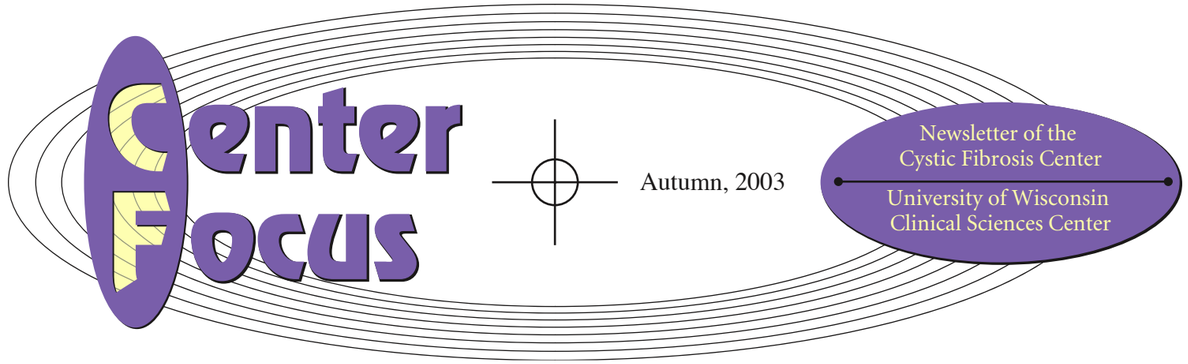

Great News!! The Pediatric CF Team has expanded its outpatient clinic visit opportunities.

We have 3 nurse practitioners that are specialized in the care of children with cystic fibrosis. They work collaboratively with our team of CF specialists including physicians, nutritionists, respiratory therapists, social workers and pharmacists. Our nurse practitioners are seeing patients and can examine and prescribe medication to manage CF care. You can schedule an appointment with any one of our nurse practitioners at anytime. (For patients in the newborn screening study on Mondays, we request that you continue to be seen in that clinic.)

What is a nurse practitioner?

A nurse practitioner is a registered nurse with a master's degree in nursing. Nurse practitioners are licensed by the state to function independently or collaboratively with other health care professionals. You may have already spoken with one of our nurse practitioners over the phone, as they have been managing CF care over the phone for years.

Times to schedule an appointment with a nurse practitioner:

- If you have special questions or concerns that you would like addressed outside of a regular clinic visit.
- If you would like extra teaching or education on any CF related care.
- If the schedule of a pulmonologist is not convenient or timely.

You may want to alternate every other visit with a nurse practitioner and physician, or you may want to see a nurse practitioner in addition to your regular appointments.

Who are the nurse practitioners?

Darci Pfeil, NP. Darci has over 14 years of experience in caring for children with CF. She joined our CF center in 1993. Darci specializes in CF care, is a national speaker at the annual CF meetings, and creates many of the new programs for the CF community.

Karen Allaire, NP. Karen joined the CF center in 1992. She has a special interest in integrative health care: combining traditional cystic fibrosis care with complementary therapies such as herbs, supplements, Healing Touch and Reiki. She also has an interest in smoking cessation.

Janis King, NP. Janis joined the CF center in 2001. She has worked for many years on pediatric and obstetric inpatient units and has many years of experience as a public health nurse. She is very interested in CF education with patients and families, breastfeeding issues and assisting families in sharing information about CF with school personnel.

To schedule an appointment with a nurse practitioner, call the Pediatric Specialty Clinic appointment desk at (608)-263-6420.

LaCrosse CF Clinic

Dr. Todd Mahr, Pediatric Allergist at the Gunderson Lutheran Clinic in LaCrosse, is beginning a CF clinic. He has assembled a multi-disciplinary team to deliver care to CF patients. The clinic will occur on the second Wednesday of each month. Patients that live in southwestern Wisconsin can receive their care in LaCrosse. To make an appointment, call (800) 362-9567 ext. 5-5848 (For patients in the Newborn Screening Study who are seen on Mondays, we request that two of the four visits still occur in Madison. Those visits would be the birthday visit and the six-month visit. The three and nine-month visits could be in LaCrosse.) In a future issue of the CF Center Newsletter, we will focus on the LaCrosse CF Clinic team members.

BIIL-284 Study

We are beginning participation in a multi-center study of a drug called BIIL-284. This is an oral medication that can decrease inflammation in the lungs by antagonizing a receptor for a chemical called leukotriene B4. This is a double-blind placebo-controlled study in which some of the patients receive the active drug and others receive placebo. Neither the medical caretakers nor the patient knows who is receiving active drug versus placebo. Criteria for being in the study is having an FEV_1 of 25-85% of predicted and to not be receiving chronic oral steroids or high dose ibuprofen. If you are approached about this study, we hope that you will participate in it.

Patient Education

Because of concerns about infection control (as noted on page 2), new ways to educate patients are necessary. A web cast occurred on Thursday, August 14, entitled "The Patient as a Partner in Research." The presentations offered the basics about what makes clinical trials succeed, as well as the latest in CF clinical research. This presentation is archived on the internet at the following website:

http://www.mediaprosf.com/clients/tvn/playback.htm

What's New at Our CF Center?
Our Transition Program!

What is a transition program? It is a program that both the pediatric and adult CF care teams developed to ease the move from pediatric to adult health care. We are committed to caring for people with CF across their lifespan. Helping young adults and their families' move to the adult care team is a big part of our commitment. Just as graduation from high school is a right of passage or a milestone that you and your family will celebrate, we believe transition to adult care is also a milestone for you to experience.

When does someone transition to the adult care team? Both the pediatric and adult CF care teams expect the majority of patients will transition between 18 and 19 years of age. You will have several opportunities to meet members of the adult care team during this time. We want you to know that you are not alone and we will be here to help you with this change.

When does someone start using the program? At the age of eight, the CF team will introduce you and your family to the transition program. Over the next several years, we will work on learning more about CF at each visit and gradually teach you how to care for yourself. As you learn more about CF, you can take charge of your life and your cares. This will help to prepare you for transition to the adult care team.

How will I know when I am ready to transition? We have created a booklet to help you learn more about CF and how to take charge of your cares. As you move through the stages of transition, you will learn new things and take on more responsibilities for your care. At the different developmental stages from eight to 18 years of age, there is a list of things you can be working on to help yourself to become an independent young adult. As you grow up and follow your heart's desire, whether that is to go off to college or begin working, you will feel prepared to take care of yourself as an independent young adult. We hope this program will build your confidence and assure your parents that you are learning to take care of yourself. We'll help you throughout your journey and promise to take good care of you along the way!

Influenza Vaccine

The fall and winter are approaching, and that means that influenza season is approaching. We are once again making our annual recommendation that all CF patients six months of age and older should receive the influenza vaccine. If you are nine years of age and younger and have never received the influenza vaccine, then two doses are given separated by a month. For everyone else, one dose is given. Influenza vaccine is usually available in mid to late October. In addition to the cystic fibrosis patient, all other household members should receive the influenza vaccine.

You may have heard that the FDA has approved a new nasal flu vaccine called Flumist. Unfortunately, the nasal flu vaccine has a number of drawbacks. It is not approved for children under age of five years and is not approved for people with underlying asthma or chronic underlying medical conditions. Additionally, it is quite expensive and many if not most insurance companies will not cover this, unlike the conventional influenza vaccine injection. For all of these reasons, we will not be administering Flumist at the University of Wisconsin Hospital and will continue with the traditional injected influenza vaccine.

Infection Control

In May 2001, the Cystic Fibrosis Foundation sponsored a consensus conference on Infection Control. This conference utilized the expertise of experts in the fields of Pediatric Pulmonology, Adult Pulmonology, Infectious Disease, Nursing, Respiratory Therapy, Lawyers, Ethicists, and included the input of cystic fibrosis patients. The final documents were published in the May 2003 issues of Infection Control and Hospital Epidemiology and The American Journal of Infection Control. This is a very far reaching document and has major implications for cystic fibrosis patients. At the University of Wisconsin Cystic Fibrosis Center, we are still in the process of adapting these recommendations to both our clinic settings and inpatient settings. This article will be the first in a series of articles discussing the implications of the infection control recommendations.

It is becoming increasingly clear from scientific studies that bacteria from cystic fibrosis patients can be transmitted between patients. We do not want to become germ-o-phobic, however we must use the available scientific evidence to minimize the chance that a patient can acquire a serious bacterial infection from another patient. Keeping these principles in mind, here are several changes that are being made at the University of Wisconsin CF Center.

1. CF Family Education Day. Many of you may be familiar with the CF Family Education Days that have been held in the past. This was an activity that alternated between the CF Center in Madison and the CF Center and the CF Center in Milwaukee. There appears to be mounting evidence that an infection control risk is present when a number of CF patients are together in a confined space inside a building. Our CF Center staff has had serious discussions regarding whether we should continue CF Education Day with CF patients and parents present, or have a CF Education Day for parents and significant others only, or discontinue CF Education Day. Although the second option appeared attractive, we are cognizant of the fact that CF affects both children and adults. Our adult CF patients may be living independent of their parents and not have a friend or family member to attend Education Day on their behalf. A CF Education Day aimed only at parents gives the appearance that this is only a pediatric CF Center, whereas we are a comprehensive CF Center that offers state of the art care to both pediatric and adult patients with CF. There are alternative measures to educate families including this newsletter and Virtual Patient Education Days over the internet. Another option is making videotaped presentations available to be checked out. Keeping all of this in mind, we have decided to discontinue CF Family Education Day for the sake of infection control. (The official stance of the Cystic Fibrosis Foundation is that there are no reports of cross infection occurring at CF Education Days. However, we are taking the conservative road for the health and protection of our patients.) Elsewhere in this newsletter, you will find an article about accessing the Virtual CF Patient Education Day that was held in mid-August.

2. Waiting room infection control guidelines. It is clear that direct contact between CF patients can be a major factor in transmitting bacteria between patients. Keeping this in mind, we discourage handshakes and any physical contact between CF patients in the waiting room. Ideally, a CF patient should remain a minimal distance of three feet from other patients in the waiting area. For children and toddlers, they should bring their own toys and not play with the available toys in the waiting room. In the future, we hope to make available a waterless antiseptic alcohol-based hand rub available in a dispenser in the waiting room. Patients and family members should use this to sterilize their hands upon arrival to the waiting room. If a CF patient needs to cough, then they should cough into a tissue, immediately discard the tissue into a trash can, and repeat hand sterilization with the alcohol-based hand rub.

   Enclosed with this newsletter is a pamphlet "Stopping the Spread of Germs" produced by the Cystic Fibrosis Foundation Education Committee.

Pulmonary Function Testing for Infants

It is routine in Cystic Fibrosis Centers that patients over age 4-5 years carry out pulmonary function tests. The most common test performed by patients is called spirometry. In this test, the patient takes as deep a breath as possible and blows out all his/her air as fast and completely as possible. The second most common test is called lung volumes. In this test, the patient sits in a sealed booth (body plethysmograph) and performs panting maneuvers. This test measures the amount of air in a patient's lungs. This allows measurement of the air in the lungs that remains after normal exhalation. By adding this to the information from spirometry, we can also calculate the amount of air in the lungs when the lungs are completely full of air.

There are many other pulmonary function tests, but spirometry and lung volumes are the most common tests used in the evaluation of patients with CF. These tests are valuable because they are currently our only quantitative measure of lung function. Also, pulmonary function tests reveal mild abnormalities that cannot be picked up by history or physical examination. Both of these factors combine to make pulmonary function testing (PFT) a very important tool in the evaluation of CF lung disease over time.

Historically, physicians have not had methods which allowed the evaluation of pulmonary function in infants and preschool aged children. In the late 1970's and early 1980's, the first infant pulmonary function systems were developed in the United States, Great Britain, and Israel. At the University of Wisconsin Children's Hospital, we have performed infant pulmonary testing for about ten years. Some children with cystic fibrosis had infant pulmonary functioning testing carried out on an annual basis as part of research projects at our center. This testing involved first generation infant pulmonary function testing equipment. This equipment provided some valuable information but the information was more limited than the information collected when a patient performs spirometry and lung volume measurements.

In May of 2002, the FDA approved the first machine that could be called the second generation infant pulmonary function testing equipment. The advent of this second generation of equipment is very exciting to physicians caring for patients with cystic fibrosis. This new equipment
allows us to make measurements in infants and toddlers that are very similar to the measurements of spirometry and lung volumes in older children and adults. Therefore, we can collect much more complete information about lung function in infants and toddlers than we could with the previous generation of equipment.

We will be testing many children with cystic fibrosis about every six months on an ongoing basis to assess their lung function. The infant pulmonary function testing is more time consuming than the testing in older children and adults because it requires sedation and careful adjustment of the equipment during the maneuvers in order to collect optimal quality data.

If your child is a candidate for infant pulmonary function testing, you will be given specific instructions about the preparation for sedation including bringing your child in with an empty stomach. Also, bring your child in when he/she has not napped because the sedation we use works much better if your child is somewhat tired. The main sedative agent is chloral hydrate which is given orally. Occasionally, other agents may be used, but chloral hydrate is the standard medication used around the country. The testing itself will take 1-1 1/2 hours. However, the whole process will take about four hours given the need for your child to fall asleep after the sedative is given and the monitoring and recovery period after the testing is completed.

We are proud to be one of the first centers in the country to offer this second generation of infant pulmonary function testing. If you have questions about this testing please ask your doctor at the CF Center.

Cystic Fibrosis Foundation Patient Registry and ESCF Studies

There are two patient registries which track health information from cystic fibrosis patients. One of these is maintained by the Cystic Fibrosis Foundation and the second is sponsored by Genentech (Epidemiologic Study of Cystic Fibrosis, ESCF). Patients at the University of Wisconsin CF Center have previously been in both of these data registries. Because of new regulations that protect the private health information of individuals, we now require a signed informed consent for these studies. You may have received consent forms about these studies in the mail or were approached about these studies during a clinic visit. We hope that by the end of 2003, that all CF patients will agree to be in both of these studies. These studies involve obtaining information from your routine clinic visits and sending them to the respective patient registries. This information is strictly confidential and protected.