New Providers in CF Clinic…

Hello, my name is Darci Pfeil. I am a Pediatric Nurse Practitioner with the UW Cystic Fibrosis Center. I have been caring for patients and families with CF for over 16 years. In 1989, I began working as a staff nurse on the inpatient CF unit here at UW Children's Hospital. In 1993, I became a Pediatric Nurse Practitioner and joined the Pediatric Pulmonary Team and the UW CF Center. My professional career focuses solely on children with cystic fibrosis. My clinic is on Wednesday mornings. I can be reached through the pulmonary office at 608-263-8555. Appointments can be made by calling 608-263-6420. It is my pleasure to serve you.

Hi my name is Kate Swenson. I am a Pediatric Nurse Practitioner for the Pediatric Pulmonary Team in the Pediatric Specialties Clinic at the UW. I started with the team at the end of April 2005. I am beginning my own clinics on Fridays. I have one hour appointments beginning at 8:15 a.m. and the last appointment at 4 p.m. I frequently see patients with cystic fibrosis, bronchopulmonary dysplasia and asthma. To schedule an appointment please call 608-263-8555.

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University of Wisconsin
Clinical Sciences Center

Center Focus

Winter, 2006

Newsletter of the
Cystic Fibrosis Center

The Importance of Good Nutrition

Over the past six months, you may have noticed changes occurring in your Cystic Fibrosis Center. Perhaps you've been coming to clinic more often, spending more time with the nutritionist, and receiving Nutrition Action Plans summarizing your nutrition status and goals at each visit. The reasons for these changes are many.

Every time you come to a clinic visit, you are weighed, measured, and examined. All of this information collected during your visit is entered into a database maintained by the Cystic Fibrosis Foundation (CFF). Each year, the CFF summarizes the data and gives a report to each CF Center. The report informs the Center of how well it performs compared to other CF Centers in areas such as lung function and nutrition outcomes.

One thing we discovered after receiving our report last year is that we are at the national average for nutrition outcomes. Being average is not good enough for us. We want to be among the best because that means you, as patients and families, are receiving excellent care.

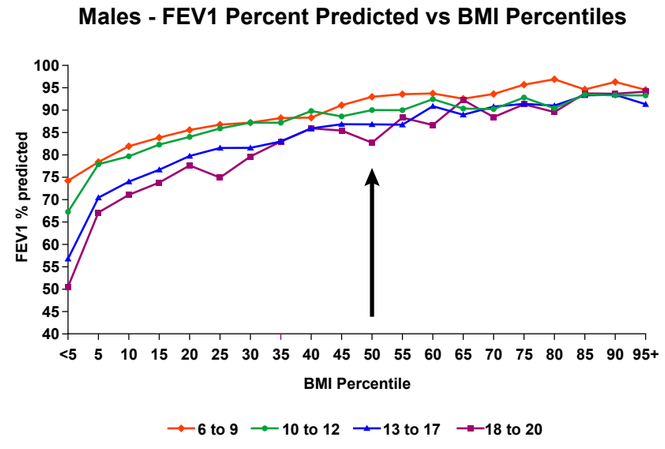
As one of the first steps to improve care, several members of our CF Center Team visited one of the Centers with the best nutrition outcomes. We learned that the patients there come to clinic more often when they are not gaining weight or losing weight. We also learned that the patients receive more supplemental feedings and tube feedings and spread enzymes out over meals instead of taking them all at the beginning of the meal. Finally, the most important thing we learned is that everyone on the team has a strong desire to improve nutrition because it improves lung function.

Recently, the CFF stated this as well. The CFF established a team of leaders in the CF Community and asked them to study all of the articles they could find on CF and nutrition. After studying those articles, this team discovered that when people with CF are at a good weight for height, they have better lung function and are healthier. To compare weight to height, a tool called the Body Mass Index (BMI) is used. In people age 2 to 20 years, the goal is to have the BMI at the 50th percentile or above when plotted on a graph. For adults (age 20 years or older), this equals a BMI of 22 kg/M2 for females and a BMI of 23 kg/M2 for males. Information on the calculation and graphs is at the end of the article. The graph below shows that the FEV1, a measure of lung function, is at its best at and above a BMI of the 50th percentile.

We hope this explains why you are seeing a greater emphasis on nutrition status when you come to clinic. We also hope you understand that we want to work with you to develop a care plan that fits your needs, lifestyle, and condition. Remember, you and your family are a very important part of the team. If we all work together, we will reach our goals and maximize care.

BMI = weight in pounds/height in inches squared x 703

For children age 2 to 20 years, the graphs to plot BMI are located at: *http://www.cdc.gov/nchs/about/major/nhanes/growthcharts/clinical_charts.htm*



Influenza Vaccine

As winter approaches, we will be entering another influenza season. There are a large variety of influenza viruses. The popular press has carried many stories recently about the Avian or Bird Flu. We do not know if this will be a problem in this country and currently, there is no vaccine for the Bird Flu. However, the traditional influenza vaccine is available and this vaccine is recommended for all CF patients and household members. One dose is required for patients over nine years of age or those who received flu vaccine in previous years. If a child is less than nine years of age and has never received the influenza vaccine previously, then two doses are required separated by one month. We are currently administering the influenza vaccine in our clinics. We encourage you to obtain influenza vaccine in CF clinic or from your primary care provider. As stated above, all other household members should also receive the influenza vaccine.

Aggressive Care Makes a Difference

Elsewhere in this newsletter, you will read about the Cystic Fibrosis Foundation Therapeutics Development pipeline and new drugs that are being developed. It is hoped that these new drugs will slow the progression of lung disease in CF patients and we are very hopeful of a cure for the lung disease in the future. In the meantime, is there anything that we can do with current therapies to slow the progression of lung disease? This has previously been studied in CF patients using the Epidemiologic Study of Cystic Fibrosis (ESCF) database. A study was published in 2003 comparing patients at CF centers who had better pulmonary functions compared to those who had worse pulmonary functions. Many factors were analyzed. Two of interest were the average annual clinic visits and cultures between those sites with better pulmonary functions compared to those lower pulmonary functions. The CF Centers that had better pulmonary functions have patients come for more frequent clinic visits and respiratory cultures were also obtained more frequently.

We strongly believe that we can utilize existing therapies to significantly improve the health of people with CF. Quarterly clinic visits may be appropriate for people with CF who have excellent pulmonary function, control of cough, and growth. However, returning in three months to clinic may not be soon enough for patients who have nutritional or pulmonary issues. We have recently begun seeing patients in one month when there are pulmonary and nutritional issues in which we have made an intervention. This one month follow-up is important to see if the intervention has helped and to make sure that there is improvement. Together, we can work jointly to significantly improve the health of people with CF.

CFF Therapeutics Pipeline

The color insert in this edition of Center Focus shows the Cystic Fibrosis Foundation's therapeutics pipeline. As you can see, there are multiple steps that are required for new drugs to be approved. The first step is research, that is, finding potential drugs. The second step are preclinical studies performed in the laboratory. If medications successfully complete the preclinical phase, then the Food and Drug Administration requires three phases of human trials. Phase One is a human safety trial. Phase Two are further studies to assess human safety and if the drug is effective. The last stage that is required prior to FDA approval is Phase Three. These tend to be larger studies and are considered the definitive trial. Phase Two and Three trials are usually performed as double-blind randomized placebo-controlled trials. This means that some of the patients receive active drug and the other patients receive inactive drug. Neither the patient nor the investigators are aware of who is receiving the active drug. This study design is required for drugs to be thoroughly investigated.

Note that there are a number of different therapies that are being investigated. These therapies range from correcting the basic defect with gene therapy to investigating new antibiotics. There are two new antibiotics that are currently in Phase Three trials. Aztreonam is an antibiotic that previously was given in an intravenous form. There is a Phase Three study of an aerosolized form of this antibiotic currently under way. Secondly, notice that there is a dry powder inhaled formulation of Tobramycin being studied. There are already asthma drugs given by a dry powder inhaler route. Hopefully, this dry powder inhaled form of Tobramycin will be as effective as the nebulized form. This will enable a much faster and convenient way to deliver this medication. There are other medications that will soon be entering Phase Three trials. We hope to participate in studies here and look forward to your participation in these studies.

CF Scholarships

There are two programs that provide college scholarships to patients with CF. One of these is sponsored by Solvay Pharmaceuticals. We do not yet have the application form for 2006. Please call our office at 608-263-8555 if you are interested in this scholarship.

The other scholarship is sponsored by the Cystic Fibrosis Scholarship Foundation. Here is information about their program: The Cystic Fibrosis Scholarship Foundation ("CFSF"), founded by a parent of a young adult with CF, is pleased to announce a scholarship program for students with cystic fibrosis. CFSF is not part of nor is it funded by the Cystic Fibrosis Foundation. The program is available to those who will be enrolled in an undergraduate college program or a vocational school in the fall of 2006.

Scholarships will be awarded based on a combination of financial need, academic achievement, and leadership. During the six years of the Foundation's existence, approximately 25% of the students who have applied have been awarded scholarships. Awards may be used for tuition, books and room and board. Awards will be sent directly to the institution that the student is attending. Both multi-year awards and single year scholarships are awarded. The multi-year awards are $1,000 a year for a maximum of four years and the single year awards are for $1,000. Students granted a single year award may apply in subsequent years for further awards although there is no guarantee of future awards. Multi-year award recipients must maintain a 2.0 grade point average or above to maintain their scholarship. All students will be considered for both the multi-year and single year awards.

Recipients of awards will be notified by April 19, 2006. Scholarship application forms will be accepted after January 15, 2006 and are due by March 17, 2006. Application forms are available via e-mail to *MKBCFSF@aol.com*, by phone at 847-328-0127 or in writing. The application forms are also available at the website: *www.cfscholarship.org*. When requesting an application, please indicate your current status in school (i.e. high school senior, freshman in college, etc.). This is important in order to be sent the appropriate application form.

2814 Grant Street, Evanston, IL 60201
Phone: 847-328-0127 FAX: 847-328-0127

Changes In Medicare

You may have heard that Medicare will soon be providing prescription drug coverage to those who only have Medicare benefits and also to those who have BOTH Medicare and Medicaid benefits. Prescription drug coverage will be available through Medicare under a new section called Medicare Part D, starting on January 1, 2006. It is important to understand how the new drug coverage will work and how these changes may affect you and your family.

Medicare drug coverage will be offered through groups called Prescription Drug Plans (PDP). An eligible Medicare-only recipient must enroll in a new prescription drug plan between November 15, 2005 and May 15, 2006. Failure to enroll in a plan during the initial enrollment period will result in having to pay higher premiums if you enroll later. However, you may decline Medicare coverage because you have other drug coverage, such as through a spouse's employer. If that coverage is comparable to PDP coverage, you will not have to pay a penalty if you later enroll in Medicare Part D.

If you have BOTH Medicare and Medicaid, you MUST enroll in a PDP between November 15, 2005 and December 31, 2005. If you do not enroll in a plan by December 31, 2005, you will automatically be enrolled in a low income PDP. This type of PDP is available to Medicare/Medicaid recipients who are low income. While premium payments will be waived for those individuals in a low income PDP, a low income PDP may not provide coverage for all of the medication needs of some people with CF. There will be low-income assistance, called subsidies, for these individuals to help with drug costs under the PDP cost sharing plan. Individuals who have both Medicare and Medicaid will LOSE current drug coverage through Medicaid on January 1, 2006.

A variety of Prescription Drug Plans will be offered under Medicare Part D. Different areas of the country will have different PDPs available. Premiums will range from about $20 a month to $35 a month. Information about what drugs the plans will cover should be available on or about October 13, 2005. A person will be able to call a toll free number at Medicare or go to the Medicare website at *www.Medicare.gov*, to inquire about the plan that might be best for them.

It is important for people with CF to know that Medicare Part B will continue to provide coverage for prescription drugs that are administered by durable medical equipment, such as Pulmozyme or TOBI. Make sure that you are enrolled in Part B so that you have coverage for those drugs. Part D Prescription Drug Plans will not cover drugs that are currently covered by Part B. When looking for a PDP Plan that will cover the medications you use, you will not find a plan that covers those drugs already covered by Part B.

Our CF care center will be receiving information from the CF Services Pharmacy about how to choose a plan that covers your CF medication needs. We will share this information with you when it becomes available. Most of these PDPs will require patients to meet an annual deductible and to pay co-payments for drugs. Each enrollee will pay an annual deductible of $250. For drug expenses after the first $250 and up to $2,250, the enrollee pays 25 percent of the drug costs and the PDP covers the remaining 75 percent. The enrollee pays 100 percent of the drugs expenses from $2,250 to $3,600, which is a gap in Medicare coverage. After $3,600 in drug expenses, coverage resumes and the enrollee pays only 5 percent of the drug costs or a small co-payment until the end of the calendar year.

If your resources are less than $11,500 (single) or $23,000 (married), you may qualify for extra help paying for Medicare Prescription Drug Coverage. You can find out more about this assistance at *www.medicare.gov*.

For Wisconsin specific help, you can receive assistance from the Wisconsin Coalition for Advocacy Medicare Part D Disability Drug Benefit helpline at 1-800-926-4862. Please let us know if you have any questions; patients seen in the adult CF Center may call Damien Wilson at (608) 263-8474 and patients seen in the pediatric center may call Craig Becker at (608) 263-8572. You may also contact the CF Legal Information Hotline at 1-800-622-0385.

Sincerely,

CF Care Center Team

Opportunities for Patients and Families as Leaders at the UW Pediatric CF Center …

At the recent North American Cystic Fibrosis Conference held this past October in Baltimore, MD, a symposium entitled Helping Patients and Families to Become Effective Team Members occurred. The symposium focused on ways to incorporate patients and families as active members into the Cystic Fibrosis Care Team. Here at UW we are excited for the opportunity to create a variety of new ways to involve patients and families as partners with staff in guiding our work throughout the CF center. Stay tuned for more information about opportunities to build a partnership with the CF team.

CF Education Day is making a return…

Due to popular demand, we are planning for the return of CF Education Day. In order to comply with the National Infection Control Guidelines for CF, our education day will be for parents and family members only. We are planning a half-day educational conference on the topics you wish to hear about. Please contact our CF center at 608-263-8555 or email *camoodie@wisc.edu* with any topic suggestions you may have for the education day. Your input is invaluable and we look forward to your suggestions. Stay tuned for further details.