Adult Cystic Fibrosis Advisory Council (AAC)

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UW Health Adult Cystic Fibrosis Care Center

In September 2012, the University of Wisconsin Adult Cystic Fibrosis Care Center will be initiating the Adult Cystic Fibrosis Advisory Council (AAC). The Organizational Meeting will take place on September 24th, attended by patients, family members, and Care Center staff. At that meeting, prudent infectious disease controls will be in place; patients with particular pathogens that may be a risk for cross-contamination, will have the ability to attend the Council via Web Conference from their homes.

The general goal of the Adult Advisory Council will be to:

1. Serve as advisory resource to administration and staff of the CF Center and its programs.
2. Promote improved relationships between patients, families and staff.
3. Provide a vehicle for constructive communication between patients, families and staff.
4. Provide an avenue for patients and families to offer input into policy issues and the development of new programs and facilities.

The Adult Advisory Council has already fielded patient and family member applications and selected officers for Chairperson, Vice Chair, and Secretary. These officers have formed a steering committee to shape the by-laws of the Council, and to organize the initial meeting in later September.

We welcome this partnership of patients and family members in providing support and feedback to improve our capacity to service our patient population.

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University of Wisconsin
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Center Focus

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Newsletter of the
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Back to School: Power Packed Meals and Snacks

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As many of you head back to school, you need plenty of fuel from meals and snacks to help you learn and grow. Sometimes it gets hard trying to think of new things to eat for breakfast or to pack in your lunch box. Here are some ideas to get you off to a good start.

Breakfast:

Make a Super Smoothie:

- Start with ½ cup whole milk yogurt (ex: Stonyfield Farms®) and ½ cup whole milk, Pediasure®, Ensure®, or cream.
- Add your favorite fruit (bananas, berries, peaches, mango, coconut).
- Choose a high calorie booster like nut butter, ground flaxseed, wheat germ, or protein powder.
- Throw in a little ice if you wish and blend until smooth in a blender.

Enjoy nut butter or nutella on a big bagel (a bagel will give you more calories than bread or an English muffin).

Add ground flaxseed powder, wheat germ, or protein powder to oatmeal, pancake mix, or muffin batter.

Choose high calorie cereals like frosted shredded wheat, granola, or raisin bran; serve with a mixture of whole milk and cream.

Add shredded cheese, crumbled bacon, and melted butter to your scrambled eggs.

Make hot cereals with cream or a liquid nutritional supplement like Pediasure® or Ensure® instead of water.

Lunch:

Make your sandwiches with bagels or sub rolls. Add butter, mayo, extra cheese, avocado, or nut butters.

Try dried fruit (apricots, raisins, apples, pineapple, mango) which is usually higher in calories.

Choose whole milk yogurt or 4% milk fat cottage cheese.

Bring peanut butter or cheese filled pretzels or crackers instead of plain.

Pack some high calorie dip for fruits and veggies.

If you eat in the cafeteria, consider talking to the cafeteria staff or principal. Many families have been able to get the school to provide extra condiments (butter, sour cream, mayo, cheese, salad dressing) for their children with CF. Other families get their own supply of whole milk or bring cases of nutritional supplements like Pediasure® or Ensure®, to have during lunch time.

Snacks:

Try the little packets of Justin's® or Sunland® nut butters (honey almond, chocolate peanut, banana peanut, chocolate hazelnut – *YUM!*). They come in little squeeze packs that you can eat in a snap. They're good plain or put on top of crackers or fruit.

Have a handful of trail mix made with nuts, seeds, dried fruit, chocolate, cereal, and pretzels.

Instead of a granola bar, have a high calorie sports bar (Myoplex®, Power Bar®, Clif®, Zone®, or Balance®). They have at least 100 more calories than a granola bar!!

Warm up some leftovers or microwave a frozen meal.

Make a milkshake with whole milk, ice cream, and crushed cookies or fruit.

Finally, if you need us to talk to someone at your school or write a letter explaining why you need high calorie meals and/or extra snacks or bathroom breaks, please let us know. Many schools are working harder than ever to promote low fat, low sodium, and low calorie meals. Most people with CF need just the opposite. We'd be happy to help educate your teachers and friends to help you have a successful school year!

The CFTR2 Project

CFTR stands for the Cystic Fibrosis Transmembrane Regulator protein. This is the product of the CF gene. CFTR2 is the Clinical and Functional Translation of the CFTR protein. If you are confused, bear with us. Here is the explanation.

Everyone has a CF gene on chromosome number 7. This gene was discovered in 1989. A change in the DNA of the gene is called a mutation. The most common CF mutation is the F508del (also called the delta F508 mutation). There have been approximately 1,900 different changes in the DNA that have been reported to a CF mutation database that is maintained in Toronto, Canada. However, most changes in the DNA are known as benign polymorphisms. These are changes in the DNA compared to the normal DNA sequence, but the change (the polymorphism) does not cause disease. Remember that in order to have CF disease, one must have two disease-causing mutations. (The two mutations come from each parent of a person affected by CF. The parents are known as carriers of a CF mutation.)

The goal of the CFTR2 project is to characterize all changes in the DNA sequence of the CF gene as either disease causing, non-disease causing, or a mutation that could cause CF-related complications (recurrent sinusitis or pancreatitis). Of the 1,900 changes in the DNA of the CF gene, it is estimated that 160 to 190 of these are disease causing mutations. At the time that this article is written, a determination has been made of 130 mutations in the CF gene.

The CFTR2 website is available for anyone to view. There are details on various aspects of the mutations such as average sweat chloride values, average FEV_1 values, and pancreatic sufficiency versus insufficiency. The website address is *www.cftr2.org*.

In the winter issue of the Center Focus, we discussed Kalydeco (ivacaftor). This is a chloride channel potentiator which improves the functioning of the abnormal chloride channels in patients with CF who have the G551D mutation. This medication was approved by the Food and Drug Administration earlier this year. Now that we are entering an era of mutation specific therapy, it is important to know of each CF patient's mutation. If you are not aware of your mutation, please ask your provider at your next clinic visit.

For some patients, we do not have test results for one or both mutations. If that is the case, there is a free program in place in which we can send blood to the DNA diagnostic laboratory at Johns Hopkins University in Baltimore, and they will perform comprehensive testing on the specimen.

Influenza Vaccine Time

It is that time of the year again in which all patients with cystic fibrosis and all family household members who live with the CF patient should receive the flu vaccine. We can administer the flu vaccine to CF patients in our clinics. Family members may receive flu vaccine from the primary care provider. Often, one can also receive the flu vaccine at a pharmacy or health department.

The Importance of Masks and Infection Control

In the 1980's and 1990's, unrelated patients with cystic fibrosis could be together in the children's hospital school or visit each other in their hospital rooms. There was also a CF summer camp that occurred annually until 1994. However, we now have the medical knowledge that bacteria can be transmitted between patients. Although we are particularly concerned about the *Burkholderia cepacia* complex bacteria being transmittable between patients, there is also evidence that other bacteria such as *Pseudomonas aeruginosa* can be transmitted between patients. Thus, we no longer sponsor CF summer camp and there are infection control guidelines for patients in the hospital.

In addition to infection control guidelines for hospitalized patients, there are also infection control guidelines for patients attending clinic. We respectfully ask that all patients with cystic fibrosis wear a face mask when they check-in at the registration desk. Also, please use the alcohol based hand gel in the waiting room. The face mask should remain on until the patient with CF is in the clinic exam room. The mask may be removed in the clinic exam room, but please wear the mask again upon departing the exam room and until the patient with CF has departed the building.

Cystic Fibrosis Family Education Day November 10, 2012

Please join us on November 10, 2012 for Cystic Fibrosis Family Education Day. The event will be held at the Health Sciences Learning Center, 750 Highland Avenue, Madison, WI 53705. This is the medical school building that is adjacent to the University of Wisconsin Hospital. Parking is available in the University of Wisconsin Hospital parking garage.

(See agenda on following page.)

This free event includes parking, breakfast, lunch and break snacks.

Due to infection control concerns mandated by the Cystic Fibrosis Foundation, we respectfully request that people with CF not attend the family education day. We will live stream all of part of the presentations at *live.videos.med.wisc.edu*

People who attend the CF Family Education Day can enter a drawing for a free one night stay at The Geneva Inn.

A registration form can be found at:

https://uwmadison.qualtrics.com/SE/?SID=SV_5nxH1Lauv9IJwmV

For questions, please email Deb Silvis, Outreach Specialist and Family Involvement Faculty at *dsilvis@pediatrics.wisc.edu*

The University of Wisconsin Cystic Fibrosis Center and Pediatric Pulmonary Center present:

Family Education Day for Cystic Fibrosis

November 10th, 2012
Room 1325, Health Sciences Learning Center (adjacent to the University of Wisconsin Hospital)

a.m.

8:00-9:00 **Registration with continental breakfast and vendor displays**

9:00-9:15 **Welcome**
Michael J. Rock, M.D., Director, University of Wisconsin Cystic Fibrosis Center
Mary Marcus, Co-Director, University of Wisconsin Pediatric Pulmonary Center

9:15-10:00 **"What Every Person with CF needs to know about Social Security benefits, Medicaid, Medicare and Health Insurance"**
Beth Sufian, JD
Director of the CF Legal Information Hotline, Director of the CF Social Security Project

10:00-10:15 **Break and vendor displays**

10:15-11:00 **"Update on Research and Drug Development for Cystic Fibrosis"**
Michael J. Rock, M.D., Director, University of Wisconsin Cystic Fibrosis Center

11:00-12:00 **"Perspectives on the Past, Present and Future; One Person's Reflections on Life and CF"**
Lindsay Shipp, Adult with CF, VX770 drug trial participant, and entertainer

p.m.

12:00-1:00 **Lunch and vendor displays**

1:00-2:00 **"Finding the Sweet Spot: Balancing CF Therapies with Living Life"**
Susanna A. McColley, M.D., Co-Director, Cystic Fibrosis Center
Ann & Robert H. Lurie Children's Hospital of Chicago

2:00-2:15 **Break and vendor displays**

2:15-3:00 **"Living with Cystic Fibrosis: Parent and Consumer Panel"**
Moderated by
Deb Silvis, Outreach Specialist,
Family Involvement Faculty

3:00-3:15 **Wrap-up and Evaluations**
(Drawing for stay at Geneva Inn)

See registration information on previous page