University of Wisconsin
Clinical Sciences Center

Center Focus

Newsletter of the
Cystic Fibrosis Center

Summer, 2009

Vaccine Information

There has certainly been much publicity about the H1N1 virus (sometimes referred to as the swine flu virus). Here is what we know as of the time of publication of this newsletter. Vaccine manufacturers are working on the production of a specific vaccine for H1N1. This will likely be a separate vaccine given in addition to the conventional influenza vaccine. You can keep track of this situation by following news on the Centers for Disease Control website: *www.cdc.gov* or by calling 1-800-CDC-INFO for more information. In the meantime, these are steps to stay healthy:

- Influenza spreads mainly person-to-person through coughing or sneezing from infected individuals
- Cover your mouth or nose with a tissue when you sneeze and throw the tissue in the trash after you use it
- After you sneeze, wash your hands with soap and water or use alcohol-based hand sanitizer
- Avoid touching your eyes, nose and mouth, as this is how viruses can be spread
- If you are a person with CF and have had contact with a person with H1N1 virus, please call your care provider

In the wake of the H1N1 virus pandemic, the CDC produced new interim guidance for use of 23-valent pneumococcal polysaccharide vaccine. Pneumococcal disease is caused by *Streptococcus pneumoniae* bacteria. During the 20th century influenza pandemics, secondary bacterial pneumonia was an important cause of illness and and *Streptococcus pneumoniae* (pneumococcus) was reported as the most common cause. Pneumococcal polysaccharide vaccine (PPSV) protects against 23 types of pneumococcal bacteria, including those most likely to cause serious disease. On June 9, 2009, the CDC produced new guidance on the use of this vaccine. The PPSV is recommended for all persons above 65 years of age and for people 2 years to 64 years of age who have long-term health problems including chronic pulmonary problems. This would include persons with CF. Therefore, if you have not yet received the PPSV, we recommend that you see your primary care provider for this vaccine.

Parent Education Programs Sponsored by the UW Pediatric CF Center

We are delighted to announce a new format to our Parent Education programs. Beginning with a kick-off event on September 30, 2009, the UW CF Center will hold quarterly evening education programs for parents. Each program will offer a presentation on a specific topic that has been suggested by parents for parents. The September 30th program, titled, Back to School: What Your Child's School Needs to Know, will include a panel of parents and health professionals. The program will begin at 6pm and will include a light dinner. Please mark your calendars and plan to attend. Further information will come in the mail as the date approaches. We already have ideas for our winter and spring meetings. The winter meeting, date to be determined, will bring you all of the latest information from the 2009 North American CF Conference to be held in Minneapolis in October. The spring meeting will focus on transition issues in the pre-teen and teen years. Please contact Kate Kowalski, MSSW, Family Involvement Coordinator, at *kmkowalski@pediatrics.wisc.edu* or 608.262.7457 with questions or topic suggestions for future meetings.

The After Visit Summary (AVS)

Like many other medical organizations, UWHealth is moving toward an electronic medical record (EMR). As part of implementing our EMR, we now have medications listed within our software application (known as Healthlink). This software allows us to directly fax prescriptions to your pharmacy. In addition to this software management of medications, we also have a feature within Healthlink known as the after visit summary (AVS). This page lists the patient's vital signs (pulse, respiratory rate, and blood pressure), height and weight and list of medications. We also can place patient instructions into the AVS. At the conclusion of your clinic visit, the last care provider will print the AVS. Please pick this up at the scheduling desk at the conclusion of your clinic visit.

Airway Clearance Feature: Acapella

Acapella is another form of an airway clearance technique. Acapella combines two different types of therapy into one; high frequency oscillation and positive expiratory pressure (PEP). Inside of the acapella is a lever with a magnet on the end. The magnet helps to intermittently interrupt the air flow across the lever which causes a vibration in the lungs. A dial at one end of the acapella adjusts the amount of resistance. When air is blown out of the lungs through the acapella valve, the air flow moves the lever back and forth. The number and size of lever movements or frequency is based on the resistance of the air flow.

The acapella comes in two colors. Green is for those who have ≥15 L/min of expiratory flow. Blue are for those who have ≤ 15 L/min of expiratory flow. The acapella does have an adapter to combine nebulizer therapy. Talk to your Doctor to see if this combination is right for you.

How to do a Acapella Treatment

- Make sure that the adjustment dial is set to the correct range identified by your health care provider.
- Sit with your elbows resting comfortable on the table or sit upright.
- Make a tight seal around the mouthpiece with your mouth. If you are using a mask, apply the mask tightly but comfortably over your nose and mouth.
- Breathe in naturally taking a deep breath.
- Hold your breath for 2-3 seconds. This allows the air to move evenly throughout the lungs.
- Breathe out slowly and forcefully into the Acapella. Keep your cheeks hard and flat while you are breathing out.
- During the time that you are breathing out, you will feel a "fluttering" sensation in your neck and lungs. If you put your hand on your chest while you are exhaling, you should be able to feel the vibrations. If you do not feel vibrations on your chest, you should blow out with more force.
- You should breathe out 3-4 times as long as you breathe in.
- Continue taking slow deep breaths while breathing out through your flutter valve for about 5 minutes or 20 breaths.
- Remove the mouthpiece or mask and perform a "huff cough".
- A huff cough is performed by taking a deep breath and holding it for 1-3 seconds. Then force the air out of your lungs with your mouth open like you would do if you were trying to fog a mirror.
- Continue steps 2-10 for a total of 20 minutes or as prescribed by your doctor.
- **NOTE: It is very important to pace yourself as you breathe out into the Acapella device. Pause for a few seconds between breaths. If you feel dizzy or get a headache while using the Acapella, you may be breathing too fast. If you develop these symptoms, stop using the Acapella for a few minutes and breathe normally. When you begin to use the Acapella again, take more time between breaths.**

Cleaning the Acapella

The Acapella should be cleaned twice weekly or more often if it is visibly soiled. One of two methods can be used to clean your Acapella.

1. Remove the mouthpiece or mask from the Acapella.
2. Place both pieces of the Acapella on the top shelf of the dishwasher and wash with your normal dishwasher soap.

Or if you do not own a dishwasher, you can

1. Wash both parts of the Acapella in warm soapy dishwater for five minutes.
2. Rinse all of the parts and then place them in a bowl with enough rubbing alcohol (isopropyl alcohol) to cover the Acapella. Soak the Acapella device in the alcohol for five minutes.
3. Remove the Acapella from the alcohol and rinse with sterile water. **DO NOT USE WATER FROM THE FAUCET, BOTTLED, OR DISTILLED WATER.** You can make water sterile by boiling it for five minutes. Use this water once and then throw it out.
4. Drain the Acapella by turning it upside down.
5. Place the Acapella on a clean paper towel to dry.

Return Clinic visits or hospitalizations

It is best to bring your acapella long with you to clinic or if you are hospitalized to have the Respiratory Therapist review your technique.

Email Distribution List

Speaking of upcoming events (such as the Infection Control webcast on August 11, 2009), many of these events occur with a short advanced notice and there is not enough time to place this in our next newsletter. In order to distribute late breaking information to you, we are starting an email distribution list. This will allow us to quickly send information to you. We are gathering email addresses in two ways: 1) we are collecting email addresses during clinic visits, and 2) if you want to be added to our email distribution list, please send your email address to:

Madison-cysticfibrosis-administration@pediatrics.wisc.edu

Please do not use this email address for individual patient related questions and remember to add this address to your address book so your system does not classify it as SPAM.

Don't Forget the Salt!

Stephanie Cronin
UW Pediatric Pulmonary Center Nutrition Trainee

With summer now here, it's important to remember that people with CF need more salt and fluids in their diets. In the warmer months, people with CF are at higher risk for salt and fluid losses because of increased sweat losses.

Salt losses are related to dehydration, which can lead to serious heat-related illness, including heat stroke. Watch for signs of dehydration, such as dark urine, fatigue, confusion, weakness, fever, muscle cramps, and vomiting. Heat stroke is characterized by loss of consciousness, seizures, and kidney failure, and requires *immediate medical attention*. Another sign that a person with CF may be salt-depleted is the appearance of salt crystals around her hairline and on her skin. She may complain that her skin feels "gritty." Infants may lose interest in feeding and appear weak and tired.

People with CF should be monitored for these signs during sports activities, intense playtime, and vacations to hot, humid locations, and in non-airconditioned places like camps, tents, and some homes. However, heat-related illness can also occur from simply being outside on a hot day. Parents, spouses, friends, and coaches should encourage fluids and salt before, during, and after outdoor activities and/or any of the above mentioned situations to prevent dehydration.

People with CF can stay hydrated by increasing their salt and fluid intakes. Most of the time, this can be accomplished by eating salty snacks (such as pretzels), adding salt to food, and drinking to match thirst. Cold, non-carbonated beverages are best for warm weather activity. Sometimes, though, eating salty foods might not be possible. Another way to increase salt intake is to add salt to beverages. For every 12 ounces of Gatorade, Powerade, water, or diluted fruit juice, try adding ⅛ teaspoon of salt to meet salt needs.

Try this recipe for a high salt, high protein snack mix to munch on this summer.

Crunchy Snack Mix

½ cup creamy peanut butter
⅓ cup vegetable oil
1 tsp. celery salt
2 12 oz cans mixed nuts
2 cups Rice Chex
2 cups Wheat Chex
2 cups Cheerios
4 cups pretzels

Mix together peanut butter, oil, and salt. Melt. Mix with rest of ingredients. Bake at 250°F for 1 hour, stirring every 15 minutes. Cool on paper towels and store in airtight container. Makes approximately 12 -1 cup servings.

Nutrition Information

Serving size: 1 cup
Calories per serving:
Protein (gm): 14
Fat (gm) 28
Carbohydrate (gm): 21
Sodium (mg): 850

Suggested Salt Supplementation for Infants and Children with CF

Age (Years)	Usual Conditions* Salt (teaspoons per day)	Extreme Conditions** Salt (teaspoons per day)
0 – ½	⅛ – ¼	¼ – ½
½ – 1	¼ – ½	½ – ¾
1 – 5	⅓ – ⅔	⅔ – 1⅓
Over 5	¾ – 1	1½ – 2

* Moderate temperature and humidity. No excessive sweating. Regular bowel movements. No vomiting or diarrhea.

** High temperature and/or humidity. Dry, desert climate. Strenuous and/or prolonged exercise, especially in heat. Excessive sweating. Fever, diarrhea, vomiting.

Note: Give salt in small portions throughout the day rather than one big dose. These guidelines should be adapted according to individual need and the particular situation. The CF Center will assist you in determining individual recommendations.

Recommended Fluid Intakes for Activities > 30 minutes in the Heat

Remember to adjust for individual needs. Some individuals and some situations may require more fluid than the guidelines recommend.

Time	Fluid
1 – 2 hours before	10-14 ounces
10 – 15 minutes before	8-12 ounces
Every 30 minutes during activity	5-12 ounces (at least 1 ounce for every 20 lbs body weight)
After activity	16 ounces for every lb of weight loss during the activity

VIRTUAL CF EDUCATION DAYS

Germs and the Impact on the Health of People with CF

Watch the 3-part Web cast

Tuesday, August 11, 2009

8 p.m. Eastern Time

(7 p.m. Central, 6 p.m. Mountain, 5 p.m. Pacific)

Join us for the summer Virtual CF Education Day Web cast to learn about germs that affect people with CF and how to avoid them. The discussion will include the basic principles of infection control and information about *b. cepacia, pseudomonas,* MRSA, and influenza. It is important to know and use the basic principles of infection control and prevent the spread of germs in the lives of people with CF.

Leading experts in infectious diseases and CF, John LiPuma, MD, Professor of Pediatric Infectious Diseases and Epidemiology at the University of Michigan, and Lisa Saiman, MD, MPH, Professor of Clinical Pediatrics at Columbia University and NY Presbyterian Hospital, will take an in-depth look at these topics and answer questions from the CF community.

Questions can be submitted during registration and the live broadcast. The broadcast will then be archived in three segments on the CF Foundation's Web site!

Pre-registration begins **July 6th** on the CF Foundation's Web site, www.cff.org or www.cfWebcast.org. Please use code **GC809** when you pre-register. You may submit questions at the time of registration. Questions also may be submitted during the live broadcast. To view this Web cast, your computer must have Microsoft Media Player, version 9.0 or higher. For free Microsoft Media Player go to: http://www.microsoft.com/windows/windowsmedia/download/default.asp. Log in about 10 minutes before the presentation **(check your time zone!)** by going to the CF Foundation Web site (www.cff.org) or www.cfWebcast.org, and click on the "Virtual CF Education Days" icon.
* The screen will remain blank until the program begins. Participation will be limited to the first 1,000 people online.

*****It is recommended that you verify the ability to view the Web cast by watching an archived Virtual CF Education Day Web cast. You can find these archived Web casts on the CF Foundation's Web site, www.cff.org or www.cfWeb cast.org. If you can't join the live Web cast, the program will be archived on the CF Foundation's Web site (www.cff.org).

This Virtual CF Education Day Live Web cast is made possible by an unrestricted educational grant from Genentech, Inc.

Columbia University Medical Center

Morgan Stanley Children's Hospital of NewYork-Presbyterian
Columbia University Medical Center

Cystic Fibrosis Foundation
...adding tomorrows every day.

Therapeutics Development Network

In order to address the need to have a coordinated network of centers that could participate in clinical trials of new therapies for CF, the Cystic Fibrosis Foundation established the Therapeutics Development Network (TDN) in 1998. This network initially was comprised of 13 centers and was later expanded to 18 centers. In the most recent expansion that began in January 2009, the network has grown to nearly 80 centers. Here at the University of Wisconsin, we are proud to have been part of this most recent expansion and to now be a TDN center (*http://www.cff.org/research/TDN/*).

The therapeutic pipeline has many drugs in development, and thus a need for more patients to participate in clinical trials. In the year 2003, approximately 1000 patients were in clinical trials. In the year 2006, approximately 3000 patients were in clinical trials. This year, the Cystic Fibrosis Foundation estimates that 6000 patients will be needed to participate in clinical trials. This is approximately one out of every 5 CF patients in the United States. Please see our Spring 2009 issue of our Center Focus newsletter for a listing of clinical trials being conducted here. Everyone is interested in new and better therapies for CF. **You** are the key to finding new treatments for cystic fibrosis.

The Cystic Fibrosis Patient Assistance Foundation

As we all know, economic times are difficult now. Insurance companies and medical assistance are also affected by the economy and these entities are striving to save money. Unfortunately, this can adversely affect CF patients when their insurance or medical assistance does not cover some of the expense medications. In order to address this, the Cystic Fibrosis Foundation has created the Patient Assistance Foundation. This is a non-profit organization that provides financial assistance to persons with CF who require certain FDA-approved medications. Currently, the Patient Assistance Foundation provides assistance with Pulmozyme® (DNase) and TOBI® (tobramycin solution for inhalation). To determine if you qualify, please contact the Patient Assistance Foundation at 888-315-4154 or visit their website, (*www.cfpaf.org*).

HERE YE, HERE YE… Seeking Parent Volunteers To Become Members Of The UW CF Center Parent Advisory Council

As part of our commitment to provide the best care possible to patients with CF, the UW CF Center is convening a Parent Advisory Council. The purpose of the Parent Advisory Council is to work in partnership with UW CF Center faculty and staff to enhance the medical care and quality of life for children with CF. The Parent Advisory Council will meet regularly with Center faculty and staff to offer a parent perspective to various issues and projects of importance to patients, families and health care providers in the care of children with CF. The time commitment involved will be 60-90 minutes 4 to 6 times per year. We hope you will consider joining this important group. Please contact Kate Kowalski, MSSW, Family Involvement Coordinator, at *kmkowalski@pediatrics.wisc.edu* or 608.262.7457 with questions or to say that you will join us!

Family Mentors Needed for Graduate Student Training Program

As part of their training experience, graduate students working with the Pediatric Pulmonary Center participate in a Family Mentor Experience. The purpose of the experience is to help trainees understand the day-to-day challenges involved with caring for a child with a chronic health condition. The Family Mentor Experience involves a trainee being matched with a mentor family, and spending two to three hours with the family in their home. The trainee talks with parents/family members about what it is like to care for a child with a chronic illness, and with the child about her/his experience of living with a chronic illness. Both the families and the trainees are provided with an orientation prior to the visit to assure that all understand the purpose and recommended guidelines. The parent orientation is typically done by phone. If you are interested in learning more about having your family become a mentor family, please contact Kate Kowalski, MSSW, Family Involvement Coordinator, at *kmkowalski@pediatrics.wisc.edu* or 608.262.7457.

What's New At CFF

Tracy Earll, State Director

On a beautiful Sunday in May, which happens to be national Cystic Fibrosis Awareness month, nearly 500 people gathered at the Monona Terrace in Madison with one goal in mind. To raise awareness and funds for CF. (In fact, that day, a record $185,000 was raised in Madison for our cause.) All across the country, this was the picture painted in thousands of cities. Raising over $30 million dollars.

While at the same time we were walking, Dr. Bob Beall, the dedicated and innovative leader of the foundation walked into a conference room of staff who were tracking funds raised throughout the country to announce what many of us have all been "patiently" waiting for... ***VX-770 has moved into Phase 3 clinical trials.***

VX-770 is one of two drugs (the other being VX-809) that are aimed at treating the basic defect of CF. VX-770 is a "potentiator" that may act upon the CFTR protein and help to open the chloride channel in CF cells. VX-809 is a "corrector" that helps move defective CFTR protein to the proper place in the airway cell membrane and improve its function as a chloride channel. This is currently in phase 2 trials.

The VX-770 trial is designed to generate data that the U.S. Food and Drug Administration can use to determine if VX-770 is safe, effective and acceptable for approval. The VX-770 registration program will consist of three different clinical trials, including:

- A primary 48-week trial for patients age 12 years and older who carry a G551D mutation of CF;
- A 48-week trial for patients ages 6 to 11 years who carry a G551D mutation of CF; and
- A 16-week trial to evaluate for the first time patients age 12 years and older who are homozygous for the Delta F508 mutation of CF, the most common mutation in CF patients.

"The initiation of the VX-770 registration program is a major advancement in our efforts to bring forward a new therapy aimed at treating the underlying cause of CF," said Robert J. Beall, Ph.D., president and chief executive officer of the Cystic Fibrosis Foundation. "This investigational drug represents one of the most promising routes to changing the course of this disease, and we are encouraged with the progress of both VX-770 and VX-809, which recently entered a Phase 2a clinical trial in CF patients."

No matter when your loved one was diagnosed with cystic fibrosis, the role of the Cystic Fibrosis Foundation has been to raise the vital funds needed to further the progress of cystic fibrosis research and care. We think it is safe to say that the foundation been extremely efficient and dedicated to this goal over the last several years. And due to our great work, we are getting closer and closer to finding a true control to this disease. Just remember, we can't do this without you. Financial resources are the only thing holding us back from reaching our goals. Please call the local foundation at 608.298.9902 or visit us at (*http://www.cff.org/chapters/madison*) for more information on getting involved.

Our Website

Did you know that we are on the web? Please visit our website at:

www.uwcfcenter.org

This newsletter and previous versions of this newsletter are on our website. We also hope to update our website frequently to inform you of upcoming events.

Farewell To Dr. Zoran Danov

One of our pediatric pulmonology fellows, Dr. Zoran Danov, is graduating from our program. He is moving to Lexington, Kentucky where he will join the Pediatric Pulmonology Division at the University of Kentucky. Our best wishes to Dr. Danov for his career in the bluegrass state.

Meet Tucker

Tucker was diagnosed with cystic fibrosis when he was 2 weeks old. We were informed by his pediatrician when we took him in for his one week check up that the newborn screening test that was completed when he was born came back positive for CF. We were then referred to the UW Children's Hospital & Clinics the very next week, where the sweat test was completed and confirmed the diagnosis. We were completely devastated when we got the news. Our devastation turned into anger, frustration, and sadness. A couple of months later after we had time to emotionally deal with the diagnosis, we came to the realization that everything in life happens for a reason. At that point, we committed ourselves to do whatever it took to raise as much money as possible for the Cystic Fibrosis Foundation (CFF), in hopes that a cure would be found during Tucker's lifetime.

Since that time, we have become very active fundraisers for the CFF. We participate in the annual Great Strides walk in Madison during the month of May. We also do a variety of other fundraisers throughout the year, with our most successful fundraiser being our annual golf tournament held in honor of Tucker, which is called "Teein' It Up For Tucker". Since we began our fundraising efforts, we have raised over $150,000 for the Cystic Fibrosis Foundation. We are truly blessed to have such wonderful and supportive family and friends in our lives who have made this possible. They are the ones that continue to keep our hope alive that a cure will be found during Tucker's lifetime! They have definitely made our family's journey with CF much less difficult to travel over these past 7 years, and for that we will forever be grateful.

Tucker is now 7 years old and in the 1st grade at Wonewoc-Center School. He loves to play basketball, baseball, golf, and football! He absolutely loves the Wisconsin Badgers, the Green Bay Packers, and the Milwaukee Brewers! He is a great kid with a huge heart, who lives each day to the fullest, and reminds us to do the same. He has blessed our lives more than he will ever know!

Every day of our lives we wish we could take Tucker's place, but unfortunately we can't. That is why we will continue to support the Cystic Fibrosis Foundation in every way possible until this disease can be cured!!! We look forward to celebrating the day when "CF" stands for ***"Cure Found"!***

Tory & Jodi Needlham
Wonewoc, WI