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University of Wisconsin
Clinical Sciences Center

Center Focus

Winter, 2015

Newsletter of the
Cystic Fibrosis Center

Immunizations

Flu season is once again upon us. This is a gentle reminder of the recommendations regarding vaccines to keep your child (or yourself, for adults with CF) as healthy as possible throughout the upcoming flu season. Here are 2 websites from the American Academy of Pediatrics regarding the 2014 immunization schedules and information regarding Pneumococcal disease:

http://www2.aap.org/immunization/IZSchedule.html

http://www2.aap.org/immunization/illnesses/pneumococcal/pneumococcal.html

The Wisconsin Immunization Registry (WIR) is an excellent resource to check your child's immunization status. It identifies all vaccines received in the state of WI and what vaccines your child is in need of. The link to the website is: *https://www.dhswir.org/PR/clientSearch.do?language=en*. If your child receives immunizations in a state other than WI, please check with your state's immunization registry.

Listed below are the vaccines that are recommended for your child with chronic lung disease:

- Annual influenza vaccine (flu shot). This vaccine is recommended for all patients seen at AFCH Pulmonary/CF Specialty clinic each year starting at the age of 6 months. The vaccine can be obtained at your PCP office, AFCH Pulmonary/CF Specialty clinic or in your community. The vaccine is listed as "influenza" in the WIR system.

- Pneumovax vaccine (PPSV 23). This vaccine is recommended for all patients seen at AFCH Pulmonary/CF Specialty clinic starting at the age of 2. If your child has already received this vaccine they will not need a booster until the age of 65. The vaccine is listed as "pneumo-poly" in the WIR system. If your child has never received this vaccine and is 6 years of age and older they should receive this vaccine this fall from your PCP office or at AFCH Pulmonary/CF Specialty clinic. If your child has never received this vaccine and is 2-5 years of age they will first require 1-2 doses of Prevnar 13 given at least 8 weeks apart. This vaccine is listed as "pneumococcal" in the WIR system. It is then further described as Prevnar (PCV7) or Prevnar 13(PCV13). This is a vaccine that most all children received in the first 2 years of life. It is typically a 4 shot series. If your child has received 4 doses of Prevnar (no Prevnar 13) they will require one dose of Prevnar 13 8 weeks prior to receiving Pneumovax. If your child received 3 doses of either Prevnar or Prevnar 13 they will require one dose of Prevnar 13 8 weeks prior to receiving Pneumovax. If your child has received less than 3 doses of either Prevnar or Prevnar 13 they will require 2 doses of Prevnar 13 8 weeks apart prior to receiving Pneumovax. Prevnar 13 can be obtained at your PCP office or at AFCH Pulmonary/CF Specialty clinic.

If you have any questions or concerns regarding the information listed above, please do not hesitate to contact the AFCH Pulmonary/CF Specialty clinic or your PCP office.

Enterovirus D68

In the summer of 2014, you may have heard about a virus that caused severe respiratory illness in patients with asthma. The virus is called Enterovirus D68. It was first discovered in 1962 and is one of more than 100 non-polio enteroviruses that can cause respiratory illness with symptoms of fever, runny nose, sneezing, cough, and body and muscle aches. There were patients across the country with a diagnosis of asthma who became very ill with this virus. To our knowledge, we are not aware that this virus caused significant illness in people with CF.

This virus most commonly circulates in the summer and fall. For this virus and all respiratory viruses, you can utilize these steps to avoid becoming ill:

- Wash hands often with soap and water for 20 seconds.
- Avoid touching eyes, nose and mouth with unwashed hands.
- Avoid close contact such as kissing, hugging, and sharing cups or eating utensils with people who are sick.
- Cover your coughs and sneezes with a tissue or shirt sleeve, not your hands.
- Clean and disinfect frequently touched surfaces, such as toys and doorknobs, especially if someone is sick.
- Stay home when you are sick.

News from the Wisconsin Office of the Cystic Fibrosis Foundation

CFF is pleased to continue Great Strides at Warner Park and the Mallards Stadium, the 'Duck Pond.' Great Strides is our favorite time of the year. It offers us the opportunity to bring together the entire CF community to bond and share our triumphs and hopes for the future. Our CF 'Team' will gather together on the 17th of May to walk for a cure and celebrate our 'home runs' in advocacy and research this past year. We look forward to meeting with all our fabulous participants and hope that each of you will choose to participate in this great event! Contact your Great Strides General Manager, Tim Schmitt at 608-298-9902 or *tschmitt@cff.org* for more details.

While we continue to search for a cure, we want to ensure that we are providing the best possible services and resources to assist all patients and families. Our Patient Assistance Resource Center (PARC) wants to help you access affordable medications, devices and the best healthcare, help navigate and understand insurance coverage, offer an online database of documents and give free legal aid. People with CF, families and care centers may contact PARC at 1-888-315-4154 or *parc@cff.org*.

Want your voice to be heard?! The Cystic Fibrosis Foundation is looking to partner with the CF community on a host of projects. If you are interested in being a CF adult or family advisor you can learn more and apply at this link:

https://adobeformscentral.com/?f=iL0dQOHUaS6Nf7LCqHf5Dg

Cystic Fibrosis Center Specific Outcomes

Here is our center specific data for the calendar year 2013. There is publicly available data on all Cystic Fibrosis Centers available at the Cystic Fibrosis Foundation website (*www.cff.org*). Data on that website is adjusted for attained age of patients, gender, pancreatic sufficiency, race/ethnicity, socio-economic status, and age of diagnosis. As of the writing of this newsletter, the CFF website has not yet been updated to include the 2013 data. We expect that data to be publicly available in the near future. In the meantime, we now have our UW CF Center data for last year, and what follows in this article is the raw data reported to us for the calendar year 2013.

Pediatric Center

A very useful measure of lung function is the FEV_1 percent predicted. For patients 6-12 years of age, the University of Wisconsin CF Center median FEV_1 percent predicted is 100.5%, which is 5.3% above the national average of 95.2%. For patients 13-17 years of age, the median FEV_1 percent predicted is 90.9% at UW compared to a national median of 86.4%. For patients 6-17 years of age, the UW median FEV_1 percent predicted is 94.8% compared to a national median of 89.2%.

Our nutritional outcomes are expressed in terms of BMI (Body Mass Index) percentile. For pediatric patients 2-19 years of age, we strive for a BMI percentile of at least the 50th percentile. The median BMI percentile for CF patients 2-19 years of age at UW was 58.7 compared to a national average of 53.3.

Adult Center

For patients 18-29 years of age, the median FEV_1 percent predicted at UW is 78.3% compared to a national average of 72.3%. For CF patients > 30 years of age, the median FEV_1 percent predicted at UW is 51.9% compared to a national average of 57.9%.

In adults over 20 years of age, one no longer uses BMI expressed as a percentile. In adult patients, BMI is expressed as the actual value with the units of kilograms per meter squared. The Cystic Fibrosis Foundation goal is that males should have a BMI of greater than or equal to 23 kilograms per meter squared and females should have a goal BMI greater than or equal to 22 kilograms per meter squared. In the UW Center, the percentage of adult patients who met those goals was 43.2% compared to the national average of 46.5%

Calories Count

Erin Seffrood, MS, RD, CSP, CD
Pediatric Dietitian

If you attended the CF Education Day back in November 2014, you got to hear a great presentation by Scott Powers, PhD, Clinical Psychologist and Director of the Center for Child Behavior and Nutrition Research and Training at Cincinnati Children's Hospital. He has done a great deal of research about getting kids with cystic fibrosis to gain weight. One simple thing that works well is adding a certain number of extra calories each day.

As a pediatric dietitian, I often suggest trying certain foods that are high calorie (avocado, nut butters) or substituting higher calorie choices for lower calorie choices (whole milk versus skim milk) but don't give a specific number of calories to add on a daily basis. After I heard Dr. Powers speak, I started incorporating his recommendation more often and IT WORKS! Simply make it a point to add 100, 200, 300 extra calories every single day in addition to what you are already eating. If you do this consistently. It will pay off. Below are some things to add each day. Pick one thing and stick to it for a few months and you will likely see results.

100 Calorie Choices (do once, twice, or three times per day - consistency is key!)

- 1 Tbsp peanut butter, nut butter or chocolate/nut spread
- 1 Tbsp butter or oil
- 1 cheese stick
- 1 Tbsp mayonnaise
- Handful of nuts, seeds or trail mix
- 2 Tbsp heavy whipping cream
- 1/4 cup hummus
- 1/4 cup dried fruit
- 1/4 cup sour cream
- 2 Tbsp honey
- 1 1/2 Tbsp ranch dressing

CF Services Pharmacy No Longer A Specialty Pharmacy for Cayston

The CF Services Pharmacy is no longer a specialty pharmacy for Cayston. For patients who have been receiving Cayston through the CF Services Pharmacy, the Cayston Access Program (*https://www.cayston.com/cayston-access-program/*) will assist in transitioning these patients' specialty pharmacy to one of the other three providers (IV Solutions in Lubbock, Texas; Foundation Care in Earth City, Missouri; or Pharmaceutical Specialties, Inc. in Bogart, Georgia).

Meet The Staff

Vivek Balasubramaniam, M.D.

Dr. Vivek Balasubramaniam received his undergraduate degree from the University of California, Los Angeles and his medical degree from the University of Pittsburgh. He stayed in Pittsburgh for a pediatric residency, and then moved to Denver, Colorado for a dual fellowship in Pediatric Pulmonology and Critical Care Medicine at The Children's Hospital. He was a research fellow from 2002 to 2003 and then joined the faculty in the Division of Pediatric Pulmonary Medicine in July 2003. Dr. Balasubramaniam joined the faculty in the Pediatric Pulmonology and Sleep Medicine Division here at the University of Wisconsin in August 2014. His research interest is in the pathogenesis, treatment and prevention of lung disease that occurs as a result of premature birth.

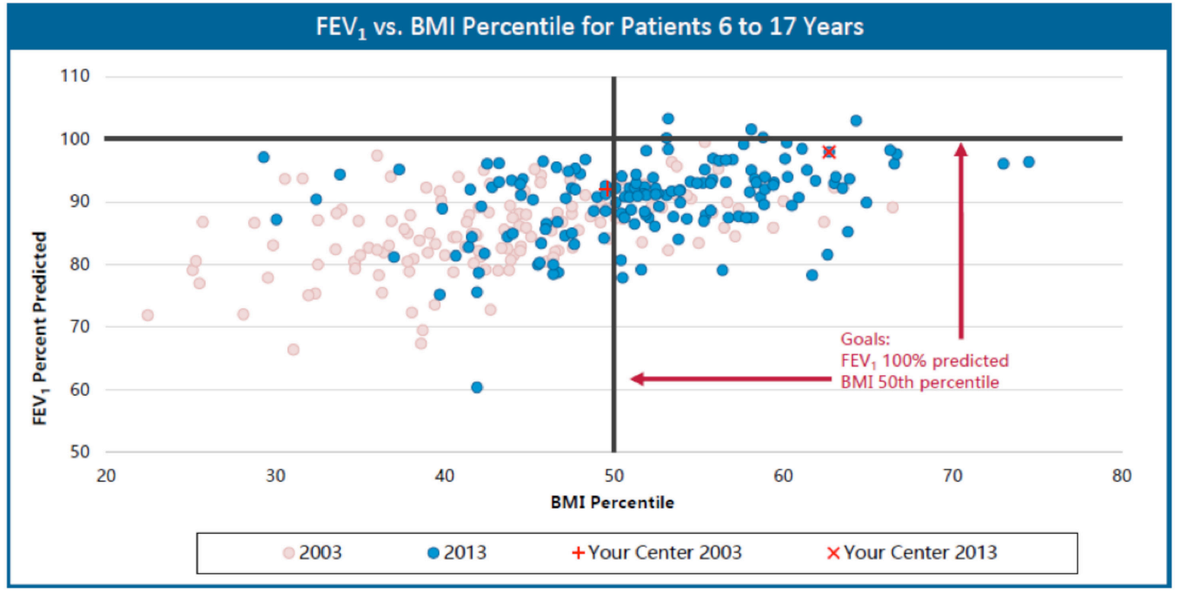
Trends over the last four years

The following table shows data for the past four years. The UW CF Center specific data is shown first, followed by the national average in parenthesis. This allows you to see trends that are occurring both on a local and a national level.

	2013 UW (national average)	2012 UW (national average)	2011 UW (national average)	2010 UW (national average)
Median FEV_1 % predicted 6-12 years	100.5 (95.2)	99.7 (97.9)	98.7 (97)	102.3 (97)
Median FEV_1 % predicted 13-17 years	90.9 (86.4)	93.1 (90.8)	92.9 (90)	90.8 (90)
Median FEV_1 % predicted 6-17 years	94.8 (89.2)	98.6 (95)	96.3 (94.5)	96.0 (94.0)
Median FEV_1 % predicted 18-29 years	78.3 (72.3)	74.8 (71.9)	76.4 (71.5)	66.3 (70)
Median FEV_1 % predicted >30 years	51.9 (57.9)	47.4 (57.9)	51.5 (57.5)	56.3 (56)
Median BMI percentile 2-19 years	58.7 (53.3)	59.3 (52.8)	56 (51.9)	56 (49.4)
Adults over 20 years of age: % males with BMI >23 and females BMI >22	43.2 (46.5)	44.7 (45.7)	46.2 (44.9)	43.9 (43.3)

Of note is that in 2013, the CF Foundation started using the Global Lung Initiative reference equations. (The previous reference equations were by Wang and Hankinson.) This change results in lower FEV_1 percent predicted values for preteens and teens.

Similar to last year, the CF Foundation provided a graph to the care centers in which FEV_1 %predicted is plotted against BMI. Here is the graph for 2013:



Each pink circle is center data for 2003 and each blue circle is data from 2013. The UW CF Center data from 2003 is indicated by the plus sign, and the UW CF center data from 2013 is indicated by the 'x". The vertical line is the CF Foundation goal of a BMI of the 50th percentile and the horizontal line is the CF Foundation goal of an FEV_1 of 100% predicted. There are 2 or 3 blue dots that are above and to the right of the UW CF Center data in 2013. Therefore, the UW CF Center has the 3rd or 4th best outcomes in the country in 2013 when combining FEV_1 %predicted and BMI.

Save The Date- November 7, 2015

Our next CF Family Education Day will be November 7, 2015 at the Fluno Center (601 University Avenue, Madison, WI 53715; *http://fluno.com*). We are pleased to announce that our keynote speaker will be Dr. Michael Boyle, director of the adult CF program at the Johns Hopkins University in Baltimore, Maryland. Dr. Boyle presented plenary sessions at the North American Cystic Fibrosis Conference in 2007 and 2014. Dr. Boyle is an outstanding speaker and we are honored that he can come to Madison for our CF Family Education Day.

Infection Control

In our Spring, 2014 issue of the Center Focus, we discussed the new CF Foundation Infection Control Guidelines. This was published in the August 2014 issue of Infection Control and Hospital Epidemiology. Within this document, there were 77 recommendations listed. In our Spring, 2014 issue of Center Focus, we have highlighted some of the global recommendations that affect our interactions with patients in the clinic and in the hospital. Here are some other recommendations that are of importance:

Nebulizers: Cleaning and Disinfecting. The CF Foundation recommends that the following steps be performed for nebulizers used in the home as soon as possible after each use:

a. Clean the nebulizer parts with dish detergent soap and water

b. Disinfect the nebulizer parts using one of the following methods:

Heat methods:

a. Place in boiling water and boil for 5 minutes

b. Place in a microwave-safe receptacle submerged in water and microwave for 5 minutes

c. Use a dishwasher if the water is more than or equal to 70°C or 158°F for 30 minutes

d. Use an electric steam sterilizer

Cold methods:

a. Soak in 70% isopropyl alcohol for 5 minutes

b. Soak in 3% hydrogen peroxide for 30 minutes

i. Rinse off the cold-method disinfectant using sterile water, not tap water; the final rinse must be with sterile or filtered (less than or equal to 0.2-micron filter) water

ii. Air dry the nebulizer parts before storage

Leisure activities

A. The CF Foundation recommends that people with CF should limit prolonged and/or repeated exposure to activities that generate dust from soil and organic matter (eg, gardening and lawn mowing) to decrease exposure to potential soilbourne pathogens

B. The CF Foundation recommends that people with CF should avoid exposure to construction and renovation activities that generate dust to decrease exposure to potential pathogens.

C. The CF Foundation recommends that people with CF can swim in pools or water parks with adequate disinfection (eg, chlorination)

D. The CF Foundation recommends that people with CF avoid activities in hot tubs, whirlpool spas, and stagnant water. (There is insufficient evidence to recommend for or against people with CF avoiding activities in natural bodies of water that are not stagnant, eg, oceans, ponds and hot springs)

E. The CF Foundation recommends that people with CF perform hand hygiene after changing the litter, handling feces, cleaning and disinfecting the cages or fish tanks of their pets, or interacting with farm animals.

F. The CF Foundation recommends that people with CF avoid cleaning stalls, pens or coops.

Studies

The following studies are in progress at the University of Wisconsin CF Center:

FIRST Study: Feeding Infants Right from the Start:

Inclusion criteria: Diagnosed with CF by newborn screening

Purpose of the study: The purpose of the research is to understand how different types of infant feedings, breast milk and/or infant formula, affect growth and lung health in infants with cystic fibrosis (CF). (This study concludes on the child's second birthday.)

FIRST Study: Feeding Infants Right from the Start- Phase 2:

Inclusion criteria: Infants who participated in the 0-2 years FIRST study.

Purpose of the study: The purpose of the research is to understand how infant feeding and childhood food intake influences growth and lung health in children with CF over 2-6 years of age.

A Long-Term Prospective Observational Safety Study of the Incidence of and Risk Factors for Fibrosing Colonopathy in US Patients with Cystic Fibrosis Treated with Pancreatic Enzyme Replacement Therapy: A Harmonized Protocol Across Sponsors

Inclusion criteria: Patients suspected of having fibrosing colonopathy (FC)

Purpose of the study: The U.S. Food and Drug Administration (FDA) has asked the manufacturers of pancreatic enzyme supplements to study the incidence of FC in Cystic Fibrosis patients to see if there may be a link between taking pancreatic enzyme medications and developing FC.

AquADEKs2: A Multi-Center, Randomized, Controlled, Double-Blind Study of the Effects of an Antioxidant-Enriched Multivitamin Supplement on Inflammation and Oxidative Stress in Cystic Fibrosis Patients

Inclusion criteria: ≥10 years of age and weight ≥30 kg; Pancreatic insufficiency documented as a fecal elastase ≤100 µg/g of stool; FEV_1 of 40-90% predicted based on the Wang (males <18 years, females <16 years) or Hankinson (males ≥18 years, females ≥16 years) standardized equations at the screening visit

Purpose of the study: To learn if taking AquADEKs-2 is safe and effective in people with CF.

OPTIMIZing Treatment for Early Pseudomonas aeruginosa Infection in Cystic Fibrosis. The OPTIMIZE Multicenter, Placebo-Controlled, Double-Blind, Randomized Trial

Inclusion criteria: age ≥6 months to ≤18 years; documented new positive culture for *Pseudomonas aeruginosa* (either first lifetime culture positive or *P. aeruginosa* recovered after a two-year history of negative cultures)

Purpose of the study: Standard care for a first *Pseudomonas aeruginosa* (*Pa*) lung infection in people with Cystic Fibrosis is to treat with tobramycin inhaled solution (TIS). This study is being done to see if adding the antibiotic azithromycin to TIS treatment will reduce lung exacerbations, help get rid of *Pa* when it is first found, prevent *Pa* from coming back after the first time, and improve health by combining azithromycin with TIS.

MRI of Cystic Fibrosis Lung Disease Severity

Inclusion criteria: ≥10 years of age

Purpose of the study: The purpose of this study is to determine the ability of MRI to evaluate CF disease severity compared with the existing standards of breathing tests, chest x-ray, chest CT, and quality of life questionnaire.

The EPIC Observational Study: Longitudinal Assessment of Risk Factors For and Impact of Pseudomonas aeruginosa Acquisition and Early Anti-Pseudomonal Treatment in Children with CF

Inclusion criteria: Previous participation in the EPIC observational study for years 6-10.

Purpose of the study: The purpose of this study is to answer questions about Pseudomonas aeruginosa (*Pa*) and other germs that cause lung infections in people who have CF.

A Phase 3 Rollover Study of Lumacaftor in Combination With Ivacaftor in Subjects 12 Years and Older With Cystic Fibrosis

Inclusion criteria: Previous participation in the VX-809 study.

Purpose of the study: To evaluate the long-term safety and tolerability of lumacaftor in combination with ivacaftor in subjects with CF, homozygous for the F508del-CFTR mutation.

The following studies will be starting soon:

CLEAN-CF (Clearing Lungs with ENAC Inhibition in Cystic Fibrosis): A Randomized, Double-Blind, Placebo-Controlled, Parallel-Group Study to Evaluate the Safety and Efficacy of P-1037 Solution for Inhalation in Patients with Cystic Fibrosis (CF)

Inclusion criteria: ≥12 years of age; FEV_1 of 40-90% predicted using the Global Lung Initiative reference equations

Purpose of the study: The goal of the study is to evaluate the safety and tolerability of P-1037 and to determine whether the combination of P-1037 with hypertonic saline or P-1037 alone has a greater effect on lung function in patients with CF than placebo (0.17% saline)

INSTANT (Innovative Sweat Test, A New Technology): A Multicenter Trial of A New Method of Sweat Testing

Inclusion criteria: A diagnosis of CF or CRMS (CFTR-related metabolic syndrome)

Purpose of the study: The primary objective is to evaluate the diagnostic accuracy of the CF Quantum Sweat Test.

If you have questions about these studies, please contact the CF research coordinator for adult patients (Chiron Stevens, 608-263-1244) or the CF research coordinator for pediatric patients (Linda Makholm, 608-262-0340)