University of Wisconsin
Clinical Sciences Center

Center Focus

Newsletter of the Pediatric and Adult Cystic Fibrosis Centers

Spring, 2018

CFF Advocacy Day

The first-ever Cystic Fibrosis Advocacy Day took place on February 13, 2018 at the Wisconsin state capitol. We focused on educating lawmakers about CF: we helped them learn more about the disease, and aimed to build positive relationships. We talked about the importance of the Wisconsin Chronic Disease Program and Medicaid. We also encouraged lawmakers to support insurance protections for people with CF. We left behind a packet of materials for legislators, which included a state factsheet.

Our group included someone with CF, parents and loved ones, and members of the UW clinical care teams for both kids and adults. Cystic Fibrosis Foundation policy experts Zoe Aldrich and Meghan Pudeler helped us prepare and learn what to expect at the meetings.

We broke up into groups of 3-5, and had meetings with state senators, representatives, and members of their staffs throughout the day. One parent showed her son's CF vest and inhaler to Senator Alberta Darling. Darci Pfeil taught Senator Jon Erpenbach's staff member about CF care needs. Senator Fred Risser taught us about the status of a bill as it made its way through the legislature.

The event felt meaningful and empowering, and we believe created good groundwork for future collaborations that will support the needs of people with CF in accessing care, and in protecting quality of life.

For more information about CF advocacy, you can go to:

https://www.cff.org/Get-Involved/Advocate/Advocate-With-Us/

https://twitter.com/CFFWI?lang=en

Save the date for 2018 CF Family Education Day…

Date: Saturday, November 10, 2018

Time: 8 am-1 pm, lunch included

Location: The Fluno Center on UW Madison Campus

Theme: Living with CF

National Speakers: Dr. Robert Beall (former CEO of the CFF) and the Partnerships for Sustaining Daily Care Team from the CFF

Keynote speaker: Adult who is living and thriving with CF

Watch for more information to come…

Pediatric Inpatient Transition from P5 to P7

AFCH is expanding!!

We are so excited, after lots of planning and preparing the nursing staff of the Pediatric Universal Care Unit (P7) will begin to care for patients with cystic fibrosis who require hospitalization as of July 2018.

The change is taking place to enhance and improve care for all our pediatric pulmonary patients, including those with cystic fibrosis.

If you would like more information or are interested in talking to the P7 manager, Laura Ahola can be reached at 608-890-7175.

We look forward to moving to our new inpatient location!!

SYMDEKO: a new CFTR modulator therapy

On February 12, 2018, the U.S. Food and Drug Administration approved a new therapy for some individuals with cystic fibrosis.

Symdeko (tezacaftor/ivacaftor) is the third medication in a class of therapies known as CFTR modulators. It is approved for individuals with CF, ages 12 and older, who have either two copies of the F508del gene, or at least one copy of another 26 mutations.

Symdeko is made up of two drugs that work together to improve the functioning of CFTR – the chloride channel protein that is defective in individuals with CF. The tezacaftor brings more CFTR proteins to the surface of an affected cell. Ivacaftor helps the chloride channel to "stay open longer." Symdeko works very similar to the already approved, Orkambi (lumacaftor/ivacaftor); it exchanges the lumacaftor drug for tezacaftor. Symdeko is taken as an oral pill twice daily: a yellow pill in the morning, and a blue pill in the evening. It needs to be taken with fat-containing foods.

In the two phase 3 studies of Symdeko, individuals showed an average improvement in their FEV1 by 4 to 6 percentage points when compared to those who were taking placebo. The effect was seen to last for 24 weeks (the length of the study). Additionally, the individuals on Symdeko had a decrease in their "exacerbations" by 35 percent, and an improvement in their respiratory symptoms.

Similar to other CFTR modulator therapies, Symdeko does have potential side effects including abnormalities in liver enzymes, a risk of developing cataracts in the EYE (this was only seen in children and adolescents), and interactions with other medications such as anti-fungal therapies, and certain antibiotics. The most common side effects included headache, upset stomach, and dizziness. Notably, there was not an increase in chest tightness, which has been seen with Orkambi.

The CF clinic has a list of all patients who are eligible for Symdeko. Your provider may discuss this new medication at your next visit. For further information about Symdeko, you can read more at the website: *www.symdeko.com*.

Foods with 20 grams of fat

2 ½ TBSP peanut butter

24 almonds (1/2 cup is 27 grams)

4 TBSP pumpkin seeds or sunflower seeds

2 oz cashews

1/3 cup peanuts

1/3 cup walnuts

1 avocado

2 TBSP butter or mayo or margarine

2 ½ oz cheddar cheese

4 TBSP cream cheese (oil based may be less fat, cream based may be more fat)

2 TBSP salad dressing (VARIABLE, check labels)

1 ½ TBSP oil

CF Pediatric Parent Advisory Group Meeting

The first CF Pediatric Parent Advisory Group Meeting (CF PAG) was scheduled for January 2018 but was cancelled due to weather. It was rescheduled and we kicked off our inaugural meeting on Thursday March 8th, 2018. We had several parents who had expressed interest and attended the first meeting. The meeting was a success and we hit the ground running with several great ideas and topics to work on. We can't wait to provide you with updates as our work progresses.

If you are interested in learning more about the CF PAG, please feel free to discuss at next clinic visit and we can provide you with more information!

Next meeting is scheduled for May 10th, 2018.

New Faces

Hi everyone! My name is Jessica La Mar, and I am so happy to be a part of the UW Adult Cystic Fibrosis team. This may not be a first hello as I have met or spoke with many of you already. I am a nurse practitioner (NP) and also the new clinic coordinator for all adult cystic fibrosis patients. I spend the majority of my time in the pulmonary clinic, though I do see patients in the hospital from time to time. You will often see me during the CF clinic on Tuesdays, but I do work Monday through Friday. I have been a NP since the fall of 2016, graduating with my doctor of nursing practice (DNP) at UW-Madison ***(go Badgers!)*** and then accepting my first position in the UW Pulmonary Clinic immediately after. Though prior to completing my graduate studies, I was a nurse for 10 years here at UW working mostly in the Trauma and Life Support Center (TLC). I enjoyed my work as an intensive care nurse but wanted to become a NP to help patients before they became so ill.

I'm looking forward to be an integral member of your care team and getting to know you. As the clinic coordinator my goal is to help ease transitions between clinic visits, home IV therapy, and hospitalizations. Additionally, to work on improving the efficacy and efficiency of your busy clinic visits to ensure you are getting the care you need, when you need it. Our goal as a team is to help you sustain daily cares with prescribed medications, airway clearance and exercise. Please feel free to reach out to me the future with any questions/concerns about your care. As always, you can call the clinic at 608.263.7203 to contact me.

A few snippets about me:

1. I love do-it-yourself (DIY) projects, especially creating home décor and personalized gifts.
2. I grew up in a very small town in central Wisconsin (Spencer) and moved to Madison after finishing my nursing degree in 2006. I immediately fell in LOVE with Madison and living in this beautiful city every day. You can usually find me at the Farmer's Market on the Square most Saturday mornings.
3. I have one cat named Gucci… he looks like he has a tuxedo on!

My name is Erin Billmeyer and I am a nurse practitioner with the adult cystic fibrosis pulmonary group. I work mostly with patients when they need to be hospitalized for any reason. Occasionally you can also find me in the clinic helping out when the things are busy or someone needs to be seen urgently. I enjoy working in the hospital because I get to know patients better and am able to build a relationship with them. I came to the cystic fibrosis group after working in the the ICU for 4 years. Prior to be a nurse practitioner, I worked as a RN in the ICU, Radiology and pulmonary research unit for 13 years.

On a personal note, I love to do triathalons and bike long distances. I like cooking and am always excited to talk about food.