University of Wisconsin
Clinical Sciences Center

Center Focus

Newsletter of the Pediatric and Adult Cystic Fibrosis Centers

Spring, 2017

Mission statement

Our Cystic Fibrosis Center's mission is to provide excellent patient care while partnering with patients and their families to enhance quality of life, promote education, increase public awareness of the many challenges that patients with CF face, and make meaningful contributions in the search for a cure by participating in clinical trials that evaluate new therapies for treating patients with CF.

CF Family Education Day 2017

CF Family Education Day is scheduled for Saturday, April 22 at the Fluno Center on UW-Madison Campus. The theme of this year's education day is "Something for Everyone". Registration begins at 8 a.m. with the program starting at 8:45 a.m. and ending at 3:30 p.m. Continental breakfast, lunch and snacks will be served. Dr. J.P. Clancy, from Cincinnati Children's CF Center is our key note speaker. To register online go to: *http://go.wisc.edu/CFFamilyDay*. If you have questions or are unable to register online, please call or email Leslie Lord, Outreach Specialist at *llord@pediatrics.wisc.edu* or call 608-262-7457 to register for the event. A room block has been established at the Fluno Center for those who wish to stay overnight. Please contact the Fluno Center directly at (877)-773-5866 or (608)-262-7457. For additional information about the Fluno Center: *http://fluno.com*

By focusing on specific outcomes, effectively using data, standardizing care processes, and empowering patients and families, we transformed the culture and delivery of care for our patients with CF. As our work continues, we continue to learn valuable lessons and insights, and we welcome any questions or comments!

CF Parent Advisory Group at AFCH… Coming Soon

Our Pediatric CF Center at American Family Children's Hospital has been partnering with families to improve the quality of the services we provide. We have been working to reduce the time spent in the waiting room, improving our infection control practices and enhancing our efforts to clean and sanitize clinic rooms.

We are looking to build on the momentum that has been building and are forming a Parent Advisory Group to help guide and lead us. You will be receiving an invitation to join in the next several weeks. We are hoping to have a wide range of family perspectives to guide us in our efforts to improve the care and services we provide.

YOUR CF CLINIC VISIT

Our goal is to see all of our CF patients on a quarterly basis at a minimum, and while we understand that sometimes these visits and associated testing may be long and overwhelming, there is a method behind our madness! As an accredited Cystic Fibrosis Care Center, your multidisciplinary team is devoted to giving you/your child the best care possible. Therefore, we recommend evaluation and treatment recommended in CF clinical care guidelines which are based on the best medical evidence and practices.

Along with at least four visits per year with a cystic fibrosis specialist who will be monitoring you closely and adjusting your treatment regimen as necessary, there are multiple other recommendations that we follow which are summarized in the charts below.

CFF Guidelines for Frequency of Evaluation/Tests for patients with CF.

Adult CF Team Welcomes It's Newest Member!

Lisa Atkins, PT, CCI

Hello patients, families, and friends of the UW Health Adult Cystic Fibrosis community,

I am happy to write this inaugural Physical Therapy segment for the University of WI Cystic Fibrosis Center Newsletter.

Last fall, Dr. Trina Hollatz, the Adult CF Program Director, learned that the Cystic Fibrosis Foundation was offering a grant that would support adjunct medical services in an Adult Outpatient Cystic Fibrosis Clinic.

Why was the Cystic Fibrosis Foundation interested in having physical therapy be part of the medical team? Because evidence continues to mount that regular exercise can improve the quality of life of those living with CF. Even more exciting is that there is accumulating evidence that an early and consistently maintained aerobic exercise program can prolong the lives of many living with CF.

I was excited to learn about this opportunity and volunteered. Dr. Hollatz and I wrote, applied for, and received this special grant.

The "I" in this article is Lisa Atkins PT. You can find my profile on *www.UWHealth.org*, search my name, and you can see me there.

I started shadowing the very experienced clinicians at the UW Adult CF Clinic on Tuesday's beginning in January 2017 and have already learned so much. My general objective is to be able to synthesize and organize a program for the adult patients living with CF and their families. This will include designing individualized exercise programs with you personally in the clinic. It will also include bringing the latest exercise science findings to you through multiple venues.

To help our patients navigate this complex arena, and find personal answers, the expertise of a physical therapist can help.

Common and important questions that we can answer together are: What types of exercise are best for me? How many should I do and for how long? Is exercise prescription different or special for those with CF?

To help you find a meaningful and enjoyable exercise program, and one with a medically informed foundation, you will find me asking you questions like:

What types of exercise do you enjoy?

Do you like team exercise, such as soccer or softball?

Do you prefer competitive games such as pickle ball or do you prefer gym classes like Zumba?

Do you have any pain that prevents you from engaging in exercise?

I look forward to meeting you and supporting you in living an active life.

See you on Tuesdays,

Lisa Atkins, PT

A few snippets about me:

1. I love yoga, biking, hiking, skiing, swimming and walking my dogs.
2. I love all animals.
3. I enjoy knitting and reading Sherlock Holmes mysteries when I am not reading medical journals!
4. I need sunlight and coffee to be happy.
5. My favorite TV program has and will always be Star Trek, all of them.

Order of Respiratory Therapies

The Cystic Fibrosis Foundation has clarified the order of therapy so that mucus thinners can now be done during ACT's (Airway Clearance Techniques). We have changed our inpatient policy to match this practice. To provide standard of care throughout the pediatric and adult CF centers, during inpatient stays, Respiratory Therapy will remain in the patient room for the duration of therapy. This will allow for the respiratory therapist to adequately assess, recommend any modifications to therapy, assist with education and improve communication to health care providers.

Order of therapy

- Bronchodilator - Prior to ACT's
- Hypertonic Saline - Can be done during ACT's
- Pulmozyme - Can be done during ACT's
- Inhaled antibiotics - After ACT's
- Inhaled Steroids - After ACT's

Exerpt from the CF Foundation:

Airway Clearance Techniques (ACTs)

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There are different ways to clear your airways. Most are easy to do. Infants and toddlers will need help from a parent or caregiver. Older kids and adults can choose ACTs that they can do on their own.

All ACTs involve coughing or huffing. Many of them use percussion (clapping) or vibration to loosen mucus from airway walls.

ACTs are often used with other treatments, including inhaled bronchodilators, medications that help thin and move the mucus, and antibiotics. Bronchodilators should be inhaled before you start ACTs. This medication helps to widen your airways (bronchi) by relaxing the muscles lining your airway walls. Mucus thinners (sometimes referred to as mucolytics) help thin and then move the mucus out of the airways so it can be coughed out. These medications can be taken through a nebulizer during ACTs. Inhaled antibiotics should be taken after ACTs are finished and the lungs are as clear of mucus as possible. This will allow the medication to reach deeply into the smaller airways to attack bacteria.

Please feel free contact your CF provider if you have any questions.

Regards,

Respiratory Therapy

Update on the Vest

Currently AFCH has a policy in place on the use of High Frequency Chest Compression Vest Treatment. It states "Patients under the age of two should not receive this type of airway clearance therapy due to the development of head and spine." Patients under the age of two should continue to receive manual chest therapy. We are in the process of reassessing the literature and may revise this policy. We will inform families when our reassessment of the current policy in complete and if any revisions have been made. Stay tuned.

New Infection Control Changes in AFCH Clinic

As some of you may have already noticed, there are a few new changes when you come to clinic at AFCH. We are now wearing masks along with our gowns and gloves. This change in policy is to help keep you healthy. Another change is that we are no longer using the paper liner to cover exam tables. This allows us to better clean the exam tables in between patients instead of simply changing the paper liner between patients. This change is also to help keep you healthy. We are also working on a "CF Passport" for you to use whenever you visit laboratory, radiology or any other healthcare provider outside of the CF Center. This will inform the staff of our infection control recommendations to keep you healthy. CF Passport coming soon…

GREAT STRIDES 2017 DATES AND LOCATIONS:

Appleton 04/29/2017

Eau Claire 05/20/2017

Fond du Lac 05/06/2017

Green Bay 05/06/2017

La Crosse 05/13/2017

Madison 05/07/2017

Marshfield 05/07/2017

Milwaukee 05/21/2017

Stevens Point 05/21/17

Wisconsin Dells 05/06/17

New Infection Control Policy when Hospitalized in either the Children's (AFCH) or the Adult (UWHC) Hospital

When you are in the hospital you will be asked daily to bathe with Chlorhexidine Gluconate (CHG) soap. While this may seem like an inconvenience, it is actually very important! Here are some facts about CHG soap.

What is Chlorhexidine Gluconate (CHG) soap?

CHG soap (Hibiclens®) is used to reduce the numbers of germs on your skin. This soap is very good because it kills germs on your skin for a longer time than other soaps and it works when other body fluids such as blood are present. It is called a Daily CHG Treatment when you are in the hospital because it is a way to bathe and a treatment to decrease infection at the same time.

Why is using CHG soap (Hibiclens®) important?

The Centers for Disease Control and Prevention (CDC) estimates that 1 out of every 25 patients will get an infection while they are in the hospital. These infections are called hospital acquired infections (HAIs). In 2002, the number of HAIs in the U.S. was about 1.7 million. The bacteria on the skin changes from normal to hospital acquired within 48 hours therefore CHG treatments are typically only needed while in the hospital.

Studies show that your chance of getting an HAI is lower when the number of germs on your skin is lower. Reducing the chance of getting an HAI is important because HAIs lead to longer hospital stays and increased costs. To help lower the amount of germs on your skin your nursing staff will assist you to use CHG on a daily basis during your daily bathing time. Daily bathing can include a shower or a bath.

CHG soap can prevent the spread of germs while in the hospital and can treat already present infections.

What can I do about how drying CHG soap (Hibiclens®) is to my skin?

It is recommended to use lotion after using CHG soap. However, the type of lotion must work with CHG soap so please check with your nurse. Most lotion from home will stop the CHG soap from working well. Please ask for hospital supplied lotion such as Aloe Vesta®.

Do I have to use CHG soap (Hibiclens®) at home?

Most of the time, CHG is not needed at home. However, sometimes patients do need to bathe daily at home with CHG soap (Hibiclens®). Your doctor or nurse would let you know.

Update on Clinical Research Studies

Clinical trials are key research tools in advancing medical knowledge and patient care. We have definitely seen the benefits in patients with cystic fibrosis with FDA approval of Kalydeco and Orkambi in the recent years. Please consider participating in ongoing and future trials. Here are some of the current trials being conducted at your CF Center:

Pediatric Clinical Research Projects:

1. CFFC-OB-11

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. We are in year 5 of this 10 year observational study.

2. EPIC-002

Longitudinal Assessment of Risk Factors for and Impact of Pseudomonas aeruginosa and Early Anti-Pseudomonal Treatment in Children with CF (EPIC-002). This is an observational study to learn about what risk factors may lead to lung infections caused by Pseudomonas aeruginosa (Pa) in children with CF and what the impact of those infections may be. The original five year study was extended for an additional 10 years and we are in year 14 of the 15 year study.

3. Quantum003

This is a multi-center, observational comparative study to compare the standard-of-care method for diagnosing cystic fibrosis (CF) against a new method of sweat testing called the CF Quantum Sweat Test System. This study is open to enrollment at our center.

4. VX15-809-110

Vertex Lumacaftor (VX-809) and Ivacaftor in Children with CF aged 6 years and older who have two copies of the delF508 CFTR mutation. This is an open-label interventional study designed to look at the safety of lumacaftor in combination with ivacaftor (Orkambi®). Length of participation is 96 weeks and this study will continue at our center through June 2018.

5. VX15-770-124

A Study to Evaluate the Safety, Pharmacokinetics, and Pharmacodynamics of Ivacaftor in Subjects With Cystic Fibrosis Who Are Less Than 24 Months of Age and Have a CFTR Gating Mutation. This is an open-label interventional study and length of participation is 24 weeks. This study will soon be open to enrollment at our center.

6. SHIP001

Hypertonic Saline in Preschoolers (SHIP) ages 3 to 5 years. This is a randomized interventional study to look at the safety and effectiveness of hypertonic saline compared to isotonic saline (normal saline) in children with CF. Length of participation is 48 weeks. This study will soon be open to enrollment at our center.

7. MRI in the evaluation of liver and pancreas involvement in pediatric patients with cystic fibrosis

The primary objective of this pilot study is to show the feasibility of obtaining useful magnetic resonance imaging (MRI) images in children to identify cystic fibrosis (CF) liver disease (CFLD) in patients aged 5-17.

8. Feeding Infants Right from the Start (FIRST) Phase 1:

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.)

9. Feeding Infants Right from the Start (FIRST) 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.

10. FIRST Breast Milk Sub Study:

Inclusion criteria: Mothers who are currently breastfeeding, and have children 5 months of age or younger enrolled in the FIRST study.

Purpose of the study: The purpose of this sub study is to measure the nutrient content of human milk, such as the amount of fatty acids, calories, and protein, in order to study the relationship between breast milk nutrients and infant nutritional status. In addition, obtaining mother's dietary intake information in order to understand how mothers' diets affect nutrient content of human milk.

11. FIRST Gut Microbiome Sub Study:

Inclusion criteria: Infant/Children enrolled in the FIRST study Phase 1 or 2

Purpose of study: The purpose of this sub study is to characterize diversity and abundance of gut microbiota in infants/children with CF and study altered diversity and/or abundance of gut microbiota in relation to antibiotic exposure, growth, and lung health.

Adult Clinical Research Projects:

1. **Standardized Treatment of Pulmonary Exacerbations II (STOP2)**

Purpose: To evaluate the efficacy and safety of differing durations of IV treatment, given in the hospital or at home for a pulmonary exacerbation in adult patients with CF. Inclusion criteria: Age 18 or older; Enrolled in the Cystic Fibrosis Foundation National Patient Registry; at the time of Visit 1 there is a plan to initiate IV antibiotics for a pulmonary exacerbation; willing to adhere to a specific treatment duration determined by initial response to treatment and subsequent randomization.

Recruiting Soon:

Protocol VX15-152-102: A Study Evaluating the Safety of VX-152 Combination Therapy in Adults with Cystic Fibrosis

Purpose: To evaluate the safety and tolerability of VX-152 in Triple Combination (TC) with tezacaftor (TEZ; VX-661) and ivacaftor (IVA; VX-770) in subjects with cystic fibrosis (CF) who are heterozygous for the F508del mutation and a minimal function (MF) CFTR), or who are homozygous for the F508del mutation of the CF transmembrane conductance regulator (CFTR) gene (F508del/F508del). Inclusion Criteria: Age 18 or older; Subjects must have an eligible CFTR genotype: Cohorts 1A, 1B, 1C: Heterozygous for F508del and a minimal function mutation; Cohorts 2A, 2B: Homozygous for F508del; FEV1 40-90% at screening visit

Protocol GS-US-404-1808: A Study to Evaluate Effect of GS-5745 on FEV1 in Adults with Cystic Fibrosis

Purpose: This study will evaluate the effect of GS-5745 on pre-bronchodilator FEV1 in adults with cystic fibrosis (CF) after 8 weeks of treatment. Inclusion Criteria: Age 18 or older; Body weight of > 40 kg (88.2 lb) at study screening; Pre-bronchodilator FEV1 ≥ 40% and ≤ 80% of predicted at screening; Negative Sputum Investigation/History of any Mycobacterium spp. or Burkholderia spp; Clinically stable CF disease and regimen

If you are interested but don't qualify for any of these studies, you may qualify for studies at other facilities for which we would be happy to refer you for, a full list of ongoing clinical trials can be found at clinicaltrials.gov. In order to expedite your search, enter cystic fibrosis into the search field and click box to see only open studies. If questions, please feel free to contact one of our research coordinators:

Adult team: Sophia Chiron-Stevens at *scstevens@medicine.wisc.edu* or 608-263-1244

Pediatric team: Linda Makholm at *lmakholm@uwhealth.org* or 608-262-0340

Update on CFF Learning and Leadership Collaborative Grant/QI Group

In the fall of 2015, The Pediatric CF Center at American Family Children's Hospital (AFCH) was awarded a Learning and Leadership Quality Improvement (QI) Grant from the CF Foundation (CFF). We embarked on this journey to improve nutritional outcomes in the 0-2 year old population. The grant wrapped up at the end of October 2016, with a presentation of our work with all 15 centers that were chosen to participate, during the 30th Annual North American Cystic Fibrosis Conference in Orlando Florida.

As we approached the end of our time with the LLC grant, we as a team realized the importance of the QI work done and the continued work needing to be done to improve the care of people with CF. We continue to meet every other week with a comprehensive quality-improvement approach directed at increasing patient centered care and improving healthcare delivery.

In January 2017, we welcomed another parent representative to the group to lend their voice, ideas, and experiences. Along with our current family representative, we continue to forge a collaborative approach between the patient/parent representatives and the health care team (MD, advanced practice nursing, dietitian, nursing, and social work) to yield some great ideas for change!

We continue to work on several projects that have already been implemented, and have begun to implement new projects to improve the care of patients with CF. Projects that are on-going include:

- Improving timeliness of rooming patients with CF.
- Continuing improvement efforts to clean and sanitize clinic rooms.
- Pre-clinic huddle.
- Enforcing gowning, gloving, and masking.
- Continuing to follow closely our 0-2 year old population length to weight growth.

New projects on the horizon include:

- The creation of CF passports.
- Improving understanding of current policies/guidelines for infection control amongst the CFF, CDC, and UW Hospital policy.
- The development of a CF Parent Advisory Group at AFCH.

As these projects unfold we will continue to keep you updated on our progress.

Welcome to 2017, CFF Supporters!

We hope that the New Year is treating you well, and that you are rested, energized, and eager to begin your Great Strides planning. As you begin to consider your fundraising plan, recruiting members for your team, or taking the first step by registering for Great Strides, please remember why your participation is so important to our CF Fighters:

- Great Strides is a nation-wide event where all CF Fighters, their friends, and families join to honor them, to recognize their strength, and to celebrate their successes.
- Great Strides is the largest opportunity to acknowledge the accomplishments of the volunteer-driven Cystic Fibrosis Foundation, to share the CF story, and to educate the community about CF.
- Great Strides is the largest fundraising initiative supporting CF in the world. Donations generated through Great Strides funds CF research, drug development, care, and advocacy, and allows us to continue efforts to support and engage the community.

We are so grateful for the support you have shown through your participation in Great Strides, and in our other fundraising events and campaigns.

BECAUSE OF YOU

- We've funded 9 FDA approved therapies, including 2 that treat the basic genetic defect in CF.
- There are now 15 therapies in development in the CF Foundation Therapeutics Pipeline.
- $36 million has been issued for CF Care Center grants in 2016.
- CF Compass now fields 1,000 (average) calls per month providing guidance and support to CF Fighters and Families.
- $164 million was issued in medical and scientific awards in 2016.
- There are currently 46 CF clinical trials,
- Over 50% of our CF Fighters are now over the age of 18.

Imagine what else we can achieve, together! Please consider participating 2017 Wisconsin Great Strides. Walks will take place at 10 locations throughout Wisconsin, and the Madison walk will take place on Sunday, May 7. To register for Great Strides, visit: *www.cff.org/greatstrides*. Contact our office at (262) 798-2060 for questions about Great Strides, or any of our other annual events!

GREAT STRIDES

CYSTIC FIBROSIS FOUNDATION

Sign up for a walk in your area today.

http://greatstrides.cff.org

We Need Your Feedback!

Patient and Family Experience of Care Survey Reminder.

Our CF Center is participating in a national Experience of Care survey and we want to hear from you!

By participating, you will help us learn what is working well and where we can improve – ***your voice can make a difference!***

As A CF Center, we have the unique opportunity to learn about your experience of care with a special survey created just for people with CF and their families – 15 minutes of your time can make a difference for you and everyone at our center!

If you are contacted by Quality Data Management (QDM), an Ohio-based company in the 440 area code, please take some time to share your experience! You will not be asked to take more than two surveys per year.

Thank you for your help

Your CF Care Team

For general information, contact your CF care center. This Quality Improvement initiative is supported by the Cystic Fibrosis Foundation. Confidentially will be observed with all participants and responses.

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