

Back to School!

Summer has flown by and it is already that time of year that children are heading back to school! As you and your child get ready for the start of the school year, do not forget to ask the school if there are any forms that need to be completed by your child's medical provider. This includes a school medication administration form for enzymes, albuterol, or any other medications your child takes during the school day. If you are seeing your provider prior to school starting, please ask for a school medication form to be completed. If you will not be seen in clinic prior to school starting, please contact our office at 608-263-6420 option 2, and request that a form be completed and faxed to the school.

If your child is newly heading off to school, please ask us about our school resource folders. These folders provide useful information for schools and teachers, including information regarding nutrition, enzymes, general CF information, and the importance of access to sports drinks during gym and other physical activity. It is important to have a conversation with school staff at the beginning of the school year to discuss the above information available in the folder, so any questions can be addressed right away.

If your school has further questions, the Cystic Fibrosis Foundation has many great resources for schools and teachers to learn more about CF. You can find this information by going to: <https://www.cff.org/Life-With-CF/Daily-Life/CF-and-School/>

If any questions arise, please contact our office at 608-263-6420 option 2 to discuss further. *Have a great school year!*

Adult CF clinic Mental Health Programming Update

Rebecca Hays, MSW APSW

As you know, physical and mental health are intertwined. The UW adult CF program has begun to implement routine mental health care assessment and support services. This is in line with national CF care recommendations and noted by many of you as important to well-being.

It's been an honor and great joy to get to know many of you in my first few months as the mental health coordinator. My first goal has been for us to get to know one another.

Since there are a million definitions of 'mental health,' I've wanted to learn your vision of its meaning in your life: how you can tell if you're in a good (or bad) spot. You have shared what gives you joy, what keeps you going, how CF fits into your daily life, and what feeling down means for you. You've also shared experiences with mental health treatment.

Here are some of the GOALS and DEFINITIONS for 'mental health' that you've described:

- Being able to roll with ups and downs.
- Feeling well enough to go hunting with Dad.
- Truly connecting with a therapist.
- Forcing myself to do clearances even when I don't feel like it.
- Getting assessment and treatment for the circling thoughts I have at bedtime.
- Watching birds build a nest – enjoying that moment.
- Having a goal.

I've been reminded of ways that mood, physical symptoms, & self-care can interact. From there, we can use mental health screening tools to further our discussion, and work together to identify goals and intervention options.

I've appreciated your openness, and look forward to working together to support your wellbeing.

Thanks!

CFF Update

The Cystic Fibrosis Foundation – Wisconsin Chapter has been busy this Spring. We would like to thank everyone who took part in the 30th anniversary of Great Strides! We were able to raise \$850,000 at 10 walk locations throughout the state and we are so grateful for the donors, volunteers, walkers, sponsors, and committees who helped make it all happen. We are also proud to announce that the second annual Swinging for a Cure golf tournament, presented by Dave Jones Inc., was held at the beginning of June at Nakoma Golf Club, and raised over \$230,000 for the CF Foundation. We are so grateful to Dave Jones Inc. for their hard work and continued generosity!

As we move into Summer, we are focusing our energy on expanding our advocacy program. We are working to build a District Advocacy Network, under the leadership of our State Advocacy Chair, Carolyn Clow. Our goal is to strengthen our political impact.

We are looking for one district captain in each of Wisconsin's 8 Congressional districts, as well as other advocates who are willing to help reach out to legislators. The district captain will take the lead on developing a relationship with the member of Congress and with helping to organize advocates in the district. We are implementing this approach to develop more leaders throughout our state and to stay in touch with the members of our Congressional delegation. No special expertise is needed. We will support and train you to make **your stories and needs** heard by **your legislators**.

Our first goal is to improve our ability to influence policy makers by developing the relationship between them and our volunteers (YOU). Starting soon, we will be working to schedule meetings with our members of Congress in their local offices here in Wisconsin. (We just recently met with the office of Senator Ron Johnson.) We are looking to gauge the interest of advocates like you who would like to be involved in these meetings as well as in future meetings with State and Federal representatives.

If you are interested in further advocacy opportunities here in Wisconsin, please reach out to me at ksalentine@cff.org, and we will make sure to include you in our developing plans. The voices of members of the CF community influenced the debate on healthcare in Washington last summer and at our first ever state advocacy day in Madison in February. We look forward to continuing opportunities to making our voices heard.

Thank you for all that you do!

Kelly Salentine

Executive Director

Cystic Fibrosis Foundation – Wisconsin Chapter

Flu season is quickly approaching!

Listed below are the vaccines that are recommended for children and adults with cystic fibrosis:

- Annual influenza vaccine (flu shot). This vaccine is recommended for all patients seen in our Pulmonary/CF clinic each year starting at the age of 6 months. The vaccine can be obtained at your PCP office, 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 our Pulmonary/CF Specialty clinic starting at the age of 2. If this vaccine has been received already, you 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 they will need to have proof of first receiving 2 doses of Prevnar 13. This vaccine is listed as “pneumococcal” in the WIR system. It is then further described as Prevnar or Prevnar 13. 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 only received Prevnar, and not Prevnar 13, they will require 2 doses of Prevnar 13 prior to receiving the Pneumovax vaccine. Prevnar 13 can be obtained at your PCP office or at the Pulmonary/CF Specialty clinic. However, your child must receive their last dose of Prevnar 13 8 weeks prior to receiving the Pneumovax vaccine. Therefore, you may want to inquire in advance as to whether your child has received at least 2 doses of Prevnar 13 due to the 8 week waiting period before the Pneumovax vaccine can be administered. If your child is in need of 2 doses of Prevnar 13 prior to receiving the Pneumovax vaccine, this can be obtained at your PCP office.

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

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

Back to School Already - High Calorie Lunches and Snack Ideas

It seems like the last school bell just rang and now it's already time to start thinking about the start of a new school year once again. Whether you bring your lunch to daycare, are starting school for the first time or twelfth time, or eating your meals at home, try some of these yummy ideas to get the most out of your meals and snacks. It might also be worth investing in sets of reusable containers in various sizes and colors as these make meals more fun for kids. The tiny ones are great for packing dips!

- Spread a flour tortilla with guacamole, cream cheese, and/or butter and layer with cheese slices and lunch meat (salami, ham, and roast beef pack a good punch).
- Alternate little mozzarella balls, olives and cherry tomatoes on a toothpick and send a small container or olive oil or dressing for dipping.
- Try egg or tuna salad (made with extra mayo) with butter crackers.
- Make a pesto and mozzarella cheese sandwich or a pizza pita pocket filled with pizza sauce, shredded cheese, pepperoni, and black olives.
- Enjoy little packs of peanut butter or nut butter with pretzel rods.
- Have a cup of 4% fat cottage cheese with fruit or butter crackers.
- Bring little bags or cups of green and black olives - add croutons for a crunch and calories!
- Try summer sausage and cheese slices with bagel crisps or pita chips - they have more calories than regular crackers.
- Spread peanut butter, honey, and banana on a bagel.
- Bring a thermos of tomato soup made with heavy cream.
- Have salted boiled eggs.
- Try a BLT - pre-cooked bacon makes this easy. You could also eat it with avocado!
- Eat warm or cold ravioli or other pasta with pesto and Parmesan cheese.
- Make trail mix with nuts, seeds, pretzels, cereals, dried fruit, and chocolate pieces.
- Toss up a Caesar salad - crispy romaine lettuce, grilled or breaded chicken, Caesar dressing, croutons, shredded cheese and diced boiled egg.
- Serve up whole milk yogurt with granola and berries.

Easy Peanut Butter Balls - makes 12

1/2 cup peanut or nut butter

1/2 cup oats

1/2 cup dry whole milk powder

1/4 cup honey

(not for children less than 2 years of age)

1/4 cup chocolate chips or raisins/craisins

1. Combine all ingredients together in a medium bowl.
2. Roll into golf-ball size balls.
3. Store in an airtight container in the refrigerator.
4. 1 ball with chocolate chips = 150 calories; 1 ball with dried fruit = 130 calories. You can get almost 300 calories with 2 peanut butter balls versus 80 calories for a small granola bar!!

Pulmonary Function Lab

A note to our pediatric parents

Like the rest of the team, the staff in the PFT lab love it when families are involved in their child's care. We want to partner with parents to help kids become independent with PFT testing. Our goal is for most kids by the age of 8 to come to the lab unaccompanied by a parent. Prior to this, we find that kids respond best to instruction when the parents are a silent support. We ask you to help us instill confidence in your child's independence by encouraging independent testing. This may also allow you the opportunity to discuss any concerns you may have with other members of the care team while your child is testing in the lab. Thank you for your support!

Silver Lining Corner

(Authored by parents of children with CF)

CF families are eligible for a Family Access membership at the Madison Children's Museum. Family Access memberships are available for a minimum of \$10, but they ask that families give as much as they are able. These memberships are available in person, by phone, or by mail. All you need to do is provide proof of a CF diagnosis, like an electronic chart printout or doctor's note.

Changes to Clinic Check-In at American Family Children's Hospital

The next time you come to your appointment in CF Clinic, you may notice that you get weighed and measured three times. Why the change? After participating in a research study where all kids have to get weighed and measured multiple times, we were seeing some big differences in results. We've also noticed that a weight or height measure will get entered into the chart incorrectly.

We know how important these weight and height measures are - for determining results of PFTs, watching progress with weight gain and linear growth, and making suggestions for when to return for a clinic visit. It is our aim to give you the most accurate information possible about your health status and not send any incorrect messages.

We have asked our medical assistants to weigh and measure each patient 3 times each. We will then take the average of the three. Will this be a flawless system? Probably not - but we hope it improves things. If you notice that you are not being weighed and measured three times and/or if the numbers do not seem accurate to you, please bring it to our attention at the beginning of your visit. Thank you for your help with this important step in the check-in process.

Happy Retirement to Dr. Christopher Green

Dr. Chris Green has been on the faculty of the University of Wisconsin since 1983. He served as the CF Center Director from 1992-1995. In the past 8 years, Dr. Green has been the Chief Medical Officer of UW Hospital. After these many years of exemplary service, Dr. Green has retired from the University of Wisconsin. We wish him well in his retirement.



Patient and Family Experience of Care Survey

The CF Foundation has launched several initiatives to bring the perspectives of people with CF and families to improve care and health. One of the initiatives is the CF Patient and Family Experience of Care survey, which we implemented in 2016. The CF Foundation has partnered with Quality Data Management (QDM, Cleveland, Ohio) to collect the survey. We have a business associate agreement with the CF Foundation that allows QDM to contact adults with CF and families of children twice per year after clinic visits to complete the survey. This arrangement meets all of the requirements of HIPAA and protected health information and has been approved by our hospital attorney.

What does this all mean for you? We need your help in completing this survey amongst all other surveys you may be asked to complete (hospital and other clinics you may visit). Over the last 2 years, we received 150 completed surveys, so thank you for completing a survey. However, the survey is not a one-time ask, as the collection is continuous so we can look at our results over time to gauge whether we made progress in our improvements.

What is the data collection process? You will be asked to complete the survey twice per year (families with more than one child with CF will only receive 2 survey invitations a year). If we have your email address, then you will receive an email inviting you to complete a survey with a link to the survey (subject line is CF Experience of Care Survey - UW Madison). If we do not have your email address or you do not complete the survey from the email invitation, then you will receive a phone call from a QDM call center agent. What appears on your caller ID depends on your local phone company and their directory. The Cleveland area code is 440. The agent will give you a brief introduction and ask you to complete a survey. The agent will send you to an interactive voice recognition system or you can request to complete the survey with the agent. People have a preference on how they like to complete the survey in which some people prefer the one-to-one interaction with the agent, some people prefer the voice recognition system, and others prefer completing the survey on the Internet (the agent can give you the URL and your access code). The call center will attempt to contact you up to 3 times. We appreciate you taking the time to complete the survey and we regularly review the results and comments shared by participants to make improvements.

Is the survey truly anonymous? QDM has your contact information and your name is not attached to any of the results. Sharing with us what is working well for you and what is not is one of the sources of information used to help make improvements.

CF Clinical Trials update

Our CF Center is participating in the following clinical trials:

CHEC-SC: Characterizing CFTR Modulated Changes in Sweat Chloride and their Association with Clinical Outcomes.

This study is examining the change in sweat chloride values in patients who are receiving CFTR modulator drugs (Kalydeco, Orkambi, or Symdeko). The study involves a single clinic visit with pulmonary function testing and a sweat test. Patients must have been receiving a modulator drug for at least 3 months.

ARRIVAL: A Phase 3 Study of Ivacaftor in Subjects less than 24 months with a CFTR Gating mutation.

- **Inclusion criteria:** Diagnosis of CF with a CFTR gating mutation, Age less than 24 months
- **Purpose of the study:** To evaluate the safety and tolerability of ivacaftor in subjects with CF who are <24 months of age at the time of treatment initiation and have a CFTR gating mutation on at least 1 allele

Phase 3 study of VX-445 triple combination drug in people with CF 12 years and older who have one copy of the F508del mutation and one copy of a minimal function mutation (Vertex VX17-445-102)

- **Inclusion criteria:** Diagnosis with CF (age \geq 12 years), with one F508del mutation and a second “minimal function” mutation. A list of the eligible mutations can be found here: <https://www.cff.org/PDF-Archive/Study-VX-445-102-Eligible-Mutations-April-2018.pdf>
- **Purpose of the study:** To evaluate the effectiveness and safety of triple combination therapy of tezacaftor, ivacaftor and VX-445. Subjects will be randomly assigned to either placebo or the triple combination treatment and receive this for 24 weeks. After this 24 week study, subjects are eligible to roll over into an open label study in which everyone receives the triple combination therapy.

A Phase 3, 2-part, Open-label Study to Evaluate the Safety and Pharmacokinetics of Lumacaftor/Ivacaftor in Subjects 1 to Less Than 2 Years of Age With Cystic Fibrosis, Homozygous for F508del

- **Inclusion criteria:** Diagnosis of CF, age 1-2 years, with two copies of the F508del mutation.
- **Purpose of the study:** To evaluate the pharmacokinetics and safety of lumacaftor/ivacaftor in patients with CF who are 1-2 years of age.

CFF Patient Registry Questionnaire

This fall we will be handing out a brief questionnaire in clinic for patients in both the Adult and Pediatric centers.

This questionnaire will help us provide the most updated information to the Cystic Fibrosis Foundation (CFF) as part of the CF Registry data. Most of the information that is collected for the Registry is clinical data, such as PFT results or medication use. However, it is also important for the CFF to have demographic information about people with CF, as this information helps the Foundation learn how best to advocate for families with CF, and where the greatest needs are.

The questionnaire will have about 25 multiple-choice questions, so it should be easy to complete during your regular visit. The questions will cover topics such as number of people living in your household, smoke exposure, supplements you may receive free of charge, insurance etc. You can always skip any question you do not feel comfortable answering.

We hope you will help us, and the CFF, by completing these questionnaires.

Thank you!

Pediatric Depression and Anxiety Screening

As most of you know, the CF Foundation has recommended that all children over 12 years of age are screened for anxiety and depression. Our pediatric CF Center supports this recommendation as we know that anxiety and depression is not something that everyone recognizes. When people struggle with mental health issues, it can be difficult to do the things we need to do each day. This includes doing your daily CF cares.

As our screening continues, we are learning from you what resources and supports would be helpful. As our work continues, we continue to support you and your family to keep as healthy as possible as you fit CF into your daily life. The mind body connection is so important. I appreciate your openness to screening and follow up.

Tips for college

You may have questions about how CF will affect your college experience (and vice versa). It's quite normal to have worry and excitement at the same time. Your parents may be a little anxious, too. That said, finishing high school required flexibility, perseverance, and resourcefulness – so you've already got skills that will help you succeed in college too.

College students with CF have shared their experiences and tips. Here are a few blogs:

Accommodations for college: <https://www.cff.org/Life-With-CF/Transitions/Managing-My-CF-in-College/Accommodations-for-College/>

10 Things I Wish I Knew Before Coming to College: <https://www.cff.org/CF-Community-Blog/Posts/2015/10-Things-I-Wish-I-Knew-Before-Coming-to-College/>

Generally, it helps to identify possible needs and available resources ahead of time. CF social workers (Craig, Tracy, or Rebecca) can help you trouble-shoot, and can help you find “Disabled Students” departments/ counselors at your school. Even if you don't identify as ‘disabled,’ these office can help with accommodations, adjustment, and support.

LEARNING, LIVING SITUATION, AND SCHOOL ACCOMMODATIONS:

Each school has programming and counselors to help meet your learning and living needs. These offices can help honor recommendations from a high school IEP. They might also be able help you be assigned a solo dorm room for infection control, a fridge for meds, or an ‘incomplete’ in the case of a CF exacerbation. They can link you to counselors with experience helping people navigate emotional adjustments of college, or fitness trainers who can help build an exercise plan.

Here are a few examples of disabled student service programs:

Madison College: <https://madisoncollege.edu/disability-resource-services>

UW Madison: <https://mcburney.wisc.edu/>

UW Eau Claire: <https://www.uwec.edu/ssd/>

HEALTH:

You will continue to transition towards taking on more of the responsibility for your own health care & move towards getting care at an adult center, if you have not done so already. *We will continue to help you do this!* Here is another nice resource: <https://www.cfrise.com/>

If you are moving away for college, you may need to sort out where to get CF care. It may be helpful to establish care with a healthcare team in your college town – at least a primary care doctor at university health services. You may want to schedule routine UW CF clinic visits ahead of time to coincide with school breaks.

FINANCES:

Here is a list of financial aid and scholarships for people with CF: <https://www.cff.org/Life-With-CF/Transitions/Managing-My-CF-in-College/Scholarships-and-Financial-Aid/>

Congratulations on this next step!

Save the date for 2018 CF Family Education Day...

- **Date:** Saturday November 10, 2018
- **Time:** 8am-1pm, 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...

Update on the Pediatric Parent Advisory Group

Our Pediatric CF Parent Advisory Group has met three times. We met with the nurse manager for our P7 inpatient unit. We have made some suggestions for the inpatient care unit as well as outpatient imitatives that will be implanted. We are always welcome to new members. We especially hope to have some parents of kids who are in the 12-18-year-old range. If you are interested, please call and let us know. You can call Craig Becker from social work at 608-263-8572.

New to the Team

My name is Tracy Kussmaul and I am part of the Adult Cystic Fibrosis team serving in the role as Clinical Nurse Specialist (CNS) for the Adult Progressive Care In-Patient Unit (D6/5). Most of my work is done behind the scenes supporting the nurses at the bedside who are providing care to patients when they are admitted to the hospital and staying on the in-patient unit. Prior to joining the team I worked as an RN in both Radiology and ICU for 10 years. Outside of work I enjoy spending time outdoors with my family and two dogs.



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