The CFF LLC Lung Transplant project

It is with excitement that we announce a new quality improvement initiative here in Rochester at our Adult CF Center! The goal of this year-long project sponsored by the national CF Foundation is to improve the care of CF patients with advanced lung disease who are in need of referral for lung transplantation. We are paired with the UPMC lung transplant center in Pittsburgh, PA to work collaboratively on improving the referral process and post-transplant care coordination of lung transplant patients. The project kickoff was in Chicago, IL in October and work will continue until the NACFC in October, 2018. Teams are having weekly meetings and monthly teleconferencing with all 20 CF and lung transplant centers involved in the LLC (learning/leadership collaborative) across the country. In-person meetings will also take place in Washington, D.C. in March, 2018 and Toronto, Canada in June, 2018.

We are committed to this project and to our CF community as a whole! If you have any questions or curiosities regarding this project, please contact your Center, or any team member!

Rochester Team Members: Steven Scofield, MD; Donna Germuga, RT; Elle Woodworth, RD; Shandell Moreno, Lisa Brady, family member

Pittsburgh Team Members: Joseph Pilewski, MD; Matthew Morrell, MD; Shannon Whiteman, RN, Eric Wright, CF patient

Team Coach: Ian McIntosh, Director of Healthcare, Cystic Fibrosis Canada

Inside this issue:

Cystic Fibrosis Foundation 2018 Events

Rochester Great Strides: Sunday, May 20, 2018
Hornell Great Strides: Saturday, June 9, 2018
Rochester Cycle for Life: Saturday, August 11, 2018
Rochester's Finest Kickoff: July 2018
Rochester's Finest Celebration Dinner: Thursday, October 11, 2018

For those visiting the clinic between now and Rochester Great Strides on May 20th, please be sure to take home a special gift from the Cystic Fibrosis Foundation!

CFF Compass-patient assistance for multiple issues

About Compass

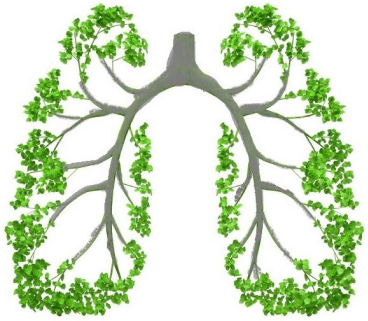
Cystic Fibrosis Foundation Compass is a personalized service to help you with the insurance, financial, legal, and other issues you are facing.

A dedicated, knowledgeable CF Foundation case manager is ready to work with you, one-on-one. This expert guidance is free and confidential. Call us at 844-COMPASS (844-266-7277) Monday through Friday, 9 a.m. until 7 p.m. ET, or email us at compass@cff.org.

Compass is available to anyone with CF, their family, and their care team, regardless of income or insurance status.

We strive to provide superior service to all individuals with CF, their families, and their care teams. However, we may have to decline assistance for certain topics outside the scope of our services.

Just Breathe Foundation:



Our mission is to make the lives of individuals and their families battling cystic fibrosis (CF) more manageable. We provide funds to applicants in Western New York struggling with the financial hardships the disease causes. We strive to improve the quality of life and to inspire others to show their support for the individuals and their families who struggle with cystic fibrosis on a daily basis. .. http://justbreathecf.og/

Cystic Fibrosis Family Connection:

The Cystic Fibrosis Family Connection (CFFC) is a charitable, not-for-profit corporation established to assist cystic fibrosis patients and their families.

Dedicated to supporting CF patients and families in their daily struggle with cystic fibrosis.

https://www.cffamilyconnection.org/

Nurses Nook

Hello CF Community!

Many of us have just returned from the North American Cystic Fibrosis Conference (NACFC) that took place in Indianapolis, IN from November 1 to November 4, 2017. There were many wonderful updates, but one of the most exciting was the continued commitment from the Cystic Fibrosis Foundation (CFF) to discover and make available CFTR modulators for all individuals with CF and to cover all CF causing mutations.

Below is a link to watch the actual presentation (Plenary 1) from NACFC titled "Matching Medications with Mutations" via YouTube. The presentation is inspiring, scientifically groundbreaking, and simply amazing. The link also includes the other two Plenary Sessions presented at the conference.

https://www.youtube.com/playlist?list=PLhoQ6vyZhgqrLqDphEWq-sFHpd1YO5Ocb

What follows is an attempt to summarize and offer highlights of Plenary 1. Some of the phrases and words are taken straight from the speakers as they said it best!

Dr. Preston Campbell, president and CEO of the CFF gave an introduction about the work that has been accomplished in the year 2017 related to CF and what is expected in the coming years. He

shared amazing news about life expectancy for individuals with CF. In 2016, the median predicted survival was age 41 years and in just one year, that increased to 47 years. Registry data has shown that those individuals with the G551D mutation who started ivacaftor (KALYDECO) early in life have a median predicted survival of 70-80 years. He shared the commitment of the CFF to keep going "UNTIL IT'S DONE" and that all individuals with CF will have CFTR modulators available, that novel antibiotics will be available to treat multi-drug resistant bacteria, and that the mission will continue forward to cure CF.

Dr. Campbell also shared some recent successes from Vertex, the pharmaceutical company that has been crucial in the discovery & implementation of the current CFTR modulators approved by the FDA: Ivacaftor (KALYDECO) for those with one of 38 approved mutations ages 2 years+ and lumacaftor/ivacaftor (ORKAMBI) for those with two copies of F508del age 6years+. Vertex is also working on Phase 3 trials of another medication for those with two copies of F508del and results are promising- a decision from the FDA is expected in early 2018. Also underway are studies looking at triple combination therapies to treat not only those with two copies of F508del, but also those with one copy of the gene.

Next to speak was Dr. Michael Boyle, Senior Vice President of Therapeutics Development at CFF and Professor of Medicine at Johns Hopkins. He reported that at the end of 2016, there were 54% of individuals who were eligible for therapy with ORKAMBI or KALYDECO and about 46% of patients had mutations that were currently without modulator therapy. Of those without modulator therapy, there are about 1,700 rare mutations and over 1,000 of those are carried by five or less people in the world with CF making clinical trials not possible. He identified five modulators needs that were identified heading into 2017 and what progress had been made: 1) better therapy for those already on modulators (progress being made with above noted Vertex study with decision from FDA by early 2018); 2) residual function mutations treatment (progress made with a total of 28 more mutations since May 2017 now eligible for KALYDECO); 3) ability to assess for rare mutations (progress made with theratyping discussed under next speaker); 4) Single F508del mutations treatment (progress made with studying 3-drug combination therapy studies by several pharmaceutical companies) and 5) Class 1 mutations treatment (these are mutations in which no functional CFTR is created). Dr. Boyle identified a goal of the CFF that by 2020, 93% of individuals with CF with have a highly effective modulator therapy.

Dr. Phil Thomas PhD, UT Southwestern was the final speaker and he is a scientist who has been involved since early on in the discovery of these CFTR modulators. He shared his story about what it is like to be a scientist working in CF and his personal experience/journey. He used a basic analogy (a chain link fence with a gate and cattle!) to portray what CF is and how these modulators work. He explained that when CFTR functions properly, it (CFTR) is a gate in a fence that keeps the cattle (chloride ions) from moving from one pasture to the other. When you open the gate (the CFTR), the cattle (chloride ions) can move through the gate. One must know how fast the cattle (chloride ions) can move and that depends on how many gates (CFTR) are in the fence, how wide the gates are, and how many of them open. In CF, there is a problem and there aren't enough working gates (CFTR) and the cattle (chloride ions) can't move and this leads to a dehydrated airway surface.

Dr. Thomas reviewed that CF causing mutations disrupt the proper process. For example, the G551D mutation makes the gates (CFTR) not open well and the medication KALYDECO (ivacaftor) helps open the gate. In F508del, there are not gates (CFTR) in the fence and you need a corrector to help you get the gate into the fence: lumacaftor helps the gate get into the fence and ivacaftor helps open it (these two drugs combined make ORKAMBI). For a Class IV mutation like R347P, there is a gate (CFTR) in the fence, but it is not wide enough to let the cattle (chloride ions) through.

Continued on page 4

Continued from page 3

Currently there is no modulator to help fix this kind of mutation.

Dr. Thomas and other scientists are now using theratyping which is the notion that one can look for a group that responds to the same treatment. As an example, it is possible that many of the rare mutations will respond to current modulator therapies. Cell work can be used to evaluate modulator response much quicker in the lab as opposed to clinical trials in humans. This is beneficial because of the challenge of rare mutations noted above (there are over 1,000 mutations that five or less people in the world carry making traditional clinical trials not possible). The CFF is supporting an effort to look at 649 of these rare mutations and a similar approach can be used to address therapies for the individuals with nonsense mutations (those that don't make the CFTR protein). Dr. Thomas did note that as humans, we aren't made just by CF mutations and that we have lots of changes in our DNA and that can complicate how we as humans respond to drugs. Personalized medicine is the future in which scientists will use a person's own cells to assess that individual's response to CFTR and amazingly, this is underway. Dr. Thomas ends with the message that the future is bright!

Bridget Platania NP (Peds Pulmonary)

News from West 7

Hi everyone! I'm Emily Stender and I'm the new Unit Educator on West 7. I've worked here as a Registered Nurse for about 5 years and I've learned a lot in that time about our unique CF population. Growing up, I was a sick kid and frequently in and out of the hospital. Before they use to isolate CF rooms, I was actually placed in a semi-private with a CF'er (who we still know and love, and treat today!). I thought I had learned a lot about the complexities of CF at that time. Even in nursing school, where we barely touched on the topic, I thought I was an old pro. I was very wrong. I'm so happy that working here has allowed me to learn and be able appreciate this unique world. The goal now is education for everyone else. I look forward to many years to come in this new role and hope to be a liaison for this wonderful community!

Things to look forward to:

Starting this spring there will be a new initiative set in placed by the hospital staff to conduct bedside rounding to help facilitate a more cohesive plan for our CF patients. Rounding will include the CCC providers, Social Work, Nurses, and the Attending Physician. More details to come on this matter. West 7 also met with the team from the University of Pittsburgh Medical Center (UPMC) to create a plan for when we have transfers to their facility regarding lung transplants. Finally, to keep Highland Hospital up-to-date with the current evidence based practice, we are changing the way we monitor aminoglycoside medications (tobramycin, amikacin and gentamicin). There will be 2 time-specific blood draws, rather than 1 we currently are doing. This also rolls out in January. Exciting times ahead!

Calling the CCC: The Triage Process

As many of you know by now, when you call the CCC there is a triage process that ensues. To some, this can be frustrating and it is our goal to help you understand why this process can be crucial in the care and follow-up you receive. Many call the Complex Care Center requesting to speak directly to their provider, and although the providers would like to speak directly to their patients, this would take them away from seeing patients in the office. So, this is where the CCC nurses come in!

The front desk has their own triage process, and upon initial contact with a patient they determine severity of needs and go from there. They will either call the nurses to speak with a patient or family member directly, or send a message to the nurses to follow up and get more information on the concern. From there, the nurses will gather additional information and make a plan of care to ensure the necessary next steps are taken. The next steps can include booking an appointment at the CCC, sending medication and follow-up instructions, or calling patients in to the hospital for higher level of care. The whole point of the triage process is so that the providers know the severity of the needs and can address concerns appropriately based on that.

We understand that when patients call the office it's usually because they aren't feeling well and our triage process can seem bothersome to explain your symptoms to multiple people. There truly is a method to the madness, and each step is to ensure our patients receive the best and most suitable care. We truly understand the complex needs of our patients, and work diligently to provide immediate care and follow up for all our patients. We encourage our patients to call with any questions or concerns regarding their health care.

Andrea Roy, RN

Meet new members of the team!

My name is Karlee Bushnell. I am born and raised in Rochester. I graduated from the Rochester Institute of Technology in 2016 with my B.S. in Nutrition Management. I then completed my dietetic internship in Boston through Simmons College before moving back to Rochester. I worked at Highland Hospital as a diet office assistant while I was an undergrad at RIT, and recently began my professional career as an Inpatient Registered Dietitian covering E5, E7, and W7. I am very happy to be back!

Karlee Bushnell, RD

Working with the cystic fibrosis population is new to me, but so far very interesting. I try to treat every patient that I meet with respect, and a careful kind of compassion to understand their needs while hospitalized. It is important for me that the patients I see realize that they are more than just a name on my list for the day, and I try to make every interaction comfortable and personalized. I am working on broadening my understanding of the cystic fibrosis population, and my co-workers have been great so far about sharing with me what they have already learned.

Heather joined the Cystic Fibrosis Foundation (CFF) with 17 years of fundraising experience. She has worked at all levels of fundraising with different types of non-profit organizations including higher education and healthcare. Her experience also extends to government leadership. She is from WNY and resides in Gowanda, NY with her husband Ryan. They have a son Lucas who is currently serving in the U.S. Navy. Heather began her new role with the CFF at the end of October and is looking forward to meeting and working with all of the amazing families involved in helping achieve the CFF's mission to find a cure for cystic fibrosis!

Heather McKeever - Executive Director, Western New York Chapter, CFF

There is now a pharmacist at the Complex Care Center! I am a new member to the team and joined this past September. I got my PharmD from University at Buffalo, completed a residency training and worked in the hospital setting before joining the center. I am available to answer any questions you have about your medications and can help ensure you have access to your medications. During your visits I will be monitoring your response to therapies, checking necessary labs and checking for drug interactions. I will also provide you with education on preventing and managing any side effects from your medications and provide counseling on any new medications we start. It is an honor to work at this center and I look forward to being at your service!

Rezarta Lako, PharmD, BCPS

Tube Feeding- Could It Improve Your CF Care?

February 4-10 marks Tube Feeding Awareness week. Tube feedings are important sources of nutrition for many of our CF patients and can provide calories without the fight or discomfort from eating large amounts of food. Even though we know the benefits of improved nutrition, tube feedings are often approached with fear, frustration, and anger from patients and families- and rightfully so! It is frustrating that eating regular food by mouth isn't enough and being different can be scary. However, anyone can learn to easily care for a tube feeding and it has the potential to greatly improve your health.

What is a feeding tube?

A feeding tube is a device placed through the abdomen into the stomach, through the stomach into the intestine, or through the nose into the stomach. Feeding tubes provide an additional source of nutrition for people that are unable to get enough calories by eating alone. Tube feedings can be given in small amounts overnight while you sleep, in 'bolus' form- one larger volume at a time, or continuously throughout the day. There are a number of different types of tubes and ways to obtain nutrition via the feeding tube. Your gastroenterologist (digestion doctor) and registered dietitian at the CF Center will be able to help you determine which tube and feeding method will be best for you or your child.

Why is a feeding tube helpful?

We know that good nutrition results in better lung function. Better lung function means better quality of life and fewer CF complications. A feeding tube can be a way to increase your energy, possibly reduce CF flare ups, allow you to exercise and increase your endurance and strength, achieve and maintain a healthy weight, and decrease the stress around meals and snacks.

What are some potential concerns?

-Insertion of the tube is a surgical procedure and some people experience pain or discomfort at the tube site for a short time after it is placed.

-Getting child care for someone with a feeding tube may be more difficult if they require feeding throughout the day. That being said, individuals and families manage feeds at home every day. Some even change their tube on their own. Daily feeding and care for the tube site is easy to learn and does not need to be done by a healthcare professional.

-It may cause emotional stress to have, or see your child have, a tube.

-People may struggle to believe you or your child need a feeding tube when appearance is normal and not 'sickly'.

TF Myths

Myth: A parent didn't try hard enough to make their child eat or the patient didn't work hard enough to eat more.

Fact: In our experience, parents try everything to get their children to eat and patients are exhausted by their efforts to eat more to gain weight. It is a natural instinct for a parent to feed his or her child or a person to eat. Cystic Fibrosis increases energy needs and decreases your body's ability to absorb nutrition so some people require extra nutrition to stay healthy. Tube feeding is often a last resort. Tube feeding allows a person to get the nutrition boost needed to grow, develop, and thrive when eating and drinking by mouth isn't enough.

Myth: Tube feeding is forever.

Fact: Some people will need a feeding tube their whole life to stay healthy. But for some, tube feeding is temporary. Children may be able slowly decrease and stop their feeding tube once they are no longer medically necessary. It is often difficult to estimate how long a feeding tube will be needed.

Myth: Physical activity will be limited by having a feeding tube.

Fact: Tube feeding itself does not limit a one's ability to roll around, play, climb, run, swim or play sports. Physical ability will not be impacted by having a feeding tube. Even children who feed continuously can wear feeding pumps in backpacks and still be active.

Myth: You must use formula for tube feeding.

Fact: You can tube feed breast milk, formula, or blended foods. The diet can be different based on medical condition and how you feel.

Resources

Your CF Center is the best resource to determine if a feeding tube is right for you. See below for additional resources that may help you make an informed decision.

Own Your Feeding Tube Podcast Series on Youtube- https://www.youtube.com/watch?v=GVoDm085E4Q

Oley Foundation- http://oley.org/

A Parent's War Against the Feeding Tube- https://www.cff.org/CF-Community-Blog/Posts/2015/My-Small-War-Against-the-Feeding-Tube/

Friend and Family Guide to Tube Feeding- http://www.feedingtubeawareness.org/wp-content/uploads/2015/06/Family_and_Friend_Guide_to_Tube_Feeding.pdf

High Salt Food

- Tomato juice
- Frozen veggies packaged with sauce
- Seasonings: garlic salt, onion salt, celery salt, lawrys season salt, taco seasoning mix, Morton sea salt rubs, season all
- Meat: Jerky, lunch meat, smoked fish/meat, pork, sausage, bacon
- Canned foods: vegetables, beans, meat, soup, tomato sauce, spaghettios, olives, pickles,
- Condiments: ketchup, soy sauce, salad dressing, hot sauce, marinades, alfredo, BBQ, broth, gravy, salted butter
- Grains: buns, tortillas, pita, dry cereal, bagel, ready made pancake mix, croutons, crunchy noodles, onion straws
- Dairy: imported blue cheese, feta, cottage cheese, processed cheeses (string cheese, American cheese), boxed mac and cheese
- Meals: frozen dinner, hamburger helper, frozen veggie burger
- Snacks: chips, salted nuts, salted nut butter, pretzels, saltines/ritz, ramen

Recipe 1: Top hummus with olive oil and feta cheese, then dip into with salty pita chips

Recipe 2: Guacamole or black bean dip with salty tortilla chips

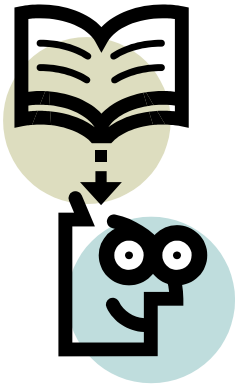
Recipe 3: Top salad with: salted nuts/seeds, cheese, bacon, croutons, crunchy noodles or sesame sticks, and a high fat/salt dressing

Recipe 4: avocado toast with 2 slices of thick, whole wheat bread and spread an entire avocado between the two and top with plenty of salt and pepper

Recipe 5: make your own trail mix with crushed up salted pretzels, nuts, and chocolate/dried fruit

Flavored salt recipe ideas: http://www.thekitchn.com/how-to-make-flavored-salts-cooking-lessons-from-the-kitchn-174397; http://www.sheknows.com/food-and-recipes/articles/1107353/flavored-salt-recipes

Buying flavored salts: https://www.seasalt.com/gourmet-salt/flavored-salt.html



Education Night

Please mark your calendars for CFFC's 2018 Education Night.
We have selected the evening of Thursday, July 19th.
The program will be an equal mix of socializing and learning.
We will be providing food and beverages to enhance these experiences.
Stay tuned as the details are finalized.

Community Voice Initiative

Community Voice provides opportunities for people with cystic fibrosis and their family members to have an active say in the Cystic Fibrosis Foundation's initiatives, ensuring the voice of the CF community is being heard to drive improvements that will benefit everyone impacted by CF. People with CF, parents, spouses and siblings are invited to join and take an active role in shaping resources for the CF community. To learn more, visit: CFF.org/CommunityVoice

The North American Cystic Fibrosis Conference Experience...

This was my first time attending the NACFC (North American Cystic Fibrosis Conference) in November. I was a newbie and not sure what to expect. The registration process was very daunting. The housing email came out in June and all the experienced NACFC attendees were making sure we all got on to get a room. What?? I have been to other national conferences before and have booked my room a month prior and usually had not issues getting a room. Boy was I glad I listened!!! Also with registering I was encouraged to register as soon as possible and when the Brown Bag sessions came out make sure I get in on them ASAP. This was great advice as I was able to get in all the programs I wanted to. Heading into the first plenary session I was overwhelmed. Every seat was full, people were standing in the back and along both sides. Apparently, there was also an overflow room. Amazing!!!!! This is a true testament to the CF Foundation, the Patients and Families and to the Caregivers. I will not lie. Going to the sessions each day and listening to all the highly experienced Providers I became overwhelmed. The amount of research currently going on and the increased survival rate is a true testament to the dedication of all involved.

I was asked to write up a little blurb about the conference and what I learned. If I included everything I learned it would be a small book. So I would like to touch on a few topics that seemed to repeat themselves throughout the conference. There was also a lot of thoughtful discussions on these topics that have made me think of what our treatment protocols are. These topics aren't currently included in care guidelines from the CFF and therefore each center may treat just a little differently.

Pseudomonas Aeruginosa is a bacteria that will affect many of our patients. Pseudomonas aeruginosa is a common environmental organism. Most initial infections are from an environmental source. There is a steep increase in infection rates as people age. In the younger population Staph Aureus is the main organism. Around age 18-24 years this changes to Pseudomonas Aeruginosa. Initial treatment of a Pseudomonas Aeruginosa infection is fairly standardized with Inhaled Tobramycin (TOBI) twice daily for 1 month. We will check cultures 2-3 weeks after completion of the first course of TOBI to see if it is resolved.

For chronic Pseudomonas aeruginosa infections there is some difference in treatment. If a child's culture comes back positive after 1 month of treatment they will go on for 6 months of inhaled TOBI on an alternating schedule of 1 month on 1 month off. Our Center considers a child chronically colonized if they are still showing Pseudomonas aeruginosa in their culture after being treated with inhaled TOBI for 6 months. At this point they will be on every other month TOBI for an indeterminate amount of time. Many centers have different definitions of chronic infection; anywhere from unable to treat first infection to after 1 year of treatment. Treatment can also vary. Some centers will use oral Zithromax with or without TOBI. There are some studies going on right now within the CF Foundation that are addressing this to decide on best practice.

MRSA (methicillin-resistant staph aureus) is another organism that was discussed. MRSA is a form of staph aureus that is not killed by typical antibiotics that can treat staph. It is very common and mostly not harmful for healthy individuals. People with CF can have MRSA and not have any symptoms. The treatment, center to center, can be different and there is no right or wrong. At our center we do not treat a positive MRSA culture unless there are symptoms. Some centers will not treat with the first positive culture but will treat if there are subsequent cultures. And some centers have a very intensive eradication policy for any positive culture. This is an inpatient stay with numerous antibiotics both IV and orally. We are always aware of the amount of antibiotics we prescribe and need to balance the positive benefit from the antibiotic versus the risk of becoming resistant to certain antibiotics which makes future infections harder to treat. So again, there are no right or wrong answers and more research needs to be done to find the right balance.

I am so humbled to be able to work with such an amazing group of patients, families and care providers. The team approach in CF management is certainly obvious and is a model for other chronic illnesses on how working together can get results. I am looking forward to continuing to learn and grow within my CF practice.

Laurie Johnson, NP

Using Self-compassion as a Tool in the Treatment of Your

Living with CF presents many challenges that take a toll on both physical and psychological well-being. CF is different for everyone and all cases of CF progress over time. Individuals with CF often report feeling frustrated, angry, or discouraged by disease exacerbations or the introduction of new procedures or treatments (e.g., placement of a g-tube). They may feel like they aren't good enough or can't meet their own or others' expectations about managing their illness. This, in turn, can exacerbate physical symptoms. It can be helpful to remember that disease progression is normal and does not mean that you are doing anything wrong with regard to CF care. If you can approach such changes with a sense of acceptance and self-compassion, you will find this can help your overall outlook and health. In fact, research shows that self-compassion is associated with decreased psychological distress, improved well-being, and better disease self-management. The following suggestions may be useful in promoting increased self-compassion:

Notice your inner critic. Some people find that giving their critic a name helps them to notice it and gain some space from the criticism.

When you find yourself being self-critical, think of how you would talk to a beloved friend or family member and use similar words or phrases to talk to yourself ("It's okay, darling. Sometimes things don't go the way you expected. What do you need to feel supported?").

Remember that you can always begin again. If you forgot to do something, remind yourself that everyone forgets things at times and allow yourself grace and forgiveness as you continue to do the best you can to take care of yourself.

Don't be afraid to ask for help. Human beings are social creatures who survive by being connected to and helping others. It isn't possible to do everything on your own so don't expect yourself to handle your CF care alone either.

Use touch to soothe your emotional distress. Put your hand on your heart or give yourself a gentle hug or caress as a soothing gesture of love and support for yourself and the distress you are experiencing.

If you're interested in learning more about self-compassion, there are many online resources and meditations that can guide you (http://self-compassion.org/ is a good place to start). If you find you need additional support, the behavioral health team at the CCC has expertise working with individuals with CF around these concerns. You can reach the team by calling the clinic at (585) 276-7900.

"Our successes and failures come and go—they neither define us nor do they determine our worthiness. They are merely a part of the process of being alive"
— Kristin Neff, Self-Compassion: Stop Beating Yourself Up and Leave Insecurity Behind

DIGEST (Developing Innovative Gastrointestinal Training)

Now is an exciting time for medical advances in diagnosing and treating gastrointestinal aspects of cystic fibrosis! The CF Foundation recognizes that CF is a complex disease, and that many of the GI symptoms greatly impact patient's lives. In order to better promote research and clinical knowledge regarding GI manifestations of CF, the Foundation has established a training grant, DIGEST (Developing Innnovative Gastrointestinal Training). I am proud to have received this grant, and I have learned so much through the Foundation's mentorship and from all of my wonderful patients. As you may know, CF can affect the GI tract in many ways, including reflux, abdominal pain, liver disease, and difficulty with growth- just to name a few. It is important for patients to know that oftentimes there are treatments that can help improve health and quality of life, so please bring up GI related concerns to your CF Center providers.

At the NACFC this year there were many sessions that focused on GI issues. International experts informed us on how beneficial some of the CFTR-modifying drugs, Orkambi and Kalydeco, can be for GI issues such as bacterial imbalance and growth. There was also a lot of excitement regarding a new product, Relizorb, an enzyme cartridge that can be used with tube feeding. Relizorb has been shown to allow better absorption of calories as well as the anti inflammatory Omega-3 fatty acids. It can also be helpful in minimizing GI symptoms associated with tube feeding. Complex issues such as abdominal pain were discussed at the conference, along with ideas regarding diagnosis and treatment. Overall there was a lot of helpful clinical information shared, as well as discussion of ongoing research that promises to make the future brighter for patient's with CF.

New CF Resource

We are pleased to announce a new resource that is available to you. "CF and Mental Health: Building Resilience – A Guide for Parents and Caregivers", is now live on cff.org! It is on three of our web pages in the downloads section:

Coping While Caring for Someone with CF

Anxiety& CF

Sexual and Reproductive Health Guides for Young Women with CF

The Boston CF Center and Center for Young Women's Health at Boston Children's Hospital would like to share their newly developed sexual and reproductive health guides for young women with CF.

This 12-guide series was developed in partnership with patients, parents, members of the multidisciplinary CF team, and adolescent medicine specialists. The guides are freely accessible online and can also be printed out for patients and families. They are written specifically for adolescent and young adult women with CF.

https://youngwomenshealth.org/cystic-fibrosis-all-guides/

Here is a list of the guide titles and topics covered:

Cystic Fibrosis (CF)-general

Cystic Fibrosis: Puberty and Menstrual Periods

Cystic Fibrosis: What Should I Know About Sex?

Cystic Fibrosis: Vaginal Yeast Infections

Cystic Fibrosis: Talking with Your CF Team about Sexual and Reproductive Health

Cystic Fibrosis: Talking with Your Partner about CF and Sexual and Reproductive Health

Cystic Fibrosis: Contraception

Cystic Fibrosis: Urinary Incontinence

Cystic Fibrosis: Sexually Transmitted Infections (STIs)

Cystic Fibrosis: Pregnancy and Fertility Basics

Cystic Fibrosis: Deciding Whether to Become a Parent

Cystic Fibrosis: Pregnancy and CF

Feel free to contact Traci Kazmerski, MD, MS, the principal investigator of this project Traci.Kazmerski@childrens.harvard.edu or the Center for Young Women's Health at Boston Children's Hospital with any questions.

Annual Kit Taylor Memorial Lecture

The Rochester CF Clinic has announced the 38th Annual Kit Taylor Memorial Lecture. The speaker will be Steven Freedman, MD, PhD, Chief of Division of Translational Research at Harvard Medical School and Director of the Pancreas Center at Beth Israel Deaconess Medical Center.

Save the Date

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ROCHESTER CYSTIC FIBROSIS CENTER
PEDIATRIC PROVIDERS AT STRONG MEMORIAL HOSPITAL:

Karen Voter, MD

Clinic Scheduling ..... (585) 275-2464
Clinic and Prescription Refill ..... (585) 275-8706
Bridget Platania, Nurse Coordinator ..... (585) 275-2464
Pat Lamarche, Nurse Practitioner ..... (585) 275-2464
Holly Torkington-Wood, RN ..... (585) 275-2464
Judy Sroka, Research Coordinator ..... (585) 275-2464
Laurie Johnson, NP ..... (585) 275-2464

Kim Bordeaux, Anna Roberts, Respiratory Therapist

Marie Bieber, Nutritionist/Dietitian ..... (585) 275-2464
Marcy Odell, Social Work ..... (585) 275-9105
Karen Voter, Newborn Screening Coordinator ..... (585) 275-2464

For Urgent Issues:
Call the office at (585) 275-2464. If after-hours, the answering system will provide instructions for your needs.

ADULT PROVIDERS AT COMPLEX CARE CENTER:

Tiffany Pulcino, MD; Steven Scofield, MD; Heather Busick, MD

Clinic Scheduling ..... (585) 276-7900
Adult Cf Center ..... FAX (585) 288-1381
Donna Germuga, Clinic Coordinator/ RT ..... (585) 276-7900
Elle Woodworth, Dietitian ..... (585) 276-7900
Kristen Davidson, Psychologist ..... (585) 276-7900
Kristine Reinhardt, Nurse Practitioner ..... (585) 276-7900
Jennifer Naugle, Nurse Manager ..... (585) 276-7900
Sarah Creek, RN ..... (585) 276-7900
Andrea Roy, RN ..... (585) 276-7900
Judy Sroka, Research Coordinator ..... (585) 275-2464
Betsy Brewer, Office Manager ..... (585) 276-7900
Tasha Denise Maldonado, Front Desk ..... (585) 276-7900
Julie Buerger, PT ..... (585) 276-7900
Nancy Dukelow, OT ..... (585) 276-7900
Adela Planerova, Director of Dental Services ..... (585) 276-7900

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Refill Requests:
Please submit your request to your pharmacy and allow 2 weeks advance notice. For urgent requests, call the coordinator line or send a message via MyChart.