

SUMMER/FALL 2018

Hello!

I would like to welcome our patients and their families to the first edition of our newly-reborn Cystic Fibrosis Center Newsletter. I wanted to update you on some recent changes; our CF Center Care Team has grown!

Golisano Children's Hospital at Upstate Medical University has contributed necessary funds to expand our group of professionals who provide care to CF patients and families. We are very proud of our "extended family" here at the Center, and would like to introduce you to some of our team members.

Dr. Aisha Baig, MD joined our CF Care team after finishing her Pediatric Pulmonology fellowship training which included CF in Omaha, Nebraska.

The Cystic Fibrosis Foundation has also generously supported the addition of three critical members to the CF team: **Jessica Ehnnot, PharmD, Matt Bowman, PT, DPT** and **Lejla Bush, LMSW**. They are our CF Care Team's Pharmacist, Physical Therapist, and Mental Health Coordinator. **Dr. Chris Fortner**, our CF Center Director, was awarded three grants from the CFF to provide this invaluable support to CF patients.

Jessica Ehnnot, PharmD ensures that all medications and prescriptions are appropriately filled and assists providers in determining therapeutic dosages when IV antibiotics are prescribed.

Matt Bowman, PT, DPT assesses and monitors patients' heart and lung functions while they are undergoing the stress of exercise and recommends ways to maintain or increase each patient's physical abilities to promote health.

Lejla Bush, LMSW assesses and supports each CF family's emotional needs, and recommends local resources for those in need of mental health care in their home area. She is also the Adult CF Social Worker.

In our second edition of the Center's yet to be named CF newsletter, I will highlight our more seasoned staff.

Although our Cystic Fibrosis Care Center Team is expanding, we realize that our contribution to your CF patient's health is tiny, compared with the assistance each patient and their family provides. With our growing CF Team, we strive to provide excellent care and support you with the many facets of CF care. Please do not hesitate to contact any of the team members for any questions or concerns you may have.

Thank you! We look forward to seeing you in our Center.

Sincerely,
Zafer Soultan, MD
Pediatric Pulmonary Division Chief



We Want to Hear From You

As part of our commitment to you, we would like this newsletter to reflect what you want to hear about. We are accepting submissions for the title of your newsletter. We hope that you will also suggest subjects that you would like to know more about. If you are interested in being featured in "**Your Voice Your View**", talk to your care team or submit an entry. Please limit your submission to less than 350 words.



Virtual Events

Designed by and for adults with cystic fibrosis and their families, Cystic Fibrosis Foundation virtual events provide the opportunity to connect, share, and learn from peers through mini-cons, virtual events featuring panels, fun activities, group chats, small-group video breakouts and open and honest dialogue.

• September 28-29:

BreatheCon two-day event for adults with CF

• November 7, 2018:

CF Adults Connect

For more info and to register:
www.cff.org/virtualevents

BREATHECON
CYSTIC FIBROSIS FOUNDATION

Resources & Research

Approximately nine months from now, your CF Center will deliver to you ... **CF Teaching Day 2019!** Mark your calendars for **Friday, May 31**. Begin nesting and preparing by seeking out necessary child care and vacation days off at work. The day will be packed with education, vendors with the latest resources, research and food with the clinic staff, followed by dinner and family networking.

David's Refuge: a respite for parents and caregivers



- An all-expense paid weekend away at a Bed & Breakfast or hotel with opportunities to be pampered
- Potential childcare funding through The Jim and Juli Boeheim Foundation
- To learn more, go to davidsrefuge.org/ or call 315-682-4204 or email davidsrefuge@gmail.com

Taste of the Tier: Oct 12, 2018 @ 5:30pm

Riverdale Banquet Hall, Endwell NY



- An evening filled with friends, food and fun for a great cause!
- Art work created by patients for auction

For more info, contact CNY CFF's Kristin Wise Kwise@cff.org

North American Cystic Fibrosis Conference 2018:

Registration is open and Live Streaming is available. Visit https://www.nacconference.org/Content/Watch_Live/

Cystic Fibrosis Patient Registry: CF Foundation Registry for all patients with CF.

Fibrosing Colonopathy: A long-term observation of pancreatic enzymes and fibrosing colonopathy. Open to enrollment.

STOP-2: Treatment of pulmonary exacerbations. Open to enrollment.

EPIC: The impact of early pseudomonas aeruginosa treatment. Closed to enrollment.

Vertex "Triple Combination" Modulator Studies: Phase 3 safety and efficacy studies of compound 659 with Tezacaftor and Ivacaftor on different CFTR variant combinations (VX17-659-102, VX17-659-103, VX17-659-105). Closed to enrollment.

Vertex 661-110 "Tezacaftor/Ivacaftor" CFTR

Modulator Studies: A phase 3 long term evaluation of safety and efficacy of Symdeko with patients >12 years old. Closed to enrollment.

Vertex 809-110 "Lumacaftor/Ivacaftor" CFTR

Modulator Studies: A phase 3 long term evaluation of safety and efficacy of Orkambi in patients \geq 6 years old. Closed to enrollment.

Vertex 661-113 and 661-116 "Tezacaftor/Ivacaftor"

CFTR Modulator Studies: A phase 3 study to evaluate the pharmacokinetics, safety, and tolerability of Symdeko patients 6-11 years old and the roll-over. Closed to enrollment.

Your Voice, Your View

You Don't Have to Do it Alone, Written by Rachelle Bloss, a CF mom

Finding the support when needed is extremely important when it comes to being handed a new and scary diagnosis. A year and half ago, I had no idea what cystic fibrosis was. My child was born with a positive newborn screen. He was also born with a meconium ileus, a blocked and twisted intestine that required bowel reconstruction. He had to spend 3 months in the NICU, 8 weeks with an ostomy bag. He has had 3 hospital admissions, several surgeries, countless blood draws, cultures, labs and x-rays. For us, cystic fibrosis has been a whole new world and at times very scary. We have had to make extremely difficult decisions and, like so many other families, we have had to learn to adjust to our new life but I can tell you that we have not had to do it alone. I can also tell you that the experience has not all been negative.

The amount of support that we have been given from our care team at clinic, our local Cystic Fibrosis Foundation Chapter, the Tammy Twitchell Crafts Foundation, and so many of our local cystic fibrosis families has been extremely humbling. We have met an amazing team of doctors, specialist and nurses along the way. They don't just see our child as a patient but treat him as if he were one of their own. Every decision that has been made regarding our child has been done as a group effort making sure that he is being given the best possible outcome with our full and complete understanding. My son has taught me so much and has shown me how resilient and strong our fighters are. The amount of knowledge that I have gained over the past year for something I knew nothing about is tremendous and I could not have done any of it without the love and support from the friendships that I have made along the way. If there is any advice that I could give to any CF parent, it would be to truly take advantage of the resources that are out there, because you are never alone.



Staff Spotlight



Lejla Bush, LMSW

My name is Lejla Bush, LMSW. I am the adult social worker and mental health coordinator at the Upstate Pediatric Pulmonary and Cystic Fibrosis Center. I have a Bachelor of Arts in Psychology from Le Moyne College and a Master's Degree in Social Work from Syracuse University. My

"Working with her has allowed for collaboration and a more complete psychosocial team."

because I see something courageous and unique in the way each individual takes on their journey of living with CF in their own way. In addition to that, I enjoy working with this population, because it allows me to advocate for the mental health needs of individuals living with chronic illness.

journey with Upstate started in 2014, when I interned in the Pulmonary Center. While working as an intern, I realized how much I enjoyed working with the CF population and how drawn I was to working in this center. After working two years in the Emergency Department, I learned that I had the opportunity to return to the CF Center. In December 2018, I was very honored and pleased to start working as a social worker in the CF center again. I enjoy working with patients with CF and their families,

In my spare time, I enjoy watching movies, reading and spending time outdoors. I value the relationships that I have built in life with my family, friends and co-workers. I am grateful for the relationships that I have built with staff at the CF center thus far.

"Lejla comes back to the CF center with passion and commitment. She is resourceful and discerning. Working with her has allowed for collaboration and a more complete psychosocial team."

"I am very impressed with Lejla. She is kind and patient when speaking to her patients. I find that she is very insightful and able to identify the issue and come up with a plan.

And she likes penguins."

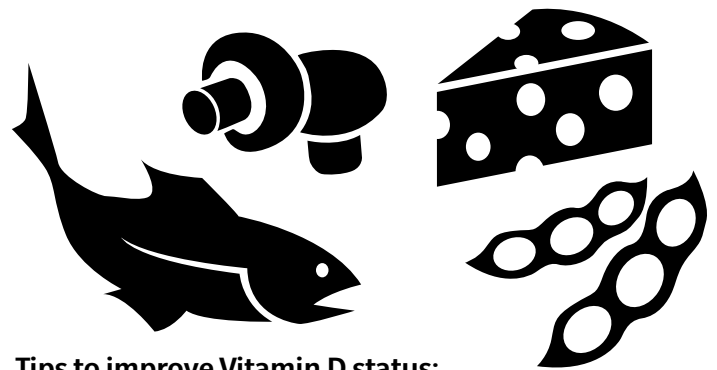
Vitamin D Update

As you know, many people with CF absorb less fat soluble vitamins, A, D, E & K. Taking your CF vitamin daily will help to improve that, but may not be enough to maintain a normal level. According to our research, less than half of our patients with CF had a normal vitamin D level.

Vitamin D is important for good bone health and can potentially help decrease airway inflammation leading to fewer CF exacerbations and better lung function.

Studies have shown that vitamin D in large doses, also known as loading doses can help to increase and sustain a normal vitamin D level. Over the last year, your CF team has been providing a loading dose to patients with low vitamin D levels based on age and current vitamin D level. It is important for your CF team to recheck this level to see how well the vitamin D was absorbed and how long the dose will last. Multiple loading doses may be needed in order to reach the goal vitamin D level. **The goal Vitamin D level is 30-60 ng/mL.** Talk to your doctor or log on to MyChart to review your vitamin D Levels.

For best results and the best health outcomes, it is important to take your CF multivitamin as prescribed in addition to the loading doses. It is our hope to ensure bone health as people with CF live longer.



Tips to improve Vitamin D status:

Always take your CF vitamins with food containing fat and enzymes (if prescribed).

Check with your pharmacist that a CF vitamin is being dispensed; if you are unsure, please bring your bottle of CF vitamins with you to clinic.

Try to increase your dietary intake of Vitamin D.

Foods high in Vitamin D are:

Dairy products that are fortified such as milk, cheese, yogurt

Fatty fish such as tuna, mackerel, salmon

Fortified non-dairy milk products

Fortified cereal and soy products

Egg yolks

Mushrooms

Beef liver and cod liver oil



3 Tips...

to get You Through Cold & Flu Season

Use Your Own Pens

Keep your own pens on you at all times. This way you won't need to use shared pens at the bank, doctor's office, or office reception.

Stop Biting Your Nails

Unless you can guarantee that your nails are completely germ-free, then biting your nails is a sure-fire way for cold and flu germs to enter your body.

Wash Your Hands – A Lot!

Aside from washing your hands after going to the bathroom, or before eating, you should also wash them after touching anything that lots of other people have touched. Carry a small bottle of hand sanitizer gel around with you if there are no washrooms.



Get Your Flu Shot

“It is best to get vaccinated before the flu begins spreading in your community. It takes about two weeks after vaccination for antibodies to develop in the body that protect against flu. CDC recommends that people get a flu vaccine by the end of October.” – Center for Disease Control, 2018.

“Influenza vaccination is generally recommended to patients with cystic fibrosis. Previous studies have shown that influenza infections cause worsening lung function, disease progression and increase tendency of bacterial infections in CF”

– Journal of Cystic Fibrosis, 2008.

Protect yourself and your loved ones, and get a flu shot.

New Indications for Modulators

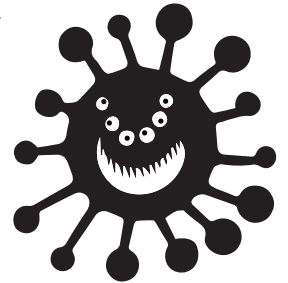
Talk to your care team about the expanded indications for Orkambi.

Now available for 2-5 years old with 2 copies of F508del.

Email List:

Are you interested in receiving newsletters, drug recall alerts, research opportunities, and other health updates from your CF Center by email?

The Upstate Hospital CF Center has created an email database specifically for our patients to communicate updates to you in a timely manner. If you are interested, talk to your care team to sign up.



Pediatric Pulmonary Center
Upstate University Hospital
Rm. 4627, 4W
750 East Adams Street
Syracuse, NY 13210