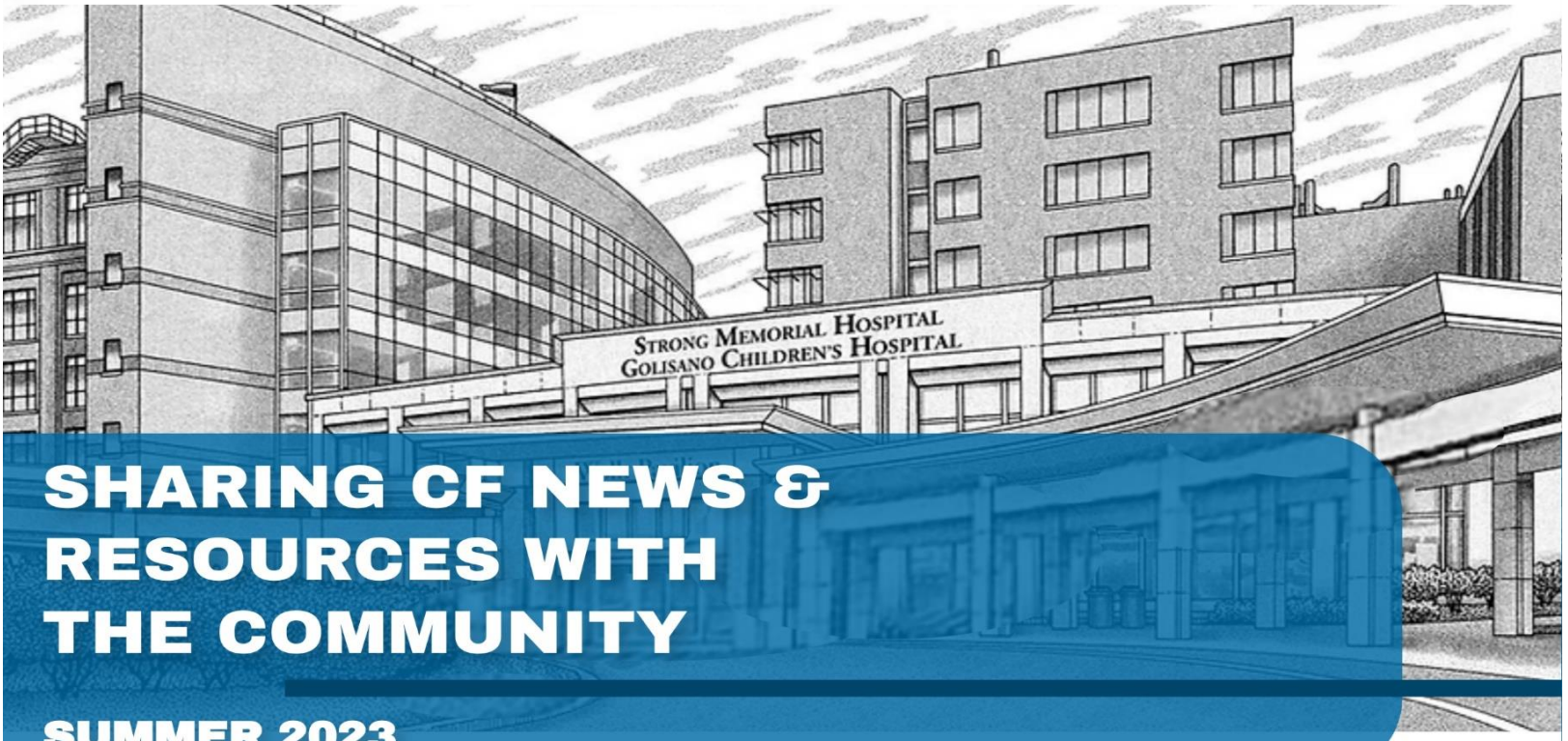
INSPIRE



SHARING CF NEWS & RESOURCES WITH THE COMMUNITY

SUMMER 2023

QUALITY IMPROVEMENT UPDATE

- Our continued commitment to improvement.

We have discussed before the efforts the adult CF program is making on quality improvement in the areas of advanced lung disease and transition to transplant. We are attending our final summit on the dissemination network this June and will be wrapping up the formal QI work but will continue to work with our team and the teams from University of Pittsburgh transplant canter and Columbia Presbyterian in NYC. This has been a very valuable learning experience and we are grateful to have been a part of it since 2017!

Please let us know if there are other Quality Improvement topics you think are in need of some attention!

CF CLINIC AT THE CCC

- The CCC Adult CF clinic has changed times and days for CF specialty clinic appointments.

The previous schedule is that CF clinic appointments are on Monday afternoon and Friday morning the first full week of the month and the third week of the month. The week schedule will remain but clinic for routine follow up and new patient visits will be on Monday afternoon and Thursday morning.

Sick or acute visits are still able to be seen every day, but to optimize the support staff in CF clinic as well as residents and fellows who are learning about CF, we believe that specialty clinics are the most beneficial for everyone.

If you want/need to meet with one of the support staff (dietician, respiratory therapist, social worker or pharmacist) they all have schedules available for in person, video, or phone visits. Just let us know what you need.

Visit the Complex Care Center Website:
https://www.urmc.rochester.edu/complex-care-center

News from the CF Family Connection:

The last few years have brought a lot of changes to the CF community and Cystic Fibrosis Family Connection (CFFC) is no exception.

The most obvious change is the total overhaul of our website (**https://www.cffamilyconnection.org**) which was completed earlier this year. On the surface, we applied a new, more colorful template. Below the surface, we have changed the focus of the site to emphasize the programs offered by CFFC. You can find each of the programs listed in the main menu. (More on programs below.) Also, our "Information" section now deals more in current events, but we do provide links to sites with more comprehensive information about all aspects of life with CF.

We are still committed to our long-standing core programs: hospital help packets, clinic baggies and Education Day. We had some hiccups with the help packets but, with assistance for the staff at Highland and Golisano Hospitals, we believe we have things running smoothly. Improved treatments have reduced the demand for hospital help packets which gives us room in our budget for expanding our programs. In addition to increasing the Emergency Fund allocation, we are exploring other needs in the community through discussions with CF Center staff (both pediatric and adult).
Stay tuned!

If you haven't already, consider signing up for the CFFC email newsletter by sending your name and email address to **info@cffamikyconnection.org**.

Produced once a month, the newsletter summarizes the information added to our website and Facebook page. We occasionally distribute significant, time sensitive information (like important surveys or schedule updates). For daily links to relevant - and interesting - information, you can follow us on Facebook.

https://www.facebook.com/CFFamilyConnection

PEDIATRIC QUALITY IMPROVEMENT UPDATE:

Our multidisciplinary Antibiotic Stewardship Team continues to work on this project which was highlighted in the last newsletter. We are getting ready to release an "Antibiotic Choice" document to our provider team. The purpose of this document is to both serve as a resource for providers to select an effective antibiotic targeting the specific bacteria grown in a sputum culture and to help with consistent prescribing practices. Ask us about this if you or your child needs an oral antibiotic! Our team has not yet formally rolled out the Pulmonary Exacerbation Score discussed in the last newsletter - this part of the project remains in process. Stay tuned!

We also have a QI workgroup looking at H2 (histamine 2 blocker) and PPI (proton pump inhibitors) use in our pediatric patients with CF. These two classes of medications are typically used to treat reflux but may also be used to make enzymes work more effectively. Our workgroup is examining how often these medications are prescribed, how long a typical course of treatment is, and the process/decision points used to consider a dose decrease or a trial off the medication. Stay tuned! You may be asked more in-depth questions about these medications if you or your child is taking one.

Bridget Platania RN, CPNP *(she/her)*

RESEARCH & NEW THERAPIES

At the URMC CF Center, both the Adult and Pediatric programs are very interested in being involved in research for new therapies for CF. If you are interested in participating, let your providers know and if a study comes up you are eligible for, we will contact you and give you the details. If our center isn't participating in something you are interested in, let us know and we will see if we can find a center that may be able to enroll you!

Our current studies here at UR:

STRONG-CF: (Not Yet IRB Reviewed)

Strength & Muscle Related Outcomes for Nutrition & Lung Function in CF (STRONG-CF)

The purpose of this study is to learn things that may help people with CF who are 18 years and older.

This study is being done because many people with CF struggle with nutrition. This can be stressful for people with CF and their families and caregivers. We would like to learn more about several measures of nutrition on individuals with CF.

There are currently two main ways of measuring nutrition in the CF population: body mass index (BMI) and laboratory values. This study plans to look at more ways to measure nutrition, and body composition, like the percentages of fat, bone and muscle in your body. One of the ways we will measure these items is by using dual energy X-ray absorptiometry (DXA) scan, which is a type of x-ray.

This study hopes to provide researchers with more detailed information about nutrition and body composition in adults with CF population. This study will also give researchers information to help with future research.

MayFlower Study: (Study Not Yet IRB Approved)

A Prospective Study Evaluating Maternal & Fetal Outcomes in the ERA of Modulators (MAYFLOWERS)

This is an observational study to follow pregnant people, 16 years and older with CF and their infants who are exposed to highly effective CFTR modulators (like Kalydeco, Trikafta, Symdeco) but will also include pregnant mothers with CF and their infants who are not on modulator medication. The mothers will be followed during their pregnancy and 2 years after the baby is born. The infants will also be followed for 2 years after delivery. The study is designed so it is as convenient as possible for the mothers and their support people by allowing data collection with virtual tools instead of a lot of in person visits. Also, importantly we must be sure the study is conducted carefully to protect both the women participating and to make sure the data is accurate. In order to accomplish these goals, the study visits are scheduled around times that match with regular OB/GYN visits and/or CF visits, including the option of telehealth or phone visits, if routine CF clinical visits are not convenient for the study participants.

COMPASS®

NEWSLETTER

CYSTIC FIBROSIS FOUNDATION

FOUNDATION COMPASS

In 2022, CF Foundation Compass assisted nearly 12,500 people living with cystic fibrosis navigate challenges related to insurance, financial and legal assistance, and other life issues. Many reported that Compass helped them feel less stressed and better able to tackle the challenges they face when living with cystic fibrosis. The best part? Compass is available to anyone with CF, their family, and care teams.

To find out how Compass can help you, call or email 844-COMPASS (844-266-7277) Monday - Friday, 9a.m. - 7 p.m. ET; compass@cff.org.

PLEASE REVIEW THE PATIENT REGISTRY RESULTS AT CFF.ORG

The positive clinical trends seen in 2020 and 2021 were sustained in 2022 with continued improvement in lung function and median survival. The number of pulmonary exacerbations and lung transplants remained low, while the number of pregnancies among women with CF remained much higher than historical trends. All of this occurred in the context of decreased use of chronic therapies, likely as a result of improved lung health for the many benefitting from elexacaftor/tezacaftor/ivacaftor (ETI).

Although life was returning to “normal” in many regards, the number of clinical encounters remained lower than pre-pandemic levels. More visits were conducted in clinic in 2022 as compared 2020 and 2021. The many positive trends in 2022 are cause for optimism, but many challenges remain. The unanticipated effects of ETI and the pandemic such as excessive weight gain and obesity as well an increase in mental health concerns underscore the continued importance of holistic CF care delivery with a multi-disciplinary approach.

EXPERIENCE OF CARE SURVEY

MAKING IMPROVEMENTS TO QUALITY OF CARE TO PEOPLE WITH CF

TAKE ACTION

Nowadays we receive a lot of surveys when we get home from the hospital or a clinic visit. Every time I complete one, I find myself wondering what happened to it and if they took any of my suggestions. I stopped filling them out for a while because it seemed futile.

Well, that is not the case with the Experience of Care Survey. This survey is intended to improve the quality of care that care centers deliver to people with CF and is a way for people with CF, as well as parents and caregivers, to provide information anonymously to participating care centers. It is different from all of the other surveys in that we know that our feedback will be shared with, and reviewed by, our care teams. The hope is that we will see results in the form of a better experience during clinic visits instead seeing our feedback disappearing into thin air. You should be receiving this survey soon after clinic visits.

We get to provide feedback on our most recent visit, timeliness of care, infection control, our care team, health care resources and our overall health. So, basically, it covers our medical appointments from beginning to end. I took the survey for adults with CF, which I received in the mail, but it is also available to take online and on the phone -- and there are separate versions for parents of kids with CF and for Spanish speakers. If you haven't been asked to complete it yet, ask your care team about it.

As patients, this survey gives us the opportunity to effect change (in 20 minutes or less!).

Care centers can access the survey results in real time. The responses will provide the insights for care teams to learn their strengths and weaknesses in the eyes of the people they serve.

When I took the survey, I initially had concerns about confidentiality but was assured the surveys are completely confidential. The anonymity of the survey helped me feel like I could be completely honest. I took my time to really think about the questions and my answers. This is our opportunity to make our health care better!

Whether you're an adult with CF or a parent of a child with CF: help make our medical visits more productive and useful. Help the clinic staff become more aware of our distinctive needs. It all starts with filling out this survey and sharing our experiences openly and honestly.

We are on the precipice of CF pivoting toward a much more manageable disease with effective, responsive health care. But it starts with us: the patients, the stakeholders. Please fill the surveys out -- online or when they arrive in the mail -- and start the process of getting more involved with your health care.

-Rich DeNagel, Adult Living with CF Excerpt from CFF.ORG

Make-A-Wish®
ELIGIBILITY CHANGES

*Starting **January 1, 2024**, there will be updates to the national Make-A-Wish policy regarding cystic fibrosis eligibility. The current eligibility requirements will remain in effect until then.*

MAKE-A-WISH OFFERED THE FOLLOWING STATEMENT:

"Medical eligibility criteria are reviewed regularly by a national advisory council of doctors and medical professionals. Given the ongoing life-changing advances in cystic fibrosis, beginning in January 2024, cystic fibrosis will no longer automatically qualify for a wish. CF will be eligible when accompanied by additional complications or factors that make the current situation critical.

At Make-A-Wish, we celebrate medical advancements and improved treatments for the wish kids we serve—children diagnosed with a progressive, degenerative, or malignant condition that is currently placing their life in jeopardy. These advances also allow us to regularly evaluate the equity of the eligibility criteria for a wish.

Given the ongoing life-changing advances in cystic fibrosis research and treatment, Make-A-Wish is transitioning our CF eligibility to qualify for a wish on a case-by-case basis. This means that CF will no longer automatically qualify for a wish and will be eligible when accompanied by additional complications or factors that make the current situation critical. Because of this, wish referrals based on CF will be reviewed case-by-case as of January 1, 2024. This is consistent with our approach to other critical illnesses that do not automatically qualify for wishes because the course of the condition varies from person to person. These illnesses include certain types of cancer, epilepsy, sickle cell, and heart disease."

Given this information, we, as a CF Care Center wanted to keep our population informed and encourage folks to consider if a wish is appropriate for you sooner rather than later.

If you have not yet received a wish and you have questions, or would like to discuss this further, please contact our CF Center Social Worker:

Marcy Odell, LMSW
(585) 275-2464

REMINDERS ON SUMMER SAFETY

Don't let cystic fibrosis stop you from having plenty of summer fun! Here are some things to keep in mind during the summer months.

STAY HYDRATED

Warmer summer temperatures can cause dehydration very quickly, especially with increased physical activity like exercise or playing. For people with cystic fibrosis, dehydration can cause the body's mucus to become thicker. Remember to have plenty of fluids to drink this summer, especially when planning an outdoor activity. Drinking water throughout the day is a great way to stay hydrated. A good rule of thumb is to drink 6-12 ounces of water or other non-caffeinated beverage for every 30 minutes spent exercising or play in the sun. Popsicles are a fun way to add extra fluids on a hot summer day.

SALT REPLACEMENT

Heat and physical activity can cause you to burn extra calories and lose salt through sweat. A great way to replace the salt that is lost is by having a salty snack. Some good examples of salty snacks include nuts, pretzels, potato chips, trail mix and sports drinks.

MEDICATIONS

As you make plans for a day at the beach or a road trip to the mountains, check with your local pharmacist about storage instructions for your medications. Some common cystic fibrosis medications must be refrigerated while others need to be stored at room temperature. Never leave your medications in the care and keep them away from direct sunlight.

AVOID ALLERGENS & POLLUTANTS

Make sure that you consider the air quality of potential vacation destinations before booking your vacation. Avoid areas known to have high levels of smog or pollution because these things contain chemical irritants that can trigger breathing problems. The Environmental Protection Agency (EPA) maintains a searchable database of air quality information that is a great tool you can use to check out the area you are planning to visit.
http://www.airnow.gov/

It's also a good idea to be aware of any environmental allergies that you may have and avoid areas that are likely to contain them. Talk with your health care provider if you cannot avoid allergens on your summer vacation and discuss if an antihistamine is appropriate for you.

TRAVELING BY AIR

If you are traveling by air, keep your medications and necessary supplies in a carry-on bag. Airlines do lose luggage on occasion and a lost bag can take days to find its way back to you. Never pack anything in luggage that you are checking in that you cannot live without for a period of time, like enzymes. Things that you will want to keep in a carry-on bag include medicines, infant formulas, nutritional supplements, medical equipment (nebulizer, vest, etc.), health care provider contact information. It's a good idea to check the airline website for any additional information about special handling of medical equipment. If a letter of medical necessity is required by the airline, please contact the CF team for assistance, allowing at least 5 business days prior to your departure.

HAVE FUN BUT STICK TO YOUR ROUTINE

Getting away on a summer vacation can be a great way to unwind and relax. It is also easy to get sidetracked from your daily CF regimen. Continue to take your medications at the same time and plan your vacation activities around any respiratory treatments that you need to continue to do while on vacation. Taking time out to do airway clearance may be inconvenient but skipping it could lead to complications that could end your vacation early.

Remember, if while away on a vacation, you need to reach your CF team, please do not hesitate to call us; we are here to help!

CYSTIC FIBROSIS FOUNDATION®

COMMUNITY EVENTS

WHAT IS ROSE UP?

Inspired by the strength of the CF community, a group of adults with cystic fibrosis founded ROSE UP, a national virtual fundraising event for people with CF, along with their family, friends, and colleagues.

ROSE UP participants are invited to come together on a single day to raise funds and awareness of the CFF mission through an act that is meaningful to them.

Anyone can ROSE UP in any way, by doing anything you are passionate about - from running, reading, dancing to baking and sharing how you ROSE UP on social media.

To learn more, contact the CFF WNY Chapter: western-newyork@cff.org

CFF IS SEEKING CF AMBASSADORS

Would you like to share your story as a CF Ambassador? As a CF Ambassador (someone living with cystic fibrosis) you will work with the CFF to share your story through social media and messaging leading up to Rose Up Day on September 21, 2023.

Your role as CF Ambassador:

- Raise awareness about cystic fibrosis and the CF Foundation
- Encourage CF community participation in events
- Inspire and thank event participants
- Recruit new audiences to the event and the CF Foundation

If interested in learning more about the CF Ambassador program, contact the CFF Chapter office: western-newyork@cff.org; or call 716-204-2535

SAVE THE DATE FOR ROSE UP

SEPTEMBER 21ST, 2023

#ROSEUPCF

STAFFING UPDATES

EMPLOYEE SPOTLIGHT!

Hi everyone!! My name is Elle. I am an RN on 8 South at Golisano Childrens Hospital. Over the past 5 and a half years working with many patient populations, I developed a strong passion for the Cystic Fibrosis population. So much so, I created a project that incorporates all the care we provide to these patients during their 2 weeks stay. This educational binder became my level 3 project and soon helped me become the CF champion on the unit. When I first became a nurse, I had no idea what CF was. In turn, becoming so involved with this population helps me to spread awareness for not only medical staff but others around us. Thanks to Etsy, I found a piece of art that illustrates growth and beauty in what can be a scary disease to those affected. By selling these shirts, I was able to raise over $1,000 which will be used for gift cards and donations to organizations (Liv, Luv, Breathe, etc.) for these families involved especially during their admission to 8 South. Thank you for taking the time to read about my journey as a nurse and helping us as a community be "CF Strong."

WELCOME NEW STAFF!

As you'll soon see, we have several new faces in at University of Rochester and would like to take a moment to welcome our new colleagues!

Pediatric Pulmonary:
Liza Ortiz- Physician Support Specialist

The Complex Care Center:
Dr. Will Hardy - Physician
Diana Johnson - Care Manager
Ty'Asia Mitchum - Med Tech

Patrick Johnson RT - Lab Supervisor
Jennifer Winkelman RT - PFT Lab

COMING SOON

SPRING 2024

We are excited to announce that our education day is transforming into an in person family fun day!! Our board members are looking forward to bringing the local CF community together at an outdoor space with activities for all ages to enjoy.
More information to come!

CFFC
Cystic Fibrosis Family Connection