



Spring/Summer 2017

Inspire Spring 2017

University of Rochester Cystic Fibrosis Center Adult and Pediatric News

New Division Chief in Pediatric Pulmonology

Dr. Augusto A. Litonjua is a physician at Brigham and Women's Hospital (BWH). He is also an associate professor of medicine at Harvard Medical School.

He received his medical degree from the University of the Philippines Manila (Manila, Philippines). He then completed a fellowship in internal medicine and a chief medical residency at The Brooklyn Hospital Center (formerly Brooklyn Hospital and Brooklyn - Caledonian Hospital). He also completed two fellowships: the first in pulmonary and critical care at West Virginia University Hospitals, and the second in pulmonary disease at BWH. He is board certified in critical

care medicine, internal medicine and pulmonary disease.

Dr. Litonjua's clinical interests include chronic obstructive pulmonary disease and asthma. His research examines the ways in which environmental exposures and genetic factors impact the development of asthma and lung function decline. He is currently studying the impact of vitamin D on lung disease, and his publications were the first to demonstrate that higher maternal vitamin D intake during pregnancy protects against lung-related illnesses in children. Dr. Litonjua has authored over 200 peer-reviewed publications and is the recipient of funding from the National Institutes of Health

Community Voice

Community Voice provides opportunities for people with cystic fibrosis and their family members to have an active say in Cystic Fibrosis Foundation initiatives, ensuring the voice of the cystic fibrosis community is helping drive improvements that will benefit everyone affected by CF.

If you are interested in learning more or getting involved, please check out: <http://www.cff.org/Get-Involved/Community/Community-Voice/>



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CYSTIC FIBROSIS FOUNDATION

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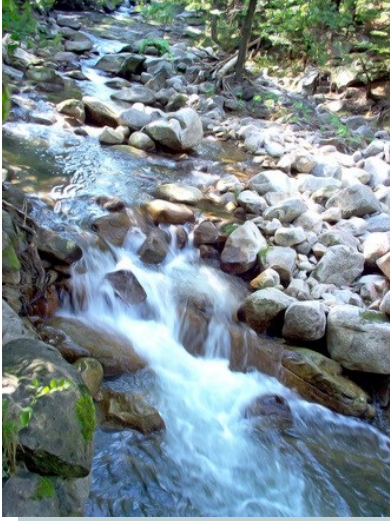
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What's in a Number and Spirometry's



CF miniCON



In addition to BreatheCon, in 2017 we are introducing two CF MiniCons—one-day, topic-specific online events with presentations, group chats, and small group video breakouts. To create a virtual space for honest and open dialogue, these events are open only to adults with CF age 18 and over (not Foundation staff), but we wanted to make you aware and encourage you to share the information below with any adults with CF who you think may want to take part.

· CF MiniCon: Young Adult Transition – July 22, Time TBD
BreatheCon 2017 – September 8-9, Times TBD

While last year’s BreatheCon was a pilot program, with 188 attendees and limited mostly to word-of-mouth promotion, this year we want to spread the word about these virtual events to all adults with CF.

Additional details will be posted in the weeks leading up to each event at www.cff.org/minicon and www.cff.org/

[breathecon](http://breathecon.org). For questions or more information, please email Danielle Lowe Cipriani at dcipriani@cff.org.



Cool News at Highland

Something cool happened at Highland Hospital on Friday, 3/31/17. Three mini refrigerators purchased by Cystic Fibrosis Family Connection (CFFC) were donated to Highland for the exclusive use of CF patients admitted to West 7. The real cool thing about this donation is the way it evolved; demonstrating that collaborative health care is alive and well in our community.

The adult clinic and Highland staff periodically ask the folks who have been admitted to the hospital for ideas to improve the in-patient experience. The idea that consistently topped the list was having a refrigerator in the room to store the extra items that frequently supplement the hospital food plan. Key players in the CF care team put their heads together to work out the details of fulfilling this request.

The staff involved included: Dr. Steve Scofield (Director, Rochester adult CF center), Donna Germuga (CF care manager at the adult CF center and CFFC board member), Jean Wingard (Director, Highland dietary services), Rachel Neilans (dietician), Linda Sirianni (dietician), Stacey Bonacci (West 7 nurse manager) and Kyrstin Baldino (nurse, CF liaison).

The last piece of the puzzle was obtaining the refrigerators for patients' rooms. When the proposal was presented to CFFC, it was an easy, unanimous vote of the board of directors to finance this request. We purchased three 1.7 cu. ft. refrigerators based on the expected in-patient counts. The units have adjustable temperature control with a small freezer space.

We played a little trick on one of our community who had checked in for a stay at Highland. Robin Fisher kindly agreed to be part of our photo op. As you can see, the idea of having a refrigerator in her room brought a smile to her face. The smile became a frown when we told her the refrigerators had to be carted off temporarily for a safety inspection.

Going forward, the management of the refrigerators will be the joint responsibility of the dietary and West 7 nursing staff. Don't be afraid to ask to have one brought to your room.

“The real cool thing about this donation is the way it evolved; demonstrating that collaborative health care is alive and well in our community”



Interested in Participating in Health Research?

The Clinical and Translational Science Institute supports a local registry of individuals interested in taking part in future research studies. If you are included on the list, researchers may contact you in the future to ask you to take part in a research study. If you are contacted, you can decide at that time whether or not you want to participate in the clinical study. To sign up, visit RocHealthResearch.urmc.edu or email researchhelp@urmc.rochester.edu. You can also register to participate in a national registry by visiting ResearchMatch.org. See more about research on page 6.



Julie Buerger, PT

Julie is a physical therapist that joined our team in December 2016. She has been with the University of Rochester for 4 years working primarily with individuals that have gait and balance problems, spinal injuries, and other neurological disorders. In her spare time she enjoys outdoor activities like hiking, paddle boarding, and skiing.



Occupational Therapy is a therapy focused on helping people across the lifespan participate in the things they want and need to do through the therapeutic use of everyday activities (occupations). For individuals with CF this may include education and assistance with incorporation of energy conservation principles (Prioritize, plan, pace, position), ways to achieve medication compliance, and completion of an occupational profile.

I have been an OT in the across the continuum of care at the URMC system for the past 13 years. Prior to that I worked in a variety of settings across the country, after graduating from The Sage Colleges in Troy NY. Currently, I am working at the Complex Care Center one day per week, in the Outpatient Occupational Therapy department 3 days a week specializing in vision, cognition, neurological concerns, and lymphedema, and the hospital/an oncology clinic 1 day per week.

Nancy Dukelow MS OTR/L



I hope this finds you all well and enjoying the change of season and able to take witness to all that is happening right now. As we approach the environmental changes, we, at the Complex Care Center, continue to grow and evolve as well!

We are now staffed with two behavioral health professions who are providing short term behavioral health services to our CF patients. Myself, as a licensed clinical social worker and Kristen Davidson, a psychologist who is providing assessment testing for people with developmental disabilities and providing short term therapy as well.

Behavioral Health Services continue to be short term in design. Our goal is to work with you to assess where you are in this big thing called life. Are you living to your fullest potential? If so – amazing, change nothing and go forth with being amazing. However, if like most of us, you are feeling that things have gotten a bit heavy or if you are not living to your fullest potential, let's talk. Accessing Behavioral Health Services begins with a new patient visit in which we will talk about your well-being. We will work together to develop a plan that keeps your emotional, social and medical needs in mind.

Sometimes meeting with us means that we connect you with supports in the community that might provide more intensive, longer term support. Or with support groups. The goal is always to work together with you, and your care team, to develop a plan that helps you get closer to total wellness whether it is in regards to managing the emotional effects of your CF, depression, anxiety or a myriad of other things.

So as you approach this season of growth and change, I hope you are able to take some time to care for yourself and take some inventory on your wellness. If we can help, let us know.

Be well,

Megan Keller, LCSW



Meet the New Team Members at the Complex Care Center



Amy Jerum, DNP
Workforce Development, Complex care Center

My work at the Complex Care Center (CCC) has been a wonderful addition to my professional life. The CCC is the brain-child of Dr. Tiffany Pulcino, a dual med-peds trained physician and one of the most dynamic people I have ever met. The CCC opened its doors about a year ago to provide primary care to people with childhood onset conditions including, but not limited to, intellectual disabilities, cerebral palsy, cystic fibrosis, and sickle cell disease. The CCC is a true medical home, providing medical and dental care, physical and respiratory therapy, and nutritional and behavioral health counseling. The dedicated providers and staff of the CCC work with families to ensure they have the best understanding of the healthcare resources that are available to people after graduating from pediatric care. The CCC provides the

services that allow every adult challenged by pediatric onset chronic health concerns to continue on a path of growth and engagement in our community. Here we have the opportunity to utilize an approach to health that involves a team and a setting designed around the needs of the patient, their support networks, the community, and their providers. I was brought on board to help with workforce development in the community. My goal is to be a resource for both pediatric and adult providers. We want to meet our patients where they are, form a plan for health management that is consistent with their own life goals, and support them on that journey. I am constantly amazed by the accomplishments of these adults, their strength of character, body, and soul is something I am truly lucky to be a even a tiny part of.

Dr. Kristen Davidson joined the behavioral health team at the Complex Care Center in January 2017. She has spent the last 10 years in private practice where she has worked with children, adolescents, young adults, and their families with a wide-range of medical and psychiatric concerns. As the part time psychologist at the Complex Care Center her role includes developmental testing for individuals with suspected developmental disabilities as well as providing short-term psychotherapy around concerns related to managing and adjusting to medical illness, treatment compliance, goal-setting, lifestyle changes, and mood and anxiety disorders. As a faculty member in the Departments of Psychiatry and Pediatrics, she also provides supervision to psychology trainees. She also continues to maintain her general private practice on a part time basis.



Dr. Kristen Davidson, psychologist



Kristine Reinhardt, NP

I wanted to take the time to introduce myself to the CF community. I am honored to be the nurse practitioner at the Complex Care Center (CCC), the location of the URM adult cystic fibrosis program at 905 culver rd. My journey in CF started in June 2016 as an NP graduate from the University of Rochester School of Nursing family nurse practitioner program. Prior to this I have 7 years as an RN at Strong Memorial Hospital, along with 10+ years of experience in health care. I was raised in the Rochester, NY area and feel grateful to be able provide health care to the same community that I grew up.

I provide two roles as the NP for the Complex Care Center. My first role is as expected, another provider who can see you at the office when you are feeling ill or are in need of an appointment. My second role is as the transition care manager, this is a brand new role that only select few primary care offices have currently embedded into the practice. In this role, I will come and visit you during hospital admissions

and provide clear communication between the inpatient doctor/team and your PCP team here at the CCC. I aim to be an advocate for you during these times and ensure that things go as smoothly as possible. It is always nice to have a familiar face during hospital admissions and I hope to provide that for you and take off some of the burden and stress of being in the hospital. In addition to supporting you during your hospital stay, discharge from the hospital is where my role really kicks in. I ensure that you will have a hospital follow up appointment within 1-2 weeks after discharge, I will make sure you have proper health care equipment, medications, and anything else related that you may need at home once you are discharged. My goal is to provide support for the following 30 days after discharge from the hospital through phone calls, Mychart messages, or appointments to ensure that you continue to improve and not decline. I look forward to meeting and working with all of you.



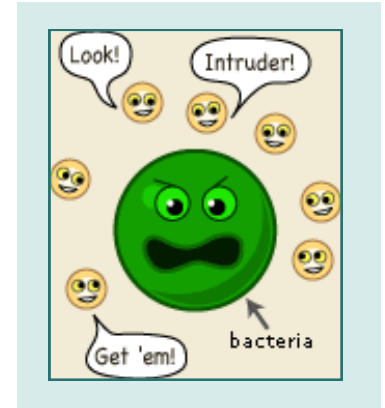
Information on Vogmasks

In response to questions about the use of Vogmasks by people with CF in clinical settings, please note that these masks are NOT approved to protect against the spread of pathogens in the clinic or hospital setting. These masks are intended for use against environmental particles such as dust, pollen, and other particles found in air pollution.

The Vogmask does NOT meet the CF Foundation infection prevention and control guideline recommendation that "all people with CF wear a surgical (procedure, isolation) mask when in a health care facility to reduce the risk of transmission or acquisition of CF pathogens."

Concerns related to the use of these masks in clinical settings stem from questions we've received about the masks and the increase of social media posts of individuals with CF wearing the masks while in clinic. We're also aware of the potential misperception that the Foundation has endorsed the Vogmask due to an article published on Cystic Fibrosis News Today that originally appeared on May 3, 2016. We have addressed this misperception with the publication.

Please let your patients and families know that the Vogmasks do not protect against infection and to urge those who arrive at care center appointments wearing a Vogmask to wear a surgical mask instead.



Community Voice: Engaging the CF Community



Community Voice, formerly known as the CF Adult and Family Advisors, provides opportunities for people with CF and their family members to have an active say in the CF Foundation's initiatives, ensuring the voice of the CF community is being heard and used to drive improvements that will benefit everyone impacted by CF.

Community Voice is open to adults with CF, parents, siblings, spouses, and children of people with CF. Members share their insights on a range of topics, based on their interests and availability. Previous efforts have included innovations in CF care, public policy and advocacy, drug development and research, and programs for people with CF and their families! These projects range in length from

taking a 5-minute survey, to participating in a focus group, or serving on a committee.

When you sign up to join Community Voice you will be asked to fill out a quick form with information about your experiences with CF and your interests and skills. By providing this information, you will get customized emails about opportunities for engagement that match your interests.

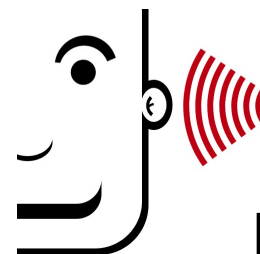
Join us in our efforts to improve the lives of all people affected by CF by sharing your perspective! Visit CFF.org/CommunityVoice to sign up. Questions? Email communityvoice@cff.org for more information.

We want to hear from you!

The Department of Public Health Sciences at the University of Rochester Medical Center is currently recruiting individuals with cystic fibrosis to learn about their experience with getting health care in the Greater Rochester and Finger Lakes region.

The aim of this project is to use interviews and focus groups to talk with and learn from individuals affected by cystic fibrosis. All participants must be over the age of 18 and must be able to read and write English. Participants will be compensated.

This project is made possible by a grant from the Greater Rochester Health Foundation. For more information about participating, please contact (585) 746-6952 or URMCSpecialhealthcareneeds@gmail.com.



CF Foundation Compass: With You Through Insurance, Financial, Legal and Other Issues

For many people with cystic fibrosis and their families, financial burdens and insurance hassles are as much a part of living with the disease as nebulizers and vests.

As part of its commitment to helping people with CF live better today and at every stage of life, the Cystic Fibrosis Foundation offers assistance through Compass. CF Foundation Compass is a personalized service that helps work through insurance, financial, legal and other issues. It's free, confidential and available to everyone, including family members and CF care providers.

"Our Compass case managers are knowledgeable and have many years of experience with CF issues. We listen closely, explain thoroughly, connect you with other resources when needed and work through issues with you from start to finish," reports Anne Willis, Director of Patient Access Programs at the CF Foundation.

Life with CF can be hard, time-consuming and expensive. A recent study by the CF Foundation showed that 80 percent of adults with CF worry about having the financial support they need to live with the disease. Nearly 75

percent worry about getting or keeping their health insurance. Plus, understanding and dealing with insurance requirements can take many hours out of days that are already full trying to manage therapies and other aspects of life with CF.

People reach out to Compass for help with everyday concerns, and they often need more support during times of change, such as receiving an initial diagnosis, aging out of a parent's health insurance or going on disability.

The good news: No one has to tackle these complicated issues alone. Working with Compass can help people with CF and their families resolve issues so they have more time for treatments and all other aspects of life.

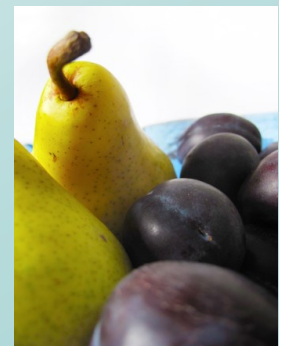
A dedicated CF Foundation Compass case manager is ready to work one-on-one with anyone who needs help. Contact them Monday through Friday from 8:30 a.m. to 5:30 p.m. ET by phone at 844-COMPASS (844-266-7277) or by email anytime at compass@cff.org. You also can learn more about Compass at CFF.org/compass.



Traveling this summer?

Try packing some of these high calorie

- Peanut butter
 - Justin's peanut/almond butter squeeze packs are 190 calories a pack and they come in a lot of flavors
 - Prepackaged PB cups at 210 calories a pack (dip pretzels in the peanut butter for a high fat, salty snack)
- Nutella
- Scandishake packets or Carnation Instant Breakfast (can buy small milk carton at rest stops), bring blender bottle
- High calorie granola bars (>250 calories per bar)
 - Cliff builder bars = 270 kcal/bar
 - Trail mix (ones with M&Ms/candy will add extra calories)
 - Dried fruit
 - Cran-raisins-infused with pomegranate juice (1/4 cup = 130 calories-from label) about 100 calories/ounce
 - Raisins: 85 calories/oz
 - Dried apples: 65-70 calories/oz depending on brand
 - Dried plums (prunes) 70 calories/oz
 - Dried banana: 98-100 calories/oz
- Dried mango: 90 calories/oz
- Dried coconut (Bakers) 130-140 calories/oz
- Dried pineapple: 90-100 calories/oz
- Dried papaya: 70 calories/oz
- Packaged nuts
 - Beef jerky
 - Pop Tarts
 - Pediasure/Ensure/Boost/etc.



Recent and Upcoming Changes for the Pediatric Center

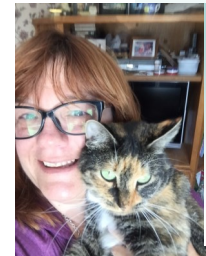
We are excited about the team at the Pediatric center, including many new additions to the team!

New people already here:

- Laurie Johnson joined us this winter. She is an experienced NP who is bringing her skills to our division to join Bridget Platania and Pat Lamarche. We are extremely fortunate to have NP's who have worked with children and young adults with a variety of chronic diseases. She is rapidly learning the ins and outs of CF care.
- Megan Gabel is a Pediatric Gastroenterologist who is working with our team on an ongoing basis. She is in CF clinic on Tuesday afternoon, and available for consultation at other times. She is getting specialized training from other Gastroenterologists across the country. Many of your children have already benefited from a more experienced approach to stomach problems.
- Marie Bieber is a dietician who joined us this spring. She has worked with patients with other chronic illnesses, including eating disorders. She brings lots of ideas for increasing calories and improving eating behaviors.
- Irene Ramos-Suarez is bringing enthusiasm to our research program. She has been working to get the STOP2 trial going for the Adult program, but is a great resource for studies that you or your children may be eligible for.



Marie Bieber, RD



Laurie Johnson, NP

New people coming:

- Tammy Cheney will be our new outpatient specialist. She will join Megan Schlindwein as one of voices you hear when you call and one of the faces you see at checkout in clinic.
- Augusto Litonjua will be joining us in July as the new Pediatric Pulmonary division chief. He has a background in Internal Medicine pulmonary and critical care. He received his medical degree from the University of the Philippines, then completed training in internal medicine at the Brooklyn Hospital Center. He completed fellowships in Pulmonary and Critical Care at West Virginia University Hospitals and another in Pulmonary Disease at the Brigham and Women's Hospital in Boston. He has been on the faculty at the Brigham and Women's Hospital. He does research on environmental exposures on asthma and lung function, and is currently studying the impact of Vitamin D on lung health. He will bring a strong research focus to the division.

Other members of the team who remain dedicated to caring for your children and your family:

- Dr. Voter will continue as CF center director and will assume the role of Clinical Director for the Pediatric Pulmonary Division
- Pat Lamarche and Bridget Platania will continue to see children with a variety of pulmonary diseases, including CF. They are making sure we are keeping up with the new guidelines as they are introduced by the CF Foundation
- Holly Torkington-Wood continues to answer phone calls with her mix of common sense and problem solving
- Marcy Odell helps us to remember the importance of mental health and social complexities
- Kim Bordeaux and Anna Roberts keep the PFT lab going and are experts in airway clearance
- Megan Schlindwein and Debbie Longbine keep the office running and help when there are new medical center initiatives. Debbie organizes events such as the Kit Taylor memorial lecture-ship.
- Judy Sroka makes sure our research focus is on track, and we are looking for new studies.

And we are all interested in working with you and your family to keep CF patients healthy and feeling good.



Bio for New Nurse Practitioner in Pediatric Pulmonary, Laurie Johnson

I began in Pediatric Pulmonary Feb 1 2017. I have been a Pediatric Nurse Practitioner for 16 years. Prior to this position I was in Pediatric hematology/Oncology and before that primary care. I have 3 grown children who all live out of the area. So one of my biggest hobbies is traveling to see my kids. (and with 2 of them living in NYC it is no hardship). Other hobbies include reading, watching movies and bad TV shows and going out with friends. I am very excited to be working in this division and with the Cystic Fibrosis patients. The group I have joined is such a collaborative and professional group that I feel honored to be a part of it.

The Oral Glucose Tolerance Test

The Oral Glucose Tolerance Test (OGTT) and Cystic Fibrosis Related Diabetes (CFRD)

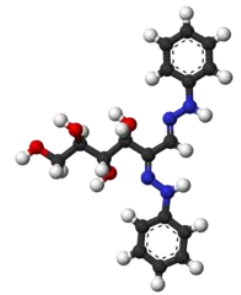
We are using the "Nurse's Nook" section of the spring newsletter to address the issue of the OGTT and explain why it is ordered as a screening test for CFRD. Here at the Pediatric CF Center, we have a goal for all of our patients, age 10 years of age and older, to have the OGTT performed each year. It helps us as the care team assess your/your child's overall health and by doing so, we also achieve the recommendation in the CF care guidelines for CFRD provided by the Cystic Fibrosis Foundation (CFF).

You may be familiar with diabetes as a general term and you may have heard of Type 1 and Type 2 classifications. As an overview, when we eat food, our body breaks it down into fat, protein and sugar. When the sugar enters the blood stream, we all have a measurable level of blood sugar which typically goes up after we eat. This signals the pancreas to secrete insulin which helps the fat, protein and sugar leave the blood stream and enter the cells in the body to use as fuel. When someone is classified as having Type 1 diabetes, their body does not make any insulin and they must receive insulin via injection (i.e. a shot) to make the body work correctly. For individuals who have Type 2 diabetes, the body doesn't make enough insulin and it doesn't respond correctly to the insulin that is present. CFRD isn't really Type 1 or Type 2 and falls into its own category. In an individual with CFRD, the body doesn't make enough insulin and sometimes lacks a normal response to insulin which is most typically caused by scars in the pancreas due to the thick sticky mucus that is characteristic of CF.

There are some hallmark symptoms of CFRD like increased thirst and urination, caused by high blood sugar levels. Other symptoms may include excessive fatigue, weight loss and unexplained decline in lung function.

The OGTT will be ordered by the NP or MD at a clinic visit. It can be combined with other anniversary labs as appropriate. You/your child must be fasting (i.e. no food or drink) for 8 hours before the

test. You/your child will have a blood draw that shows a fasting value and then you/your child will be provided with a beverage to drink called glucola. This is a very sweet drink that provides the body with a lot of glucose. The repeat blood draw at 2 hours shows how your body processes all that glucose and the 2-hour result will show if CFRD is present or not.



Tips to remember about an OGTT:

- Call the lab prior to test to schedule a time. This is to ensure that there will be a supply of glucola on site the day you go for your test.
 - Some families choose to have this done at a time outside of a clinic visit for ease of scheduling (i.e. during summer, a Saturday, or a school break)
 - Another option would be to schedule the OGTT at the outpatient lab inside of the med center/hospital (off the main lobby), have a fasting level done and drink the glucola and then have a clinic visit with the CF Team with plan to return to the outpatient lab for the 2-hour blood draw
 - You can find a listing of lab locations at this website: <https://www.urmc.rochester.edu/urm-labs/service-centers.aspx>
- The individual having the test must be fasting (i.e. no food or drink) for 8 hours before test
- The individual having the test should feel well on the day the test is done; the test should be rescheduled if the individual with CF is not feeling well or not at baseline health
- You may want to bring some food /snack to eat after the test is complete (don't forget your enzymes!)

Find out how to interpret your results on the next page...

... How to interpret 2-hour OGTT results:



Test	Time	Blood Glucose Level	Diagnosis	Action
OGTT	Done in the morning after an 8-hour fast	2-hour glucose greater than or equal to 200 mg/dL	CFRD	Referral to Pediatric Endocrinology
		2-hour glucose Between 140 – 199 mg/dL	Impaired glucose tolerance	Higher risk of developing CFRD; repeat OGTT once each year unless symptoms occur and then do earlier
		2-hour glucose Less than 140 mg/dL	Normal	Repeat OGTT once each year

If you are diagnosed with CFRD, the CF care team will refer you to pediatric endocrinology (a team of care providers with special training in the treatment of diabetes). Working with you, this team will design a treatment plan to help you manage your diabetes while also maintaining your health with CF. The goal for treating CFRD is to keep blood sugar (glucose) at normal or near-normal levels. Doing so will help you gain weight, maintain muscle mass, feel better and have more energy. Maintaining normal glucose levels also lowers the risk of problems caused by diabetes. CFRD can be well managed with insulin along with monitoring your blood sugar levels, eating your usual high-calorie diet and staying active.

Reference:

<https://www.cff.org/Life-With-CF/Daily-Life/Cystic-Fibrosis-related-Diabetes/> and <https://www.cff.org/Life-With-CF/Daily-Life/Cystic-Fibrosis-related-Diabetes/Managing-CFRD.pdf>



Upcoming Events

Join us for the CF Cycle for Life

Saturday, August 12, 2017, at Mendon Ponds Park

This annual bicycle event will take you through some of the most scenic terrain in the Rochester region. CF Cycle for Life features well-stocked rest stops every 10 – 12 miles, bike mechanics along the route, support vehicles, great food, entertainment and much more.

Choose from three routes: a 10-mile route perfect for beginners, a 35-mile route for any skill level or the 61-mile route for a more challenging ride. Whatever route you choose, you can cycle at your own pace. This is not a race, so enjoy the ride!

For more information or to register, visit: 222.fightcf.cff.org/RochesterCycle

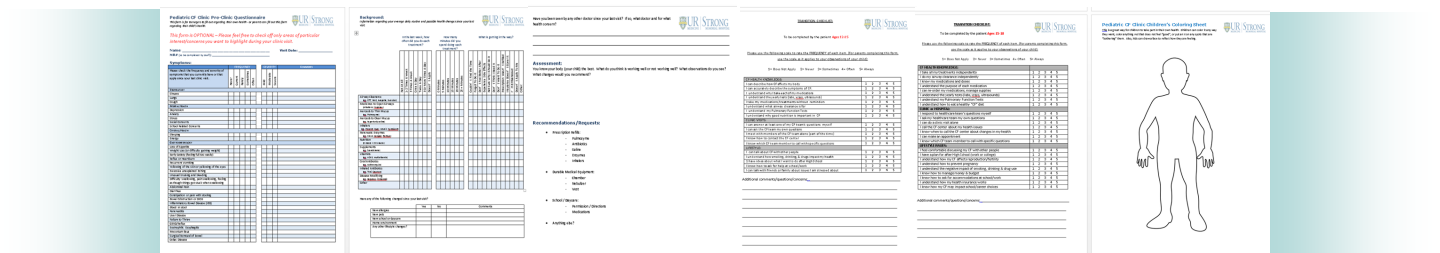




New Tool for Pediatric Clinic Visits:

If you have been to a Pediatric clinic visit in the past few months, you may have been asked to complete a pre-clinic questionnaire. This tool was created in response to feedback from patients and families at last year's CFFC Family Education Night. The idea behind the questionnaire was to provide a way for patients/families to highlight and remember to discuss any recent, pertinent questions/concerns they have had. Clinic visits can often be long with many things to discuss and it can be difficult to remember things. This tool is intended to be a way to organize important topics. Please know that this pre-clinic questionnaire is **OPTIONAL**. It is

several pages long - please know that you are not required to complete the questionnaire. You may, if you choose, check-off or comment only on certain areas. Moving forward, this questionnaire will be offered to you when you check in - please use it as you see fit. Please also know that the questionnaire is available on our CF Center website (<https://www.urmc.rochester.edu/childrens-hospital/pulmonology/cystic-fibrosis.aspx>) and can be printed off and completed prior to coming to clinic. Please don't hesitate to continue to provide feedback to us on this tool. As always, our goal is to make your experience at your CF center the best it can be.



1ST ANNUAL CONFERENCE OF THE UR MEDICINE COMPLEX CARE CENTER *Transforming the Care of Childhood Onset Chronic Health Conditions:* An Interdisciplinary Approach - Friday March 24, 2017

This conference represents the culmination of a successful first year of the UR Medicine Complex Care Center (CCC). The CCC model of care includes a workforce development initiative in order to provide education and training to both formal and informal caregivers. With over 300 primary care patients and over 1000 dental patients to date, we were ready to share our knowledge and success stories.

Attendance of over 80 people was a genuine confirmation of the community's receptivity to the CCC model. We were thrilled to have the diversity of subject matter experts and topics for our first annual conference:

- Tiffany Pulcino, MD – the interdisciplinary model: the Complex Care Center
- Donna Berryman, MLIS, EdD – patient education and health literacy
- Donna Nasca & Regina Dennis – suc-

cesses in health home care management

- Suzie Noronha, MD – navigating transitions in sickle cell disease
- Joel Kent, MD – balancing risk and pain relief in opioid prescribing
- Adela Planerova, DDS, MS – use of desensitization with adults in special needs dentistry.

Continuing education credits were available for physicians, nurse practitioners, physician assistants, nurses and social workers. An evaluation form was provided for participants to give feedback on the speakers, venue, food and topics. We also requested a list of future topic ideas which will be used to plan for next year. Highlights of this year's comments included:

- Thankful/very glad to have CCC as a resource

- Fantastic topics with health care examples
- Expert-level presentations, all knowledgeable, caring, compassionate advocates for this population

The CCC thanks and appreciates support from the Del Monte Institute for Neuroscience. Kathleen Jensen worked with our committee behind the scenes to plan this day. Director John Foxe, PhD, gave a fascinating overview of some of the research being done. We are hoping he'll give a longer presentation next year, stay tuned!

Mark your calendars – next year's conference is set for Friday March 23, 2018. Email your ideas for topics to Laura Robinson at lauram_robinson@urmc.rochester.edu We hope to see you there.



Establishing a Culture of Research- A Research Coordinator's Perspective

A tad over two months ago I started working as a coordinator for research with Cystic Fibrosis (CF)

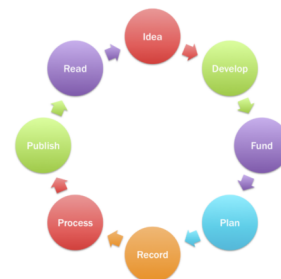
at the University of Rochester. Although I had worked coordinating research for other diseases I did not know much about CF except that it was an inherited disease. I was determined to learn all I could and was soon taken aback by the fact that there are over 1800 mutations affecting the CFRT gene, and counting. I was most of all impressed with the progress made through research during the past decades.

Soon enough, I found myself attending this year's Cystic Fibrosis Foundation Therapeutic Development Network (CFF TDN) meeting in Nashville. There I met face to face with the CFF TDN research coordinators and others offering their help and support, and hundreds of people with the same goal: to improve and extend the lives of people with CF.

I was ready to go on the morning of April 4, concerned about leaving for the airport on time to catch my flight. As I sat waiting in the last event of the meeting with Dr. Scofield and Donna Germuda, I could not help but become absorbed on the big screen in the room. The young mother of a CF patient was being interviewed. She described how overwhelmed she felt on the day she and her husband received the news of her newborn being diagnosed with CF. The thought of the emotions she had gone through was powerful. And then this young mother did something amazing. She reached out for help and the only hope

that could be offered to her at that time. She and her family moved across the country to participate in a clinical trial offered for infants at Seattle Children's Hospital. The newly developed drug turned out to be a great help for her baby and others with the same CFRT type of mutation. I was glad I stayed a few minutes longer.

During the past three weeks I have been meeting adult CF patients at the Complex Care Clinic (CCC) and introducing myself and our new CF research study known as STOP 2. Dr. Steven Scofield, Principal Investigator and Dr. Karen Voter, Co-Principal Investigator, have recently started enrollment in STOP 2 from among the adult CF patients. We encourage all CF patients and their family members to go on the CFF website, <https://www.cff.org/Trials/Pipeline> and look up this research study as well as others. We would love to know your opinion about STOP 2 and about what types of studies you think would be most worthwhile for us to engage in in the future. We welcome your comments and questions. Please do not hesitate to contact me at irene_ramosuarez@urmc.rochester.edu. The success of our research starts with you, the people we care about.



What's up with research at the University of Rochester?

I'm sure everyone knows that the CF foundation has been a powerful voice and force behind research in Cystic Fibrosis. There are so many research goals and trials underway, it's an exciting time to be a CF provider because of the hope that we all have about the future treatments that we expect to see. The CF foundation not only supports researchers and pharmaceutical companies but provides financial support for CF centers like ours who want to be part of their research network.

It's called the Therapeutics Development Network (TDN) and they provide support so programs can hire research coordinators to help increase participation in research trials. They also have a yearly conference dedicated to research and educating your providers and research coordinators. Our team just returned from the latest conference and it's certainly was exciting to feel a part of that energy. Our team has been meeting regularly about research and it's safe to say we have a lot of energy and conviction to increase our involvement in CF research. Between our medical providers, clinic coordinators and research coordinators we have developed a plan to make sure that we talk more regularly with you about research trials that are available at our center and other centers.

Our center has a long history of participating in research trials and is already participating in some trials that are ongoing. However we are most excited to announce that we recently got approved for a new trial called STOP2. It's a trial for persons 18 years and older who need IV antibiotics for a pulmonary exacerbation and it is studying what is the best length of time to treat with IV antibiotics. We are hoping to find 10-15 persons who are interested in participating in the study. Our team will talk with you about this study and others in clinic.

If you interested in learning more in general about the CF foundation's research efforts go to:

<https://www.cff.org/Research/Developing-New-Treatments/Clinical-Trials/>

What's in a number?

Numbers tell us a lot of things! What time is it! Where you live! How old you are! How tall you are! How your lungs are doing! How do you ask? Well, let's start with the basics of breathing. The purpose of the lungs is gas exchange- good air (oxygen) in, bad air (carbon dioxide) out. Air travels in the nose and mouth down the air pipes (bronchi) to the air sac (bronchioles) which has many capillaries wrapped around it. This is where the gas exchange is made between the good air and bad air.

In Cystic fibrosis there is a defective gene that causes the lining of the lung to produce a defective protein which leads to a thickening of the lining of the lung and produce sticky mucus that blocks the air tubes. Bacteria and germs like to hide out in moist, dark places. Your lungs are the perfect breeding ground for this! If left untreated mucus can build up in the lungs, get infected, and lead to a lot of inflammation and obstruction. This is where the numbers come in....Are you following?

At your clinic visit you will do a test called spirometry. We put in your birth day to tell us your age, we put in your height and weight- that helps us find out what number you should be compared to other children and adults your age and height. Then we start the test- we have you take a great, big breath in and blow out as hard as you can. We can tell if you are not doing your best because all values should be exactly the same. That's why you have to blow many times in the same test! We look at the numbers....

- FVC-Forced Vital Capacity-this is the total amount of air that can be measured and tells us how much air your lungs can hold.
- FEV1-Forced Expiratory Volume in 1 second- this is how much air you can blow out in 1 second and tells us how the large and medium size airways (bronchi) are doing.

Interpretations of values for FEV1 are as follows:

Normal: 80-100%

Mild obstruction: 70-79%

Moderate obstruction: 60-69%

Moderately-severe: 50-59%

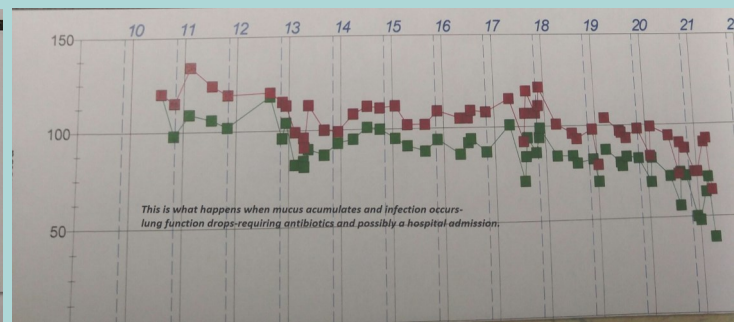
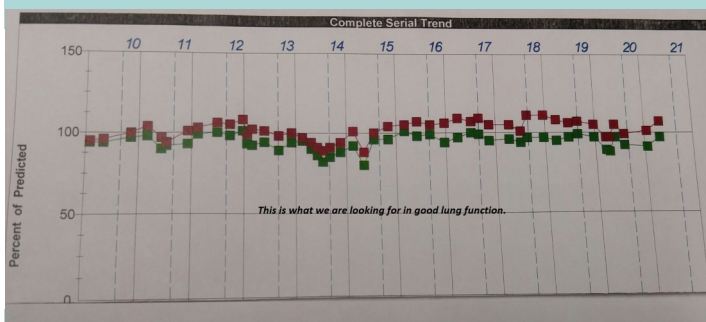
Severe: 35-49%

Very severe: < 35%

- FEV1/FVC-This is the ratio of how much air you can blow out in 1 second compared to how much air your lungs can hold? A decrease in this number is specific for obstructive lung disease and inflammation in the lungs.

What number is probably most important-that's your FEV1. When your airways get clogged with mucus that affects how fast you can blow the air out. The more infected your lungs are, the longer it takes for the same amount of air to get past the mucus and inflammation in your lungs. When your lungs are working well your FEV1 should be above 80%. When you are not feeling as good this number drops.....Sometimes the doctors will give you some antibiotics to help clean out the bacteria as well as other medicine that you may breathe in to help. If really decreased- you may have to make a trip into the hospital for some stronger medications to help clean out your bacteria in your lungs (sometimes called a "tune-up").

What can you do to help keep your numbers good- take care of your lungs. It's important to do your airway clearance and exercise every day-even if you are feeling great! As well as take the medication that the folks in the CF center told you to take. Everyone is different and everybody's numbers are different too! So-what's your number? Ask the next time you are in....



Cystic Fibrosis Resources



Cystic Fibrosis Foundation – Rochester Chapter - <https://www.cff.org/>



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." - www.cffamilyconnection.org/



CysticLife.org - a social network just for the cystic fibrosis community. This positive, uplifting web site is the central location for the CF community to share tips, questions, ideas, experiences and encouragement. <https://cysticlife.org/index.php>



CysticFibrosis.com is one of the world's largest social media networks dedicated exclusively to the cystic fibrosis community. <http://cysticfibrosis.com/forums/>



CF2Chat – A forum and chat website for people with cystic fibrosis. <http://www.cf2chat.com/e107/news.php>



CFcooking - Healthy Cooking for People with Cystic Fibrosis. Watch videos at this site: www.cfcooking.org or on [YouTube](https://www.youtube.com/)

Have a Great Summer



Rochester Cystic Fibrosis Center

Pediatric Providers at Strong Memorial Hospital

Karen Voter, MD
Clinic Scheduling.....(585) 275-2464
Clinic and Prescription Refill..... FAX (585) 275-8706
Bridget Platania, Nurse Coordinator.....(585) 275-2464
Pat Lamarche, Nurse Practitioner.....(585) 275-2464
Laurie Johnson, Nurse Practitioner.....(585) 275-2464
Holly Torkington-Wood, RN.....(585) 275-2464
Judy Sroka, Research Coordinator.....(585) 275-2464
Kim Bordeaux, Anna Roberts, Respiratory Therapist
Marie Bieber, Nutritionist/Dietician.....(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.....(585) 276-7900
Donna Germuga, Respiratory Therapist.....(585) 276-7900
Elle Woodworth, Dietician.....(585) 276-7900
Megan Keller, LCSW Mental Health Provider.....(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
Julie Buerger, Physical Therapist.....(585) 276-7900
Nancy Dukelow, Occupational Therapist.....(585) 276-7900
Jusy Sroka, Research Coordinator.....(585) 275-2464
Irene Ramos-Suarez, Research Coordinator.....(585) 275-2464
Elizabeth Vazquez, Office Manager.....(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.



University of Rochester
Pediatric Cystic Fibrosis Center

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Adult Cystic Fibrosis Center

Complex Care Center
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CYSTIC-FIBROSIS