MICHIGAN MEDICINE
UNIVERSITY OF MICHIGAN

Cystic Fibrosis Center
NEWS & NOTES

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NEWS FROM THE FRONT

Vision for the Future:

Changes in our Clinic Structure During the Pandemic and Beyond

By Samya Nasr, MD, Director, Cystic Fibrosis Center

With restrictions dictated by the global COVID-19 pandemic, our clinic was forced to make drastic changes to patient care delivery. We had to act quickly to change our clinics from in-person to virtual visits. The hospital provided immediate support for all of us to start telehealth visits and to prevent disruption of patient care. On March 16, 2020, in-person visits for people with CF were suspended and virtual visits started the week after.

As part of our center care, we usually have a meeting once a week in person (before clinic huddle) to prepare for the clinics and review the needs of our patients. With the stay home order, our weekly CF huddles became a secure virtual meeting with noted information shared electronically between team members. These huddles are continuing virtually and will change to in-person meetings when the staff can be back fully on site.

Before the pandemic, to prepare for transition, the adult CF team would come to the last pediatric appointment to introduce themselves, answer questions, and ensure a smooth handoff. That transition process was put on hold. But, after a discussion with the adult teams, we are now offering virtual visits with the adult care team in place of the face-to-face meet and greet, done before the young adults are transferred to the adult CF clinic.

Patients started gradually coming back to clinic in June 2020, to be seen by their doctor, nurse, and respiratory therapist. The rest of the team (dietitian, social worker, psychologist, and pharmacist) have continued virtual (hybrid) visits. These visits are usually done within a week before or after the face-to-face clinic visit. Some clinic visits have been done fully virtual if parents requested it, for close follow-up, and sometimes following hospitalization.

Although the virtual model was started out of necessity, it will continue moving forward in some capacity. Some of the team members felt it improved patient care, the dietitian, social worker, and pharmacist, since they can do more with their visits and offer more frequent follow-ups. These virtual visits have provided additional opportunities for working with the patients to improve their care, and do more education, without worrying about the rush of the clinic visits. These virtual visits have also offered a more open conversation with some patients, especially as children are more comfortable in their homes.

Moving forward, virtual visits will continue in some capacity, but not replace in-person care. Throat/sputum cultures to help tell us what bacteria is growing in the lungs are usually done in clinic every 2-4 months. These were suspended during the virtual visits. Also, routine annual CF laboratory work and chest x-rays were suspended until in-person clinics resumed. Measuring lung function in clinic is very important for follow-up. Even though most of our patients have home spirometry now, it is not as accurate or as easy to perform as the in-clinic one. The home spirometry helps us monitor the patients remotely, but should not replace the in-clinic ones. Once the pandemic is over, we believe that we will continue using a hybrid model, with some visits in person and some virtual. We can use virtual visits for most of the dietitian, social worker, and pharmacist visits. Physician's visits will be mainly in person, except if it will be difficult to come to clinic (for example because of bad weather). Also, virtual visits can be used for between-visit follow-up and following hospitalizations. Using the hybrid model, as outlined above, will allow flexibility for patients and families while still allowing for close monitoring and adhering to CF care guidelines.

NEWS FROM THE FRONT

Adult Cystic Fibrosis Clinic Update

By Tom Sisson, MD and Shijing Jia, MD

Another year has passed, and the COVID-19 pandemic is still with us. The continuing presence of the virus in our lives makes daily decisions challenging for everyone, especially those also living with a chronic respiratory disease. In our Adult Cystic Fibrosis Clinic, we continue to strive to provide the best care for our patients. Although our goal of providing optimal care has been made even more difficult by the retirement of our respected clinic director, Dr. Richard Simon, we are very excited about new staff and quality improvement projects in our center. We believe these additions will improve patient experiences and outcomes in 2022.

Before we provide an overview of what is new and exciting in the Adult Clinic, we wish to pay tribute to Dr. Simon as he begins his well-deserved retirement. It is safe to say, that without Dr. Simon's commitment to CF care since 1994, there would be no established Adult Cystic Fibrosis program at the University of Michigan. Not only has Dr. Simon been a role model and teacher for the other adult CF doctors in our center, but he has also contributed to the development of many of our most effective treatments through his role on the Data Safety Monitoring Board at the CF Foundation. His incredible impact was recently recognized at the 2021 North American CF Conference where he received the Richard Talamo Distinguished Clinical Achievement Award, one of the highest honors given by the CF Foundation.

With Dr. Simon's retirement from his leadership position, Dr. Shijing Jia and Dr. Thomas Sisson have assumed co-director responsibilities for the Adult Clinic. Both Dr. Jia and Dr. Sisson were trained at the University of Michigan under the mentorship of Dr. Simon, and his guidance has led to a seamless transition. Dr. Simon's retirement also meant we needed to add another physician to our adult center. We were excited to hire Dr. Sarah Brown. Dr. Brown is originally from Michigan and went to medical school at the University of Michigan before going to Boston for residency training. We were lucky to bring her back to Michigan for fellowship and even more happy to interest her in cystic fibrosis care. In addition to Dr. Brown, we have recently expanded our care team by hiring Becca Aiello as a full-time respiratory therapist and Krysta Walter to help support Linda Fitzgerald in providing clinical pharmacist expertise. Their energy and enthusiasm have already had an impact on clinical care. Katie Hall

continues to work tirelessly in growing the program as our adult program coordinator.

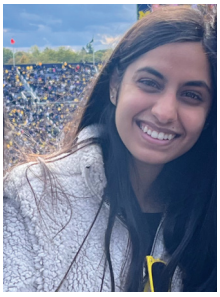
As we begin 2022, our adult program will continue to provide the best care possible for our patients. To meet this goal, we have started several quality improvement projects. Enhancing the process of lung transplant education and evaluation was our team's first goal (and this work is ongoing). We have begun a second project with the goal of using the best length of antibiotic treatment for cystic fibrosis exacerbations. We continue to connect in monthly patient advisory board meetings with the goal of enhancing the patient experience at our center, and we have recently expanded the membership of this passionate group. Our program is also committed to educating the next generation of cystic fibrosis doctors, and we are excited that the Cystic Fibrosis Foundation has supported the development of a fellow's clinic within our adult center for training doctors to become pulmonologists.

Our team in the Adult Cystic Fibrosis Center is happy to leave the challenges of 2021 behind and to begin 2022 with renewed optimism. Dr. Simon will be greatly missed, but we are excited that our new leadership, new physician and staff, and our commitment to improving CF care will make our program better than ever.

STAFF INTRODUCTIONS

Adult Staff

Dr. Sarah Brown obtained her medical degree from the University of Michigan Medical School and completed her Internal Medicine residency at Brigham and Women's Hospital. She returned to U-M for Pulmonary and Critical Care fellowship. She is now a clinical assistant professor in Pulmonary and Critical Care at U-M.



Mehak Gulati is a part-time undergraduate assistant in the adult CF clinic. She will be graduating from U-M with a BS in Molecular Biology in May 2022.

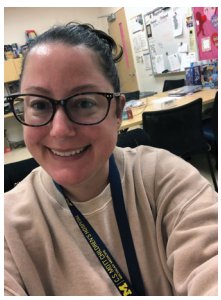
Krysta Walter, PharmD, BCTXP, is a solid organ transplant clinical pharmacist specialist that joined the adult CF team this past August. She completed pharmacy school at U-M and then two years of pharmacy residency training at the University of North Carolina. In addition to caring for patients with cystic fibrosis, she also assists in the management of adult lung, liver, and kidney transplant recipients.

Pediatric Staff

Gita Gupta MD, MS joined our Pediatric Pulmonology clinic in our Fellowship program. She completed Med-Peds residency at Wayne State University followed by a fellowship in Sleep medicine here at Michigan. She recently completed a Master in Clinical Research Design and Statistical Analysis.

Heather Johnson, MSN, RN, earned both her undergraduate and graduate degrees at U-M and has 11 years of inpatient pediatric nursing experience on 12 East (the pediatric CF home unit).

Annette Kortz joined the Pediatric Pulmonology clinic as a Patient Service Associate. She has been with Michigan Medicine for 13 years, after 12 years at another hospital system. She is married and has a German Shepherd and three cats. She looks forward to this new adventure in her career.



Molly Pandrea, RRT, BS, has been a respiratory therapist for five years. In her spare time she enjoys reading, traveling, and playing with her dog.

Courtney L. Prochaska, MSN, RN, CPNP-PC, joined the Pediatric Pulmonology clinic nurse team in February 2022. She has been a nurse for 10 years and recently graduated from U-M with her master's degree in nursing. She has three children, a girl and two boys. Her husband owns a farm where he mostly grows vegetables and flowers.

Gabby Quintana, LLMSW, earned her MSW from the U-M School of Social Work. She joined the Pediatric CF Clinic in early 2022. Gabby previously worked in Adult Nephrology and with hemodialysis patients at Beaumont Royal Oak. She is a former intern with the Michigan Medicine Child Protection Team and Pediatric Trauma and Injury Prevention program at C.S. Mott Children's Hospital.

Meredith Ramm, Clinical Subjects Coordinator, has worked at U-M for the past nine years and has recently transitioned to a new role as a Clinical Subject Coordinator in the CF Center's research area. She loves to travel to national parks and spend time with her husband and daughter Nora.

WHAT'S NEWS

New Respiratory Therapy Role in the Adult CF Clinic

Becca Aiello, RRT

This role is exciting for me because I am the first person to fill it. I am a few months into this position now and I have already learned so much. Primarily, the focus has been and always will be to ensure that all patients have the best care and resources from me as possible. While the role is continuing to develop and evolve, I have been so fortunate to meet with some outstanding colleagues who have given me ideas. So far, I have been able to help with home spirometry and home respiratory equipment and meet with patients during clinic visits. I also recently joined the quality improvement team, and I look forward to getting more involved with clinic improvement projects. In the future, I hope to provide quarterly education opportunities for patients, attend conferences to bring back new ideas and do whatever it takes to provide support to people with CF and my team. There are so many possible opportunities, and I cannot wait to see what the future holds.

Aiello has been a respiratory therapist for six years and previously worked in Mott hospital. When not working, she's dreaming up her next vacation, dragging her husband to the local rock-climbing gym, or taking her spoiled dog for his third walk of the day.

Revamping CF R.I.S.E.

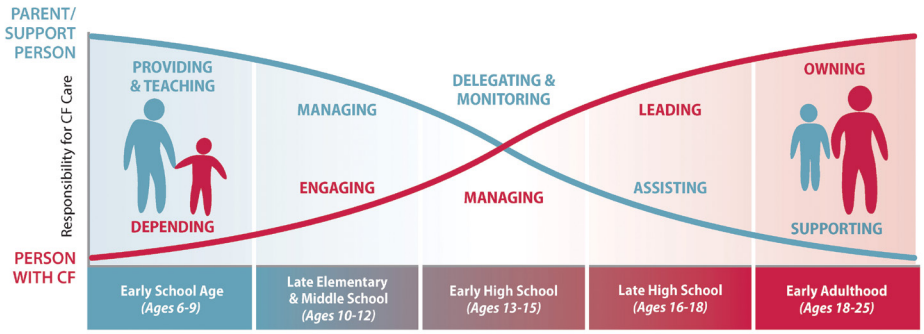
By Julie Lehrmann, LMSW and Jourdan Stiffler, BSN, RN

In our cystic fibrosis (CF) program, we refer to transition in terms of parents transferring CF care responsibilities to their child, as the child grows up. We also refer to it as transitioning from pediatric to adult health care. Sometimes, transitions can come with feelings of uncertainty and apprehension. Our goal, however, is to create feelings of confidence and self-empowerment. One way we can do that is by working together through a program called CF R.I.S.E. (Responsibility. Independence. Self-care. Education.). CF R.I.S.E. is a combined online and in-person program designed to facilitate CF knowledge growth and provide a toolset for one's transition. We paused in 2020 but are excited to announce that we have returned to using the CF R.I.S.E. program — and in a new way!

This year, it is also part of a clinic quality improvement project where the health care team, primarily the clinic nurse and social worker, review Knowledge Assessments with patients 16 years and older in clinic. Your social workers are continuing their involvement, with the newly added expertise of nursing. The goal of CF R.I.S.E. is to provide some structure to better prepare each individual as they transition to greater and eventually sole responsibility of their own care. In clinic, the person with CF can plan to choose a topic, complete Knowledge Assessment questions and then review their answers with the clinic nurse. The Knowledge Assessments are set up as starting points to ask questions and seek clarification on any knowledge if needed. Check out the different topics at: www.cfrise.com.

Another part of the CF R.I.S.E. program is reviewing Responsibility Checklists, in which the person with CF and their parent/caregiver each show how much CF care they each do. This is a helpful way to see what steps might be next to slowly transition care. The picture shows how responsibility is slowly shifted from parent to child/adult over time.

Since we transition individuals to the adult CF clinic around 21 years of age, we still have a lot of time and opportunity to prepare them. We hope that restarting this program (initially with 16 years of age and older) will help reduce any feelings of uncertainty for both the individual and their caregivers. We look forward to many fun conversations with you!



WHAT'S NEWS

Home Spirometry

By Breanna Bell, RRT and the RT peds/adult team

Since the start of the pandemic, our pediatric CF program has worked with the Cystic Fibrosis Foundation to provide 209 eligible pediatric CF patients a home spirometer. The purpose is to allow patients to monitor lung function from home. This initiative has been a great success for our patients who are seen virtually. With a short respiratory therapist (RT) Video Visit to learn how to use the device at home, patients can perform spirometry ahead of their physician Video Visits. Patients are expected to upload results for their pulmonologist to review 1-2 days ahead of their virtual visit. Some of the values measured in percentage (%) may not match what you see when performing spirometry in clinic due to differences in the reference sets within the machines. The pulmonologists will compare the absolute values instead of percentages. So don't be alarmed when values look different. For more information about this, please talk to the RTs performing spirometry in clinic or your pulmonologist. As we navigate through the pandemic, virtual appointments may be necessary at times, and being able to perform spirometry at home allows us the benefit of seeing your lung function.

Patients along with their caregivers can and should schedule an Education Video Visit to learn proper testing technique if you haven't already done so. The RTs in the Pulmonary Function Lab are responsible for education on the home spirometer in the clinic. In addition, we can do a review session for anyone who needs a refresher on how to use their home spirometer. The data is only usable when the test is done with proper technique. Proper technique includes performing three good trials before you send a result to clinic. The RT can show you in person if you prefer, and can schedule an appointment for you the same day as your next clinic visit. It takes approximately 20 minutes to do the scheduled education visit.

The home spirometry program is still active for new patients to CF Clinic; however, this is a limited program, and unfortunately no replacement devices are available. At this time, all eligible patients have received their devices. Current patients using the device who are having a problem with their home spirometer can reach out to their clinic via phone or portal message to troubleshoot with an RT. We are fortunate to be able to monitor lung function for so many patients during this unprecedented time with the use of this great tool. Remember...Blow and Upload!

For Pediatric CF patients please upload results screen to your portal.

For Adult CF patients please email results to UM-CF-PFT@med.umich.edu

Pharmacy "Key Words"

Pharmacists have a language all their own. You can always call your pulmonologist's office for help, but learning these key words will help:

"On File" – When your doctor sent a refill to your pharmacy but you cannot fill it, it will have a new prescription number and is not attached to your old prescription. Ask your pharmacy if it is "on file.

"Co-Pay override" – When you have a primary insurance that pays more than 50 percent of the cost of a medication, and you have Medicaid as secondary insurance, but you still have a co-pay. Your pharmacy can call Medicaid and get the override.

"Specialty Medication" – Your insurance may require that you use a specific specialty pharmacy. Most are mail order, and will deliver to your home.

"Prior Authorization" – Prior Auths (PA) usually take your doctor 48-72 hours to process. Insurance can take 1 to 14 days, but urgent PAs can be handled in 72 hours. If you are almost out of a medication and need a PA, call the CF clinic during office hours and we can expedite it. Most insurances allow you to fill 5-7 days before your refill is "due."

Medication Cost Too High?

"CSHCS" – Medicaid's Children's Special Health Care Services (CSHCS) can cover of co-pays and is available for all people with CF, regardless of age. Contact your CF social worker for more information, or call the CSHCS Family Phone Line at 1-800-359-3722.

"CFF Compass" – The CF Foundation's Compass program works with you, your insurance, and your pharmacy, and your care team to help with medications, insurance and financial concerns, and legal issues. Visit CFF.org for more information.

"Assistance Programs" Individual drug companies can help with co-pays. Search the drug company's website for patient assistance details. ***NOTE:*** *If you have a Medicaid or CSHCS plan you are not eligible for assistance programs.*

Another option is the Healthwell Foundation for patient assistance. Visit their website for eligibility and more information:

Treatments: healthwellfoundation.org/fund/cystic-fibrosis-treatments-2

Vitamins and supplements: healthwellfoundation.org/fund/cystic-fibrosis-vitamins-supplements

PATIENT SPOTLIGHT

From Athlete to College Graduate: *Q&A with Cierra Busby, an adult with CF*

What are your athletic talents and why do you love it?

My athletic talents are mainly motocross (dirt bikes) and softball on occasion. I started at 7 years old. I raced motocross for 16 years and professionally for 4 years in the Women's Motocross (WMX). What drew me into it was my Dad. He had motorcycles and I was too young to have one, so I tried motocross and (come to find out) it was my natural talent! I loved it because it was my "happy place" growing up and it was the one place I knew that I had obstacles to overcome there.

Do you still ride professionally or for fun?

I retired in 2016 and haven't ridden for fun since 2019. It makes me sad, but since I've gotten older and finished school, motocross hasn't been a priority. I do still have a "happy place" when I'm not working. I vinyl wrap cars and make them look good and fancy. It's my happy place that makes others happy when it comes to cars and car shows.

Was it challenging to balance your everyday CF care with sports?

It was super challenging to maintain sport life and regular life. The sport I did was really good for my CF because it was lots of cardio and strength exercise. But when I was sick a lot of the time when I was younger, it was hard to maintain the sport. Sometimes I had to still race when sick with pneumonia or other infections. I overcame by not giving up, as much as I wanted to, as I did the sport because I loved it and it made me feel good overall — helping me to get better quicker. I've learned over time and still now that you, yourself, does come first. You just have to let an illness takes its course. You can't rush healthy. I've learned that when I am sick, just rest and do what is intended so you can get back to normal activities. I don't like to, but you have to!

What has having CF done to help you in your education goals?

Hmmm...I guess more so just not giving up. Going to college for my degrees and having CF has taught me to never give up—and being strong to fight and overcome anything is a great trait to have.

What was your education in?

I earned my bachelor of Science with majors in Biology and Forensics and a minor in Chemistry.

What are you doing for work?

Currently I work as a Medical Assistant in a family practice. I also work on cars on the side.

Where do you see yourself in 10 years?

In 10 years, I see myself with a master's degree in biology or working in a Forensic Lab helping solve crimes.

What advice would you give to someone who is struggling at balancing sports or school with their CF care and health?

My advice to give them would be don't give up. We know CF and school and even sports are a difficult combo, but it can be done. Put your mind to it despite the obstacles. It can be overcome with determination!

PARENT TO PARENT

Our First Hospital Stay, a Blessing in Disguise

By Kirsten Kulik, parent of a child with CF

The words most CF parents dread to hear is "Hospital Stay." This is the story of our first hospital stay that set the standard for every day after.

Our situation is not really that unique, but it is something that does not occur very often. In September of 2016, we took my 10-year-old son, Chase, to a routine doctor's appointment for a cough. That doctor appointment led to a diagnosis of pneumonia, chest x-rays, an emergency room visit, and the first time hearing the words cystic fibrosis. The next day, after a sweat test and frank conversation with the doctors, we headed home with an Aerobika, antibiotics, and a binder on cystic fibrosis. To say that we were floored is putting it mildly, but we were determined to take this under control and help him stay healthy. By day 3 of our new CF world, our boat would be rocked with the dreaded words "hospital stay," but in the long run it was a blessing in disguise.

Unless you are in the world of CF, not many people have an understanding of what goes into keeping a child healthy all while maintaining a normal life. Having just entered this world of CF, I had no reality of what really needed to happen to keep Chase healthy. I remember asking the attending doctor in the emergency room if they thought Chase would need to stay a couple of days, and still can see the look on his face when he stated that he would be in for at least two weeks. It was a look of disbelief and regret at having to share this with me. This moment was the first time I realized that I was in way over my head and had really no clue.

The first day of our hospital stay, against my normal personality, I sat back quietly feeling overwhelmed and concerned that I would sound uneducated and frankly stupid with as little as I knew about CF. The doctors, nurses, dietitians, respiratory therapists, and physical therapists came and left and it felt like they were speaking a second language. Everything I felt so confident about a couple of days ago just flew out the window. Then one of the social workers stopped by. I am not sure if it was look on my face or what, but it was like she automatically knew I was overwhelmed and lost. We had a very hard discussion on what I was feeling and how everyone here could help. It was like a switch flipped, and she helped me realize that no one here was going to think that I was stupid or have a problem if I asked questions. She helped me realize that this was the perfect opportunity to learn as much as possible from the experts in CF. So, with this new mindset and with the help of the nurse, RTs, PTs, and Child Life we set up a detailed schedule for Chase on the door that showed what medications were being given at what time, treatment times along with the order of any medications. When the RT came in, I started to ask questions and learned what each medication and therapy did for him and how each worked. When PT came in, it was the start of learning new techniques, possible alternatives to clear his lungs, and the proper way to do a huff cough. When the doctors rounded the next morning, I listened and learned from the teaching session, trying to gather as much information as possible. I asked questions that seemed remedial but were welcomed and encouraged. I learned about different bacteria, different treatments, what each therapy does, and mostly I learned that the doctors were not just there for Chase but for us also.

By the end of the hospital stay I came home grateful for the team of experts we have, for the knowledge that they were willing to share, and for the nurses who took the time to help and ask questions we did not know to ask. I came home more confident in the knowledge I needed to take care of Chase, but most importantly I came home with a new mind. We will do everything possible to avoid the hospital but I will no longer fear or dread each hospital stay. I will take it as the chance to learn, advocate, and grow.

TEAM UPDATES

Adult CF Advisory Board Update

By Katie Hall, LMSW, Adult CF Program Coordinator

Our Adult CF Advisory Board has continued to help guide our team as we continuously encounter new hurdles due to the COVID-19 pandemic. Some of the highlights over the past year have included one of our advisory board members writing about their experience making decisions around the holidays, guiding pandemic clinic practices, and helping with generating descriptions/expectations for new advisory board members. Last year, we sent out a call for new members to the advisory board and have onboarded several new members to increase the patient voices represented on the board. Our group meets once a month, typically the second Tuesday of the month. If you have an interest in participating in the advisory board, please email Katie Hall at aultkath@med.umich.edu.

Quality Improvement (QI) work is something that both the CF Foundation and Michigan Medicine value. For the last 1.5 years, the Adult CF Center and Transplant Center have partnered with the CF Foundation to complete a QI collaborative. While our work continues as a joint CF/Transplant QI team, our CF center is now moving toward process improvements within the CF clinic. We want to take a moment to highlight the tremendous work our patient representative Meagan Tenyer has done. Meagan has attended weekly meetings all throughout the pandemic to make sure the patient voice was always at the table. She has shared her story with CF centers across the country and not only been a voice for CF patients at Michigan Medicine, but also heard at the CF Foundation and other centers across the United States. For the work Meagan has accomplished, she was nominated for and awarded the Joy Award at Michigan Medicine. The Joy Award is given to one patient/family member each year who dedicates their time to make positive impacts for patients at Michigan Medicine. Congratulations, Meagan!

Pediatric Program CF Parent Advisory Board Update

By Catherine Enochs, BSN, RN, Pediatric Program Coordinator

Our pediatric CF parent advisory board has been engaged and active in 2021. We're looking forward to the work we'll do together in 2022, continuing to bring light to clinic improvement needs and make an impact on CF patients and families.

The parent advisors are currently working on some of our new patient binder materials, making it less overwhelming and more user friendly. Two parents also are working more closely with us as Patient/Family Partners on our team as we engage in the CF Foundation's national quality improvement network, the CF Learning Network. Rebekah and Brandi have devoted time and energy to our improvements, including our transition to telemedicine, and have attended conferences, trainings, and many, many meetings to help us improve the care we provide to all people with CF. Their perspectives and input have been invaluable, and we look forward to continuing working together in 2022.

One of our parent-initiated projects that we will be continuing into 2022 is a video resource library of helpful instructional and informational videos for people with CF and their families. The first video of the series is live on YouTube now and has one of our sweet patients showing young children and their parents how to learn to swallow pills (https://youtu.be/yjqU9ZxUcHo). We're working on spirometry, home spirometry, and various airway clearance techniques as well. As a center, one of our goals is to better represent the entire CF population in our print and video materials, including all races and ethnicities. If you and your child/ren might be interested being featured in one of these videos or being involved in future print or video projects, please reach out to UM-Peds-CFCenter@med.umich.edu.

CFPEERCONNECT

CF Peer Connect is a peer mentoring program for people with cystic fibrosis and CF family members age 16 and older. Through this program, you'll be matched with a peer mentor who has experience with topics that are important to you. Together, you can connect over video, phone, or email.

Visit: cfpeerconnect.com/about

TEAM UPDATES

Want to see your lab results?

Sign up for the My U of M Health Portal to view your U-M labs and tests. Children 11–18 years old have restricted parental views, which do not allow labs to be seen by proxy. You can message your doctor, request refills, and even reschedule appointments all via the secure patient portal. Visit myuofmhealth.org to get your access code and sign up!

After Hours Calls

We have an on-call pulmonary doctor available 24 hours a day to assist with urgent medical needs. Unfortunately, insurance companies are not available after hours, so even urgent prior authorization and insurance problems need to be handled during business hours through the office staff. Forms and other paperwork are not generally available to the on-call doctor but should be handled by the office staff during business hours.

For urgent medical needs, call your CF Center: Pediatric at 734-764-4123 or Adult at 734-647-9342, and follow prompts to contact the on-call Pulmonologist through the operator.

The Cystic Fibrosis Foundation – Michigan Chapter holds several fundraising events including Metro Detroit's Finest, Evening with the Stars, and Great Strides walks throughout Michigan and Northwest Ohio. For more information on how you can get involved in the fight against cystic fibrosis, please call the chapter office at 248-269-8759 or email us at Michigan@cff.org.

Are you interested in opportunities to advocate to state and federal lawmakers in Michigan? Contact Shelly Francis at the CF Foundation Michigan Chapter at sfrancis@cff.org for more information.

RESEARCH UPDATES

The CF Foundation's Therapeutics Development Network (TDN) is a driving force in CF research. Michigan Medicine is a CF TDN Center, which helps us get involved in clinical research so we can contribute to making improvements in CF treatments and therapies. However, we can only accomplish that with the participation of our patients! If you have questions about our research program, you may contact Marisa Linn at mlinn@med.umich.edu and Dawn Kruse at dmkruse@med.umich.edu.

In order to help you better understand some of the studies open to enrollment, below are brief summaries of research we are conducting at Michigan Medicine.

Antibiotic Studies:

1. STAR-ter: cycled antibiotics for eradication of new cases of MRSA **(Recruiting Ages 2-45)**

Anti-inflammatory Studies:

1. Swarm-PA: A Phase 1b/2a, Multi-Center, Double-Blind, Randomized, Placebo-Controlled, Single and Multiple Ascending Dose Study to Evaluate the Safety and Tolerability of AP-PA02 Multi-Phage Therapeutic Candidate for Inhalation in Subjects with CF and Chronic Pulmonary Pseudomonas aeruginosa (Pa) Infection **(Not Yet Recruiting)**

2. 20-0001: A Phase 1b/2, Multi-Centered, Randomized, Double-Blind, Placebo-Controlled Trial of the Safety and Microbiological Activity of a Single Dose of Bacteriophage Therapy in Cystic Fibrosis Subjects Colonized with Pseudomonas aeruginosa **(Not Yet Recruiting)**

3. INS1007-211: A Phase 2a, Single-Blind, Placebo-Controlled, Parallel-Group Study to Assess Safety, Tolerability, and Pharmacokinetics of Brensocatib Tablets in Adults with Cystic Fibrosis **(Not Yet Recruiting)**

Modulator Studies:

1. VX18-445-110: open-label extension for participants in the VX18-445-104 study **(Enrollment Closed)**

2. VX18-445-113: open-label extension for participants in the VX17-659-105 study **(Enrollment Closed)**

3. M19-530: A Phase 2 Study of ABBV-3067 Alone and in Combination with ABBV-2222 in Cystic Fibrosis **(Not Yet Recruiting)**

4. VX20-121-102: A Phase3, Randomized, Double-blind, controlled Study Evaluating the Efficacy and Safety of VX-121 Combination Therapy in Subjects With Cystic Fibrosis Who Are Heterozygous for f508del and a Minimal Function Mutation (F/MF) **(Not Yet Recruiting)**

5. VX20-121-103: A Phase 3, randomized, Double-blind, Controlled Study Evaluating the Efficacy and Safety of VX-121 Combination Therapy in Subjects with Cystic Fibrosis Who Are Homozygous for f508del or Heterozygous for f508del and a Gating (F/G) or Residual Function (F/RF) Mutation **(Not Yet Recruiting)**

Observational Studies:

1. SIMPLIFY: Removing hypertonic saline and/or Pulmozyme therapies on patients taking Trikafta **(Recruiting Ages 12+)**

2. Prospective Study of Cystic Fibrosis (CF) Patients by Lung Magnetic Resonance (MRI) Technology, CT scan of the Chest and Clinical Measures of Pulmonary Function **(Age 6-11 Years Old)**

3. CHEC-OB-17: CFTR Modulated Changes in Sweat Chloride and Outcomes- for patients currently taking an FDA-approved CFTR modulator **(Recruiting All Ages)**

4. NTM-OB-17: Evaluation of a standardized approach to diagnosis (PREDICT) and treatment (PATIENCE) of nontuberculous mycobacteria (NTM) **(Recruiting Ages 6+)**

5. NOPRODCYS0001: Prospective Phase 0 Study to Investigate Pseudomonas aeruginosa and Staphylococcus aureus Bacterial Load, Patient Characteristics and Exploratory Biomarkers in Adult Patients with Cystic Fibrosis or Non-Cystic Fibrosis Bronchiectasis **(Enrollment Closed)**

6. BEGIN: A Prospective Study to Evaluate Biological and Clinical Effects of Significantly Corrected CFTR Function in Infants and Young Children **(Enrollment Paused)**

7. Readiness to Transition to Adult Healthcare and Pre-Transition Experiences Among Parents/Caregivers and Adolescents and Young Adults with Cystic Fibrosis **(Enrolling)**

8. PROMISE: Evaluating the effects of CFTR modulators on airway inflammation and microbiology **(Enrollment Closed)**

9. HERO-2: Home Reported Outcomes in People with Cystic Fibrosis Taking Trikafta **(Pediatric Program- SITE 184)**

10. Mayflowers: To characterize changes in FEV1 over the course of pregnancy based on cumulative CFTR modulator use while pregnant along with defining other factors that may influence changes in pulmonary function including duration of pre-pregnancy modulator use, baseline FEV1, genotype, history of exacerbations, and pre-existing co-morbid conditions. **(Adult Program- SITE 215)**

Infection Control Studies:

1. Assessing effectiveness of Infection Prevention and Control in Cystic Fibrosis **(Recruiting)**

CLINICIAN'S CORNER

Modulator Therapy: *Breakthrough Medications, but What About Side Effects?*

By Hanna Phan, PharmD and Amy Filbrun, MD, MS

Since 2012, with the introduction of ivacaftor (Kalydeco®), the availability of cystic fibrosis transmembrane regulator (CFTR) modulators continues to expand with now nearly 90 percent of people with CF expected to be potentially eligible depending on age. The currently available CFTR modulators are listed in Table 1. The CF Care Team considers various factors in determining if a person is eligible for a CFTR modulator including age, CF genotype (i.e., CF-causing mutations), current medications (i.e., looking for possible drug interactions) and organ function (e.g., liver).

Table 1.
Currently Available CFTR Modulators (as of February 23, 2022)

CFTR Modulator Generic (Brand®)	Current Ages Approved	How it Comes
Ivacaftor (Kalydeco®)*	4 months and older	Granules/sprinkles Tablet
Lumacaftor-Ivacaftor (Orkambi®)	2 years and older	Granules/sprinkles Tablet
Tezacaftor-Ivacaftor (Symdeko®)	6 years and older	Tablet
Elexacaftor-Tezacaftor-Ivacaftor (Trikafta®)*	6 years and older	Tablet

**Highly effective modulator therapy*

When you are thinking about starting a CFTR modulator, it is important to consider both benefits and potential risks or side effects. Each CFTR modulator can have slightly different side effects and each person can respond differently to any given CFTR modulator. Some people may have minimal or no side effects, and others may have some notable side effects. Unfortunately, it is not possible to definitively predict if or when side effects may occur. However, we do recommend routine screens when taking modulators; keeping up with these tests can help identify and manage problems early. Below is a list of examples of possible side effects from CFTR modulators, when the effect may commonly appear, and what to expect for monitoring.

We encourage you to contact your CF care team if you have any of the symptoms listed that are either worsening, affecting your daily care or quality of life, or are worrisome to you. We will work closely with you to assess and manage symptoms and CFTR modulator use. Some potential approaches include dose changes or a pause in CFTR modulator therapy with an individualized, slow ramp-up after symptom improvement.

Making sure to attend all quarterly visits with your CF team, including visits with the pharmacist when starting modulators, is important. Also, let the CF Care Team, especially the CF pharmacist and your doctor, know about any new medications and/or supplements as they may interact with a CFTR modulator. Please reach out to your care team if you have further questions about CFTR modulator therapy.

Table 2.
Examples of Possible Side Effects and Approaches to Management

How and When It May Appear	Necessary Monitoring
Liver Function Changes	
• Mostly seen within first year of starting medication but may occur any time • Can be monitored by lab draws and symptoms • Watch for new abdominal pain with nausea/vomiting, jaundice (yellowing of skin, whites of eyes), dark-colored urine (looks like iced tea color)	Blood/lab draws • Every 3 months in 1st year of medication • Annually, in most cases, after 1st year of medication
Increased cough, sputum production, nasal/sinus drainage (aka "the purge")	
• Mostly seen in first 1-2 months of medication • Seen more with some CFTR modulators than others (e.g., more so with Trikafta)	Usual monitoring at home as per your CF care plan for any changes in symptoms
Abdominal/stomach pain	
Mostly seen in first 1-2 months of medication but can occur any time	Usual monitoring at home as per your CF care plan for any changes in symptoms
Change in stool (constipation or diarrhea)	
Most commonly seen in first 1-2 months of medication but may occur at any time	Usual monitoring at home as per your CF care plan for any changes in symptoms
Headache, dizziness	
Most commonly seen in first 1-2 months of medication but may occur at any time	Usual monitoring at home as per your CF care plan for any changes in symptoms
Rash	
• Seen more with Trikafta versus other CFTR modulators • Commonly seen in first few weeks of medication • Can be all over the body or certain areas in some people	Watch for any new rashes or itchiness after starting medication
Behavioral Health Change	
• Reports in the CF community suggest possible mood/behavioral changes after starting a CFTR modulator • May include new or worsening depression and/or anxiety	Monitor for any changes in mood/behavior after starting medication
Cataracts	
May occur at any time	• Monitor for any vision changes • Annual eye exam ***including a cataract exam***

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