CYSTIC FIBROSIS CENTER
UNIVERSITY OF MICHIGAN
HEALTH SYSTEM

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NEWS AND NOTES

SPRING 2016, VOL. XXX

NEWS FROM THE FRONT

Newborn Screen

By Samya Nasr, M.D.
Director, Cystic Fibrosis Center

Michigan's Newborn Screening (NBS) Program began screening for cystic fibrosis (CF) in 2007. The screening is a two–step process; the first step is to screen for the IRT value for all babies born in Michigan (Immune Reactive Trypsinogen). The second step is done if the IRT value is very high (96 percent or higher). This step includes checking for CF mutations (screening is done for 40 mutations). Infants with high IRT and one or two mutations are referred to the NBS office for the State of Michigan, which is located at the University of Michigan. This office is led by me as the CF NBS Coordinator for the State of Michigan. The NBS office communicates with the baby's primary physician and family to choose a CF Foundation accredited Center for the sweat testing. Babies can be referred to our Center or to one of the other four CF Foundation accredited Centers in Michigan. All CF Centers report the results of the sweat testing to the NBS Program. Outcomes for all positive screens are maintained in a database and screening performance metrics are reviewed by the CF NBS Quality Improvement Committee.

Our CF Center has been leading significant quality improvement research efforts in the area of CF NBS. The research projects have been focusing on improving the NBS process and reducing the families' anxiety associated with positive screening. These research projects have been funded by the Cystic Fibrosis Foundation (CFF). Here are summaries of some of these projects:

1- Ways to reduce the need for repeating the sweat testing. We have to repeat the test if we are not able to collect enough sweat from the baby. We were able to include all the other CF Centers in Michigan in this project. After evaluating the sweat testing process in all Centers in Michigan, we developed several steps to reduce the need for repeating the test. This project has been very successful in reducing the need for repeating the test.

2- Another project was to evaluate and address families' anxiety after they hear about the positive results. In that project the focus was on the babies who were diagnosed as carriers. We were able to come up with a few steps to reduce the parents' anxiety.

3- A third project was aimed at evaluating the primary physicians' knowledge about CF NBS and their comfort with the process and with being the first ones to tell the parents about the results.

4- This project was followed by another one to provide resources for the primary physician to use and refer to when faced with a positive CF NBS test.

5- The latest project is just starting. It will be focusing on families of babies diagnosed with CF in the state. It will be done in two steps. The first step is to evaluate families' experience with the way they received the news about the diagnosis, how they dealt with the news, and what they thought of the information they received from their Center. The second step will be to modify the information we provide to our families to be able to meet their needs better. We will include input from our Family Advisory Board members and from other families attending our Center and the other CF Centers in Michigan. Once we modify the information and material we provide to parents, we will share them with other CF Centers in Michigan as well.

We will continue to work on all aspects of CF to try to improve the care for our kids and adults with CF and their families and caregivers. If you have any ideas that you think we should focus on in our work, please share them with us.

Please contact Dr. Samya Nasr at 734-764-4123 for more details.

INSIDE THIS ISSUE...

NEWS FROM THE FRONT

Increased CFF Support

By Richard H. Simon, M.D.

Over the last year, there has been a great increase in the money that has become available to the Cystic Fibrosis Foundation to support its many missions. This unexpected situation arose from arrangements the CF Foundation had made many years ago with the drug company Vertex Pharmaceuticals. The background is that for years, the CF Foundation had been channeling a large proportion of its resources toward laboratory-based research to better understand the basic problems caused by cystic fibrosis. This research led to a number of promising ideas for the development of treatments, but it became clear that a major roadblock was finding pharmaceutical companies who would use their resources to test the new treatments. Understandably, most drug companies were more interested in tackling diseases that affect larger numbers of people so success would bring larger profits. The CF Foundation solved this problem by providing money directly to pharmaceutical companies to offset the cost of developing CF treatments. Vertex Pharmaceuticals was one of these companies. The financial arrangement was that if the studies did not lead to a new treatment, the company did not need to reimburse the Foundation. But if the work was successful and the company was able to market the drug, the Foundation would receive royalties that the Foundation could then use to support further research and its other missions. With the FDA approval of ivacaftor (Kalydeco) and lumacaftor/ivacaftor (Orkambi), the CF Foundation received a large payment from Vertex Pharmaceuticals. After careful deliberations, the CF Foundation has begun using this money for a number of different purposes.

Because the main mission of the Foundation is to find a cure for cystic fibrosis, a large amount of the new money is being invested in new research areas. The Foundation has been successful in partnering with many more pharmaceutical companies using the same approach that worked with Vertex. The infusion of money is being used to develop new, exciting, innovative strategies to discover better drugs. This model for financing drug discovery, which the CF Foundation pioneered, is being commended by many people including Dr. Francis Collins, the Director of the National Institutes of Health.

Fortunately the new funds that have become available to the Foundation are large enough to expand programs beyond the research arena. The Foundation has significantly increased the size of the grants that it sends annually to all the CF Clinical Centers across the county. The Centers have been using these funds to staff positions for which there is insufficient support from their institutions. This has allowed many CF Clinical Centers to hire additional social workers, dietitians, and other medical personnel to provide care to patients with CF.

The Foundation has also been targeting areas that it recognizes need special attention. A prominent example is the awarding of grants to support mental health coordinators at the CF Clinical Centers. Previous surveys had shown that living with cystic fibrosis causes anxiety and depression in many patients and their families. Most CF Centers do not have the resources to adequately address this situation. In response, the Foundation assembled a large committee to study the problem that included psychosocial experts, caregivers with expertise in cystic fibrosis, and representatives of patients and their families. The committee recommended increasing the mental health services within the Centers. The Foundation encouraged the Clinical Centers to apply for funds to support a mental health coordinator who can oversee programs to screen for anxiety and depression, provide education about psychosocial problems, and arrange for treatment when needed. Separate applications were submitted by the pediatric and adult CF programs at the University of Michigan CF Center and both were accepted and funded.

To identify other high-priority needs, the CF Foundation set up a "strategic investment taskforce" made up of representatives from a broad range of disciplines including medical directors of both pediatric and adult programs, nurses, dietitians, social workers, psychologists, respiratory therapists, pharmacists, and physical therapists. Perhaps most importantly, patients with CF and their family members were part of the taskforce. The committee met at the national CF Foundation offices in November 2015. They generated a priority list that the Foundation is using to direct their resources. Among the many initiatives that will likely receive support are adding pharmacists to the CF team, helping patients/families find their way through the complex and burdensome systems that are part of medical care in this country, increasing the availability of exercise programs, and setting up web-based systems to better connect patients/families with each other and with the CF Centers.

In summary, these large investments of funds into all aspects of cystic fibrosis are very encouraging. The effects will be both immediate and long term. All of the CF Care Centers will be able to expand what they can do for people with CF. And there is no doubt that the increased money being channeled into research will speed the development of better treatments.

WHAT'S NEWS

Staff Introductions

New RN Coordinator Role Cathy Enochs, BSN, RN, A-E

Cathy has been named CF Nurse Coordinator for our CF Center. In addition to Donna Genyk, LMSW, our CF Center Coordinator, Cathy brings a wealth of experience to this position: she has been a Registered Nurse at C.S. Mott Children's Hospital since 2007 and previously worked as an inpatient nurse on 6 Mott until she joined the CF Team in the outpatient Pediatric Pulmonary clinic in 2011. Since then, Cathy has developed a love for the CF population and has worked to educate new staff and improve communication regarding CF patients and their care. Cathy will continue to play a role in the clinic, but with her role of CF Nurse Coordinator will be able to provide more support and focus specifically to the CF Center, including attending meetings and assisting with the CF Family Advisory Board, outpatient and inpatient communication, quality improvement initiatives, communication with the Adult CF team, liaison to other CF Centers, education of team members regarding CF care guidelines, and monitoring that care along with the CF team. Cathy and Donna Genyk, CF Center coordinator, will work together, each bringing their individual professional expertise to joint CF Center initiatives.

New Adult CF Center Coordinator Role, Katie Hall, LMSW

Katie Hall, LMSW, began working at University of Michigan in July 2009 covering different inpatient services. In 2011, Katie joined the inpatient adult CF care team, where she remained until this year. Recently, she started working full time in the outpatient clinic. You can now find Katie in the adult CF clinic where she has dual roles as the clinic social worker and adult CF program coordinator. In her spare time, Katie works off hours in the adult emergency department.

Eeyeen Ong, MS, RD

Eeyeen Ong received both her undergraduate dietetic and master's degrees in Human Nutrition from Eastern Michigan University. She has worked as an adult clinical dietitian for more than 10 years and began working at the UMHS in 2012. Eeyeen enjoyed working with patients with pulmonary disease and began working in the Adult Cystic Fibrosis Clinic in April 2015. With more time to devote to her patients, Eeyeen is looking forward to getting to know everyone, following their nutritional status, and completing clinical assessments.

Katie Hall, LMSW (left), and Eeyeen Ong, MS, RD (right)

Maria Nedanowski, RN

Maria received her nursing degree from Schoolcraft College in 2006. She attended Eastern Michigan University for her undergraduate program. Maria started working as a Registered Nurse on 5 West Mott/ 12 East Mott in 2006 and remained on that unit for nine years. She transferred to the Pediatric Pulmonary Division in July 2015.

Trisha Vess, BSN, RN

Trisha received her undergraduate nursing degree from Michigan State University. When she started working here at the U, it took a little adjustment getting used to the maize and blue colors! She started on 12 East Mott as a bedside nurse and worked there for three and a half years, so she knows a lot of our CF patients and families well. She is very excited to be part of the Pediatric Pulmonary nursing team!

Ashley Lager, RD

Ashley received her bachelor's degree in Dietetics from Michigan State University in 2012 and completed her dietetic internship here at UMHS in 2013. Ashley has been working as an adult clinical dietitian for two years and recently started her position here at UMHS in February 2015. She works with patients on the Medicine Pulmonary service and those on a general medicine and a surgery service.

Adult Inpatient Social Work, Lauren Barris, LMSW

Lauren Barris, LMSW, received her bachelor's degree in Psychology and Spanish and her master's degree in Social Work from the University of Michigan. In 2015, she joined the team here at UMHS as the Inpatient Clinical Social Worker for 6C. Lauren provides social work services to all of 6C, supporting both the General Medicine patients and the Pulmonary patients. She finds working with the Cystic Fibrosis population particularly rewarding due to the complex challenges these patients face and the opportunity to build relationships with them over time. As a new addition to the CF Care Team, Lauren is looking forward to getting to know all of our patients, helping them to navigate the healthcare system and community resources, and supporting each of them as they work to balance their physical and emotional well-being!

WHAT'S NEWS

Mental Health Screening

By Katie Hall, LMSW
Adult CF Center Coordinator

Taking care of your physical health, like taking enzymes before meals, using your vest before bed, or even brushing your teeth in the morning, becomes a routine part of your day. Often times, when we think about our health, we only consider the physical aspect, without recognizing mental health as an integral part of our overall health. Depression and anxiety contribute significantly to the overall well-being of an individual and without treating one you cannot adequately treat the other.

The Cystic Fibrosis Foundation has recognized the importance of Centers screening their patients for depression and anxiety on an annual basis. They are providing select Centers with additional funding to implement the guidelines which include screening all patients annually for depression and anxiety. In the adult clinic, we are looking forward to implementing these guidelines in the early part of 2016.

As a patient, you will be receiving two questionnaires during your outpatient clinic visit. The PHQ-9 will help us to assess for depression and the GAD-7 will be used to screen for anxiety. We intend to discuss the results with you during your appointment and provide referrals as appropriate.

Our goal is that screening for depression and anxiety become part of the normal routine. Next time you are in Clinic, feel free to ask us about the mental health screenings.

Preparing for Transition – CF Rise Update

By Julie Merz, LMSW

The thought of transition from Pediatric to Adult care can cause some anxiety; however, we are focused on successfully preparing patients for this graduation to the adult care. We usually transition patients between 18 and 21 years of age, so that we are available through other life transitions (e.g college, career, moving out).

We have been utilizing the CF RISE program (CF Responsibility. Independence. Self-care. Education.) since July 2015 with great success. The CF RISE program is a national program that is supported by the CF Foundation and Dr. Samya Nasr helped develop the program. This program is designed to prepare youth for managing their own CF care. We have started with patients from 16 to 21 years old (in the pediatric clinic), with this program to continue once patients transition to the adult center if desired. So far, our youth have reported a sense of increased self-esteem in knowing aspects of their care, as well as recognizing the idea of needing to take on more responsibility in their care (e.g. cleaning equipment, learning how to order medications, etc). Parents have also responded favorably, stating being pleased with knowing that their child has learned necessary components of CF and their care.

This program is designed as an ongoing process involving knowledge assessments and skills/responsibility assessments. The knowledge assessment portion is designed to make sure that each patient gained the necessary information on topics shared throughout their life (e.g. Why do I actually need to take my vitamins? Which PFT numbers am I supposed to pay attention to and why?). The skills assessments section is designed to help youth and their parents break down aspects of care that the youth could start to take on. The idea of this is to prepare youth for independent living, so they do not have to learn all of the responsibilities overnight. This can be a great starting point, especially when the idea of transferring CF care to the individual patient seems overwhelming.

The program is designed to occur during clinic visits or during hospitalization so that patients have opportunity to ask the medical team relevant questions. In this respect, we hope to have increased meaningful conversation and assist in each patient gaining more confidence in managing their own care. Our goal in the near future is to start this program with patients 12 years old and above—increasing opportunity for discussion surrounding their CF knowledge.

Congratulations to Us!

The Cystic Fibrosis Center at C.S. Mott Children's Hospital was awarded the Make-A-Wish® Michigan 2015 Shining Star Award. This award was given because of the Cystic Fibrosis Center's commitment to ensuring eligible children are provided the opportunity for a wish to be granted. We were also awarded the 2015 Golden Sneakers Award for CF Center participation and fundraising efforts by the Cystic Fibrosis Foundation.

ADULT PERSPECTIVE

10 Things I Wish I'd Known Before Coming to College

By Hannah Buck

Looking back at my freshman year, I realize that putting college before my CF wasn't the only thing I had all wrong.

As I sat down at my new desk and tidied my wobbling stack of pencils, I thought I had it all figured out. My parents had just driven away with tears in their eyes, but I was doing just fine: dresses in the closet, rug on the floor, and purple power PICC in my right arm.

I was finally a student at the University of Michigan and it was time for my life to begin. I mean, sure, I had just been discharged from the hospital two days earlier despite advice from my doctor to stay, but I had friends to make and dreams to chase.

Looking back, I realize that putting college before my cystic fibrosis wasn't the only thing I had all wrong. Why did I bring so many dresses to school—did I really think I'd be wearing anything other than black leggings and sweatshirts? Why (as I was fully aware of my "adorable" tendency to spill apple juice and pasta sauce) was my rug white? And for the love of all things beautiful, why did I bring so many pencils to school? Note taking? That's what a laptop is for, not that mine was ever charged.

Chronic illness aside, college is an amusement park of mistakes. The great thing about it, though, is that everyone around you is riding the same rides.

Hearing about my freshman year experiences with CF won't keep you from messing up, and that's the way it should be—"oops" is the best teacher. But in situations where being a full-time student could put the state of your oxygenation at risk, here are a few tips from someone who's been there.

1. Early classes? Not a good idea.

When you have cystic fibrosis, sleep is one of the best things you can do for your body, and despite what society likes to make you think, sleeping in doesn't make you a lazy person. Schedule classes that start later in the morning (or afternoon, even) and allow your body more time to fight infection while you sleep. With more time to get ready, it'll also be easier to stay on top of your morning treatments.

2. An all-nighter will last longer than one night.

Remember that thing I said about sleep? I can't tell you how many times I procrastinated writing a paper, stayed up all night to write it, and spent the next week with fevers and chest pains. Do your best to keep up with schoolwork and avoid losing sleep. Your lungs (and your GPA) will thank you.

3. Showing up really is half the battle.

In grade school, my mommy dropped me off ten feet away from the front doors; now, my campus is 3,245 acres wide, and I've found that the trek to class can be difficult. Some days, even though I felt healthy enough to sit through a class, there was no way I could physically get to it. Many schools have shuttles for injured students and students with disabilities. Either way, don't beat yourself up about having a body that doesn't always listen to you. It's not your fault.

4. It's Exercise, not extracise. (Was that funny? No? No.)

The day I came to college was the day I stopped dancing competitively and playing soccer, and since then, my baseline lung function has seriously dropped. The fact that you probably won't gain the freshman fifteen (thanks, dysfunctional pancreas!) doesn't mean that you don't have to exercise. Try intramural sports or going on daily walks, and just keep moving.

5. The dining hall is your best friend.

It's your nutritionist's dream: all-you-can-eat chicken breasts and soft-serve ice cream. Make her proud. Stuff your face.

6. Get a Gamecube for your dorm room.

When all of your friends are hanging out and having fun, the last thing you'll want to do is leave to sit alone and inhale your mucus thinner. Having an inexpensive TV with "Super Smash Bros." capabilities means that your squad will be more than comfortable chilling in your dorm. And trust me, no one will care if you throw on your Vest while dominating Donkey Kong.

7. Accept the germs you cannot change.

People surround you constantly in college. It really is unavoidable, and you'll notice that even your healthiest friends will get sick more often than they used to. Carry hand sanitizer with you everywhere and repeat after me: "I tried."

8. You'll become even more addicted to the internet.

Now, you live on your own, and unfortunately, the healthcare world sees you as a goofy teenager who doesn't know what they're talking about. Save yourself the trouble of explaining your awesomeness to the phone clerk and use your clinic's online patient portal. With one click, you can schedule an appointment, renew a prescription, and request medical advice. (Don't tell your professor I said this, but if you need to, you can even check your lab work in class! It's so easy.)

9. Call your mom.

She gave birth to a baby with a scary disease, and to her, you're still that baby. Tell her you're doing okay so she can sleep tonight. Seriously, she has work tomorrow.

10. Know that you are one spectacular human.

It takes a lot of guts to go to college with cystic fibrosis, and for that, I give you kudos. You are not allowing the course of your life to be decided by your genotype, and that's a more impressive feat than anyone could ever see from the outside. You're doing good, kid.

PARENT TO PARENT

Newborn Screen

By Erin Boillat, Mother of Elouise, 2 years old

About two and a half years ago, our world was shaken in so many ways. Our beautiful, bright-eyed, red-haired daughter was born! Elouise came into our world with so much excitement. She is our youngest of three, with her siblings five and eight years older.

Before we could leave the hospital for home, Elouise had to be screened for a few rare diseases….Our nurse offered us more information about the screening, and I declined, naively assuming that everything would be fine. She handed me a brochure listing the diseases that the screen covered, and I quickly scanned by the words cystic fibrosis. There isn't any family history of CF and we have two healthy children already. It didn't even cross our minds as an option. Elouise received the heel prick and away we went.

Our moments at home were stressful, both good and bad. The good stress was having our new little person home! The bad stress was having a new little person home that always seemed starving and fussy! I was nursing her, as I had with our other children, and it seemed like she was never satisfied with a feeding. Something in my heart was telling me that something was not quite right. It was painful to make a decision to supplement her feedings. My husband, Aaron, fed her the first bottle. She sucked it down so quickly as if she hadn't eaten before! More red flags were tugging on my heart. So, we started supplementing her feedings and noticed a slight change in her demeanor.

In the next day or so, my mom, our three kids, and myself, went shopping for back to school supplies at our local store and my phone rang. That phone conversation is imprinted deeply into my memory and will not be forgotten. The woman on the other end asked if I was the mother of a newborn and asked if I had any contact with our pediatrician. We had our first appointments set up, but Elouise was only a couple of days old at this point. The woman introduced herself as working with the newborn screening office, and informed me that she was having difficulty locating my pediatrician. She asked if I had received any letters from the newborn screening office as of yet, which I hadn't. The next words dropped me to my knees right there in the aisle with pencils, notebooks, and binders. "Ma'am, did you know your child has tested positive for cystic fibrosis? Ma'am, it does not mean that your child has CF, just that they are probably a carrier and more testing needs to be done. Ma'am, could you verify your pediatricians' information?"

I spoke with the pulmonary lab staff to set up her sweat test within an hour of receiving the life-changing phone call from the newborn screen office. The woman who answered the phone was like an angel. She provided me with a sense of peace and hope. Because Elouise was born two weeks early, we had to wait until she was 42 (I believe) weeks gestational age for the test to be done. The pulmonary staff offered to be available to us at any time during our wait. I also had an acquaintance whose child has CF. She too was a voice of reason and hope. These two women, along with family, friends, and faith kept us looking at the joy that Elouise brought to our family instead of focusing on the doom and gloom of a CF diagnosis.

I am not sure what day we got the official news; the day doesn't matter, it's what we have done moving forward that does. First and foremost, I didn't bother to read anything more than what her pulmonologist provided us. Elouise was about six months old when I was ready to visit cff.org. Once I was ready, the information wasn't overwhelming. We allowed family and friends (and even some strangers) to be a part of anything they were willing to be. My sister created a health binder in order for me to take detailed notes of each appointment. My mom was and still is my second chain of command. She rarely misses one of Elouise's appointments at U-M. I think it's very helpful to have a second adult in the exam room. I would pass Elouise to my mom so that I could pay complete attention to her specialists. We would have struggled through her first year without everyone.

We still look at each day as something to be thankful for, but not because our daughter has CF, because life is short for all of us. I like to think the grass is green all around us and we were put here together to laugh, play, and enjoy one another. We still have moments when sadness sets in, then Elouise runs by laughing and the sadness is long forgotten.

TEAM UPDATES & INFORMATION

SUPPORTING RESILIENCY IN OUR PATIENTS AND FAMILIES

By Jennifer Butcher, Ph.D.

Merriam-Webster defines resiliency as "the ability to become strong, healthy, or successful after something stressful happens." For those of you who attended or watched online as Evin Green spoke at our Family Education Night in April 2015, you saw a wonderful example of resiliency in action. During his talk, Mr. Green spoke of how his family strengthened his resiliency from a young age as he faced the challenges of growing up with CF. His talk made many of us providers in your CF Center reflect about additional ways that we can help support resiliency in our patients and families. The timing of these reflections matched well with the CF Foundation's recent initiatives to support adaptive CF self-management and positive mental health among individuals with CF and their families. As a result, we will be offering several new programs through our CF Center focused on strengthening resiliency. Some of these programs are described in more detail in other parts of this newsletter, but here is a snapshot:

1- Yearly mood, anxiety, and behavioral screening for all patients with education and treatment recommendations provided if needed.

2- Yearly family psychosocial screening to help to support family members as they care for loved ones with CF.

3- Clinic program designed to reduce stress related to medical procedures likes throat cultures.

4- Interventions designed to promote adaptive management of CF daily care.

5- Program focused on helping adolescents successfully transition to adult CF care.

6- Developing innovative ways to provide quality psychosocial interventions to our patients such as using telemedicine.

We hope that these new programs will complement the high-quality medical care that our program strives to provide to achieve our CF Center goal of caring for the whole patient.

Below are some tips for our families with children on how to teach resiliency provided by the American Psychological Association Resilience Guide for Parents and Teachers. For more information, please visit: apa.org/helpcenter/resilience.aspx.

1- Make connections – Help your child build strong friendships and family and community networks for support during stressful times.

2- Help your child by having him or her help others – Helping others can empower your child.

3- Maintain a daily routine – Routines are comforting for children, especially during stressful times.

4- Take a break – Teach your child how to manage worries and to take a break from things that are troubling him or her.

5- Teach your child self-care – Make yourself a good example and help your child learn balance with a daily schedule that includes fun and relaxing time.

6- Move toward goals – Teach your child to set reasonable goals and work toward them one step at a time. Praising your child for meeting small goals will help him or her learn to keep moving forward even when faced with difficulties.

7- Nurture a positive self-view – Remind your child of difficulties that he or she has overcome in the past and that these challenges build strength to face future challenges.

8- Keep things in perspective and maintain a hopeful outlook – An optimistic outlook enables your child to see the good in life and keep going during hard times.

9- Look for opportunities for self-discovery – Tough times are often the times when children learn the most about themselves.

10- Accept that change is part of living – Change can be scary for anyone, but help your child see that change is part of life and how new goals can replace goals that become unattainable.

TEAM UPDATES & INFORMATION

NURSING UPDATES

By Cathy Enochs, BSN, RN, A-E
RN Center Coordinator

Pharmacy "Keywords"

Having trouble getting medications from your pharmacy? You can always call your pulmonologist's office for assistance, but some of these keywords may help:

- "On File" – When your nurse or doctor told you they already sent a refill to your pharmacy, but you still cannot fill it: Sometimes the prescription is "on file." Since it is a "new" prescription, it has a new number and is not attached to your empty prescription number. Ask your pharmacist if there is a prescription "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: Likely this needs a "co-pay override" from Medicaid. The Pulmonary Office cannot obtain this sort of override, but your pharmacist can call Medicaid and get the override.
- "Specialty Medication" – Some medications are costly and have been placed in a "Specialty Medication" classification. Your insurance may require that you use a specific specialty pharmacy. Most are mail-order pharmacies and will deliver to your home, but this could be a new pharmacy to you.
- "Prior Authorization" – Prior Auths (PA) take 48-72 hrs for your doctor's office to process. Insurance can take one to 14 days. Urgent PAs have 72 hrs for insurance processing. If you need a PA and you are almost out of that medication, please call your doctor so we can expedite the PA. Refilling as soon as the insurance and pharmacy allow can provide extra time for processing. Most allow you to fill five to seven days before your refill is "due."

Medication Cost Too High?

- "CSHCS" – Medicaid Children's Special Health Care Services (CSHCS) can take care of the co-pay. Contact your doctor's office for more information, or call CSHCS Family Phone Line at 1-800-359-3722.
- "CFF Compass" – The Cystic Fibrosis Foundation has relaunched the Patient Assistance Resource Center as Compass. They can work with you, your insurance, your pharmacy, and your care center to assist in many ways. Compass helps with medication issues, insurance, and financial concerns, as well as legal issues. Visit cff.org for more information.
- "Assistance Programs" – CSHCS may not be cost-effective for some patients and families. The CF Foundation has recommended use of the Healthwell Foundation for patient assistance. Those with Medicaid plans are not eligible for assistance programs. Visit their website for eligibility and more information: healthwellfoundation.org/cystic-fibrosis

Transferring a Prescription

- Call the pharmacy where you want the prescription filled
- Ask them to transfer the prescription from your usual pharmacy
- Provide the medication name, usual pharmacy's name, and phone number
- Be sure the pharmacy has your insurance information
- You can transfer only one fill or all remaining fills

Want to See Your Lab Results?

Sign up for the My U-M Health Portal. Your U-M labs and tests are viewable via this portal. Results have an intentional delay before releasing to the portal to allow your care team to contact you if needed. Children 11–18 years old have restricted parental views, which do not allow labs to be seen by proxy. You can send messages to 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 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.

ANNOUNCEMENTS/UPDATES

SAVE THE DATE

University of Michigan Health System

Cystic Fibrosis Center

Presents

CF Family Retreat

Wednesday, April 20, 2016 • 3:30–8 p.m.

*** NEW THIS YEAR***
MEET AND GREET AT 4 p.m.
with snacks and drinks
You can meet Family Advisory Board Family Members

**

Complimentary Dinner and Presentation for CF parents and adult relatives at:
Kensington Court, 610 Hilton Blvd, Ann Arbor, MI 48108

**

RSVP to Cheryl Evans, Event Coordinator
by email: UM-Peds-CFCenter@med.umich.edu
or by phone (734) 936-1755 by April 1, 2016.
Thank you in advance.

Because of risks to people with CF, no people with CF should attend this event apart from our speaker, Meagan Tenyer, who has CF.

GREAT STRIDES 2016

What is Great Strides?

Great Strides is the Cystic Fibrosis Foundation's largest national fundraising event. Each year, more than 125,000 people participate in hundreds of walks across the country to support the Foundation's mission to cure cystic fibrosis. Last year, the Michigan Chapter's Great Strides program raised an amazing 1.5 million dollars!

Where can I walk?

Walks are held at nearly 500 locations nationwide and are open to the public. This year, we are holding 14 walks in the State of Michigan alone! The walks take place statewide so there are plenty of opportunities and ways to get involved.

Why should I walk?

Simply put, to make a difference in the lives of those living with cystic fibrosis. The funds raised from Great Strides helps provide people with CF the opportunity to lead full, productive lives by funding research and drug development, promoting individualized treatment, and ensuring access to high-quality, specialized care.

Where do I start?

Visit fightcf.cff.org to register your team today and start recruiting family, friends, students, and co-workers! Whether you are collecting money for a jeans day at work, selling home-grown flowers at your local CF Care Center, or holding a bake sale at your school, every fundraising dollar counts!

We are lucky to have a staff at the University of Michigan Care Center who not only take care of CF patients, but who also go above and beyond in their fundraising efforts. We cannot thank our volunteers and donors enough for their continued support in our mission to find a cure. Although we have made great strides thus far, we will not stop until we have found a cure for all patients living with CF.

We hope you'll join in the excitement and get involved by walking with us in 2016!

This year we'll be walking toward a cure in:

Ann Arbor	Davison	Detroit (Zoo)	Edmore
Findlay, OH	Frankenmuth	Fremont, OH	Grand Haven
Grand Rapids	Hartland	Kalamazoo	Lansing
Ludington	Mt. Pleasant	Port Huron	Rochester
Toledo, OH	Traverse City		

For more information, or to register your team and begin fundraising, please visit ightcf.cff.org or call the Michigan Chapter, Serving Michigan Northwest Ohio at (248) 269-8759.

Understanding Yearly Labs

By Amy Filbrun, M.D.
Associate Director of CF Center

The Cystic Fibrosis Foundation patient care guidelines state that CF patients should be seen at least quarterly. These guidelines also recommend that laboratory tests are evaluated at least yearly. We thought it would be a good idea to explain what these labs measure, so you can understand why they are so important.

A complete blood count with differential (CBC with diff) is a blood test used to evaluate your overall health and detect a wide range of disorders, including anemia. A CBC with diff measures several components and features of your blood, including: red blood cells, which carry oxygen, white blood cells, which fight infection, platelets, which help with blood clotting, and hemoglobin, which is the oxygen carrying protein in red blood cells.

- If red blood cells or hemoglobin are low, you have anemia. Anemia causes fatigue and weakness. Anemia has many causes, including low levels of certain vitamins or iron, blood loss, or an underlying chronic condition.

- If your white blood cell count is higher than normal, you may have an infection or inflammation. This is pretty common in CF. A high white blood cell count can also be a reaction to medication, but certain medications also can cause white blood cell counts to drop.

- A platelet count that is lower than normal is called thrombocytopenia, and is often a sign of an underlying medical condition (sometimes CF-related liver disease), or it may be a side effect from medication. Higher-than-normal platelet counts are less specific, and are often seen with acute illnesses or inflammation.

The comprehensive metabolic panel (CMP) is used as a broad screening tool to evaluate and check for conditions such as diabetes, liver disease, and kidney disease. The CMP may also be ordered to monitor known conditions, such as hypertension, and to monitor people taking specific medications for any kidney- or liver-related side effects. Some of the specific tests measured with a CMP include:

- Glucose, a type of sugar used by the body for energy. Abnormal levels can indicate diabetes or hypoglycemia (low blood sugar).

- Calcium, which plays an important role in muscle contraction, transmitting messages through the nerves, and the release of hormones. Elevated or decreased calcium levels may indicate a hormone imbalance or problems with the kidneys, bones, or pancreas.

- Albumin and total blood protein, which are needed to build and maintain muscles, bones, blood, and organ tissue. Low levels may indicate liver or kidney disease or nutritional problems.

- Sodium, potassium, carbon dioxide, and chloride (electrolytes), which help regulate the body's fluid levels and its acid-base balance. They also play a role in regulating heart rhythm, muscle contraction, and brain function. Abnormal levels also may occur with heart disease, kidney disease, or dehydration.

- Blood urea nitrogen (BUN) and creatinine, which are waste products filtered out of the blood by the kidneys. Increased concentrations in the blood may signal a decrease in kidney function.

- Alkaline phosphatase (ALP), alanine amino transferase (ALT), aspartate amino transferase (AST), and bilirubin; ALP, ALT, and AST are liver enzymes; bilirubin is produced by the liver. Elevated concentrations may indicate liver dysfunction. We also measure gamma-glutamyl transferase (GGT), which is another test that helps detect liver disease and bile duct obstruction.

PT stands for Prothrombin Time and PTT stands for Partial Thromboplastin Time. A PT test may also be called an INR test. INR (international normalized ratio) stands for a way of standardizing the results of prothrombin time tests, no matter the testing method. So your doctor can understand results in the same way even when they come from different labs and different test methods. In some labs, only the INR is reported and the PT is not reported. These are measurements of how quickly a blood clot forms. These tests may be ordered when people present with unexplained bleeding or bruising or to check for chronic conditions such as liver disease.

- Prothrombin, or factor II, is one of the clotting factors made by the liver. Blood clotting factors are needed for blood to clot (coagulation). Vitamin K is needed to make prothrombin and other clotting factors. Prothrombin time is an important test because it checks to see if blood clotting factors (factors I, II, V, VII, and X) are present. An abnormal PT is often caused by liver disease or injury or by treatment with blood thinners.

- PTT measures other clotting factors. This test is affected (elevated) by heparin, so may not be accurate if drawn from a catheter or port that contains heparin. This test is often done at the same time as the PT to check for bleeding problems.

Vitamins are needed to help your body grow, function, and fight off infection. People with CF need extra vitamins for good general health. In particular, people with CF have trouble absorbing vitamins A, E, D, and K, the fat-soluble vitamins.

- Vitamin A many roles in health, including normal vision and bone and tooth formation. It also works to help fight infections and to keep the intestines healthy. Vitamin A is found in eggs, liver, tomatoes, milk, and some fruits and vegetables.

- Vitamin D helps build and maintain strong bones and teeth. Without enough vitamin D, bones can become thin and brittle. People with CF are at risk for bone problems. So it is important to get the right amounts of vitamin D every day. Vitamin D comes from two sources. It is made by the skin when it is exposed to sunlight, and it is found in the foods we eat. It helps your body absorb calcium, and helps move calcium from your blood to your bones. While foods such as eggs and cereals contain vitamin D, the main source is milk.

- Vitamin E is an antioxidant, which means that it protects compounds in the body from combining with oxygen. When compounds become oxidized, they become harmful to the body. Vitamin E helps keep red blood cells healthy. It also helps fight infection and maintain the health of the intestines. Foods that contain vitamin E include plant oils, margarines, and some fruits and vegetables. Diet alone cannot prevent a vitamin E deficiency in people with CF, therefore a supplement is usually recommended.

- Vitamin K is best known for its role in helping blood clot. It also helps keep bones healthy. Most of the vitamin K we need comes from our diet. It is found in green vegetables, plant oils, and margarine. We also have healthy bacteria in our intestines that produce vitamin K. Even though the bacteria in your intestines is healthy, it can be destroyed by the antibiotics you may take for a lung infection. This can affect the amount of vitamin K you get on a daily basis.

An oral glucose (sugar) tolerance test (OGTT) is used to screen for Cystic Fibrosis-Related Diabetes (CFRD) or Impaired Glucose Tolerance (IGT). During the test we measure how your body removes glucose from the blood. First, you will need to fast overnight. When you get to the lab, your blood is drawn to measure your fasting blood glucose level. Next, you drink a liquid that is high in sugar. Over the next two to three hours, the lab technician will be checking your blood glucose levels (timing of draws may vary in different centers). The CF care guidelines for CFRD recommend that people with CF ages 10 and older be tested every year for CFRD with an oral glucose tolerance test (OGTT). The OGTT is the best way to diagnose CFRD.

We hope you now have a better understanding about the tests that we, and the CF Foundation, recommend. If you have questions, or would like more information about any of the blood tests that we order, please feel free to speak with your CF doctor or nurse at an upcoming visit. The importance of annual labs in your health and well being cannot be overstated!

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