

Living As Long As Moses: A CF Story

By Rod Spadinger

Some on the outside of our CF community may view having cystic fibrosis as a curse, but those who hold that mindset do not know us. This is my CF Story.

Despite living in Honolulu, Hawaii, I was introduced to Dr. Robert Kramer in Dallas, Texas, after becoming critically ill at age four. He provided me with my first lease on life. In 1976, exercise was not seen as the CF elixir of life, as it is now. Nevertheless, in order to keep me comfortable, Dr. Kramer enjoined my parents to make me run daily at the age of four. Under the threat of sanctions that would limit my viewing of Sesame Street, I ran for at least five miles a day, every day, which eventually led me to compete in the Cross-Country National Junior Olympics from the ages of seven through nine, and in track events held in Finland and in the USSR upon high school graduation, despite being diagnosed with multiple sclerosis at 17. I

ceased running as my MS diagnosis initiated a slow but steady spiral in my health. In 2012, 23 years later, my PFTs hovered around 25%. Despite this, I was convinced that a transplant would never be a necessity for me as I expected to live forever.

“After an emotional half-hour discussion, we agreed that I would fly to Texas in a number of days to be formally listed for transplant.”

Even so, I met my Kryptonite in the form of a five-day trip that began with a 25-hour Alaskan experience, followed by visits to Seattle and San Diego in December 2018. That trip proved to be the turning point for my lungs. During the months following, I was admitted for multiple hospital stays. The doctors and nursing staff at Straub Hospital were quickly becoming my second family. I remember

telling a coworker during that time that I had finally reached the level where my lung functions would no longer improve after each hospitalization; rather, they would decline slowly over time. I still maintained I would never die, that I was going to live lon-

ger than Moses.

In late July 2017, my lung function was 17%. During one of my appointments, my CF doctor told me that if I didn't leave Hawaii by that weekend to receive a transplant in Texas, there was not much more he could do for me. After an emotional half-hour discussion, we agreed that I would fly to Texas in a number of days

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EDITOR'S NOTES

Summer is in full swing! And with it, at least here in Texas, the 100-degree temperatures. This issue we're focusing on a hot topic—diversity of all kinds in the CF community. One of our directors, **Tré LaRosa**, expresses his views on the Black Lives Matter movement and what we can do to further inclusion within the CF community. We're also featuring a story about being trans in the CF community as well as a story from an African American man chronicling his experience in the CF community as a Black man. **Tannaz Motevalli** details her story growing up as a queer person of color with CF. **Teena Mobley** talks about living her best life regardless of her skin color, while **Ashley Wilson** details her own experiences within the CF community, as both an individual of mixed race and a person of color. **Dr. Xan Nowakowski** talks about the importance of sex identity awareness within the healthcare community. **Daniel Gonzalez** relays his experiences with limited healthcare resources growing up in a different country. In that same vein, **Isa Stenzel Byrnes** writes about the challenges we all face right now and how those challenges are especially difficult for racial minorities and/or underprivileged individuals.

We're excited to welcome former LMK scholarship winner, **Grace Knight**, to our growing board of directors. You can read her story on page 37. You'll also find **Andrea Eisenman's** review of Tim Wotton's memoir on cheating death. In the "Voices From The Roundtable" section we're featuring an interview with Mohamad Elsayyed, a Muslim Palestinian refugee living near Beirut, Lebanon, who is the lucky recipient of a vest donated by the CF Vests fO'r Life foundation. You can read the backstory about this foundation and one of the co-founders in the cover story by **Rod Spadinger**. You can also read about **Andrew Kirby's** journey with sobriety and **Laura Mentch's** work with the Cystic Fibrosis Reproductive and Sexual Health Collaborative.

Beth Sufian breaks down the Family and Medical Leave Act (FMLA) in her column "Ask The Attorney." **Molly Pam** talks about diversity in family and what growing a family means in her column. **Lara Govendo** writes about the importance of unity and truly listening to each other in a time when emotions run high. You can read about tips for managing your CFRD during these hot summer days and backyard BBQs in **Kat Porco's** column this issue. **Kathy Russell** writes about the many rewarding changes since starting Trikafta. **Dr. Julie Desch** lists various tools for controlling the fight-or-flight response when processing how we experience CF. **Dr. David Gudis** answers questions about best practices with an ENT visit during COVID-19.

In our "In The Spotlight" segment this issue we interview Nathan Tallent, a man with multiple degrees who absolutely loves guest services in the hotel industry. He talks about his incredible work-life balance and his passion for helping others.

Enjoy this summer issue! In the words of Effie Trinket from *Hunger Games*, may the odds be ever in your favor, Sydna.

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Information From The Internet...

Compiled by Laura Tillman

A Newly Discovered Disease May Lead To Better Treatment Of Cystic Fibrosis

Researchers have discovered a novel disease that might lead to a better understanding of cystic fibrosis and new treatment options in the future. The cause of cystic fibrosis are mutations in the cystic fibrosis transmembrane conductor regulator gene (CFTR). The researchers discovered a new disease that is caused by defects in another chloride channel, TMEM16A. This channel is also present on the surface of airway cells. The team evaluated the cellular effects of the disorder that is caused by a total loss of TMEM16A function. Surprisingly, they discovered that not only TMEM16A but



LAURA TILLMAN

also CFTR is not functional in these patients. Patients with TMEM16A deficiency don't have any respiratory symptoms at all. A loss of CFTR function due

to lack of TMEM16A does not lead to clinical symptoms of cystic fibrosis. Taken together, these results raise an intriguing question: Could the pharmacological inhibition of TMEM16A improve the respiratory symptoms of patients with cystic fibrosis? A significant reduction of mucus production and secretion as a consequence of TMEM16A inhibition has previously been shown under laboratory conditions. Thus, potent and well-tolerated TMEM16-inhibitors, which have FDA-approval for other diseases, should be further examined in preclinical and clinical studies to be used in CF lung disease and other inflammatory airway diseases.

<https://tinyurl.com/y7kfm9wn>

AND

<https://tinyurl.com/ybaql97c>

AND

<https://tinyurl.com/yd88vmvf>

For CF, Promising Antibiotic Developed For Drug-resistant Lung Infections

Continued on page 9

LOOKING AHEAD

Please consider contributing to *CF Roundtable* by sharing some of the experiences of your life in writing. Read the Focus topics listed below and see if there are any about which you might like to write. In addition, humorous stories, articles on basic life experiences, short stories, artwork, cartoons, and poetry are welcome. We require that all submissions be original and unpublished. With your submission, please include a recent photo of yourself as well as your name, address, and telephone number. Photos will be returned. Email all submissions to: cfroundtable@usacfa.org. Or go to our website: www.cfroundtable.com/newsletter.

Summer (August) 2020: Diversity in the CF Community. (Current issue.)

Autumn (November) 2020: People with CF Who Have Started A Business. (Submissions due September 15, 2020.) What type of business have you started or are planning to launch? Was it geared toward the CF community or to the general population? What has been helpful? And what have you learned from your experience?

Winter (February) 2021: Late Diagnosis of CF. (Submissions due December 15, 2020.) How did your CF diagnosis help you with existing, untreated health issues? What acceptance or resistance have you experienced within the CF community? What resources have helped you in processing your diagnosis?

Spring (May) 2021: Sexuality and Sexual Health in CF. (Submissions due March 2021.)



ASK THE ATTORNEY

Pandemic, Workplace Rights, And Cystic Fibrosis®

Family And Medical Leave

By Beth S. Sufian, J.D.

The Family Medical Leave Act ("FMLA") became law in 1993. Some individuals with CF have recently used FMLA to obtain 12 weeks of leave from work because of a need to take care of themselves. Others have been given inaccurate information regarding the ability to take FMLA leave to care for oneself, a child, or a parent who has CF. Some employers have confused the requirements for Public Health Emergency Leave ("PHEL"), a new type of leave, with the regular FMLA criteria.

The information provided below is not legal advice about a specific situation and is only meant to be legal information. If you have questions, please contact the CF Legal Information Hotline at CFLegal@sufian-passamano.com or call 1-800-622-0385.

Q1: Is it true some employers do not have to provide FMLA leave?

A1: Yes, not all employers have to provide FMLA leave. Employers who are covered by FMLA leave:

(a.) Private employers with 50 or more employees (during 20 or more work weeks in the current or preceding calendar year); and

(b.) Public employers (such as local, state, or federal government agencies); and

(c.) Public or private elementary or secondary schools.

Q2: Are all employees working for a covered employer able to receive FMLA leave?

A2: To be eligible for FMLA leave, the employee must meet the following criteria:

(a.) Employee must work for a covered employer; and

(b.) Employee must have worked for the employer for at least 12 months prior to requesting leave; and

(c.) Employee must have worked at least 1,250 hours during the 12 months immediately preceding the request for leave; and

(d.) Employee must work at a location where the employer has at least 50 employees within 75 miles.

Q3: Can I take FMLA leave because I want to avoid the risk of contracting COVID-19?

A3: Employees are permitted to take FMLA leave for a specific qualifying reason only. The employee may request leave to care for their own serious medical condition, the serious medical condition of a family member, or to care for a newly arrived child.

FMLA does not require an employer to approve FMLA to avoid the risk of

becoming infected with a virus. An employee can always ask an employer for unpaid leave even if the FMLA requirements are not met. However, the employer is not legally obligated to provide the leave.

Q4: What are qualifying reasons for FMLA leave?

A4: An employee eligible for FMLA leave may request leave for any of the following qualifying reasons:

(a.) Employee needs to care for the birth or placement of a son or daughter; or

(b.) Employee needs to care for his or her spouse, son, daughter, or parent with a serious health condition; or

(c.) Employee needs to care for themselves and has a serious health condition; or

(d.) Employee qualifies for the Military Family Leave Provisions.

For purposes of the FMLA, "serious health condition" is defined as an illness, injury, impairment, or physical or mental condition that involves inpatient care or continuing treatment by a healthcare provider. According to U.S. Department of Labor guidance, a person who needs to perform daily medical treatments to prevent incapacity is considered someone who has continuing treatment by a healthcare provider. See FMLA regulations §825.113, 825.114, 825.115.

Q5: I have CF and I have child who is five years old who does not have CF. This summer his day care is closed because of COVID-19. Can I take FMLA leave for the next 12 weeks?

A5: FMLA leave can be taken if there is a need to care for a child with a serious health condition. If the child does not have a serious condition, and you are not able to show you cannot work due to your serious health condition,



BETH SUFIAN

then the FMLA requirements will not be met.

However, under the Families First Coronavirus Response Act (“FFCRA”), Congress amended the FMLA to create a new form of leave, PHEL, to address the COVID-19 pandemic. PHEL is available when:

(a.) Worker is employed by an employer with 500 or fewer employees.

(b.) Employee has worked at least 30 days for the employer.

(c.) Employee is unable to work due to need to care for a minor child because the child’s school is closed (or childcare is unavailable) due to emergency declaration by federal, state, or local authority related to COVID-19.

(d.) Healthcare providers and first responders are excluded.

(e.) A small employer under 50 employees can elect not to provide PHEL if they can show granting the leave would jeopardize the viability of the business enterprise.

(f.) The first two weeks (or 10 work days) of PHEL are unpaid. After the initial 10 days, employer must pay the employee on PHEL 2/3 the employee’s regular compensation rate for regular number of hours. Paid PHEL is capped at \$200 per day and \$10,000 aggregate for the whole leave period.

(g.) PHEL can be up to 12 weeks of leave, but it could end sooner. If the qualifying reason for PHEL ends (no longer caring for minor child whose school or childcare is closed due to COVID-19), the leave will also end.

(h.) PHEL was adopted by Congress to address the COVID-19 emergency and the statute expires on December 31, 2020.

Q6: I asked for FMLA, but my employer says that since FMLA is unpaid I could just quit and be rehired when I am ready to return. Why do I need FMLA leave?

A6: Although federal FMLA is typically unpaid (unless there is a state law that

provides paid leave), there are important protections that come with FMLA leave:

(a.) Employee on leave retains their status as a current employee during a period of FMLA leave.

(b.) As a current employee, the worker remains enrolled and eligible to receive employee benefits, such as employer-sponsored health insurance.

(c.) At the end of the FMLA leave period, the employee is entitled to return to the same, or substantially similar, position that they held when they requested the FMLA leave at the same rate of pay and with the same benefits.

Q7: My employer granted my request for FMLA but says that I must use my accrued paid time off (like sick leave and vacation time) during FMLA leave. I want to save my paid time off for when I need it. Can the employer make me use accrued paid time off as part of my FMLA leave?

A7: Yes. Leave taken under federal FMLA is unpaid leave. However, the employee may use, and the employer may even require the employee to use, any amount of accrued paid time off (such as sick leave, vacation leave, or other paid leave) as part of the FMLA leave period.

Although the federal FMLA does not provide for paid leave, some states have adopted their own version of family and medical leave which do provide for paid leave. Some states have state laws that cover smaller employers. Google your state name and the words “paid family leave” to find out if your state provides paid leave under state law. ▲

Beth is 54 and has CF. She is an attorney who specializes in disability law and is the President of USACFA. Her contact information is on page 2. You may contact her with your legal questions about CF-related issues at CFLegal@sufianpassamano.com.



What is the Boomer Esiason Foundation?

In 1993, NFL Quarterback, Boomer Esiason, learned that his son, Gunnar, was diagnosed with the incurable genetic disease cystic fibrosis (CF). Never ones to back down from a fight, he and his wife, Cheryl, founded BEF and decided then and there to fight for a cure and for the cystic fibrosis community.

Cystic Fibrosis is an inherited chronic disease that affects the lungs, digestive system, and reproductive system of about 30,000 Americans by causing a thick build-up of mucus that leads to blockage, inflammation, and infection.

What does BEF do?

In addition to assisting the CF community with the following programs, we also support CF clinics and research centers:

- Educational Scholarships
- Lung Transplant Grant Program
- Team Boomer
- Jerry Cahill’s Cystic Fibrosis Podcasts & Wind Sprints
- Breathe In Podcast
- CF Patient Disaster Relief Program
- CF Step-by-Step Video Series
- Gunnar Esiason Blog
- Tru Heroes Nursing Program
- You Cannot Fail Hospital Bags
- CF Education Days & CF Speaking Engagements

www.esiason.org



SPIRIT MEDICINE

The Spirit of Unfairness With CF Across The Globe

By Isabel Stenzel Byrnes

All around the globe, people are enduring the consequences of the COVID-19 pandemic. For the first time in human history, we are all together, joined in the same cause—protecting human health, wearing masks, and focusing on priorities. While we all appear to be in the same boat, we are, in fact, not. The poor and economically marginalized are at much higher risk of catching this disease and dying from it. And, this June, our country is facing another devastating fight against social injustice and racial inequities. Life is challenging for everyone, due to uncertainty and insecurity, and even more so for the underprivileged and for racial minorities.

There are aspects of these circumstances that are parallel to living with cystic fibrosis. All around the globe, young people and their families are struggling to survive with CF. Like the disparities in the United States, the disparities in CF care and access to medication are vast and heartbreaking. Life expectancy is dramatically lower in developing countries. When I was getting listed for a transplant, I heard about a woman whose daughter in Japan was fighting CF. She did not have proper enzymes and the chance of getting a lung transplant was almost zero. She looked like she was 10 years old, but she was 18—the oldest known child with CF living in Japan. She died in 2004. My heart broke for the unfairness of her situation in this wealthy, advanced country.

I've known people with cystic fibrosis from all over the world—my friend

Mette in Norway, my friend Sandra in Spain, my friend Akihiro in Japan, and my friend Fabian from Germany. I have been blessed to meet friends with CF who are from Turkey and Saudi Arabia. There is something comforting to know that we Americans are not the only ones facing this disease. It has been fascinating to learn how certain countries care for their CF patients—with nationalized health care, the focus is more aggressively on preventative care rather than exacerbation care. And it has been tragic. In some countries, there is little that

can be done for CF patients. In families with limited resources, parents with many children simply cannot give their CF child the care and focus needed since their healthy children—the ones with a chance—must be prioritized.

My sister Ana, who was a genetic counselor and had CF, gave a talk on CF genetics at CFRI in 2002. She shared that the deltaF508 gene evolved 52,000 years ago. She joked that, in fact, we CFers did have a common ancestor and we really are all a family. Sadly, though, our health and survival

rates are determined by our location. Some of us are privileged to be born in countries where CF care is available. This country's medical system is far from perfect, but we are given a chance.

Though our country has a long way to go, in general we are also privileged to have a culture that supports

equality of opportunity, despite our health status. In some cultures, kids with CF cannot go to school. In others, patients cannot challenge their doctors. In many cultures, women are valued for their reproductive potential; having CF dramatically reduces their worth. People with CF do better with exercise; but, in some cultures, girls aren't even allowed to ride a bicycle! In some cultures, illness is shameful and cannot be shared outside of the home. Every day I thank God I was born with CF in the United States in 1972, at a time when opportunities were unfolding for a normal life for people with disabilities.

As a half-Japanese, half-German CF patient, I've always looked different from

Sadly, our health and survival rates are determined by our location. Some of us are privileged to be born in countries where CF care is available.



ISABEL STENZEL BYRNES

my CF peers who are mostly Caucasian. I've known a handful of CF peers who are African American or Hispanic. I've only met one other half-Asian child with CF. The beauty of this community is that I've always felt included, welcome, and equal to my CF peers. I've been embraced and loved dearly. This has done wonders for my self-development. I cannot even imagine discrimination in the CF community! I think it is because the common struggle of fighting CF and all the mental health and social consequences far outweighed any differences perceived by race and background. If only others were so lucky.

In the last few weeks of social unrest, I've talked to many elders and people with chronic health conditions who feel guilty that they cannot do more to attend the anti-racism and police brutality protests. We talk about being passive bystanders, yet want to do more for people impacted by inequality. The same holds true in the CF community, not just our global community. Even in the U.S., there are so many people with CF who do not have enough money for their copayments or the extra calories they need. There are people with CF who are uninsured, or working in high-

risk environments, or who have lost jobs. Life is so insecure for so many. It is overwhelming.

Meanwhile, I'm blessed to work from home and am both secure and relatively healthy 16 years post-transplant. I know of many CF families in the U.S. struggling with poverty, especially those who are persons of color. How do I live with such privilege while others are so much less fortunate? How do we live life without feeling weighed down by guilt? The emotion of guilt can motivate me to do something, but it can also bring me down. One thing I know is that we are all just trying to survive. There is so much inequity and each of us can only do so much to make a difference. Today, while sheltering in place, I wish I could join the courageous protestors. I wish I didn't have to worry about my health. But I cannot. I can only applaud the protestors' efforts, bless, and support them with my heart, talk about these issues with people in my life, and support these causes with my wallet. And, I can vote.

Life with CF is full of limitations. I wish I could help more people with CF across the globe, but I appreciate what Jana Stanfield says: "I cannot do all the

good that the world needs. But the world needs all the good I can do." What good can I do to cope with the unfairness? I practice kindness to all. I honor the life blessings I have received by taking as good care of myself as possible. I seize opportunities to make a small difference for those less fortunate, including donating to international CF causes. I send positive energy and prayers to those with CF who are less fortunate in the world. I learn a language so I can communicate with my CF peers in Japanese and Spanish. I connect intentionally with racial minorities with CF, knowing we are a small bunch in this club. Mostly, I thank God for the gift of life for all. I can hope that all people with CF, across all corners of the globe, can cherish the positive experience of life no matter how short life is or how full of suffering their lives may be. Isn't that what we all try to do? ▲

Isabel Stenzel Byrnes is 48 and has CF. She lives in Redwood City, California, and is happily married to Andrew Byrnes. She loves to hike, play the bagpipes, and be a slow triathlete. Her email is isabear27@hotmail.com.

An Update On The Speakers Bureau

We relaunched the Speakers Bureau last fall. So far, in 2020, we have delivered five talks. We are proud of our ability to roll with the punches as we've had to do since COVID-19 has forced us to suspend in-person speeches for the calendar year. One of our directors gave a speech to Translate Bio, Tré LaRosa spoke to Integrated Genetics, Ella Balasa spoke to Corbus Pharmaceuticals and Translate Bio, and Isabel Stenzel Byrnes spoke to Walgreens.

Our Speakers Bureau is able to speak on a far-reaching range of topics, from daily living with cystic fibrosis, working with CF, going to college with CF, legal measures and CF, mental health, grief, struggling with clinics and/

or adherence to treatments, and just about anything else you can think of. We are proud of our ability to sponsor our speeches for companies that are looking to host and better understand the patient experience. We are also thrilled with our ability to educate clinics, research organizations, or just about anybody else. It is our belief that we when we tell our stories, we improve the world for others with CF, and, hopefully, we encourage a bit more compassion in the world.

Please let us know if you would like to request a speaker—or, if you have CF, if you'd be interested in joining our Bureau! <https://www.cfroundtable.com/speakers-bureau>



SPEEDING PAST 50

An Alien No More

By Kathy Russell

We've made it through another winter. Hooray! Here in Oregon we were so fortunate. We had a very mild winter with only about a half-inch of snow in total and only a few days of quite cold weather. Also, the east wind, which can be so treacherous, blew for only a few days. The low amounts of precipitation, which ultimately left us a little short for the water year, were the only hiccup we encountered. I hope we will be able to make up the rainfall shortage this spring and early summer.

So, we got through winter only to then get news of the dreaded COVID-19. I don't even want to talk about that. Fortunately, my husband, Paul, and I have both been healthy. We have been staying at home, except for necessary outings to get food and to keep some doctor appointments. I was able to meet with my pulmonologist via teleconferencing. Since I am doing so well, that was all that was needed.

As I mention "doing so well" I need to elaborate. In March, I changed from taking Orkambi to taking Trikafta and I haven't felt this well since I was in my 50s! I am able to live as a normal person of my age. I still have arthritis and still have to use supplemental oxygen but have been able to lower my flow rate to only 1 liter/minute. In the past, I have had to use oxygen at rates up to 15 liters/minute, so this is quite an improvement. I am delighted.

For most of my life I have felt as if I came from another planet and was an alien here. I remember when I was young I felt really different because of my "sweaty" hands and feet. If I said that I was sweating, some older person

would tell me that "horses sweat, males perspire, and females glow." I didn't glow; I sweat! It was so embarrassing to have hands that were wet always—even when they were cold.

My feet sweat so much that I dissolved leather shoes after just a few weeks of wearing them. In warm weather, when it was normal to go barefoot out of doors, my sweaty feet would look as if I never washed them because of how the dust would adhere to my wet feet. I would have mud between my toes, even though they had been clean when I went outside to play. My much

older brother used to call me Dirty Foot! I didn't appreciate his humor.

When I started in hospital nursing, I had to buy new work shoes about every six weeks. I was putting out so much salt that I had to take salt pills at the added rate of about 2 grams per day. I had to wear white shoes and my sweat degraded the white dye and made the leather crumble. I was so happy when I could start wearing clogs as nurses did in Europe. My feet could breathe and they stayed a little drier. I saved a lot of money on shoes after that.

My sweaty hands caused me embarrassment, too. I remember a time when I was in a production at a local fraternal meeting. Each of us who were participating was handed a 3" by 5" card with the words that we were to speak. After holding my

card for only the duration of the production—just a few minutes—it had almost dissolved. When the group leader asked us to hand back the cards, I could have died of embarrassment because my card was so damaged. It was practically unreadable. The leader looked at me as if I were some sort of lower life form. I don't remember what she said, but I remember how bad I felt.

I remember having terrible earaches when I was young. The pain was so bad that my daddy would pick me up and carry me in his arms as he walked out from our house across the front yard and to the mailbox, which was about a city block away from the house. He would sing "You Are My Sunshine" to me as he walked and he would hold me so firmly that I felt safe and all right. It didn't take away the pain, but I felt better. The earaches were caused by my sinus disease. I was

For most of my life I have felt as if I came from another planet and was an alien here.



KATHY RUSSELL

in my 50s before I got rid of the sinus disease and, believe me, I don't miss the terrible headaches that were caused by my sinuses.

Other things that made me feel like an alien when I was young were the coughing that would cause me to turn beet red and gag from the effort; the unpleasant troubles that CF guts can cause; the looks I got when I had to stop running or playing; the shortness of breath; the ever-present sinus problems and the runny nose that went with that; and so many other little "fun" problems that CF can cause. I missed so much school that I was always having to catch up. It seemed as if everything was trying to make me feel odd.

I was short and little. I could, at eight years of age, wear clothes that my older sister had worn when she was five. In school I was always the last person chosen for teams at recess and the first target of the ball in a game of dodge ball. I learned how to not care what others thought of me when I was pretty young. I decided that only peo-

ple I cared about could hurt my feelings. Also, they would be the only people whose opinion mattered to me. (That still is my way.)

Now, for the first time in my life, I feel as if I belong here. On my 76th birthday, within the first four weeks of starting Trikafta, I realized that I woke up alert and rested. I didn't feel as though I needed to take a nap as soon as I got up. I could breathe easily and was able to use my chest muscles to breathe instead of having to use my diaphragm to get air in. My brain was functioning at a much better level than it had been for years, and I was able to stay up and awake all day. This might not seem like much to some people, but it was a major positive change for me.

Another thing that has changed for the better for me is that I am enjoying cooking. I have been a cook since I was young and had gotten to the point that I was fixing meals that took very little effort and were pretty simple. Now I am pulling out favorite old recipes and enjoying the fixing of them. I bake bread at least once a week and am

loving it. I like to cook from scratch so that I know what we are getting in our meals. I am able to do that and don't mind the time that some recipes take.

I have more energy so that I can exercise more, too. I do at least 50 steps each day on a stair climber. This may not seem like much to younger folks who are active and healthy, but when you have been mostly inactive for years it is a really big deal. I also am able to walk more than a few steps without desaturating. Wahoo!

I don't know if this feeling will last, but I surely do hope that it will. If it doesn't, at least I have had several weeks of feeling like I am an alien no more.

Until next time, please stay healthy and happy. ▲

Kathy is 76 years old and has CF. She and her husband of 55 years, Paul, live in Gresham, OR. She is a past director of USACFA and served as President and Treasurer and was Managing Editor of CF Roundtable for several years. You may contact her at krussell@usacfa.org.

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A man-made antibiotic able to fight drug-resistant bacteria in the lungs with minimal toxicity to a person's cells may be a promising approach to eliminate lung infections, such as those caused by resistant *Pseudomonas aeruginosa*, that trouble people with cystic fibrosis (CF). The antibiotic molecule was tested in the lab and in mice with lung infections, where it effectively cleared the resistant bacteria without notable side effects. The findings warrant its clinical testing as an aerosol against multi-drug resistant bacteria in the lungs. Antimicrobial peptides (AMPs) are natural small proteins that the human body uses as a first line of defense against bacteria, disrupting their membrane even in cases of resistance to more conventional antibiotics.

But AMPs work only in specific circumstances, and are susceptible to degradation when used outside their optimal conditions. Modifying AMPs in a way that makes them less vulnerable to degradation and that further increases their antimicrobial activity has been widely investigated, and some of these synthetic molecules are now reaching clinical trials for specific infections. This is the case of an engineered cationic AMP (eCAP), named WLBU2. Aiming to improve WLBU2, researchers changed the molecule — basically, they constructed near-mirror versions of its amino acids (the building blocks of proteins). The researchers made several new versions of WLBU2, but the molecule that worked best contained eight flipped amino

acids, and thus received the name D8. The D8 molecule was found to retain the antibacterial activity of WLBU2, and also killed bacteria by breaking their membrane, but was much more stable than WLBU2 in the blood, which contain proteases (proteins that degrade proteins). This suggested D8 was less vulnerable to degradation. When the stability of a certain medicine increases, it remains longer in the body, which may increase its toxicity. But to the researchers' surprise, the D8 antibiotic was much less toxic to blood cells than WLBU2. The team is now exploring D8's potential as an aerosolized treatment for CF patients burdened with drug-resistant lung infections and for those developing

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FAMILY MATTERS

What Is A Family? Why Does It Matter?

By Molly Pam

“Family” is your blood. Well, it can be for some. It can also be chosen, constructed, and modified over time. I have two sisters, only one of whom shares my DNA. We “adopted” my other sister into our family, but not in the truest sense of the word—she’s not my biological sister. We grew so close that the bond we shared was the same bond I have with my biological sister. It is so tight that I can test the limits and know they will still be there for me. That is what makes some one family. It is the people you want to spend Thanksgiving with because you are thankful for having them in your life. For me, family are the people who are there for me, and I’m there for them, whether chosen or bonded by DNA, and that’s what matters.

In the CF community, we have people of multiple races and ethnicities, sexual identities, religions, and abilities. We also have, layered on top of these characteristics, further diversity within our community. We have different mutations that determine treatment options; a range of lung functions and body mass indices that are markers of disease severity; related conditions like ABPA, CFRD, or CF liver disease; some of us have had multiple lung transplants, while others are thriving on the lungs they were born with; some are on disability, while others can work full time; and almost equal numbers are on government insurance programs as those who have private insurance.

All of these characteristics contrib-

ute to how we think about whether to have kids and the options available for us to do so. Adoption, surrogacy, fostering, pregnancy, and living a life without raising kids are all valid methods of building a family. The words we use to describe issues about family *matter* because we cannot leave out people whose only family is chosen, whose given families are a source of pain, or whose inability to grow their family causes grief. How do we, diverse in so many ways, choose how to build our families?

People with CF tend to approach

family building with more intention than many because we are often reminded that we have short lifespans. Due to difficulties getting pregnant, and questions about whether our bodies can sustain a pregnancy, more people in the CF community adopt, use surrogacy, or foster than in the general population. We ask “how can we have a baby if we cannot get pregnant?” or “will I be discriminated against during the adoption process due to my CF?” We may need to use assisted reproductive technologies due to trouble conceiving or because we are using a sur-

rogate. We might foster or adopt so that we can control the age of the child we raise so that we don’t have to chase a toddler. We might only have one child when our social group expected us to have many. Our CF families are diverse!

We used to think there were too many barriers to having kids, but that is now changing. Trikafta has given many people with CF more energy and stability. Modulators have increased life spans into the 40s; Trikafta will probably increase it further. Its effects extend beyond stability and longer lives. While it has not been studied, anecdotally Trikafta is increasing fertility, most likely due to thinned cervical mucus (just like it thins out lung mucus!) and stabilized hormones, leading to a Trikafta baby boom (side note: please use contraception if you are not actively trying to conceive, especially when starting a modulator).

This ease of pregnancy and added stability is still not available to everyone with CF. I have noticed in conversations about CF fertility struggles an

“The words we use to describe issues about family matter because we cannot leave out people whose only family is chosen...”



MOLLY PAM

absence of people who have tried and were unable to have biological children. People who had multiple miscarriages, went through round after round of in vitro fertilization (“IVF”) or intra-uterine insemination (“IUI”) who ended up having a child through pregnancy are visible. People who tried to pursue parenthood and ultimately decided against having children are not heard from as frequently. People who adopted, fostered, or did surrogacy are more visible, but doctors are often ill equipped to help patients seeking information about these options. I had a doctor who did not know surrogacy was illegal in their state. More resources and understanding of local agencies would be a great place to start. CF Peer Connect and Facebook groups like CFReSHC (Cystic Fibrosis Reproductive and Sexual Health Collaborative) are great places to connect with others. To all of the people out there in our community who haven’t been successful, I see you. The person who decided to stop after multiple miscarriages...the couple who struggled with infertility...your stories matter, too. Those of us organizing conferences, panels, articles, etc., need to do better at including these stories when we present the narrative of what it means to have a family with CF. We need to hear these stories, because they

matter just as much as the successes. And when we hear them, it helps those who have traveled a different path feel like they belong here, too.

I have been thinking a lot about the words we use as I write a guide about sexual and reproductive health with CFReSHC. I am writing the family-building chapter with a team—a resource to help people with CF, their partners, and caregivers have conversations about our reproductive intentions. We have all experienced different ways of building our families—some of us have had heartbreak about changing our plans, but all of our experiences have helped us grow as individuals and we can celebrate that. I am also working with a team of researchers to write a family-building decision aid tool that will be available for beta testing soon, eventually being hosted on a website accessible from home.

So why do I keep saying “building” a family instead of “having” a family, “forming” a family, “growing” a family, “creating” a family, or many other ways of discussing this issue? Because words matter. When thinking through what to call the process, we wanted to find a word that implied that a family can both be added to and taken away from, implying that a family is always changing. Additions can be a first child, second child, chosen family, or a pet.

Changes come when kids move out, parents get divorced, a family member dies (including a pet!), or family members become estranged from one another. “Building” includes all of this, and also acknowledges that if you do not have children, you are still part of a family. Having a child does not make you a family, but it is hard to see that the way society discusses the issue.

I built out my family when I gained a sister, and again when I married my husband. For now, we are not adding any children to our family of two, though we did add our betta fish, Sally, last summer. We are enjoying being a hands-on aunt and uncle to our local niece and nephew, seeing them weekly, going on vacations with the larger family, and having them over for sleepovers. No matter where you are in building your family, know that you are not alone, and that there are as many ways to build your family as there are members of our CF community. And every one of those paths is normal, valid, and special. Someday our family will change, because we are always building it. ▲

Molly Pam is 32 with CF. She and her husband, Adam, are currently sheltering with her parents, sister, brother-in-law, toddler nephew, and brand-new niece. They live in San Francisco.

Cystic Fibrosis Mothers

Cystic Fibrosis Mothers is a website dedicated to providing information on parenthood to women with cystic fibrosis around the world. Our aim is to provide a central online resource for the global cystic fibrosis community. It includes personal stories, research articles, advice and links to further sources of information built up over time.

We also provide a private support group on Facebook with more than 500 members worldwide. To visit our website go to: www.cfmothers.com.

If you would like to join our Facebook support group, please go to: <https://cfmothers.com/cfmothers-forums/>.



WELLNESS

Diversity Of Thought

By Julie Desch, M.D.

One of the benefits of aging with cystic fibrosis is that I have been able to watch the natural evolution of the way that I think of my illness and be a witness to the consequences of this evolution. I am approaching 60 years of living in a body that doesn't function normally and, as a result, there has been a divergence from what living with CF used to feel like and what it feels like now as I type this sentence. Of course, there has also been a divergence during all of those years in how it is treated. When I was a child, CF was treated with mist tents and chloramphenicol, a nasty antibiotic with life-threatening side effects that is only rarely used today. Now, as we all know, the advent of modulators, better mucolytics, and antibiotics have changed the game for many, although it goes without saying that for those who have mutations that don't respond to modulators, this is less true.

As a presupposition for this article, I'd ask that you consider, just for a few minutes, that the thoughts that we pay attention to and hold to be true very much color how we experience life. I don't mean this in a "woo-woo" way wherein we create our own reality and a shift in thinking erases having CF. That is dangerous thinking that places blame where blame doesn't belong. I simply mean that thinking has the ability to change how we feel, and thus how we experience life. I've lived long enough now to notice that this is, in fact, very true, and, in seeing, this I feel more comfortable facing the inevitability of the consequences of CF.

There can be diversity in the way we think about illness. This doesn't occur to us very often, at least it didn't for me, until one day it did. I used to think that there was only one way to approach CF—that it and everything about it sucked, that fear was always lurking around the corner in my mind, and the only way to deal with that fear was to be in fight-or-flight mode. The fight-or-flight response is a way of describing the stress hormones released by the sympathetic nervous system. The biological opposite of this response is the rest-and-digest response of the parasympathetic system. Together, these make up the autonomic nervous system.

Whether we experience life through the sympathetic nervous sys-

tem or the parasympathetic nervous system is not in our control. It is a purely instinctual occurrence that is automatically determined by our perception of our environment. Are we feeling safe? If yes, then the rest-and-digest system activates. If we are sensing danger, our fight-or-flight system activates. In animals, we can see how this functions optimally. A giraffe sees a tiger and immediately shifts into fight-or-flight mode. It runs away, and, if it is successful, in 90 seconds or so, the mechanism turns off and the giraffe goes back to feeding.

Humans, on the other hand, think and therein lies our downfall. Our brains have evolved to place us at the top of the food chain by enabling us to learn from the past (memory) and plan

for the future. We do this through thinking. The problem arises when we don't *stop* thinking about what just happened, which serves to keep us in fight-or-flight mode. It takes 90 seconds for an animal to process fight-or-flight

response; whereas, in humans, that response time is stretched out to nine hours, nine years, or sometimes an entire lifetime. At best, it's a feedback loop that serves no valuable purpose. At worst, we can't stop imagining a terrible future, ultimately throwing us into the fight-or-flight state of chronic anxiety or chronic depression. Like so many things in life, this thinking process is a double-edged sword.

The perspective of living via the fight-or-flight sympathetic system is very, very different from living from a rest-and-digest parasympathetic perspective. While the automatic and instinctual response to stress is not in

“Having CF has most definitely brought with it several intimate encounters with fear.”



JULIE DESCH, M.D.

our control, the thoughts we give attention and energy to can either trap us in the fight-or-flight mode, or allow us to move to the opposite mode. In that sense, we actually do shape our experience of reality. I've selected a few ways that I've seen my thoughts change over the years that have allowed me to lean away from the fight-or-flight response.

There is an obvious way of thinking about living with CF that is embedded in our language when we talk about CF. We are battling CF. I often find myself thinking and talking about CF in terms of war. I "fight off" an infection or I "struggle" with digestion. Of course, it's true that the cells of the immune system really do try to fight off infection. But "I" don't. I just infuse antibiotics and allow them to do their magic. "I" just rest, and let nature heal herself. As you read that, is there a different sense of how a response to infection could be viewed, and thus felt? Similarly, one word used to describe the hours spent in discomfort on the toilet is "struggle." Perhaps another way to view this is saying "well, I guess I have more time to read" or "wow, isn't it great that I can watch Netflix on my phone?" I don't have to envision a war going on in my body and, when I don't, there is a lot more relaxation and much less contraction of the body and mind around "winning" a battle.

Before my back and knees let me know in no uncertain terms that they were done with high-impact training, I used to run. I loved the feeling that came at the end of the run. The feeling of exhilaration and accomplishment combined with perfect fatigue to a post-exertion blissful relaxation. My motivation to run wasn't really about that feeling. The motivation for me was that I was running *from* something, and that something was death. Lurking in my subconscious mind was a message that running was keeping me alive, and therefore, if I didn't run, well, I would

cease to be alive. I'm still here three years after having stopped running so, clearly, that message was incorrect. Looking back now, I see that running from something bad completely took the fun out of running and was entirely unnecessary. It was a way of thinking about exercise that caused me to live in a narrative that was 1) untrue, 2) fear-based, and 3) not fun. I regret that I saw too late that running could have been a more joyful experience of being alive. Now, I bring that understanding to walking instead of running and all is good. Fear. Fear is the giant on switch for the fight-or-flight response. Having CF has most definitely brought with it several intimate encounters with fear. Fear is a normal and natural response to danger. It is nature's way of saying "pay attention to this!" With the emotion of fear come thoughts that are separate from the emotion, but continue to feed it and keep it alive, like water and sunlight to seeds planted in a garden. Several years ago, when I was at high altitude, I had my first episode of hemoptysis. Of course, our first response was to drive back home, closer to medical care. I spent those several hours driving back home immersed in thoughts of fear. "What did this mean? Should I go to the hospital? What if it starts again and doesn't stop before I get there?" Of course, these thoughts kept me trapped in sympathetic overdrive, long after it was necessary. The anxiety grew with each mile as I let my thinking about the danger I was in grow more and more real, even though the bleeding had long since stopped. Thinking is like this much of the time. We get lost in thoughts, trapped in their message of doom and gloom as they proliferate. The danger of that happening is perfectly reflected by what I did when I arrived home. Carried away by fear, I immediately hopped up on my treadmill and proceeded to run three miles just to prove

to myself that I could; that I wasn't, in fact, dying.

More recently, I've experienced a different way of being with fear regarding the COVID-19 pandemic. Of course, when it first became clear that this was a dangerous time to be out in public, I heeded the fear message and completely secluded myself. Once this was done and there was nothing left for me to do to take appropriate action, I spent a fairly peaceful three weeks (mostly alone) meditating, hanging out on Zoom, reading, eating, sleeping, and not perseverating on danger. Comparing this to the hemoptysis response is like day versus night. Completely different thinking led to a completely different experience of life. The hemoptysis response was cyclical, manic thinking taking the lead, and truly unskillful action following like a shadow. The COVID response was simply just the appropriate response, followed by peace.

So, the invitation is merely to look into your thinking process about living with CF and ask yourself what is the experience of living with this particular way of thinking. Is it a helpful way of thinking that leads to appropriate action? Or, is there a way that the thought process is keeping you trapped in a flight-or-flight mode? If it is, what does fight or flight feel like? Is it pleasant? Once it has sent its message, is it helpful to stay there? Is there a way of thinking that gently allows you to relax, to rest and digest? If so, what does that feel like? Exploring in this way can result in naturally dropping unhelpful modes of thinking, and with this can come a more relaxed and peaceful way of being. ▲

Julie Desch is 59 years old and lives in San Rafael, CA. She enjoys meditation, reading, writing, exercise of every variety, and hanging out with her partner, two boys, and three dogs. She can be reached at Juliedesch@gmail.com.



CFRD EDUCATION

Enjoying Summer While Managing CFRD

By Kat Porco

Despite COVID-19, summer is here. The days of picnics, BBQs, and having a libation in the sunshine is upon us. However, blood sugar management remains especially critical. Often, we do not drink enough hydrating fluids in the summer, we are hot, we perspire more, and we run the risk of becoming dehydrated more easily due to CF. This, in turn, elevates blood sugars, further concentrating the sugars in our blood. So, how can we strategize our intake, cover the carbs, and eliminate insulin stacking?

First and foremost, it is important to remember the rules around insulin usage. Insulin is active for approximately three to four hours, so if you are having a social-distancing BBQ and may potentially elongate a meal, plan to take your insulin at the best

time to cover the most carbs. If you have an InPen or pump, there are strategies that allow you to dose more frequently. The key is understanding how much insulin is still on board and available for intake, thus titrating the coverage amount to include the insu-



KAT PORCO

lin that is already circulating. However, if you do not have these tools, and you choose to snack without insulin, drink lots of water and go for a walk to minimize hyperglycemia.

Exercising has two beneficial byproducts for managing diabetes. First, your muscles need energy (in the form of glucose) to work. When you exercise, you start to burn sugar as an energy source, which will help to lower glucose levels in the blood. Exercise also lowers resistance to insulin (i.e., you use the insulin available more efficiently), allowing for the sugar in the cells to be utilized more readily as energy, effectively diluting the sugars.

If you start exercising in a hyperglycemic state, keep in mind that if blood sugars are over 300, your blood viscosity is close to the consistency of ketchup, versus 100, where the consistency is closer to water. Asking your body to

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pneumonias associated with mechanical ventilation.

<https://tinyurl.com/y8n5vszf>

Microbe DNA Discovery May Lead To Noninvasive Test For CF Infections

People with cystic fibrosis (CF) have higher levels of DNA from common microbes that cause CF-related infections circulating in their blood compared to healthy controls. These findings support the development of a simple blood test as a noninvasive diagnostic tool to identify chronic lung infections in CF patients. A new type of biomarker in infectious disease diagnosis comes from DNA released into the blood by decomposing cells called cell-free DNA (cfDNA). Most cfDNA comes from a

person's own cells, but a small amount of microbial DNA has been detected effectively in the blood during acute infections, particularly from microorganisms that directly invade the bloodstream. Given that some CF-related microbes can be invasive and inflammation can lead to tissue damage, cfDNA originating from lung microbes may enter the bloodstream of CF patients during chronic infection. Researchers collected blood samples from 21 people with CF, ages 19 to 62. Blood from 12 healthy people of similar age was collected as controls. cfDNA was isolated and sequenced to identify its microbial source. The analysis revealed that the amount of microbial cfDNA — from organisms that commonly infect the

lungs of CF patients — was nearly 10 times higher in the blood of CF patients compared to healthy controls. Microorganisms also were cultured and identified from sputum samples collected from patients to compare each method. From these, a total of 44 identified microbial infections were found, and 15 of those statistically matched the cfDNA results. For some bacteria, the cfDNA and the sputum results matched perfectly, while with others it did not. In particular, out of 13 *Pseudomonas aeruginosa* infections identified in sputum, eight were confirmed by cfDNA, while 12 infections by *Staphylococcus aureus* were found in sputum samples, compared to four by cfDNA. Conversely, 17 identified microbial infections (from 11 patients)

circulate “ketchup” throughout the body is very taxing, so it may be beneficial to drink a couple pints of water first to try to bring that down into the 200s to ensure safety while exercising.

Seeing high blood sugars can be frustrating and even scary, but it’s important to remember that correction factors are not intended to be dosed between insulin doses during mealtime—they are supposed to be added onto the existing insulin dosage at the next meal. Dosing an insulin correction within three hours of a prior correction is considered stacking and can lead to hypoglycemia. If sugars are high one to three hours after a meal, increasing water intake and exercise are the best route, rather than dosing more insulin! Correction factors should be added if a state of hyperglycemia exists at the start of mealtime and insulin is already being dosed.

Drinking alcohol safely is more challenging with diabetes, but not impossible. However, there are a few things to be aware of if you have

CFRD and drink alcohol. Drinking alcohol increases your risk of hypoglycemia. It is critical to be very cognizant of your blood sugars if you are imbibing and make sure those around you are aware that being drunk can mimic the symptoms of being hypoglycemic, which can become very dangerous. Why does alcohol tend to lead to hypoglycemia? The liver stores glycogen and releases it when your blood sugars become low; however, when alcohol is on board, your liver starts to detox rather than manage glucose, so it does not release the glycogen when your blood sugars are trending down. This can last for up to 12 hours after drinking. It is critical to know that glucagon will NOT work for alcohol-induced hypoglycemia, as the mechanism for glucagon is to activate the liver to release glycogen, which is suppressed due to alcohol. So, keep fast-acting carbs with you if you want to treat yourself to a summer drink. Set an alarm to check your blood sugars two to four hours after your last drink,

when you are most likely to experience hypoglycemia.

Enjoy the summer and be safe, friends! ▲

Kat Porco is a diabetes educator and the co-founder of Attain Health Foundation. She received a Bachelor’s in Social Work followed by a Master’s of Science in Health Communications. Her work over the past ten years has been solely focused on supporting and advocating for the cystic fibrosis community. Throughout these years, she has seen the disconnect between the recommendations of the medical community and applicability for the patient community. Because she does understand this complex relationship, Kat felt that she could assist in bridging the gap to reach ultimate health goals through health coaching and diabetes education. She is a Duke Certified Integrative Health Coach, as well as being Nationally Board Certified in Health & Wellness Coaching through the National Board of Medical Examiners. Kat is a Certified Diabetes Care and Education Specialist (CDCES) through NCBDE.

using cfDNA were not identified by sputum culture, including *Burkholderia cepacia* and *Mycobacterium* species.

<https://tinyurl.com/y99srr4o>

Growing Lung Bacteria In Lab May Predict Antibiotics’ Effectiveness Better

Antibiotic susceptibility tests that determine how people with lung infections will respond to antibiotics may work better if researchers grow the pathogenic bacteria in the lab along with other commensal bacteria found in the lungs, because it more accurately mimics their real environment. The reason for this is that some infectious bacteria, such as *Pseudomonas aeruginosa*, are resistant to many antibiotics but rely on other bacterial species to sur-

vive. If antibiotics are able to eliminate the other bacteria, it is likely the pathogenic (harmful) one also will die. Currently, to determine the most effective antibiotic treatment for CF patients, a sample of the patient’s mucus is taken to the lab, where the disease-causing bacteria are isolated and tested against a range of antibiotics. But data is showing that these tests are not very accurate at predicting responses in the clinic, and some clinicians are no longer using them to guide treatment decisions.

A problem with the tests is that they fail to mimic the intricate environment seen in the lungs, from oxygen gradients to the presence of other bacterial species in the mucus, which often work as a community and rely on each other for nutri-

ents. *P. aeruginosa* is one such species that appears to work in cooperation with other bacteria. While it creates an anaerobic environment for the other bacteria to grow, the anaerobic bacteria produce nutrients from mucin (the main component of mucus) that *P. aeruginosa* would not be able to obtain on its own. This means the bacteria species are dependent on each other to survive and that targeting the most susceptible one (the weakest link) could be enough to eliminate the entire bacterial community.

<https://tinyurl.com/y6uwwsrl>

CF Lung Function Decline In Those With Infection Slowed By Azithromycin

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to be formally listed for transplant. Receiving a transplant is not how I saw my future—I did not need a transplant then, nor would I ever because I was going to live longer than Moses.

I received the call for both new lungs and a new liver on August 25, 2017. I knew when I got the call that, more often than not, the call for lungs turns out to be a dry run, whether due to an organ size mismatch or the donor organs being in worse condition than predicted and therefore unsuitable for transplant. I was convinced that would be the case—I would wake up and go home with my original lungs because neither the lungs nor liver would be a match for me. I was going to live longer than Moses, after all, despite having a lung allocation score (“LAS”) of 43. These were my last thoughts, lying on the operating table, as they administered the anesthesia for my procedure. I awoke from my medically induced slumber a day later with my angel donor’s lungs and liver. Joni Marie gave me a second lease on life. I was fortunate to have been matched with Joni Marie, as one of my liver transplant surgeons told me her liver was the cleanest he had ever seen. He shared with me those thoughts as he shed tears at the side of my bed.

It was during my nearly six-week flawless recovery and rehab period that I knew I wanted to give back as a CF patient after taking from the community for the past 44 years. It was my time to pay it forward!

In August 2019, I met CF Warrior Ashleigh Suggs. This young lady was, at the time, on palliative care. Nevertheless, I found that her spirit and will to live cast a bright shadow over the noticeable green oxygen container she trailed behind her. That spirit which was so joyfully fluorescent made her container largely invisible. Ashleigh was an immediate inspiration to me—she sadly passed away two months and two weeks following my introduction to her. In her

honor, I established Ashleigh’s Life Foundation in January 2020, with a mission of providing financial support to those who are battling end-stage cystic fibrosis. To date, over \$3,000 have been raised, providing eight grant awards to other CF Warriors fighting CF.

January was a pivotal month for me. In the middle of the month, a young lady from Chile reached out to me on Facebook, asking if I had a therapy vest (such as a Hill-Rom Vest or Afflovest) that I could donate to her. Within days, my Hill-Rom Vest was in transit to her 5,000 miles away. Two months later, I



ROD SPADINGER

was contacted by another individual via social media. From 7,000 miles away in Beirut, Lebanon, Mohamad and I began a dialogue. Following several FB messenger conversations, I posted a plea on that same social media platform that this young man was also in need of a therapy vest. A young lady with CF, living in Ohio, heard my call and responded with a donation of her Respirotech inCourage Vest, of which Mohamad now has possession.

I met Mark Tremblay, another CF Warrior, through these charitable acts of kindness and eagerly shared with him these two stories of providing vests to CF warriors in need. Mark was as inspired as I by these chance circumstances, facilitat-

ed through social media. We both felt that these two individuals from different parts of the world could not be the only ones in need of these CF resources, which are both valuable and rare in many other countries around the globe. As such, Mark and I cofounded the CF Vests fO²r Life Foundation, with a mission of transferring both therapeutic vests and oxygen concentrators from those who no longer need them to those who do. We are currently working to build this foundation to present the gift of life-extending CF medical equipment to those in need.

I believe in miracles more than I do coincidences. Was it a coincidence that my family found Dr. Robert Kramer in Texas when I was ill at the age of four? Was it a coincidence that I was blessed to be matched with my donor, Joni Marie, who had the cleanest liver my surgeon had ever seen? Was it a coincidence that a young lady from Chile contacted me by chance to request my unused Hill-Rom Vest? Was it a coincidence that I met Ashleigh Suggs and Mark Tremblay, both of whom inspired me to establish two charitable foundations to benefit those in need within the CF community? To me, those situations were a series of miracles. These miracles are what brought me to my third lease on life. ▲

Rod Spadinger is 47 years old and has CF. He was born and raised in Honolulu, Hawaii, but later relocated to Dallas, Texas. In August 2017, Rod underwent a successful double lung and liver transplant thanks to the generosity of his donor, Joni Marie. As a means of giving back to the CF community, Rod cofounded CF Vests fO²r Life Foundation, with the mission of providing therapeutic vests and portable oxygen concentrators to those in the CF community who do not have access to these therapies and equipment. He also established Ashleigh’s Life Foundation, whose mission is to provide financial grants to those who are at or near end-stage CF. He can be reached via email at rspading@yahoo.com.

Prolonged use of the antibiotic azithromycin resulted in a 37% slower rate of lung function decline in cystic fibrosis (CF) patients with persistent *Pseudomonas aeruginosa* infection over a three-year period. Because many CF patients experience chronic infections, the antibiotic azithromycin is commonly prescribed for long-term use. Roughly 70% of CF patients with *P. aeruginosa* infections take azithromycin regularly. Support for a long-term regimen of azithromycin treatment comes from clinical trials demonstrating health improvements in CF patients with *P. aeruginosa* over a six-month period. However, little is known about the health benefits of azithromycin over longer periods of time. Thus, researchers collected data from the U.S. CF Foundation Patient Registry. They used data from pulmonary function tests to compare the lung functionality of CF patients who were on azithromycin with those who were not. In the *P. aeruginosa*-positive group, the researchers identified a significant reduction in lung function decline, over a three-year period, in the group of patients who had been taking azithromycin. The findings indicated that long-term azithromycin treatment can slow the decline of lung functionality by 37% in *P. aeruginosa*-positive CF patients. No such reduction in lung function decline was observed in the CF patient group who was *P. aeruginosa*-negative, suggesting that azithromycin only provides long-term benefits in lung function in patients with recurring *P. aeruginosa* infection. Researchers then looked at data on the use of antibiotic injections to treat acute pulmonary exacerbations. They found that azithromycin seems to have no significant impact on the risk of pulmonary exacerbations, in both *P. aeruginosa*-positive and negative groups. The team also investigated a recent finding that azithromycin could have an adverse interaction with tobramycin, an inhaled antibiotic also prescribed to treat lung infections in CF

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FOCUS TOPIC

DIVERSITY IN THE CF COMMUNITY

No Limits With Teena

By Teena Mobley

I was adopted at 21 months, along with my two younger, biological siblings. Unfortunately, our biological mother couldn't take care of us and we were taken away from her. Overall, this was a huge blessing in my eyes and later I found out that my adoption saved my life. My adopted parents took great care of us. From a young age, I had aspirations of one day impacting the world. At the time, I didn't know how but I knew in my heart this was something I wanted to do.

I live in Long Island, New York. Currently, I am an advocate, entrepreneur, and media mogul. Persistency, consistency, and the drive to get things done helped me achieve my goals and mold me into the person I am today. My passion grew from the small seeds in my local community. I cultivated my greatest personality traits from the cystic fibrosis community, where my aim is to one day leave my legacy in the world.

Growing up, I had a passion for sports but also knew from a young age that I was different from most children. However, I did not expect to be considered different due to my physical capabilities. I was first misdiagnosed with severe asthma at a young age. One day I became really sick and, ultimately, the hospital became my home. After several tests I was finally correctly diagnosed with cystic fibrosis. From that moment on, this diagnosis only gave me the strength and resolve to know what I wanted to do with the rest of my life.

A Person of Color with Cystic Fibrosis

Cystic fibrosis is not as common in African Americans; thus, my initial diagnosis of severe asthma, as opposed to cystic fibrosis. I can't even begin to

imagine the number of people who may have had a similar situation or were diagnosed later in life due to their ethnic background. When it comes to cystic fibrosis, I think more research needs to be done, especially within other ethnic backgrounds. As we know, CF is more common in Caucasian Americans and less common in other ethnic groups. Cystic fibrosis affects people differently and it's important to understand that, especially for healthcare professionals. I also believe there is a systemic gap within the healthcare system. I think doing most of my own research, as well as hearing different stories from others within the CF community, has helped me learn about diversity within our community. Again, more research needs to be done from an ethnic background standpoint.

No Limits with Teena

When it comes to cystic fibrosis,



TEENA MOBLEY

doctors would say that our capabilities are fairly limited. I have always been ambitious with a competitive nature from being an athlete all of my life. I was told to take it easy at practice, to not overwork myself, and to not give 100% all the time because they didn't want me to overdo it. Well, if you know me then you know that I can be hard-headed at times.

Coming out of high school, I was named New York State's top long jumper in track and field, with my furthest jump at the time being 18 feet 9 inches. I graduated with athletic honors and received a track scholarship from Monroe College in New Rochelle, New York, where I attended for two years. My mornings started at 4:30 am. I was able to balance my school work and CF treatments while also being ranked one of the top long jumpers in the nation in 2014. I was an All-American in the 4x4 meter relay. This was the life I had to adapt to. It has not been easy being a cystic fibrosis athlete, but I think the overall motivating factor that pushed me was not only my support system but also wanting to push past my potential. And I did. The last thing that this illness would do was to stop me from competing and training.

After obtaining my Associate's Degree in Business Administration. I then received another D1 athletic scholarship at Long Island University, where I graduated with my Bachelor's Degree in Sport Management. Living with cystic fibrosis is exceedingly challenging for me and many others. It is hard for others to understand what we go through on a day-to-day basis. I believe being an athlete has prepared me for the real world—even when I was in school training for track twice a day, I still had to wake up earlier to do my treatments.

Unmasking Cystic Fibrosis

For many years, I was able to mask this illness in public. At first, I didn't tell others that I had this illness because it was embarrassing, especially with all of the medications I had to take. I didn't want people to treat me any differently, as if I were fragile. Being physically active and playing sports has helped make my heart stronger and clear my lungs. Being an athlete has for sure helped my general health.

Striving to be a positive role model and leave my mark on the world, I have been a CF advocate and created the Redefining Life – Cystic Fibrosis movement in December of 2016 as a way for me to give back to the CF community. I wanted to redefine the outlook people had on this illness and encourage others not to limit themselves with their dreams and goals in life. I truly believe that this illness does not define us—we can still achieve our dreams no matter the obstacles that come our way, as long

as we put our mind to it. It starts with a positive mindset. The doctors say that our capabilities are fairly limited, but I believe we can go past that.

After the launch of Redefining Life – Cystic Fibrosis, I built a pedestal to stand on and speak to others to raise awareness for this illness. It gave me the voice to become a guest speaker at schools and programs as well as leading to features on websites, podcasts, and interviews. I was able to collaborate with a scout for the Brooklyn Nets to assist with his youth basketball program; I spoke at an event hosted by Adelphi University (netting over \$6,500 in donation to the Cystic Fibrosis Foundation); I hosted the Five Feet Apart Screening at CBS Corporation; and I was featured on Jerry Cahill's Cystic Fibrosis Podcast and the Team Boomer website, just to name a few. I follow the mantra of one of my biggest influences, Sean "P. Diddy" Combs—can't stop, won't

stop!—and later grew my list of talents to include sales, marketing, events, media manipulation, and various other entertainment industry skills.

I will always continue to be a role model in my community. I have hopes to one day start my own non-profit organization and to share my talents with the world with the launch of my own company and public speaking. Having an illness will for sure not take away or define what I want to do in my life. If anything, it serves as a motivating factor for me to push the limits to what others may seem to be "impossible" for a person with a chronic illness. ▲

Teena Mobley is 25 years old and has cystic fibrosis. She lives in Long Island, New York. She loves working out, shopping, going to events, and photography. She also loves spending time with her family and friends as well as raising awareness for cystic fibrosis during her free time. She can be reached by email at mobleyteena@gmail.com.

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patients. This is also being investigated in CF patients enrolled in the TEACH clinical trial (NCT02677701). Azithromycin treatment was not associated with a slower lung function decline in the tobramycin group, whereas it was in the aztreonam lysine group. The results suggested that azithromycin was not providing the same health benefits identified in the study to patients who were also taking tobramycin.
<https://tinyurl.com/yaupra3n>

Antibiotic Treatment For Stenotrophomonas maltophilia In People With Cystic Fibrosis.

Chronic infection with *Stenotrophomonas maltophilia* has recently been shown to be an independent predictor of pulmonary exacerbation requiring hospitalization and antibiotics.

However, the role of antibiotic treatment of *Stenotrophomonas maltophilia* infection in people with cystic fibrosis is still unclear. This is an update of a previously published review. The objective of this review is to assess the effectiveness of antibiotic treatment for *Stenotrophomonas maltophilia* in people with cystic fibrosis. The primary objective is to assess this in relation to lung function and pulmonary exacerbations in the setting of acute pulmonary exacerbations. The secondary objective is to assess this in relation to the eradication of *Stenotrophomonas maltophilia*. This review did not identify any evidence regarding the effectiveness of antibiotic treatment for *Stenotrophomonas maltophilia* in people with cystic fibrosis. Until such evidence becomes available, clinicians need to use their clinical judgment as to whether or not to treat

Stenotrophomonas maltophilia infection in people with cystic fibrosis.
<https://tinyurl.com/y75m5bvf>

S. Aureus In CF Airways Change With Antibiotic Use, Age, Study Finds

Different types of *Staphylococcus aureus* can be found persistently in the airways of cystic fibrosis (CF) patients, but this bacteria population is influenced by age and prior antibiotic treatment. *S. aureus* is one of the earliest bacteria present in the airways of people with CF and is known to persist in the airways of CF patients for extended periods, and to proliferate by creating clones of itself. Over time, these clones accumulate slight genetic variations and originate different *S. aureus* types. However, there is a lack of knowledge about the

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How CF Shaped My Life

By Daniel Gonzalez

Have you ever wondered what your life would have been like if you had been born in a different country other than the United States? Both your life and medical care would have been extremely different. The doctors in the U.S. have greater access to medical knowledge and the U.S. is one of the first countries to get new medications and treatments, which is a huge advantage for those in need of more specialized care. I was born in Mexico and, while it is a great country that I enjoyed living in, moving to the U.S. was the best thing for me.

When I was about two years old, it was clear to my parents that I had some type of health issue. I did not have any respiratory symptoms, but I had major digestive problems. They took me to the hospital and were told I had liver cancer and may not have long to live. My parents were surprised but not fully convinced. My pediatrician told them it might be a good idea to travel to The Children's Hospital in Dallas for additional testing. It is here they found out I had CF. The initial misdiagnosis of liver disease may seem strange from the outside looking in, but my most severe CF symptom in childhood presented as liver disease rather than lung disease. Unfortunately, many places outside the U.S. lack the resources for researching and understanding CF, much less the tools to diagnose and treat it as effectively. I might not have been diagnosed until much later in life had my parents not sought out a different opinion, which could have led to more serious health problems.

Between the ages of two and eight, I was seen in Dallas for my CF. We would travel a couple times a year from Mexico to see the doctors. Every trip cost lots of money, between the lack of insurance and

“Not too long ago, I went back to my hometown to visit family and I had a small glimpse at the care I would have received had I stayed in Mexico.”



DANIEL GONZALEZ

trip expenses. It was clear this was becoming a financial burden; luckily my grandparents were able to help my parents with some expenses. As my CF worsened, we started talking about the next steps as far as my care was concerned. There were no options in terms of CF doctors in the town we lived in, so we would need to move to get better CF care. Once we realized the U.S. provided a better opportunity for my care, we decided to make the move to Austin, Texas.

Once I got here, I had some trouble assimilating—the shift in my environment, a new language, and the people I interacted with, as well as the move itself, affected my daily lifestyle. In Mexico, both of my parents worked;

however, our new visas didn't allow for my mom to do so. We also moved from a big house to a small apartment. I attended a new school, where I received help to adjust and learn the language, but it took me a while to get comfortable. Luckily, my CF was not too much of a problem at this point with my FEV1 being near 100 for many years, something that my doctor erroneously attributed to my race having larger torsos and lungs than others.

About four years ago, I received a liver transplant, which further emphasized the benefit of the move. After the transplant, I was in and out of the hospital for the first two months. Later, I was once again hospitalized; this time for six months. During this hospitalization, I had about six specialists visiting me during my hospital stay to check on me. I was told by multiple doctors that I was a very complicated case. If the doctors who saw me here in the U.S. thought this, it made me wonder how my care would have been in Mexico.

Not too long ago, I went back to my hometown to visit family and I had a small glimpse at the care I would have received had I stayed in Mexico. Many precautions were taken to avoid infections or illnesses for both my family and me because of my transplant. My extended family had to get flu shots (while normal in the U.S., it is not as common for people in Mexico to get them) and we all had to take a few additional precautions: being extra clean and preparing food

more carefully, avoiding tap water, and mainly drinking bottled water. All of these precautions were necessary because of my immunosuppressants, which are critical to avoid rejection after the transplant.

I know that in any country, there are doctors with knowledge and experience. However, in the U.S., doctors can choose from a wider range of specialties and have access to more education. For example, my infectious disease doctor gave me a medicine to treat the fungal infection in my lungs. After a few weeks, I was unable to move and had debilitating nerve pain whenever I tried. At first, she said it was unlikely the medicine was causing the nerve pain; however, the next day she circled back and said she had done research and found that there were only two other documented cases in the world with a similar side effect. She immediately stopped the medicine and prevented what could have led to permanent nerve damage. She was able to do so thanks to the access to information that she had. She then found a better option that would not only avoid similar side effects, but was well absorbed by people with CF.

Compared to many other countries, the U.S. has excellent medical care. Here we can be treated for our CF, in addition to any other problems that may arise. There is easier access to information and research from all over the world, coupled with access to more medications. As most CF patients already know, less than a year ago, a groundbreaking modulator, Trikafta, was approved by the FDA. Today, about 90% of people in the United States with CF are already benefiting from it. Unfortunately, people in Mexico and the EU do not have easy access to Trikafta. As the world globalizes, we are starting to see greater access to information, equipment, and newer treatments in other countries.

Having CF has allowed me to experience and meet a diverse range of cultures, situations, and people. Before, the only aid my family received was from other family members, but today I have access to many different organizations that provide financial aid, scholarships, information, and a lot of other resources. I have become a part of CF communities that have helped me grow and become more open minded, communities where other CF

patients listen and provide support for one another. I have also learned about other people with CF and their situations, including their unique struggles.

CF has had a large impact on both my life and my family. I struggled to find an ideal care team that suited me and that understood the many aspects of my CF. I also had to learn how to live in a new country with a different culture and people. I had to learn a new language; however, this has proved to be a great thing for both me and my career. I have become a greater person in spite of, or maybe even because of, my CF. It is important to remember that, while we enjoy our normal life, we must also welcome change when necessary as it can bring opportunity, whether it is throughout your daily life or in your CF care. ▲

Daniel Gonzalez is a 24-year-old college student with CF who is currently working on his Bachelor's Degree in Business Administration through Penn State World Campus. Daniel was born in Mexico and moved to Austin, TX, at a young age. He enjoys spending time with friends, playing video games, and relaxing. He can be reached via email at danygonzalez96@hotmail.com.

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different *S. aureus* types present in the airways of CF patients, and the dynamics of their long-term presence in the airways.

In this study, scientists investigated the *S. aureus* types present in the airways of people with CF over a 21 month-period, and their association with the patients' lung disease, age, and history of antibiotic treatment. Researchers identified 265 different types of *S. aureus* from 1,929 patient samples where the bacteria was present. CF patients carried on average 3.21 *S. aureus* types in their airways. Eight types of *S. aureus* were found in

more than 10 people, making them the most common types among CF patients in the study. Depending on the types of *S. aureus* found in their samples, patients were divided into four groups. The first group included patients in whom a prevalent type of *S. aureus* was identified in samples from at least half of their visits. In the second group, patients in whom only one type of *S. aureus* was identified during the entire study were included. Patients in the third group had a dominant type of *S. aureus* which was present in samples from all visits, while other types of *S. aureus* were only present

in samples from some visits. Finally, the fourth group included patients who had a mix of genetically-related *S. aureus* types in more than half of their visits. None of the different groups had a higher risk of pulmonary exacerbations or worse lung function. There was not a special *S. aureus* type, identified that was associated with a more severe lung disease. Transmission of different *S. aureus* types within centers was found to be low, and any possible *S. aureus* types transmitted between patients did not persist for long. Antibiotic resistance was generally

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Living With CF While Black

By Anonymous

Due to COVID-19, the past few months have been challenging for many people with CF, including me. At the beginning of 2020, I felt better than ever thanks to a new treatment I was eligible to start in November, 2019. I know that many African Americans who have CF are not eligible to take modulators because they do not have one copy of the delta F508 mutation. I felt I had won the lottery when I heard the FDA had approved the newest modulator for people who had one copy of the delta F508 mutation.

I started the new year with a new hope for my future. I had never been able to go to college or work due to the demands of my daily CF medical treatments and frequent hospital stays each year. Once my health improved at the end of 2019, I decided I wanted to try to work and then, in a year, apply to college. My plan was to start college in the fall of 2021.

I had heard from friends and relatives that it would be hard to find a job. I told them I knew that because I had never had a paying job, it would be a more difficult. However, I was hoping that my volunteer work over the years would show an employer I had useful skills and I could be a great employee. My African American friends explained that it would not be my lack of work experience but the color of my skin that would be the biggest obstacle. I had never tried to find a job so I thought they were exaggerating. How hard would it be to find a job as a Black male?

I started to apply for a variety of jobs in January, 2020. I am a friendly person and try to have a positive outlook. I lined up five interviews with prospective employers. All the jobs were entry-level jobs in the service industry and none of the jobs required

a college degree. This was pre-COVID-19 and so I went for face-to-face interviews and talked about my volunteer experience for different groups. I spoke about my excitement about working at the respective business. I was disappointed when I received email after email explaining that I was not what the business was looking for in an

“ I wondered if other young men with CF were asked about being a member of a gang. ”

employee. I kept searching for other job opportunities and was surprised to see some of the same listings for jobs I had interviewed for in the prior weeks. I was discouraged and started to wonder if my friends had been right. Was the color of my skin keeping me from landing a job?

I remembered the obstacles I had encountered when hospitalized—the bias many medical residents and doctors seemed to have when they entered my room. I intuitively knew that healthcare workers who are used to seeing white people with CF in the hospital were surprised when they saw an African American male with CF as the patient. I wondered if other young men with CF were being asked so many questions about drug use. They were unusual questions since there was no evidence of prior drug use. I wondered if other young men with CF were asked about being a member of a gang—also an unusual question since there was no evidence of gang association. I wondered if other young men with CF were viewed with suspicion by people at the hospital. I had a great relationship with my CF team. But when I was hospital-

ized, it was a different story. When I spoke to other white men with CF men online, it seemed that the frequent inquiries about drug use or gang membership were not the norm.

My job search abruptly ended on March 6, 2020, when I spoke to a friend with CF. He told me he was going to start staying home to avoid

being infected with COVID-19. He encouraged me to do the same. I read some articles online about the lack of any test kits in the city I live in and decided I would start staying home.

A few weeks later, I was talking to another friend who told me her daughter had come home from college and was now taking her college courses online. A light bulb went off in my head. I wondered if I could enroll in a few online classes at a college. At first, I thought it must be too late to enroll in college for the summer. But then I decided it could not hurt to ask.

It was clear my plan to work for a year before starting college was not going to be possible. I wondered if I could change my plan and start college now. As luck would have it, a local college was interested in adding students since their summer classes would be online and they weren't restricted by in-person classroom sizes. I ordered my high school transcript, completed some paperwork and, two months later, I received an exciting email letting me know I had been accepted and could register for classes the next day.

I have read stories from others call-

ing something that has come out of this national emergency a “silver lining.” I realized that being able to start college was my silver lining. After completing one week of college, I can tell you it is terrific. My plan is to stay put in order to stay safe, so being able to

take online classes is perfect. I have met some really nice people already and enjoy the classes. I am also getting involved with the racial justice movement by participating in events online.

I am hopeful that the racial justice movement will improve the lives of

people of color in the United States. I dream of employment opportunities for people of color improving. Until then, I will stay safe at home, enjoy my summer college classes, and look forward to reading stories in *CF Roundtable* about others living with CF. ▲

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low in the *S. aureus* types isolated from patient samples. About 2% of *S. aureus* were resistant to oxacillin, an antibiotic similar to methicillin, which indicates a low proportion of methicillin-resistant *S. aureus* (MRSA). A higher number of past antibiotic treatment cycles was seen to be associated with a higher chance of carrying only one type of *S. aureus*. According to the researchers the fact that antibiotic treatment affected the number of different clones [*S. aureus* types], indicates that CF patients are highly susceptible for the acquisition of new *S. aureus* strains, if not treated with antibiotics. Another finding of the study was that older CF patients were more likely to either have a mix of genetically-related *S. aureus* types, or a dominant type of *S. aureus*.
<https://tinyurl.com/yc27fqgz>

Long-lasting, Low Toxicity Antimicrobial Peptide Fights ‘Superbug’ Lung Infections

Researchers reduced the toxicity of a potential antibiotic against the most feared drug-resistant bacteria, while also improving its stability in fighting infections. The new antibiotic ~ administered via the windpipe to target lung infections ~ proved more effective than its experimental predecessor and traditional last-resort antibiotic therapies in fighting drug-resistant bacteria in laboratory cell cultures and mice. And it did so without notable toxic side-effects. The experimental drug that the team developed is built from an engineered

cationic antimicrobial peptide, or “eCAP,” which is a synthetic and more efficient version of naturally occurring antimicrobial peptides that form a first line of defense against infections in humans. eCAPs work by “punching into” bacteria, thereby destroying them.
<https://tinyurl.com/ycddhydr>

Drug-resistant *S. Maltophilia* Genetically Diverse, Widely Spread, Study Says

The antibiotic-resistant bacterium *Stenotrophomonas maltophilia*, a dangerous bacteria for people with lung diseases such as cystic fibrosis (CF), is widely spread around the world, with most strains carrying virulence and resistance genes. *Stenotrophomonas maltophilia* is increasingly recognized as a dangerous pathogen, with the World Health Organization listing it as one of the leading drug-resistant pathogens in hospital settings. The bacterium most frequently causes infections in the airways, in the blood (bacteraemia), or in catheters. Despite the worrisome clinical scenario associated with *S. maltophilia* infection, little is known about the bacterium’s infectivity and its transmission worldwide. To address this question, an international partnership with scientists from eight countries established a standardized method to analyze the genome of the different strains of *S. maltophilia*. The team analyzed a global collection of newly sequenced *S. maltophilia* strains together with publicly available whole-

genome data. In total, they collected data of 2,389 strains from 22 countries on four continents. The researchers found that the global population of *S. maltophilia* comprises a total of 23 lineages. To determine the infection potential of *S. maltophilia* strains, the researchers screened the data for genes related to virulence and resistance. They found that resistance genes were broadly present in *S. maltophilia* strains. Analysis of strain transmission in hospital outbreaks showed that genetically similar strains were isolated in short intervals of days or weeks in the same hospitals.
<https://tinyurl.com/y7yyopzk>

CF Candidate MRT5005 From Translate Bio Gets FDA Fast Track Status

The U.S. Food and Drug Administration (FDA) has granted fast track designation to MRT5005, an investigational RNA-based treatment for cystic fibrosis (CF). MRT5005 is a first-in-class RNA-based therapy that works by delivering CFTR messenger RNA (mRNA) — the template cells use to make the CFTR protein that is faulty among those with CF — to cells in the lungs. The therapy is delivered to lungs via nebulization. By providing the correct instructions to make the CFTR protein to cells and directly addressing the root cause of the disease, MRT5005 is expected to work in all CF patients, regardless of the mutations they carry. The investigational therapy is

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Making Space For Sex And Gender Diversity In CF Care And Community

By *Xan Nowakowski, Ph.D., M.P.H.*

A lot of recent writing on cystic fibrosis has, quite appropriately, celebrated how the new CFTR modulator drug, Trikafta, will likely give many patients the opportunity for long, adult lives. In the past, many of us had to put important and social and emotional goals on hold because of more pressing threats to our lives from CF complications. As we live progressively longer and healthier lives with the help of innovative therapies, we can now bring those goals more to the forefront. This process will likely be both exciting and challenging for many of us. It will bring new experiences for which we may feel unprepared, and thus require substantial adjustment. This also means it may be more difficult for those of us who have had to put goals that are more social or emotional in nature on hold in the past because of more pressing threats to our lives from CF complications to further delay going after what we want. Embracing an authentic sex or gender identity is one set of goals that often gets put on hold because of profound illness early in life. So, as our community begins to thrive more and live longer, we will likely see more people with CF opening up about who they are with respect to sex and gender.

To offer a quick overview for folks who are new to thinking about these kinds of diversity, sex and gender are different but related aspects of who we are. Sex describes our relationships with our own bodies and the various things that make them look and function as they do. Gender describes our social identities related to other peo-

ple's perceptions of our bodies. Both of these terms invoke the body but are not a direct description of the body. Both of these terms are also hard to define precisely, which can make understanding exactly how to address them in clinical and community settings seem tricky. So, it helps to understand what kinds of sex and gender identities we might see in our patient community.

Let's start with sex. When babies



XAN NOWAKOWSKI

are born, generally they get "assigned" a sex on their birth certificate. Sometimes this reflects the actual person's sex identity and sometimes it doesn't. When sex assigned at birth matches a person's sex identity, this is called "cissex." A person whose sex identity differs from the one assigned at birth is "transsex." We can also think about sex in terms of characteristics the person has—for example, what their genitals look like or what chromosomes they have. Some characteristics are associated with maleness and some are associated with femaleness. And,

some people have a mixture of the two groupings! Folks who have both female and male sex characteristics are "intersex." People with just female or just male characteristics are "endosex." Being intersex and endosex are both normal, as are being transsex and being cissex. Think about how some people have two different colored eyes and some people have two similarly colored eyes or how some people are diagnosed with one health condition at birth but turn out to have another. That last bit has happened to a fair amount of us in the CF community!

As birth care providers tend to do with babies, we all tend to think about the apparent sex of people we encounter in daily life because we are socialized to do so. And often we guess correctly, both by looking at people's bodies and by paying attention to social clues they are giving us. But sometimes we get it wrong or oversimplify things. This moves us more into the territory of gender identity. A person's gender still has something to do with their body but is a layer more removed from all of that. Gender is really more about social roles we feel comfortable playing, and how we respond to others' expectations of the social roles we *should* play based on what our bodies look like. As with sex, there are different groupings of gender characteristics that we describe as "feminine" and "masculine." Some folks largely have one set or the other, whereas others have many characteristics from both sets. When people have mixed gender characteristics, we can broadly describe them as "nonbinary." People with more streamlined gender characteristics from one set or another are likely to identify as "men" or

“women.” And if a person’s gender identity matches what has been expected of them since birth, we describe them as “cisgender.” People whose true genders diverge from early life expectations are by contrast “transgender.”

Broadly speaking, folks of transsex, intersex, and/or transgender identity are described as “sex and gender minorities.” This term refers to the fact that although our individual experiences may be very different, we share a broader vulnerability to various forms of marginalization. All of the specific identities that fall within this broad “sex and gender minority” category are far too numerous to name. Each of these identities also has nuances; indeed, it is in these finer details where really wonderful support can flow between different members of a patient community.

I’ve given a general overview of what sex and gender mean in society as a whole. But how does this relate to CF specifically? On a basic level, healthcare interactions tend to be more effective if they help people feel seen, heard, and valued. In a world where people who are transsex, intersex, and transgender often face terrible stigma and violence, simple actions that communicate safety and value can go a long way toward improving care. Showing patients that we care about who they are and what they need is a basic principle of good healthcare, and equally as important for sex and gender considerations as for any other aspect of CF care.

These same kinds of simple welcoming actions also invite creative thinking about how CF may intersect with sex and gender with respect to the kinds of services we receive and how we respond to those elements of our care. Sometimes we forget that people with CF are often more medically complex than just that one condition, which is complex enough on its own. As such,

we may not necessarily think about what happens, for example, when a transsex patient needs to balance medications for their CF with other drugs they may benefit from taking to help their body reflect who they are inside. What about what happens when gender nonbinary patients get called the wrong pronouns several times during the space of a single clinical encounter? Consider also the ability of intersex patients to work with providers who really understand their reproductive characteristics and associated care needs—often a tricky issue in integrated CF care for endosex patients as well.

So what can folks working with CF patients do to create space for the considerable sex and gender diversity of

you do to change it? Just knowing that we don’t have to clue people in to our existence, or the fact that our being in the world is a very normal and unremarkable thing, goes a long way for those of us juggling the already challenging demands of managing CF along with other forms of hardship. And being a sex or gender minority is definitely difficult, even for those of us who have considerable privilege on other fronts. We constantly have to remind people that we are real and valid—when we’re not fighting for our legal rights or avoiding physical violence, that is. Do the work for us and show us you’re aware of our existence, even if you don’t know that much about the particular hardships any of

“What about what happens when gender nonbinary patients get called the wrong pronouns several times during the space of a single clinical encounter?”

our community? My own experiences have given me a few ideas on this front. I’m an “agender” adult with CF who works in clinical education. Being agender means I don’t have a specific gender identity. It’s one of many types of nonbinary gender identities. I also do research and evaluation about healthcare services for a variety of chronic conditions. (You can read my last article in the Spring 2020 issue of *CF Roundtable* if you’d like details about my job!) My personal background has given me a lot of insight, through both my own eyes and those of others, into what people who work with CF patients can do to facilitate positive experiences.

Think about us! Reflect actively on who may be left out of particular aspects of CF care, or not feel fully included. Why is this so, and what can

us might face.

Make us visible! Use language on forms that allows people to give you accurate information about their sex and gender identity. And let patients know that you’re open to feedback about how to improve any forms your organization is using! I’ve worked with several different practices to help them update forms with more inclusive language. This process is pretty easy once you commit to doing it, at least if your organization has the latitude to update terminology. Sometimes you have to use forms required by particular government agencies or other entities. That’s okay, too—we often have constraints on our choices, and nobody understands this better than folks who experience some form of social marginalization. In those cases, provide a supplemental

Continued on page 26

form where people can tell you more about themselves than what gets captured in forms for regulatory purposes. Let us know you see us.

Ask us questions! I'll let you all in on a little secret about the trans community: many of us struggle to keep up with changing and expanding terminology about sex and gender, too. Change takes time, and those of us who are approaching midlife now have lots to learn even if we've been part of a marginalized sex and/or gender community for ages. Many of us also may not have realized that we ourselves were sex or gender minorities until we were well into our adult lives! Not everyone who is trans in some way is "born this way"; often figuring out that we are transsex and/or transgender is a long process. In some cases the same might be true for intersex people, too—for example, if a person's hormonal characteristics change substantially with aging. Bottom line: If you want to know something, ask. We'll tell you, and we probably won't make you feel embarrassed for asking, either.

Assess us! Sometimes sex and gender minority CF patients may have particular needs for our healthcare that relate to these aspects of our experience.

Other times, we may simply want to know that anyone working with us knows who we are. It is exhausting when people try to cram us into boxes where we do not fit. For people with CF, this challenge gets compounded because we already spend so much time trying to explain our disease to people unfamiliar with it. So, be proactive and give us the opportunity to tell you about what the best healthcare looks like for our unique needs. This will also help you learn about the immense diversity of our patient community, as no two of us who check both the "CF" and "sex/gender minority" boxes will be the same—or even very similar at all. For example, a new provider might wonder if the particular hormonal medication I take is an indication that I am working on sex transition. It isn't, but that's certainly a good question to ask! I take an older, higher-dose combination hormone pill to help protect my bones from the inhaled steroids I use to keep my lungs healthy. The hormones also help thin my mucus, and likely assist with weight maintenance. In this particular case, my use of hormones has nothing to do with my gender minority status—but for another patient, it might!

Listen to us! This is really the most

important tip of all. Working effectively with sex- and gender-diverse CF patients—and just people in general—depends foremost on actually listening to our voices and taking our contributions seriously. When we take the time to share our perspectives with you, let us do that sharing without being interrupted. Many of us get interrupted enough by having to cough frequently or make an urgent restroom visit! Believe us when we tell you about our experiences of hardship, and about what you can do to make staying healthy easier for us. Your attention, and the respect it communicates, can go a long way even if you have very little technical knowledge about sex and gender minority issues in healthcare. ▲

Dr. Alexandra "Xan" Nowakowski is a medical sociologist and public health program evaluator. They currently serve as an Assistant Professor in the Geriatrics and Behavioral Sciences and Social Medicine departments at Florida State University College of Medicine. They also founded the Write Where It Hurts project (www.writewhereithurts.net) on scholarship engaging lessons from lived experience of illness and trauma with their spouse, Dr. J Sumerau.



Bene factors

BRONZE

Amy Baker
Julie Bombeck
John and Pauline Dinello Trust
Pepper Hartney
Susan Mitchell (in memory of Raymond Corwin)
Nicolas Morency
Nancy Moseley
James and Ann Nash
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Judith Riley (in honor of Steven and Douglas Riley)
Bill and Sharon Thompson (in memory of Melissa Thompson)
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Defiance

This cord that binds me also frees me —
it feeds me oxygen so I can breathe, yet
keeps me closely tethered to the tank.

With this leash, I've become bound, but
I am not trapped.

I feel more vulnerable, however
I refuse to be frightened.

My weakness is now exposed, yet
I will not be conquered.

I am coming to terms with my life,
That means coming to terms with my death —
and finding the beauty in both.

Through it all, I remain steadfastly
D E F I A N T

-M. Compton, 1997



PHOTO BY DEREK POWAZEK

FROM OUR FAMILY PHOTO ALBUM...



ASHLEY WILSON AND HER FRIEND, MILLA, AFTER ENJOYING A DAY OF SURFING.



NATHAN TALLENT; FE TALLENT HOLDING SAMMY; SANTA CLAUS HOLDING SNOWKING; AND NATHAN'S MOTHER, WANDA CARVER TALLENT.



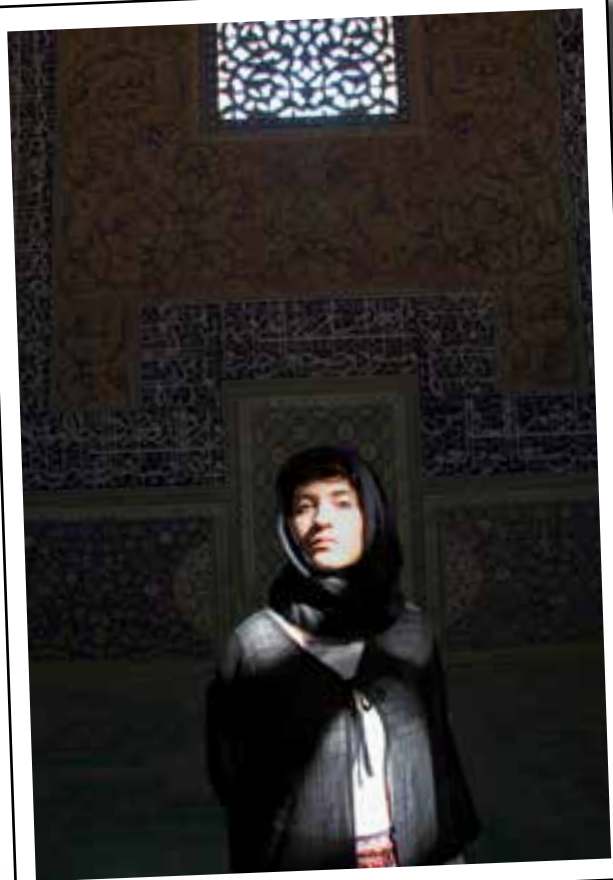
KATHY YODER IN TODOS SANTOS, MEXICO, JANUARY, 2020.



MICA TROTting ON DUKE.



ANDREW KIRBY AND HIS MOTHER, KIM KIRBY.



TANNAZ MOTEVALLI



DANIEL GONZALEZ; MOTHER, IRMA DAVILA; FATHER, RICARDO GONZALEZ; AND BROTHER, RICARDO GONZALEZ.



GRACE KNIGHT



An Interview With Mohamad Elsayyed

By Mark Tremblay

I met Mohamad Elsayyed through a mutual friend on Facebook and was immediately struck by his overwhelming optimism in the face of his illness, as well as the many other challenges he faces. Like most of you, I have met many folks in the CF and substance use recovery communities who have truly inspired me. I have, however, rarely met someone like Muhammad, who is able to genuinely find joy in the moment while simultaneously holding out hope for a better life in the future. After a few Facebook messages back and forth, I knew I wanted y'all to get to know him and his story. Please welcome Mohamad Elsayyed.

Where do you live and how old are you?

I am 21 years old. I'm a Muslim Palestinian refugee with cystic fibrosis living in Bchamoun (near Beirut), Lebanon.

How is your health currently?

My health goes up and down. I currently have 22% lung function. Last year, I was admitted for emergency surgery to remove part of my right lung. I have never received inhaled or intravenous Tobi or Cayston, despite culturing *Pseudomonas aeruginosa*. Also, because of COVID-19, I have to stay in my apartment and am not allowed outside so that I don't get sick. (Note from the author: since this interview, Muhammad was again admitted to the hospital for hemoptysis and released three days later without having received any IV antibiotics to treat his lung infection.)

When were you diagnosed with CF and how did you react when you found out?

Since birth I was shuffled from

“Where I live there are very few patients with CF and no CF specialists.”

doctor to doctor because I was sick all the time and no one knew what was wrong with me. In 2014, when I was 14 years old, I was finally diagnosed with CF based on a sweat test. It came as a huge shock when they told me I had CF. After searching the web and learning more about it, I didn't believe I had it. However, the doctor confirmed the sweat test was positive and I eventually accepted my diagnosis. Where I live there are very few patients with CF and no CF specialists, so I was referred to a doctor with an office nearby who had previously treated another CF patient.

What is your plan of care and what do you do to manage your CF on a daily basis?

We do not have a lot of treatment here so we do what we can. As a Palestinian refugee, I have very limited access to medical care and equipment, so the only treatments available to me are nebulized inhaled saline solution, creon enzymes with meals, and oral

azithromycin three times per week as a preventative measure for lung infections. Roughly six weeks ago, due to the generosity of a not-for-profit organization (CF Vests fO'r Life) operating out of the U.S., I was given a Respirtech inCourage therapy vest which I do twice a day in conjunction with my inhaled medications.

Have you considered or are you on a lung transplant list?

My doctor told me about lung transplant surgery and said if I do not need it now I will eventually; however, it is not available for me in Lebanon. He said in order to have that as an option I would have to travel to another country.

I sought asylum to the U.S. but was denied. I also applied for a visa to go to Belgium through non-conventional channels because refugees are unable to get travel visas. Last year, I finally saved up enough money to pay someone to get me a visa to go to Belgium, where I could apply for asylum through an expedited process because of my refugee status, which would give me access to better health-care and potentially a transplant. After paying for the Belgian visa, I was unexpectedly admitted to the hospital, so I missed my opportunity to go to Belgium and lost all my money.

How has your life changed because of the current COVID-19 crisis?

My doctor told me I am more susceptible to this virus and recommended that I try to stay home as much as possible. Although I do not like staying at home, I know it's best for me to do so in order to stay safe.



MOHAMAD ELSAYYED

My brother helps me get stuff I need.

What are your goals in life? Are they tempered by the fact that you have this disease?

I want to have a good life, house, job, and family. I'd like to have a wife but with all that I read about CF, I'm not sure about a wife.

What motivates you to keep pressing on?

I try to do everything people around me do but in my own way. I try to prove myself. I stay positive and tell myself I can do it and I have the power to this or that.

What do you think about when you're facing your hardest challenges?

My family helps me get through the big challenges I face in life.

What role does religion play in your life?

When we were kids, they told us about Islam in school and left it up to us to choose. Islam is a big deal to me. It tells me that I am strong and I can do it. We all know that God is looking over us and that he will take this disease from us.

What do you do to help manage your stress?

I used to visit my family and friends and play outside. I like social media—Facebook specifically, because it helps connect me to other people.

If you had to say one thing to other CF patients and one thing to your loved ones what would it be?

If I had one thing to say to others with CF, I would say stay positive and don't get discouraged. To my family I'd say a thousand times over, thank you for sticking by my side. ▲

Mark Tremblay is 50 and has CF. He is a director of USACFA. He lives in Albany, NY, with his wife MaryGrace and stepson. His contact information can be found on page 2.

Inaugural CFRD Summit



The Inaugural CFRD Summit meeting is planned for July 6, 7, & 8th, in 2021. The event will be hosted at The Lodge at Whitefish Lake, in Whitefish, MT, 26 miles from the entrance to Glacier National Park. The purpose of this gathering is to bring together thought leaders in the Cystic Fibrosis Related Diabetes (CFRD) community who are advancing research as well as promoting more progressive therapy of diabetes. The Summit program will be comprised entirely of endocrinologists (both pediatric & adult) who are a part of the Cystic Fibrosis Foundation's EnVision CF program, of which there are thirty members.

The mission of the EnVision program is to fund training for physicians interested in developing expertise in the endocrinologic care of patients with cystic fibrosis. The speakers will present their latest research and findings to the attendees. The objective of this program is not only to create a space for the EnVision endocrinologists to come together and share their wisdom with each other, but also to expand the knowledge of endocrinologists who partner with CFRD patients, but who are not fully immersed in a CF care center, thereby enhancing their understanding of the many nuances and variances of CFRD versus T1DM/T2DM.

The Summit will also encourage

the attendance of CF Center directors, with the goal of developing more cohesion, shared understanding, and fluidity in the treatment of the CFRD in the CF Clinic. The Center Directors have a rich opportunity to enhance the teams understanding of CFRD and the critical need for a strong continuum of care as it relates to diabetes management.

Our goal for the Summit is that current EnVision members walk away inspired by their colleagues' research and motivated to move their research and others' research into practice. We hope that endocrinologists who are not currently connected to a CF care center find inspiration to integrate the newfound critical knowledge into practice, as well as understand the need for communication between the providers. CF Center directors should leave with a better understanding of the pulmonary implications associated with CFRD, providing motivation for more expansive communication with patients and care team members, aggressive observation of diabetic potential in each individual they serve and a shared partnership of understanding with the endocrinologist who mutually serves their patient population.

For more information or to receive updates on the virtual training program, sign up here: <http://attain-health.org/cf-endocrinology-summit>



Calling for Social Justice in the CF Community

By Tannaz Motevalli

Disclaimer:

As one of the co-chairs for BreatheCon 2020—an online conference for and by adults with CF through the Cystic Fibrosis Foundation (CFF)—I’m writing this article to share some of my own personal thoughts on diversity and inclusion in the CF community, as well as some insight on how Kasey Raffensperger (my fellow BreatheCon co-chair) and I have been framing this year’s event specifically around promoting and encouraging diversity and inclusion in our community. The thoughts and the opinions shared in this article are my own and not a direct reflection of the CFF or others involved in planning BreatheCon 2020.

The specific events that inspire each of us to pursue social justice are unique to every individual and it is a private journey. Usually, if we are paying attention, a series of events happen and then something, or multiple things, push us over the edge and we are faced with a choice to act or stay silent. For many people recently, it was the death of George Floyd on May 25, 2020. For me, it was two particular events: the release of dashcam footage on November 24, 2015, of Laquan McDonald’s death in Chicago thirteen months prior, followed by the death of Freddie Gray on April 19, 2015, in Baltimore. Laquan McDonald was shot 16 times while walking away from former officer Jason van Dyke. Freddie Gray died seven days into a coma caused by his spine being nearly severed 45 mins after being arrested and placed in a Baltimore Police Department transport van. In 2015, I was living in Chicago pursuing my undergraduate degree. I remember watching as both cities and communities I loved took action and the Black

Lives Matter movement had reached about a year of consistent protests and demonstrations nationwide. For the first time in my life, I was willing to fight for something that did not directly affect me. Laquan McDonald could have been a teen I would later interact



TANNAZ MOTEVALLI

with while working for the Chicago Parks District. Freddie Gray could have been an old friend or former classmate. Or they could just be strangers who, despite me not knowing them, have as much right to live a full and joyous life as me. I am not free until we all are.

So, what does all of this have to do with the CF community? Racism and systemic oppression permeate everything (hence the word systemic) and the work to be anti-racist must be done every day, internally and externally. As a queer person of color with CF, a disease historically known to impact predomi-

nantly White people, I have felt like an outsider to the CF community often. Only in the past several years have I come to recognize and grapple with the fact that racism and other issues surrounding identity have big roles to play in that feeling. From one CFer to another, if you don’t believe racism is a problem in the CF community, you’re not paying attention and you need to listen to POC CFers, especially BIPOC CFers. Race negatively impacted me as a person with CF early on in my life. My mother is a nurse of over 28 years and, by the time I was born, she had taken care of several patients with CF. Within three months of my birth, she knew I had CF. She kissed my forehead on a hot summer’s day and it tasted salty. Along with the saltiness, I was not digesting my food well. She told the doctors and they did not believe her. It took two years of my mother’s persistence and a GI specialist to finally suggest a sweat test. Remember, this was the 90s and, at that time, the majority of CF patients were White. And though things have changed, the assumption that CF was only a White folk’s disease was partially to blame for delaying my diagnosis a full two years. For those with CF or those who have close family or friends with CF, you know that two years without treatment could, in some cases, have been fatal or have severely detrimental health consequences in the long run.

I’m not writing this to tell you all the times I have been negatively impacted by racism and stereotyping, because the truth of the matter is, I can never compare what I’ve experienced to the racism any of my Black friends and colleagues have experienced. I don’t know that reality and I’ll never know. So, as a nonBlack person of color in the wake of another resurgence of the Black

Lives Matter movement, I want to discuss ways the CF community can participate in anti-racist work and the pursuit of social justice.

It's important to consider how the CF community overlooks marginalized identities within the community. Affinity groups built from a shared diagnosis or health status (such as the CF community is) can be extremely dangerous when there is not an intersectional approach to identity. Intersectionality, the framework of identity that sees each individual as the intersection of their identities rather than siphoning each off into separate boxes, is key to recognizing the complexity of discrimination throughout all aspects of life. Without intersectionality, disease-based affinity groups run the risk of policing what qualifies someone as healthy or ill enough to be part of the group. Without intersectionality, the group can consciously or subconsciously align other specific identities to that diagnosis that have nothing to do with the diagnosis itself, rather than see the diagnosis as the common denominator amongst many diverse identities that interact with being a CFer. And this is where Whiteness comes into play.

For years, the physical representation of a person with CF has been predominantly White. This is changing. But because of this, I spent many years not only feeling like an outsider, but actively avoiding anything that had to do with CF and the CF community. When I attended my first BreatheCon event, I remember being shocked by how akin I felt to some of the other CFers I met. Meeting other people with CF who also took issue with some of the ways the community had been represented helped me feel less alone and more interested in participating. Around this same time, I also was making a concerted effort on my part to keep an open mind about the community and the people I was meeting.

Even after all this effort, I still sometimes feel like an outsider. To be very clear, I have not experienced many overt forms of racism and/or stereotyping among other CFers. What I have experienced are covert forms of racism and/or stereotyping, such as the lack or minimal number of resources and opportunities tailored to non-White and non-straight people with CF, hetero-centric discussions around sexuality within the community, and simply not seeing a face or a name that looked like mine.

I feel extremely thankful that I pushed myself to get more involved in this community. I am extremely thankful that I have the opportunity to co-chair BreatheCon this year. And, in the wake of so much call for change and social justice, I have to acknowledge that I am tired of having to push myself and encourage myself to become a part of a community with which I may not always feel aligned. I'm happy to do it, but, at the same time, I wish I didn't have to try so hard. CFers who feel like outsiders should not have to convince themselves to be on the inside—the community should actively welcome and include them.

I believe it is true that some are more aligned to change, whether through personality or instilled beliefs. This is not an excuse for complacency or to reason with bigotry. It is a recognition that each person has different steps to take to enact change in themselves and in the world around them; meanwhile, the duty to make change relies on us all and must be met.

In trying to strike a balance that gives everyone space to take their private journey toward social justice and still insist we all must do our part, planning BreatheCon has been an amazing opportunity to think critically about what it means to be different together. At this year's BreatheCon, we hope to ignite conversation about what community means to all of us and brainstorm

how to build a more welcoming environment for people of all experiences and backgrounds. We will also create spaces for people to connect through mutual interests and experiences rather than just a mutual diagnosis. We recognize that race and ethnicity, sexual preference and gender identity, as well as socio-economic status are heavy topics to work through and can be triggers for some. We welcome people to think about the ways those identities shape someone's unique experience of life with CF and recognize that we may not all be able to understand one another completely. The truest form of empathy is not just putting yourself in someone's shoes, it's putting yourself there and recognizing that's still not enough to really know what it's like to be them and that's okay.

This is just the beginning. There is so much work to be done, and I hope that for some folks with CF BreatheCon can activate you to not only connect with other people with CF, but to also think about what social justice can look like in the CF community and start building some change.

For adults with CF who wish to join the BreatheCon 2020 workgroup, please contact virtualevents@cff.org. ▲

Tannaz Motevalli is an Iranian-American artist, writer, and researcher living in Baltimore, MD. She works full time as one of the exhibition coordinators for the History of Medicine Division's Exhibition Program at the National Library of Medicine in Bethesda, MD. She is also pursuing a Master's Degree in Library and Information Science at the University of Maryland, College Park. Outside her career, Tannaz enjoys singing, crafting, cooking, and spending time with her human and feline friends. To reach out to Tannaz, you can reach her via email: motevallitannaz@gmail.com. You can also follow her on her Instagram, @human_grl, where she occasionally posts about social justice, disability rights, and life with CF, but also just life-stuff.



Being A Transgender Person With CF

By Mica

Inhabiting a body is a tricky thing for a person with cystic fibrosis. The discomfort and pain of disease, the hassle of doing intense daily treatments, the anxiety of what to do when you get sick—it's a lot to handle. Our bodies are medicalized, pinched, and prodded on a regular basis. Identifying as a "sick person" has its own existential questions. Having to face one's own mortality from a very young age, as many of us do, can be challenging as well as spiritually enlightening. I have long referred to the hospital as my "vacation home." And yet, it has taken many years for me to feel safe and respected there. I used to be called the wrong name. I used to be "miss'd" instead of "sir'd." I sometimes feel ashamed to ask that I be called by my correct name, or to be referred to using the correct pronouns. Sometimes I'm asked erroneous, invasive questions based on the fact that I had an F in my chart next to my name instead of an M. Once I was even verbally assaulted by a hospital orderly when I asked him, politely, to respect my identity. For transgender people, especially people of color or disabled people like myself, the medical system can be a terrifying, even dangerous, place, requiring immense skill and bravery to navigate. Being in a body is all the harder when society creates so many barriers to being seen and feeling safe.

But I am one of the lucky ones. I live in a progressive place (Vermont), I have received the gender-affirming care that I've needed, and I have an amazing medical team that respects and even celebrates my identity. It's been a long and difficult journey to acquire the supportive environment that I have now. As such, I want to briefly discuss



MICA

a few things that CFers and their caregivers should know about living with this disease and being transgender and offer some suggestions for how to improve the care that we receive. My health has improved tremendously since my physical transition, and I want to reassure other trans CFers and their caregivers that not only is this possible, but it is safe, and it is important.

Being Transgender

Being transgender is a natural state of existence, as is being homosexual, and both are ubiquitous in the plant and animal kingdoms. It is only human societies, with their arbitrary definitions of what is "right" and "wrong," that have any issue with it. Being trans is not a disease nor a disorder; it is not a "sin" nor a crime; it is simply a natural state of being. Transgender people have existed in every time, in every place, and in every culture on earth, though we have often been made invisible by social discrimination.

Because of our unique positions in between the so-called sexual binary (which, even now, [the latest science is debunking](#)), many traditional cultures have given us roles as holy people to advise the spiritual health of their com-

munities. Some contemporary examples include the Lakota Winkte, the Hawai'ian Mahu, the Hijra of India, the Waria of Indonesia, and so on. An excellent book on gender diversity is *She/He/They/Me: For the Sisters, Mistresses, and Binary Resisters* by Robyn Ryle. Also, check out [this map](#) of traditional third genders around the world. I am writing a [historical fiction series](#) on this topic, as it's something I believe to be important and inspiring. I hope that Western societies will someday regain their lost acknowledgment of the spiritual importance of the transgender experience.

In addition to social discrimination, trans people can be in danger due to gender dysphoria (the experience of feeling uncomfortable in one's body as a result of one's sexual features not matching one's gender identity). The suicide rate among untreated transgender teens in the U.S. is scarily high. Dysphoria cannot be helped in any other way besides gender-affirming medical care (which includes hormones and surgery) and social inclusion. Psychotherapy, religion, or anything else cannot be successful by itself (trust me, I've tried it!), though these can be supportive to trans-affirming medical care. My own struggle with dysphoria caused me immense emotional and physical suffering until I finally received the medical care I needed at age 26. I am so, so glad that many more trans kids growing up these days will have better access to the care they need, so that they might avoid the kind of suffering I endured early in my life.

Navigating the Medical System

When it comes to being trans and having CF, there are some extra complications. Many trans people avoid medical care due to the common, painful discrimination we experience in hospitals

and doctors' offices. But as a CFer, I cannot avoid medical care. The way that modern Western medicine pathologizes and objectifies the human body is particularly unhelpful, and it is why complementary and holistic medical care can be much more accessible to trans people (I myself became a clinical herbalist). Some things at my hospital have improved since I transitioned, but if I go to other specialists who are not trans-literate, it can be a jarring experience.

Some very basic things that need to be done to support trans people in the medical system include: noting in the medical chart and using the person's preferred name (if they have not changed it legally); noting and using their correct pronouns; using sensitive and respectful language related to gendered body parts; asking and recording the person's gender identity (which is different than sex); and avoiding making medical assumptions based on the M or F next to the patient's name. It is important to ask medical questions related to the organs and hormone dominance a person *currently* has, not just questions related to the M or F designation in the chart (some trans people don't want to or are unable to change the M or F for other reasons). Also remember that many trans people (myself included) are non-binary, meaning that they do not identify as either man or woman, male or female. In some states we can now choose a third gender option on the birth certificate and/or driver's license; therefore, medical records and insurance companies should also reflect this additional choice. These things will hopefully encourage larger changes in mainstream medicine, moving away from statistical medicine toward more personalized care. Medical providers cannot rely on the M or the F alone to make assumptions about a patient's sex, gender, physical health, social status, or medical risk factors. It requires greater nuance and asking better ques-

tions, both of which mainstream medicine needs to improve upon for all patients. Medical record software must be updated to reflect the patient's pronouns, preferred name, and gender identity. As gender and sexual diversity increases, in cisgender and transgender people alike, these changes will make medical care more respectful and more precise for everyone.

There are [excellent resources](#) out there now to help educate medical professionals on how to best care for trans patients. It is important to note that every medical professional is responsible for educating themselves on these issues because providing uneducated care can, and does, cause harm. This harm is both unnecessary and avoidable. I believe that these changes will benefit every single patient of every gender identity. When healthcare providers make it a habit to think twice about making gendered assumptions and judgments about patients, I believe this will also help tackle the misogyny inherent in the medical system.

Surgery and Hormones

When I first decided to pursue physical transition, my family and healthcare providers had some concerns. Initially, some family members were concerned about "top surgery" (i.e., removal of mammary tissue and chest masculinization) due to its proximity to my lungs. However, for me this wasn't a choice—I had been waiting for this opportunity for almost 15 years. Luckily, my doctors understood my need to do it (one of my docs is trans himself) and they helped me jump through the many bureaucratic hoops. I knew how much this surgery would improve my life. A difficult part of the process was trying to explain my decision to family members who didn't understand. While no one's opinion was going to stop me from doing it, it was hurtful when some expressed their concerns or dissent. However, my immediate family was supportive (my

sister was my strongest advocate and I don't know how I could have done it without her!) and eventually I won everyone over. Many trans-masculine people like myself bind their breasts to slim the chest profile. I knew that as a person with severe lung disease, anything like a binder that restricted my breathing would be a very bad choice, so I had to do without it. Chest masculinization surgery may be extra important for CF trans-masculine people for that reason, because binding can be so harmful to the lungs. In addition, I find that exercise and chest physical therapy is much easier and more comfortable without breasts (side note: the Vest should make a more comfortable model for people with breasts!).

A few months before my top surgery, I started injecting testosterone ("T"). Using T was less important to me than the top surgery, but I decided to try it and see how it felt. Turns out, I loved it! It gave me so much more energy, enhanced my mood, and helped me gain weight in muscle mass. With the new energy that I gained, I could exercise more, which led to greater muscle mass, which made exercise easier and further boosted my energy, creating this amazing positive feedback loop that really helped my lungs stay clear. I do believe both testosterone and the top surgery have helped improve my CF. My functional life capacity has increased significantly, not to mention the improvement in my mental health! After being on T for a few years, my exercise tolerance increased to levels I had not experienced since high school! Even though my lung function was slowly declining (until starting Trikafta last fall), my functional capacity was improving. Remember friends: FEV1 is not the sole indicator of your health! Don't let the number bum you out!

I began my transition about 4.5 years ago. The day after my top surgery

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Diversity In Healthcare

By Ashley Wilson

Because I am mixed race, I have been asked about my experience as a person of color within the CF community. I know several people over the years would be surprised to learn that I have cystic fibrosis because I am half-black. At one time, CF was thought of as a Caucasian-only disease because it originated in Europe. Through social media, I personally only know about six people who are both black and have cystic fibrosis. I believe cystic fibrosis is prevalent in other races today because interracial marriages are becoming more accepted. CF doesn't recognize skin tone. In the end, we are all a melting pot of different races from around the world.

As far as healthcare, I do not feel that I have been treated differently due to my race. I have been treated fairly as a patient. I was diagnosed with cystic fibrosis at birth in Denver, Colorado. We moved to California when I was nine years old and I've lived there ever since. I currently have a team of specialists here in California. I have not had a black doctor; however, I have had several doctors of many varied ethnic backgrounds and they have all treated me with respect and professionalism. I have had one respiratory therapist who was black and a few nurses during hospital stays. I think having a diverse healthcare team has opened my eyes to understanding and appreciating different cultures within my community. More diversity in healthcare is helpful and comforting—you can relate to someone who looks like you and possibly has a better understanding of your background. I admire people of different races and I am especially proud of those who are black and studying to go into the medical field because it is broadening

“ I appreciate when my team takes the time to get to know me as a person first, understanding that I am someone who has a variety of interests outside clinic appointments. ”



ASHLEY WILSON

the diversity of races in healthcare.

Diversity in healthcare has come a long way from where it used to be, but there are always improvements that could make patients feel better understood. Most importantly, for a patient with chronic illness, is to have a team of professionals who are genuine in nature, patient, and who are good listeners. I appreciate a team that explains things clearly and has expectations of me to follow protocol. I also appreciate a team that understands the com-

plexity of this disease and recognizes that I am a person who desires to live life, despite the fact that I live with CF. In other words, I appreciate when my team takes the time to get to know me as a person first, understanding that I am someone who has a variety of interests outside clinic appointments.

I have found that social media and cystic-fibrosis-related-foundation websites have been the greatest resources outside of my healthcare teams. I have been extremely fortunate to have parents who strive to learn about this disease and support me in making wise decisions about my health. They have provided me with excellent health insurance and the ability to see highly skilled doctors. I do not take this for granted as I know that healthcare and insurance are not available nor affordable for all. ▲

Ashley Wilson is 22 years old and has CF, CF-related diabetes, and CF liver disease. She lives in central coast California. Ashley is currently pursuing a Bachelor's Degree in Cinematic Arts and Technology and hopes to share others' stories through documentaries. Outside of school, Ashley enjoys surfing, adventures with friends, and working as a YouTube creator. She also is a producer and contributor on various podcasts. You can find her personal YouTube channel at Ashley's Roses.

Meet A New Director — Grace Knight

I am so excited to join the USACFA board this year! For the past few years, I have enjoyed reading *CF Roundtable* and am a recipient of the Lauren Melissa Kelley (LMK) scholarship. I am 22 years old and have recently earned a B.A. with distinction in English from the University of Pennsylvania. In the fall, I will be attending University of Texas School of Law in Austin, TX. I spent many months in college fighting nontuberculosis mycobacteria (NTM) and am grateful to have made it to the other side. I am currently on Trikafta and am constantly amazed at the improvement I have seen in my health while on the drug. It has opened the door to a brighter future for CF patients.

I was diagnosed with CF when I was one-and-a-half years old. At that time, there was very little knowledge

about CF in my hometown of Tyler, TX, and we did not even have a local CFF chapter. My family and I have



GRACE KNIGHT

worked hard to create awareness of the disease in Tyler and are proud to now have multiple local fundraising events throughout the year associated with the CFF. I have two younger twin brothers, both of whom are CF carriers only. After my diagnosis my mom did in vitro with genetic testing to get pregnant with them.

While at Penn, I took many classes in creative writing and am really excited to be able to use that knowledge to help keep the CF community informed. In my final year of college, I completed an honors thesis which focused on telling the stories of CF patients. It was through that project that I learned of the wisdom and strength present within our community. I am constantly inspired by this community and am honored to fight this disease with you. ▲

MICA continued from page 35

I felt reborn! I wish I had done it *way* sooner, but I am so grateful that I finally received the care that I needed. My life is so much better since my transition and I am extremely grateful to all the people—healthcare providers, family, friends, and community—who helped me make it happen!

Trikafta and Hormones

Last fall, I started Trikafta and experienced another rebirth! My FEV1 is higher than it's been in six years! I've regained at least 10% of my lung function. I basically don't even cough anymore. I can't tell you how miraculous it's been. I feel like I might live decades longer.

With that said, everything has its complications. The combination of elexacaftor, tezacaftor, and ivacaftor in Trikafta results in strong inhibition of the liver enzyme CYP3A4. That liver enzyme is responsible for metabolizing many drugs, herbs, and sex hormones

(i.e., estrogen and testosterone). Both Trikafta (and Symdeko before it) resulted in my serum testosterone level rising way too high, so I've had to get my testosterone level checked regularly and adjust the dose. I've had to cut my testosterone dose in half since starting Symdeko and Trikafta. I've also gotten more acne recently as a result of the hormone fluctuation. But both testosterone and Trikafta are too valuable to me to stop either, so I just have to check on my testosterone levels periodically. For trans CFers and their caregivers, be aware that you will have to keep tabs on hormone levels and reconfigure the dose while on CFTR modulators.

Gratitude

I am so grateful to so many people who have allowed me to thrive as I do now. I am grateful to my transcestors who have fought so hard for the freedom of transgender people in the U.S. for so

many decades, through such strife and hardship. I am grateful to my health care providers who have helped me gain the care I've needed. I am grateful to my family and community for supporting me in my transition. I'm grateful to the scientists at Vertex who have created a medicine that has totally transformed my life. And lastly, I'm grateful to CF Roundtable for providing this space to discuss the issues that are important to queer and transgender people with CF. I am so happy that we do not have to be invisible anymore!

If you're trans and have CF, let's be in touch! I'd love to know more people with this unique experience, and maybe help support each other in our struggles. I hope to hear from you! ▲

Mica (they/them) is 32 years old with CF. They are a clinical herbalist and author living in Vermont. You can find them on their website at www.cfnaturalhealth.com.



My Views On Racial Inequality

By Tré LaRosa

Spend five to ten minutes searching the literature for Black people with cystic fibrosis and their health outcomes. To say it is wrong that there hasn't been more extensive research to understand the disparities for Black people with CF is a massive understatement. It is more than wrong; it is a demonstration of racial inequality and inherent racism in the healthcare system in the United States. We, as a community, must acknowledge our privilege. Cystic fibrosis is typically regarded as a white disease. Look at federal funding and outcomes for those with sickle cell disease, typically regarded as a Black disease. The similarities between the incidence, prevalence, even life expectancy, clinical severity, and underlying Mendelian genetics between these diseases are remarkable. While there very well may be many explanations, it's hard to skirt around Occam's razor. Outcomes in the Black CF subpopulation are poorer than in the white population, and sickle cell disease is not nearly as well known or understood as cystic fibrosis. It stands to reason that in order to improve outcomes for these communities of people, we must find the causes and root out systemic biases that disproportionately harm Black people.

As we see nationwide protests about racial inequality, I feel compelled to say this: I stand with you and I am listening. I am fortunate to be part of the only nonprofit in the United States led solely by people with CF. USACFA provides resources and support for everyone in our community regardless of their ethnicity, race, sexual orientation, gender identity, religion, political affiliation, or anything else for that matter. This very issue of our CF Roundtable newsletter is dedicated to

diversity within the CF community. It is incumbent on all of us in the CF community to work for and with each other to improve the disparities within marginalized subpopulations in the CF community so all of those with CF have equitable healthcare access, outcomes, and expectations until the day when CF is cured for all.

Merely pointing out these disparities isn't enough. Understanding of



TRÉ LAROSA

the root causes is critical in order to propose solutions for mitigating these disparities. Poorer care and access to care that Black people in the U.S. face, combined with Black patients' underrepresentation in clinical trials and other factors, makes clear that systemic racism impedes the improvement of health outcomes witnessed across the spectrum of those with CF.

In the CF community, the "final ten percent" consists of a subpopulation of patients who are not currently eligible for approved modulators. These people either have an unknown mutation, experienced adverse effects of

approved modulators, or they have mutations that will require more complicated therapies to fix. To draw a slight comparison, those of us that have at least one delta F508 mutation are fortunate as it is the most common mutation, and therefore we have benefited from the most research and funding, effectively reaping the most health benefits. Thus, the phrase "F508del privilege." While it's true that clinical severity can vary across all different mutations, this privilege is hard to deny. Combine this privilege with being white in the U.S., and it only confirms my suspicions that the glaring disparities among white people with CF and Black people with CF is at least partially due to systemic racism in the U.S. healthcare system, in the way the way federal funding is allocated, and in the way CF awareness is spread.

The first step to improving these disparities is to bring the conversation to the forefront. Our community, as much as we don't want to admit it, is not immune to racial inequality. Improving these disparities is only the first step. We must seek out to entirely mitigate them. *I am honored to be a part of the USACFA board. I like being part of an organization that is passionate about highlighting the diversity in our community. I believe that the world will not be better until **every member of our community has an equal and equitable chance at the best life possible, especially one free from cystic fibrosis.*** ▲

Tré LaRosa is 26 years old and has CF. He currently lives in Cincinnati, OH, but will soon be moving to Pensacola, FL. He enjoys writing, reading, exercising, playing basketball, and exploring with his golden retriever/basset-hound mutt Duncan. His contact information is on page 2.

BOOK REVIEW

How Have I Cheated Death?

A Short And Merry Life With Cystic Fibrosis

By Tim Wotton

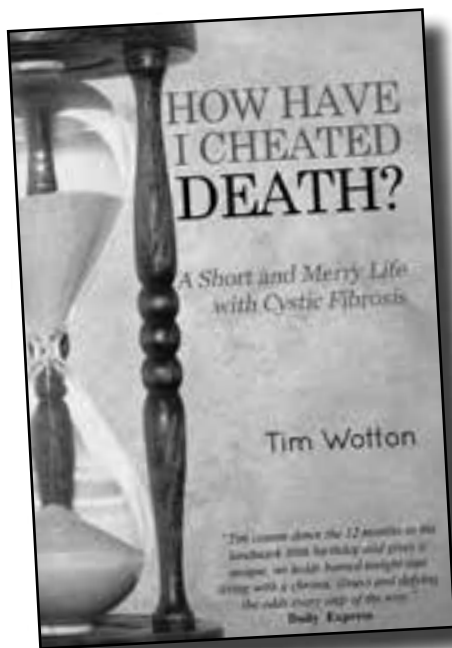
Reviewed by Andrea Eisenman

Welcome to a year in the life of Tim Wotton, an Englishman with CF, whose goal is to reach the age of forty. His book is a compendium of his personal week-by-week written with flashbacks to his past. After seeing so many people in England who don't live long enough to celebrate their 40th birthday, we learn that he is 39 and has set his sights on achieving this seemingly unlikely milestone birthday. Early in the book he says, "[f]or most of my life, thirty seemed unachievable and forty felt an impossible age to reach."

Throughout, he discusses the many friends with CF who didn't make it into their 30s, all the while wondering why he was still alive when they didn't survive. Despite his fixation on death, he also focuses on life, or so it seemed to me. If I had to guess why he is still alive, I would say he never gives up and tempers everything with a good dose of humor. He definitely pushes himself physically when it comes to exercising and, when he was younger, drinking and partying.

We go on this journey with Tim to see the highs and lows of his everyday life. Many with CF themselves will relate to the trials and tribulations of doing treatments, being diagnosed with diabetes, being hospitalized for intravenous antibiotics, and trying to balance a social life and work, all while being married and having a son.

In his narrative, he gives some background history growing up with both a fraternal twin and an older brother. Neither of his siblings has CF. His father brought all of his sons up



“ He is open about his struggles and is pragmatic about his health.”

playing the sport he himself loved, field hockey. To this day, Tim still plays hockey whenever he can and jogs to stay in shape. He hopes to pass his love of the game to his son when he is old enough. He briefly mentions his excessive drinking after many hockey tournaments and how he ultimately learned from his overindulgence.

How Have I Cheated Death? opens with how many pills Tim takes in a day, how he does his nebulizer treatments, and how he coughs so hard he vomits before he's even had breakfast! Then, he's off to work a full day only to return home and do more treatments before bed. Rinse and repeat. He compares it to the movie, *Groundhog Day*. He is open about his struggles and is pragmatic about his health. It is a relatable book about living with CF—all it takes from us

and what it gives in return: perspective and knowledge and sometimes a spouse and child to make the struggle even sweeter. For Tim, that was the case.

Spoiler alert: Tim achieves his goal of reaching 40. Along the way, he shares his feeling of working full time to provide for his family, going on family vacations, how his parents raised him, getting married, and playing hockey with a few teams of varying ability levels. He delves into the difficulties in conceiving children and ultimate success through IVF. He recounts the emotional and physical toll the process took on him and his wife. He shows great sensitivity on

how it was harder for his wife who was taking fertility hormones each time they tried (unsuccessfully) for a sibling for their son.

This book might be for you if you like reading about other people's journeys with CF for contrast or feelings of unity. I felt Tim's story was universal to those of us with CF, especially those who, like Tom, wonder if they will live to their goal milestone birthdays. It is inspiring and reminds us that, while we all have the same disease, we all cope in our own ways at various ages and levels of maturity.

Those unfamiliar with the author, Tim Wotton, will enjoy his blog, *Counting Up to 50* (<https://timwotton.wordpress.com/>). Tim is at it again, writing about trying to reach his next goal of a half-century. My bet is, he will make it and well beyond. ▲



IN THE SPOTLIGHT

With Nathan Tallent

By Jeanie Hanley and Andrea Eisenman

Nathan Tallent is one of the sweetest people. He lives in Lexington, Kentucky, with his wife Fe, whom he married after a whirlwind romance. He's very close to his mother, who has played a key role in his life and health. He has a number of degrees—an A.A. in Computer Information Systems, a B.A. in Christian Ministries, and a Master's of Science in Hospitality Administration Management. His passion lies in his work in the hotel industry, and he has often held down several jobs at once. Nathan has one of the strongest work ethic I've ever seen. Unfortunately, he was recently laid off from his job as a Guest Service Champion due to the COVID-19 pandemic. Despite this and other challenges in his life, he maintains a great attitude and a very positive outlook on life. We were thrilled to interview him and learn about his life experiences and coping. Meet our newest star. Spotlight please!

How old are you?

I'm 39 years young and coming up on my 40th birthday in July, which is a milestone for everyone, but especially for those living with cystic fibrosis. As I approach 40, I don't associate that number with the "over the hill" saying, but as an incentive to keep staying healthy and living life to its fullest!

Where were you born and how were you diagnosed with CF?

I was born in Kentucky and grew up in my hometown of Albany, KY. Soon after birth and because I did not gain my birth weight back, as is the norm, I was diagnosed with failure to thrive, milk allergies, and chronic ear infections. The doctor tried to change my formulas to no avail. I continued to vomit as soon as I ate and cried continuously because I was hungry and not



NATHAN TALLENT WITH HIS LATE DOG, SAMMY.

able to get the nourishment I needed in order to gain weight properly.

After four months of not improving, I was taken to a family doctor. Mom told him I tasted of salt when she kissed me and that I vomited and cried immediately upon drinking the formula that I could not properly digest. He sent me to a hospital in Nashville, TN, to assess my health situation, where they shrugged it off as if I had a common cold! They gave me too much medicine that, in turn, caused more complications. As a result, we went back to my family doctor who referred me to the University of Kentucky (UK) CF Clinic in Lexington. I was given a variety of tests, including a sweat test that confirmed my CF diagnosis. I was then four-and-a-half months old. All future follow-up treatments thereafter were with CF doctors and we continued all follow-ups, suggestions, and consultations as prescribed by my family doctor.

What health problems have you had with CF?

I have two rare mutations: c.1585-1G>A (f/k/a legacy mutation 1717-1G>A) and c.982delA. Fewer than 1,200 patients worldwide have the first mutation, while the latter mutation is so new that it has yet to be identified in the CFTR mutation database. This combination of mutations, at least for me, results in seemingly less severe CF complications—I haven't been hospitalized for a CF tune-up. However, I've always had allergies. I've been on a medication for many years for liver disease prevention, enzymes to digest my food, and many fat-soluble vitamins (e.g., vitamin D with calcium). I've had issues with joint pain over the years, especially as I age. Many years ago, I cultured *Pseudomonas aeruginosa* in my lungs and therefore started inhaled Tobi for many years to combat the *Pseudomonas aeruginosa*. I also take Pulmozyme to break down and thin the sticky mucus in conjunction with using the Vest for CPT. Unfortunately, my two rare mutations are not responsive to modulator therapies, like Trikafta.

Who helped you with your CF care growing up?

My mom went above and beyond in caring for me throughout my childhood and into my teenage years and beyond. She always made sure I had the proper nutrition, took all my medicines, and did all my breathing treatments, including percussion on my chest and back so as to easily get the mucus out of my lungs. I was able to have a Vest machine to do this for me later in my childhood.

What is your occupation?

I worked for Walmart from March 2010 until August 2019, when the company eliminated my job role at all

stores. I was the Training Coordinator and always loved helping to train people. However, I've worked in the front office of various hotels since 2003 and that's my true career passion, because I can always help guests!

Growing up, I stayed at The Campbell House Hotel overnight for all of my trips to Lexington for routine CF clinic visits. It was such a coincidence when I landed a job there in May 2016 as a front desk agent, just a few months after it became a Hilton Curio.

I regularly held two jobs for many years since I entered the workforce as I love people! I received a promotion to Guest Service Champion when I became a full-time employee after leaving Walmart in August. Shortly after that, COVID-19 quickly became an immediate and serious threat, especially in early March in Kentucky, where we live. I tried to take a leave of absence/short-term disability on the orders of my CF doctor, but everything went so fast with layoffs in the hospital industry that I was laid off instead.

How difficult was it to fit in your CF treatments with work?

It was sometimes challenging to get all my prescribed treatments in while holding two or three jobs at a time. Yes, you read correctly! At one time, I worked at Walmart, at a hotel, at the U.S. census bureau as an enumerator, and as an on-call substitute teacher—all during a four-month time period. Recently, when working at The Campbell House, it was hard to fit in my Vest treatment in the mornings, since I had to be at work at 7 a.m. It was much easier to do it at night, before bed, with my other treatments, such as my nebulizer.

How did you meet your wife?

We have been married since June 7, 2009. I met my wife on eharmony.com in January 2008. After communicating extensively with each other during the first few months via chat,

phone, and video, I flew over to Taipei, Taiwan, with my mom, to visit her in September 2008. Fe, who is Filipina, was working in Taiwan at the time. Our love is a true love story—I proposed atop the world's tallest building at the time: Taipei 101. Mom and I flew back to Kentucky, where I began the fiancée visa petition process to help Fe immigrate to the U.S. from the Philippines. She was approved fairly quickly and I traveled to the Philippines in April 2009, returning to Kentucky with her.

Did you want to have children?

We had discussed wanting children when we got married. However, we couldn't naturally conceive a child thanks to CF. We prayed and talked about it, trying to decide between IVF or adoption. We finally decided the adoption route was the best route, as we didn't want to risk having a child with CF as well.

What happened when you tried to adopt?

We wanted to adopt my wife's niece, Nora, since she was living in poverty in the Philippines and we knew her background well enough to know she didn't have any underlying health conditions. She was nine years old when we started the process in July 2017 and was the youngest of nine children. My wife and I were approved for adoption; however, the Philippines' Department of Social Welfare conducted home studies at Nora's home and ultimately declared her ineligible for adoption because her parents were still alive. We appealed the decision twice over a two-year period, but the adoption was denied both times.

What is your relationship with your niece now?

Our relationship with Nora now is difficult to maintain, as communication is limited due to spotty wifi, but she is also busy with her schooling. We keep in contact via phone and Facebook

as much as possible. We do pray one day, when she graduates high school, that she will come to Kentucky and go to college here. Hopefully, this will lead to her being able to immigrate to the U.S. and we can finally be together. For now, she is considered our godchild and our "unofficial child."

Tell us about your fur child.

We actually went through an adoption in our first year of marriage, much earlier before we actually tried to adopt our niece. We adopted a loving Bichon Frisé, named Sammy, in 2010. He was born in 2004. He became more like our son, since we didn't have a human child. He gave us so much love over the years and we took him to as many places as we could. He got to experience the ocean and many road trips. We celebrated his Sweet 16 birthday on March 28 of this year with KFC chicken. Sadly, Sammy passed away on April 8, from seizures that began the prior day. We miss him so much and it's even harder not to have him around during the coronavirus pandemic to cuddle with and love!

Do you think about what work you might do in the future that would minimize your exposure to COVID?

I will feel safer when I receive the vaccine. I really miss my hotel and doing what I do best, but I understand the high-exposure risk working at the front desk with guests directly in front of me. I'm not sure when I will return, as it depends on the progress of the vaccine, but I do know I want to go back. If I did have the opportunity to work from home, I've thought about being a reservation agent for the Hilton chain.

Where is your favorite place to travel? Where would you be right now if you could be anywhere?

I've been to 19 countries and 43 states. Since my wife is from the Philippines, I've been there five times

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LIVE OUT LOUD

Unity Under Tension-Filled Times

By Lara Govendo

Unity during hard times can be challenging when we're all divided.

The approach changes when the goal is unity.

I see trees of green, red roses, too
I see them bloom, for me and you
And I think to myself...
What a wonderful world
—Louis Armstrong

When's the last time you paused and looked around? The world is busy distracting us—it's become more challenging to drown out the noise. Media draws us into the latest saga. Influencers tell us how we "should" think, feel, or respond. Division runs deep as we are continually labeled into categories of political parties, religion, and value systems.

Most of the world is in despair and overwhelmed with the magnitude of current events. What can we do on an individual basis? Is there a solution when it appears that we have no personal power? Our minds are being pulled in numerous directions. Where is our focus? How do we come together when we are so divided?

It's becoming more apparent these days how much judgment runs rampant in our society. Everyone is a so-called "expert" on social media. Personally, I've witnessed others attacking each other when they disagree. My heart breaks with every hurtful word thrown at one another.

Through the lens of mental health and personal experience, I've learned the power of conversation with the

intent to understand, rather than trying to win someone over to my side. I've found that listening with undivided attention to someone else's experience rather than anticipating the conversation, and therefore my reply, has proven more effective when the goal is unity.

We all have a point of view. Our perspectives are different given personal life experiences, values, and influences. This can be a beautiful difference, if we allow it to be. It's easy to fall into the trap of assuming our perspective is the right one and there is only way to view the world. Getting along with each other is possible even when we disagree. Calm, meaningful conversations are more effective when discussing issues with others who have a dif-

fering viewpoint. As more events unfold throughout this season, we have two choices: we can jump on the group bandwagon—stating personal opinions and becoming defensive when someone disagrees or we can use our voice to connect with others, invite conversations from different points of view, and create a safe environment for each other to share our hearts.

Our words have the power to hurt or heal others with a simple sentence. Unfortunately, our society has been desensitized to the effects our words have on others and ourselves. Additionally, it's become easier to say what we want, often without consequence. Often times, we use our screens as a means to say anything that comes to mind without thinking about the consequences. We aren't held accountable for our words because social media is essentially an unsupervised playground.

Our kneejerk reaction is thinking the world needs to change. That's partly true, but it's not the whole truth. Upon self-examination of our own hearts, we need to look within before we look around us. Assessing our thoughts, words, actions, and reactions is where we need to start. It's easy to point fingers at others; however, it's personal growth to look at what needs to change within us first.

Everything we hear needs to come through an internal filter. What is the source? Can it be trusted? Is it biased? Gathering information without partiality is a challenging research project, one that we each have to perform on our own. Depending on others and their own personal filters is not hold-

Unfortunately, our society has been desensitized to the effects our words have on others and ourselves.



LARA GOVENDO

ing ourselves accountable.

We have a moral obligation to ask ourselves, what is the goal of sharing our opinion? Is it for hostility or harmony? If we are handing out hate, hate will return to us. On the flipside, if we are handing out love, love will return to us. The choice is always in our hands.

The challenge for all of us is to open our eyes to that which is happening outside of our own circles. Tunnel vision of our personal circumstances leaves us disconnected from the world. What we've witnessed is people's cruelty toward one another. Nobody cares until it affects us or someone we love. How can we expect other people to care about our cause if we don't care

about theirs, too?

It takes community to accomplish any great feat. We were created to be a part of one another's lives. Not just the celebratory moments, but the struggles, too. Creating communities that welcome everyone is where we begin to make room for others to bring their individuality in.

Let us humble ourselves, leave judgment at the door, and approach the doorway with love. In doing these simple acts, we welcome others into our lives, rather than shutting them out. Together, we accomplish far greater than we ever can alone. Unity during hard times can be challenging when we're all divided. However, we

also have the ability to come together to accomplish the same goal: love all of our neighbors. ▲

Lara Govendo is 33 years old and has CF. She lives in Vermont as a wild, adventure enthusiast who holds a Master's Degree in Mental Health Counseling. She writes about living out loud and develops educational programs to restore hope to those in need. Thanks to her double lung transplant in 2017, you can now find Lara traveling on the regular, exploring the glorious outdoors, and belly laughing with her loves. You can find her online at www.laragovendo.com (and on Facebook and Instagram at "Lungs4Lovey." Her email is lgovendo@usacfa.org.

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currently being evaluated in a Phase 1/2 trial (NCT03375047) called RESTORE-CF. The study aims to enroll at least 40 adults with CF, who will be randomly assigned to receive either a placebo or one of several doses (8–24 mg) of MRT5005 administered by nebulization. The trial's main goal is to assess the safety and tolerability of MRT5005. Effects on lung function (as measured by forced expiratory volume) will also be assessed.

<https://tinyurl.com/ydgxchrb>

AND

<https://tinyurl.com/yb6nqss6>

CF Infection Candidate AB569 Kills Resistant Bacteria, Study Finds

AB569 is safe and effective at killing bacteria resistant to traditional antibiotics, in particular *Pseudomonas aeruginosa* (*P. aeruginosa*). AB569 is a combination of acidified nitrite and disodium ethylenediaminetetraacetic acid. Both compounds are believed to work together for a more potent bacteria-killing effect. Researchers tested AB569 in human cell lines and found the agent was not toxic when used at the concen-

tration needed to kill bacteria. The compound effectively killed *P. aeruginosa* biofilms, bacteria aggregates that act in a coordinated way to evade a host's defenses and acquire resistance to treatments. The therapy received orphan drug status from the FDA for the treatment of CF patients infected with *P. aeruginosa*.

<https://tinyurl.com/y88gwf98>

CF Foundation Awards Armata Up To \$5M To Advance AP-PA02 To Trial

The Cystic Fibrosis Foundation has awarded up to \$5 million to Armata Pharmaceuticals to advance its potential treatment AP-PA02 — a phage therapy against multidrug-resistant *Pseudomonas aeruginosa* infections in cystic fibrosis (CF) patients — into a Phase 1b/2 clinical trial. Phage therapy harnesses the properties of bacteriophages (or phages) — viruses that infect and kill bacteria — to fight bacterial infections that are difficult to treat. The new trial will test the safety, tolerability, and effectiveness of AP-PA02 in CF patients chronically infected with *P.*

aeruginosa. AP-PA02 is a novel, second version of the company's candidate AP-PA01, which was previously shown to successfully treat a CF patient. After 100 days of treatment with the compound, combined with antibiotics, the patient was free of *P. aeruginosa* pneumonia and CF flares. AP-PA02 is a mixture of multiple phages, which was developed to be inhaled, selected from Armada's phage library, and tested in hundreds of *P. aeruginosa* samples from CF patients. With this approach, the candidate compound is thought to have a wider targeting range, increased potency, and potential to prevent the development of resistance.

<https://tinyurl.com/yd7r8k2j>

Topline CF Trial Data for ELX-02 Expected In First Half Of 2020, Eloxx Says

Eloxx Pharmaceuticals anticipates reporting topline data from the Phase 2 clinical trial program for ELX-02 — its lead therapeutic candidate to treat cystic fibrosis caused by nonsense mutations.

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ASK THE ENT

Your ENT Questions Answered

By David Gudis, M.D.

Q: After my third sinus surgery, it seems like I have lost my sense of smell. What treatments are available for this? Will Trikafta help?

A: Olfaction disorders (disorders of the sense of smell) are common in patients who have CF. Unfortunately, olfaction disorders can have a significant impact on a patient's quality of life. We all want to enjoy the wonderful smells of a homecooked meal or the foliage on a walk outside. We also notice the scents of our children and loved ones, sometimes without even realizing it. The inability to appreciate these scents can be very difficult for patients. Moreover, it has been demonstrated that anosmia (the loss of sense of smell) is associated with feelings of depression and loneliness. It is not clear why olfaction disorders are common in CF patients, but it is probably because CF patients also have a high incident of chronic sinusitis. Chronic sinusitis can impact olfaction in two main ways. First, polyps and swelling of the mucus membranes can prevent odorants from reaching the upper part of the nasal cavity where olfaction occurs. Second, chronic inflammation around the olfactory nerve can impair normal function (this second reason is probably the main issue for CF

patients). The first—and safest—treatments to try to improve your sense of smell is a combination of using nasal saline irrigation (in a sinus rinse bottle or Neti pot) and doing olfactory training with essential oils, which is essentially physical therapy for the olfactory nerve. There are several techniques and protocols available for olfactory training, which can be found online. It is not known yet whether Trikafta will improve olfaction, but our early data shows that it significantly improves sinonasal symptoms in general, so we are optimistic!

Q: What are the best ways for CF patients with sinus disease to stay

safe during the COVID-19 pandemic? Is it safe to see an ENT in the office, or is Telehealth better?

A: Social distance, wear a mask, and keep your hands clean! Regardless of whether a CF patient has a history of sinus problems, SARS-CoV-2 (the virus that causes COVID-19) can be dangerous. We do not know if added measures, such as nasal saline irrigation, can provide added protection. I would recommend that you arrange a Telehealth visit with your ENT before scheduling an office visit. ENT is a high-risk specialty because the highest concentration of the virus is probably in the nasopharynx, which is all the way in the back of the nasal cavity. ENT patients may also cough during exams, which could increase the amount of exposure in the office setting. Therefore, it would be worth considering a Telehealth visit first, and if an inperson visit is then also required, appropriate safety precautions can be taken in the office.

Have more questions? Send your CF ENT questions to us at cfroundtable@usacfa.org. ▲



DAVID GUDIS, M.D.

Dr. David Gudis is an Ear, Nose, and Throat doctor at Columbia University Irving Medical Center in New York City.



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and would have to say it has become my favorite place, especially being able to visit her family and our niece, Nora. If we could be anywhere right now, as it's almost June 1 (the week of our anniversary), we would be sailing on a cruise to Alaska. However, the pandemic curtailed those plans, so we've been enjoying our backyard oasis. My wife and I do intend to spend some time on my dad's houseboat on Lake Cumberland, but will do it in quarantine style for safety.

Who inspires you and what motto do you live by?

My wife, parents, and family all inspire me, but God, along with his Son, inspires me more, because I know I can do all things through Him that

give me strength, including managing my CF care. Celine Dion is an inspiration as well because of everything she went through with her husband and having a niece with CF.

I live by two mottos, both verses from the King James Version of the Bible. Luke 1:37 says "[f]or with God, nothing shall be impossible." I relate to this, as I strive to keep moving forward, knowing God is always with me and not to give up, even in the hardest of times. My other motto comes from Romans 8:28: "[a]nd we know that all things work together for good to them that love God, to them who are the called, according to His purpose." I know without a doubt that all things

work together ultimately for good, as He knows best, even though I may not understand His plans for my life. I just keep my faith that everything will be good, no matter what, as long as I put Him first in my life. ▲

Jeanie Hanley is 58 and is a physician who has CF. She is a Director and the Treasurer of USACFA. Andrea Eisenman is 55 and has CF. She is a Director of USACFA and is both the Webmaster and Executive Editor of CF Roundtable. Their contact information is on page 2.

You may contact Nathan at: nathantalent@yahoo.com. If you would like to be interviewed for "In The Spotlight," please contact either Andrea or Jeanie.

ELX-02 is a small molecule designed to restore the production of full-length functional CFTR protein in patients with a nonsense mutation. It is currently in the early stages of clinical development for CF. Eloxx believes that data from both pre-clinical and clinical studies that have been collected to date support the continued development of its program. Eloxx announced earlier this year that enrollment was nearly complete for two Phase 2 trials (NCT04135495, NCT04126473) testing ELX-02 in CF patients.

<https://tinyurl.com/y9d2v6nc>

Microbion Corporation Announces Orphan Drug Designation Granted for Pravibismane Suspension for Inhalation by US FDA

Microbion Corporation and Microbion Pharma Corp. announced that the US FDA has granted orphan drug designation for inhalation delivery of pravibismane (BisEDT antimicrobial suspension) for treatment of pulmonary infections in patients with cystic fibrosis. Pravibismane (bismuth-ethanedithiol) belongs to a new class of antimicro-

bial medications that have a different structure than that of clinically utilized antibiotics. It also works in a different way than traditional antibiotics, which typically kill bacteria by stopping their multiplication or interfering with the formation of the bacterial cell wall or cell components. Pravibismane is classified as a microbial bioenergetic inhibitor, which means it disrupts the bacteria's ability to produce the energy needed for essential cell processes like DNA, protein, and cell wall production, causing the bacteria to die. Pravibismane exhibits broad-spectrum antimicrobial activity against carbapenem- and multidrug-resistant *Pseudomonas aeruginosa*, as well as other multidrug-resistant pathogens that are recognized as serious threats to CF patients. Pravibismane also demonstrates potent activity against the microbial biofilms formed by these bacterial pathogens. Microbial biofilms play a key role in the chronic nature of the pulmonary infections in people with CF and are known to contribute to the antibiotic-resistance of these infections. Addressing the biofilm component of these infections represents a

unique strategy in the treatment of chronic, resistant infections. Critically, lab tests have shown that pravibismane retains its activity in CF sputum.

<https://tinyurl.com/yadwav5r>

AND

<https://tinyurl.com/ychbjt35>

Arrowhead Pharmaceuticals Files For Regulatory Clearance To Begin Phase 1/2a Study Of ARO-ENaC For Treatment Of Cystic Fibrosis

Arrowhead Pharmaceuticals Inc. announced that it has filed an application for clearance to begin a Phase 1/2a clinical trial of ARO-ENaC, the company's investigational RNA interference (RNAi) therapeutic being developed as a treatment for patients with cystic fibrosis (CF). ARO-ENaC utilizes Arrowhead's proprietary Targeted RNAi Molecule (TRiM™) platform and is the company's first inhaled RNAi candidate to target pulmonary epithelium. The study will evaluate the safety, tolerability, and pharmacokinetic effects of ARO-ENaC in normal healthy volunteers and will evaluate the safety, tolerability, and

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What Sobriety Taught Me About Acceptance

By Andrew Kirby

I once read “...for most normal folks, drinking means conviviality, companionship, and colorful imagination.” Not for me, though. As an alcoholic, I drank simply to escape whatever “hardship” I felt life had unjustly thrown my way. My life was one big pity party, and alcohol became a way for me to cope with how I felt inside. It took me a very long time to come to terms with having cystic fibrosis. Nowadays, I view my diagnosis as a blessing, rather than a curse.

Yes, I have this illness and life can sometimes get a bit tough. However, the only sickness I truly suffered from was what I would call a “spiritual malady,” characterized by feeling restless, irritable, and discontented with life. Though only having a very mild case of CF, I was miserable and unable to see the joys in my life. Learning this life lesson took me down a long road filled with depression, anxiety, and a generally angry disposition that eventually landed me at my first Alcoholics Anonymous meeting at 17. Six long years later, I finally got sober.

It was at a very young age that I learned from a healthcare worker of my short life expectancy. “You know, people with CF die young.” Now, I’m sure the intent was nothing more than a blunt (and careless) foot-in-mouth moment, not intended to cause any harm. Nevertheless, at four years old, this was not the appropriate time to learn about this, and I unknowingly carried the pain of this shocking and grim prognosis on my shoulders for many years.

To be honest, though, despite this revelation, I didn’t have a bad childhood. Except for problems with my schooling, I managed to stay out of trouble. I was fairly active. I (reluctant-

ly) attended church and Sunday school, I was in Cub Scouts, I played with friends after school, and I rode bikes around my neighborhood. I was just like any other ordinary kid my age. However, I didn’t feel like a normal kid. I felt different from the others. I was smaller and less athletic than other kids. Bullies honed in on me—I was picked on for my size often. This, coupled with always having to take my



ANDREW KIRBY

daily medications and nebulizer treatments, caused me to develop serious self-esteem issues. I felt out of place and inferior. No matter how hard I tried I always felt like I never fit in. Even among my best friends, I felt completely alone.

I hid my CF from others out of fear of being pitied or ridiculed. A great deal of resentment began to build up, toward my parents, my doctor, even God. Depression began to creep in as I felt I was fighting an uphill battle. Anxiety centering around when the next exacerbation and hospitalization would happen crippled me. I lost my temper with increasing frequency. I would see others with CF living happy

lives, which further angered me. In hindsight, I despised them because I was jealous. Anger became my go-to reaction for any situation that felt uncomfortable. I began to rebel against my parents and often refused to do my treatments.

The older I got, the angrier I became. Punching holes in walls became a normal occurrence. Those “voices” inside my head grew louder and louder, telling me I would never be good enough, until I finally picked up my first drink. I was 16 years old.

I’d prefer not to dive too much into my “war stories.” Omitting the more personal details of my first time being drunk, what I can say is, in that moment, I did not have CF. I was not restless, irritable, or discontented. Those “voices” in my head ceased. I felt completely free. The truth, though, is that I was far from being free and that “phenomenon of craving” kicked into overdrive the instant that first drop of alcohol touched my lips. What followed was a spree of nonsense in which I caused so much wreckage that I was finally convinced by my history teacher to attend my first AA meeting at 17 years old.

I stayed sober for an entire year, but I was still miserable because I couldn’t accept my own life. I was nothing more than a “dry drunk.” And you can only whiteknuckle your way through life for so long. Needless to say, it wasn’t long before I relapsed. I spent the next year bouncing in and out of AA. Each time I went back to drinking, my CF got worse. As time went on, hospitalizations became more frequent and my struggle with depression and anger intensified.

By the age of 19, I had no intention of quitting. In fact, I was convinced I didn’t have a problem and that the

wreckage of my youth was simply due to my immaturity. Now, being much older and in my mind more mature, I could certainly control my drinking.

But, as an older mentor of mine taught me, control is a fantasy.

In fact, I had completely lost all control of myself. The night of my 21st birthday party, I drank so much I almost had alcohol poisoning. Unfortunately, things did not slow down from there. After having surgery to repair a shoulder injury, I was prescribed painkillers. These, mixed with alcohol, marijuana, and some newly prescribed antidepressants, would eventually bring me to my knees.

Time went on and my depression reached an all-time high. My drinking increased and my overall health declined rapidly. By this point, I didn't really care about dealing with my CF—I neglected my nebulizer treatments and exercise for months at a time. I stopped taking my antidepressants, my weight dropped, and my lung function deteriorated to the point of developing a chronic cough.

Eventually, my pulmonologist of nearly 20 years confronted me about my mental health issues and I finally broke down into tears in his office. I was admitted into the hospital yet again for a tune-up, but this time for the full two weeks to make sure I took my antidepressants as prescribed.

However, the two-week stay did little to get my head on straight and soon enough I was back to drowning my sorrows in alcohol. Months later, my heavy drinking had finally defeated me, a mere eight months after my 21st birthday. Completely broken and on the verge of suicide, I entered the halls of AA for one last attempt to get sober—I had no other choice but to surrender to life.

Despite going to AA meetings several times per week, I couldn't manage to stay sober more than a few weeks at a time. I wanted to stay sober—not for my friends, my loved ones, or even for my job. I wanted (and needed) to stay

sober for myself. However, I didn't have the mental tools to do so.

So, in March of 2019, at almost 23 years old, I checked into rehab as my last-ditch effort to turn things around. Now, almost a year and a half later, I am proud to say that I haven't had a drink or any drugs since. Let me tell you, life has gotten so much better. Life hasn't necessarily been easier, but I am happy and now I am truly free and able to manage my own life.

Rehab gave me the space to do some soul searching and accept all those things about myself that I was unable to accept in the past. Rehab was both an inpatient and 12-Step program and introspection was a key component of the program. Meditation, reading, and writing became a healthy outlet, in addition to working out and eating healthy. I spent much of my time studying different spiritual concepts such as Sikhism and Taoism, but mostly Buddhism's Four Noble Truths and the Buddhist concept of suffering. What was difficult at first was learning to have gratitude and practicing acceptance, but these are key to having a new outlook. I needed to change what I wanted, rather than getting what I wanted.

And acceptance is the answer to all my problems today.

The program gave me the tools to cope with whatever curveball life threw my way, regardless of the circumstance. More importantly, I can now cope with any dark feelings inside my own mind. My relationships, be it work, family, or my loved ones, have all gotten stronger and I am now healthier and physically stronger today than I have ever been before. My diagnosis of cystic fibrosis no longer upsets me.

And I owe this all to my sobriety. ▲

Andrew Kirby is 24 years old and lives in his hometown of Exeter, NH. In his spare time, he enjoys reading, hiking, practicing martial arts, and spending time outdoors. He is also an animal lover and a practicing Buddhist. andrewokirby11@yahoo.com

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efficacy in patients with CF. Exploratory objectives in patients with CF include assessing the effects of ARO-ENaC on changes in lung clearance index (LCI) and evaluating changes in forced expiratory volume (FEV1). ARO-ENaC has the potential to be a genotype-agnostic therapy, which may prove useful in combination with existing or new CFTR-targeted therapies. In addition, preclinical studies have indicated that ARO-ENaC has an extended duration of action that enables once every 3-4 weeks or even less frequent administration, which may minimize treatment burden for patients with CF.

<https://tinyurl.com/y76o2tn5>

AzurRx Announces Manufacturing Agreement with Delpharm for MS1819 Clinical Drug Product

AzurRx BioPharma, Inc., announced that it has entered into a manufacturing agreement with Delpharm ("Delpharm") for the clinical drug product supply of its MS1819 therapy for exocrine pancreatic insufficiency (EPI). Under the terms of the agreement, Delpharm will manufacture AzurRx's MS1819 cGMP batch that will be used in the Phase 2b OPTION 2 Clinical Trial for the treatment of EPI in patients with Cystic Fibrosis (CF). As preparation for the cGMP batch, the drug product manufacturing process was transferred to Delpharm and they have successfully produced a non-GMP batch. This marks the first time in which MS1819 has been manufactured with enteric capsules, which will be used in the OPTION 2 Trial to deliver MS1819 drug product during the duodenal phase of digestion. MS1819 is a recombinant lipase enzyme for the treatment of exocrine pancreatic insufficiency (EPI). MS1819, supplied as an oral non-systemic biologic capsule, is derived from the *yarrowia lipolytica* yeast lipase and breaks up fat molecules in the digestive tract of EPI patients so

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Patient-Centered Outcomes Research: A New Approach For The CF community

By Laura Mentch

Four years ago, I met Emily Godfrey at the CFRI National CF Conference. She introduced herself as a family physician and reproductive health researcher interested in learning how CF impacts reproductive health and choices for patients. Waiting for an appropriate pause during the Q&A session, I jumped across the room to meet her as I've been a health educator in sexual and reproductive health centers for many years. With my delayed diagnosis I was learning how sexual health is different for people with CF. Friends, knowing my background, would ask me questions: "will my birth control pills work while on IVs?" or "why is my CF worse around my period?" I wanted to share accurate and helpful information but couldn't find the resources I needed. Dr. Godfrey was the first person I'd met from my professional community who was also interested in CF.

She had applied for, and received funding from, the Patient-Centered Outcomes Research Institute ("PCORI"), an organization that funds various patient-centered outcomes research ("PCOR") projects to bridge the gap between patients and healthcare providers. Emily's project aimed to bring women with CF together to engage in discussions and share their sexual and reproductive health concerns. Emily, now my colleague and friend, invited me to join the CF reproductive health project initiated by a core team of researchers, including Associate Professor Sandy Sufian, another adult with CF. The door for me to participate in PCOR opened and, without hesitation, I walked in.

In 2010, under the Affordable Care Act, Congress launched PCORI.

The mission statement on their website states: "PCORI helps people make informed healthcare decisions, and improves delivery and outcomes, by producing and promoting high-integrity, evidence-based information that is guided by patients, caregivers, and the broader healthcare community." For



LAURA MENTCH

this to happen, patients are welcomed into the fold of research and become active members of the team.

People with CF are familiar with research. We

- are subjects in clinical studies,
- respond to surveys,
- participate in focus groups,
- help with clinic Q/I projects,
- serve on research steering committees, and
- review research grants.

Have you ever wanted to be more involved? PCOR makes this possible. Read on!

On a PCOR team, patients,

researchers, and clinicians work together to create or define research that can address the questions expressed by patients and their families. Patients bring their concerns and ideas to the table; researchers share a variety of approaches to address these issues. From this exchange, patients begin to more fully understand the research process. Close working relationships facilitate patient partners stepping into all phases of research. All team members work together to write proposals and articles, prepare posters, and deliver presentations. The goal is to bring results that make a difference to the community.

Our PCORI funding led to the creation of The CF Reproductive and Sexual Health Collaborative, CFReSHC. Our Governance Board includes researchers, healthcare providers, and women with CF. More women than men with CF join the Patient Task Force. Virtual meetings with patients create a safe space for women to discuss our unique and unmet needs for sexual and reproductive health. Together, we identify questions we want CF providers and researchers to address. There are many!

Successful PCOR research happens when each team member actively embraces the following principles of patient engagement. Our commitment to these principles guides our continuing work and outreach into the CF community.

Reciprocal relationships.

All roles are defined collaboratively, clearly stated and shared among team partners. Decision making is shared.

Responsibilities are defined and

¹<https://www.pcori.org/about-us/our-vision-mission>

rotated among all members. Over time, each of us will create the agenda, facilitate, and record notes for our meetings. Each of us participates in guiding the direction of CFReSHC

Co-learning.

Patient partners are supported in learning and understanding the research process. Researchers take time to learn about PCOR and the experience of living with CF. Researchers actively engage patients and other stakeholders in the research process. Talent, experience, and contributions of all partners are welcomed and honored.

Patient partners learn the basics and process of research. Time is given for team members to ask questions when we don't understand something. After writing a grant, for example, Molly wanted to more fully understand the process. Emily sat down with Molly to review and explain some of the changes that were made in the writing.

We want all partners on the same page moving forward. Researchers were surprised to learn that our questions were not only about contraception, menstrual cycles, and pregnancy. We also wanted to explore issues of body image, BMI, incontinence, and family building.

Partnership.

The contributions of patient partners are valued and compensated fairly. Reasonable expectations of time commitments of all members are honored. The unique needs of people with cystic fibrosis are respected and accommodated.

Patient-partners are not volunteers. To be a part of the research team, we must understand our responsibilities, time commitment, and compensation from the start. Fair compensation is essential and guided by PCORI. Compensation varies according to the level of involvement a patient has with the project.

Trust. Transparency. Honesty.

A Clinician-Scientist Steps Out Of Her Comfort Zone

By Emily Godfrey



As a clinician-scientist trained in clinical research, I had heard the term “patient-centered outcomes research” being used, but I didn’t have a firm understanding of what it truly meant. So, when I first attended an “Introduction to Patient-Centered Outcomes Research” workshop I was surprised to see patient partners speaking about their topics with such confidence and ability. I also observed, for the first time, how everyone else in the

room—researchers, clinicians, community members—welcomed and honored all of the comments that came from the patients. After seeing that kind of mutual respect in action, I overcame my previous judgments about how only researchers or scholars should be part of a research team, sat back and listened, and, by the end of the workshop, I realized how much we can learn from people who experience their disease in everyday life.

What I witnessed at the workshop matched what I wanted for my professional career. For me, the PCOR process started with making new connections at CFRI: with Laura, other patient-partners, and with CF researchers. This small group eventually blossomed into what is CFReSHC today. CFReSHC is the work of the patient-partners. The patient-partners help clinician-scientists see what matters to them most as patients. As a clinician-scientist, I work to bring sound research methods to the patient-partners’ unanswered, but important questions. We collaborate in writing protocols, collecting data, and reviewing our findings.

Our core PCOR team is inspired to spread the word throughout the CF community about the power of engaging patients and care givers as authentic partners on the research team. While CF patients are naturally valued for their “lived experience” with the disease, I have discovered how each individual brings special skills and insights that contribute to the mission and purpose of our work. The research process may not be faster, but it is much more fulfilling to see important new questions, ones that matter to patients, addressed through well-vetted research protocols that incorporate PCOR principles.

Major decisions are made within the group and information is shared readily with all partners. We take time to learn about one another to support our working relationships. We are committed to open and honest communication.

We begin each meeting with a check-in or icebreaker. This helps us get to know each other better and build

rapport. When one of us has a concern, we are given space to discuss it in a meeting so we can work through it. We use first names for everyone, an early challenge for the patient partners.

In 2018, CFReSHC was awarded a Eugene Washington PCORI Engagement Award to build PCOR

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capacity within the CF community. From the beginning, we have met only online, making it possible for many with CF to participate. Our experience encouraged us to also create a guide for other disease groups on how to operate virtually. We've reviewed many platforms for virtual meetings, project management, and communication for this guide.

To create a PCOR training for CF providers, researchers, caregivers and patients we began with a needs assessment to develop learning objectives. Together, the team planned and delivered training in four 90-minute sessions. 27 patients/caregivers and 33 researchers/providers attended at least one of the following trainings:

- *PCOR 101 with patients, caregivers, and researchers;*
- *Research 101 with patients and caregivers;*
- *Participating in and Maintaining a PCOR Team with patients, caregivers, and researchers; and*
- *Designing and Implementing a PCOR Study with researchers.*

We developed this training to share the benefits of PCOR with the CF community and encourage others to include it in their work. Our Best Practice User Guide for interacting online will also be available.

Looking to the future, we intend to continue to address the sexual and reproductive health concerns of the CF community, including those of men with CF. We are devoted to bringing many patient voices to the future of research in CF and strive to work with national CF organizations to do so. What ideas do you have that could make a difference for you and others with CF?

Please contact CFReSHC for more information about our work and the application of Patient Centered Outcomes Research in the CF Community. info@CFReSHC.org ▲

Becoming A Researcher

By Molly Pam



I came into this project sideways, through a research study looking at contraception in CF. After that study, I was invited to attend a CFReSHC patient task force meeting run by the same research team. I was struck by how much these researchers cared about, and listened to, my personal experience. I started attending more meetings, and, by the summer, was asked if I wanted to join the governance board.

While I had many questions about contraception and family building, I initially assumed that because I had no personal experience with pregnancy, incontinence, hemoptysis, or exacerbations around my period, that I would be unwelcome. This couldn't have been further from the truth. I stayed not only because I was curious about future issues that might one day affect me, but also because of the passion of the researchers. I saw how deeply the researchers looked to the patients for guidance and treated us as equals and experts in our own lives and diseases. For me as a patient, this was a unique opportunity to be heard, and to listen to others whose experiences are vastly different than my own.

The relationships I have made with researchers in CFReSHC are relationships I want to be available to every person with CF, if they want them. Early on in my partnership with Dr. Traci Kazmerski, I discussed my fraught choice over whether and how to have children. I had talked with many people with CF in the same position, stuck in the decision-making process and frustrated with the lack of help or understanding from our healthcare providers. Over the course of many conversations, I noted the absence of good resources available to help the CF community think about these decisions, and identified it as a large area of need as patients live longer and healthier lives. Traci listened and changed the course of her research. She is now leading a team of patients and researchers, using PCOR principles to create a decision aid guide for patients with CF who are struggling to make family-building decisions.

This ability to change the focus of research extends beyond the individual to the community. When I approached the CFF about hosting a mini-con on sexual and reproductive health, nobody else was discussing it. This mini-con had the highest ratings of any virtual conference they had hosted to that point. The issues that were brought to the forefront are now talked about throughout the CF community.

I am a researcher at heart; that is what I studied as an undergraduate and wanted to do with my career before my health made that impossible. Emily, Traci, CFReSHC, and PCORI made it possible for me to engage in research. I consider many researchers not just colleagues, but friends. The benefits of creating these strong relationships shows in the quality of the research. They listen to us deeply to understand our ideas or concerns, so that *we can change the course of research.*

Your voice matters. • You have the ability to change the course of CF research
• Start speaking.



MILESTONES

Please share the milestones in your life with our readers. Your successes and achievements may serve as a source of motivation for others in need of an infusion of “positive mental attitude” in the pursuit of their goals. Send us a note specifying your “milestone.” Include your name, age, address and phone number. Mail to: **CF Roundtable, PO Box 1618, Gresham, OR 97030-0519.** Or email to: **cfroundtable@usacfa.org**

ANNIVERSARIES

Birthday

Kathy Russell

Gresham, OR

76 on April 17, 2020

Zachery Hays, 31

Vernon, CT

Bilateral lung transplant

3 years on February 3, 2020

Transplant

Andrea Eisenman, 55

New York, NY

Bilateral lung transplant

20 years on April 25, 2020

Wedding

Kathy and Paul Russell

Gresham, OR

55 years on March 27, 2020

NEW BEGINNINGS

Roberto De La Noval, 31

South Bend, IN

Doctorate in Theology from

University of Notre Dame

April 2, 2020



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that they can be absorbed as nutrients. Unlike the standard of care, the MS1819 synthetic lipase does not contain any animal products.

<https://tinyurl.com/y75ng49x>

AND

<https://tinyurl.com/ydyzj6v8>

Lumacaftor-Ivacaftor May Improve Glucose Tolerance In Cystic Fibrosis

Patients with cystic fibrosis (CF) who were treated for 1 year with a combination of lumacaftor and ivacaftor saw improvements in glucose tolerance abnormalities. Of the 40 patients with CF included in the study, 78% had glucose intolerance and 22% had newly diagnosed diabetes. After 1 year of treatment, 50% of patients had glucose tolerance classified as normal, 40% had glucose intolerance, and 10% had diabetes. Glucose tolerance improved in 57.5% of patients, with a significant decrease in both 1 and 2-hour oral glucose tolerance test glycemia.

<https://tinyurl.com/yatxbtmr>

Symdeko and Orkambi Seen To Also Ease Inflammation In CF Patients

In addition to restoring CFTR protein function in cells, the cystic fibrosis (CF) therapies Orkambi (lumacaftor/ivacaftor) and Symdeko (tezacaftor/ivacaftor combo) significantly reduce the excessive inflammation that damages patients' lungs. Symdeko

seems to be more potent than Orkambi at dampening inflammatory responses. This inflammatory response seems to be triggered by the inflammasome, a multiprotein complex inside immune cells that detects pathogenic (harmful) microorganisms. After being activated by microbes, the inflammasome triggers the production of major pro-inflammatory molecules, such as interleukin-1 beta (IL-1 beta) and IL-18 initiating a cascade of inflammatory responses. After three months of treatment, both Orkambi and Symdeko were seen to significantly lower blood levels of IL-18 and tumor necrosis factor (TNF). But Symdeko also reduced IL-1 beta levels, and was more consistent in terms of efficacy across patients. Notably, Orkambi and Symdeko's effects on inflammation lasted only 36 hours, but re-exposing cells to the treatments reduced inflammation as effectively. The data found in this research has suggested that systemic inflammation plays a major role in the pathogenesis of CF and that this pilot study is the first to demonstrate that CFTR modulators have potent innate anti-inflammatory properties.

<https://tinyurl.com/yahhjn37>

Tobacco Smoke Exposure Limits Benefits Of CFTR Modulators In Cystic Fibrosis

Tobacco smoke exposure inhibited the therapeutic benefit of the cystic fibrosis transmembrane conductance regulator

(CFTR) modulators in pediatric patients. Tobacco smoke has been shown to reduce CFTR functional expression in vitro and contributes to acquired CFTR dysfunction in animal models. Therefore, researchers sought to determine whether tobacco smoke exposure also inhibits the clinical benefit of CFTR modulators, specifically, tezacaftor/ivacaftor. They performed a retrospective analysis of the CF Foundation Patient Registry comparing lung function changes, pulmonary exacerbations, and hospitalizations after tezacaftor/ivacaftor initiation in patients who were exposed to smoke vs patients who were not. Smoke exposure was determined by annual caregiver self-reports. The mean baseline forced expiratory volume in 1 second percent predicted (FEV1%) was 83.3%, and 27.6% of patients were exposed to smoke. The FEV1% of children who were not exposed to smoke increased by 0.72%, whereas the FEV1% of children exposed to smoke decreased by -1.03%, according to analysis. Tezacaftor/ivacaftor contributed to 0.5% FEV1% improvement in children not exposed to smoke, but provided no benefit to children who were exposed to smoke. In addition, smoke exposure increased the odds of children being hospitalized ≥ 2 times annually and of experiencing ≥ 2 pulmonary exacerbations annually by 31% and 47%, respectively.

<https://tinyurl.com/y95hga8v>



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Nanoparticles With Plant Sterols May Aid Gene Therapy For CF

Tiny particles, or nanoparticles, work better as transport and delivery vehicles for the RNA molecules central to some forms of gene therapy when plant-based relatives of cholesterol, called phytosterols, are included, scientists report. Some of these treatments use special nanoparticles made up of lipids (fatty molecules) that encapsulate, protect, transport, and deliver RNA molecules to cells. (RNA is the template cells use to make proteins.) One such therapy is being developed as a potential treatment of cystic fibrosis (CF). The idea behind the experimental CF therapy is to load specialized nanoparticles with the RNA containing the information to make a functional CFTR protein. After being inhaled, these nanoparticles would then transport and deliver this RNA to cells in patients' lungs, restoring the production of the CFTR protein, as well the transport of compounds in and out of cells. They found that incorporating phytosterols — found in plant cell membranes and similar in ways to cholesterol — made nanoparticles more stable and effective transport vehicles. According to the researchers, this happens because phytosterols can change the shape of these nanoparticles from spherical to polyhedral, making them more ergonomic and able to move faster. This change is important, because once inside cells, nanoparticles need maneuverability to reach the cell's cytosol — the liquid-like substance that fills the interior of cells — where RNA molecules must be delivered to perform their intended function. Their experiments showed that compared to spherical nanoparticles, those containing phytosterols had higher cellular uptake, retention, and diffusivity (ability to spread). All of these features contribute to increase their ability to transport and deliver their content to its intended location. The researchers now plan to create inhalable nanoparticles containing phytosterols to use in their investigational therapy for CF.

<https://tinyurl.com/y7sv68b5>

CF Stem Cell Mutations Fixed Via CRISPR Gene Editing Tool, Study Says

A new variation of the gene-editing technology CRISPR-Cas9 can correct mutations in the CFTR gene in stem cells from CF patients. The new approach has the ability to correct mutations without the need to excise the affected region. CRISPR-Cas9 is a system used by microbes to defend themselves against invading viruses. The first version of the CRISPR technology, developed in 2012, cuts a mutation out of a gene, but a 2018 version, called base editing, is able to repair a mutation without cutting the DNA. This makes the gene-editing tool more precise and safer. In the new CRISPR technique, called base editing, the Cas part is altered in such a way that it no longer creates a cut, but still detects the mutation. So instead of creating a cut and replacing the faulty DNA, the mutation is directly repaired on site, making this a more effective genome editing tool. Results showed that the technique was able to repair the CFTR mutation without causing additional errors in other parts of the genome. These so-called off-target effects are a known reservation in the standard CRISPR technology. With the new base-editing technique, the mutation in the CFTR gene can be detected and repaired without creating further damage in the genome. While the results are promising, more research is needed before the strategy is available to patients. To use the CRISPR technology, scientists must first design a way to deliver the CRISPR tools to the appropriate cells. <https://tinyurl.com/ybzqfj7w>

Gene Therapy, KB407, Seen to Restore CFTR Protein in Cell Studies

A potential gene therapy for cystic fibrosis, KB407, is able to induce the expression of normal CFTR protein in cell models of CF. A gene therapy for CF aims to deliver a non-mutated version of the CFTR gene into these cells, allowing

for a working CFTR protein. An advantage is that this approach could work equally well regardless of a person's particular CFTR mutation. KB407 is delivered to cells using an engineered version of herpes simplex virus 1 (HSV-1) based on Krystal's STAR-D platform. Viruses are useful vectors for gene therapy because they have evolved to be very efficient at delivering genetic material into cells. The particular type of HSV-1 used in KB407 is reported to be replication-defective; that is, it is able to get its genetic payload (i.e., a full-length CFTR gene) into cells, but it will not replicate and infect other cells. Krystal will continue its preclinical studies of KB407, and plans to file an investigational new drug application, requesting trials in people, in 2021.

<https://tinyurl.com/yczast6p>

More Training Needed In CF End-of-life Care, Small UK Survey Shows

Most healthcare professionals are not fully prepared for cystic fibrosis (CF) end-of-life care, and further training and coordination between CF and palliative care teams would be helpful. The optimal timing for discussions regarding end-of-life care, as well as how and by whom this type of care is best provided, remains unclear. Patients are often treated by the same CF team over the long term, and close relationships may exist between them. But these team members may be limited in end-of-life care training, and feel inadequately prepared. While palliative care teams have extensive experience in such care, they may lack knowledge and familiarity with CF-specific issues. A combination of both specialty cares has been suggested for routine CF management, but helpful information on how these teams should interact over the course of CF progression is lacking. Researchers set out to evaluate the knowledge, experience, and preparedness for end-of-life care among both CF and palliative care teams. Results showed that more than 60% of CF team members reported having some

Continued on page 54

to extensive experience with CF end-of-life care, while 63% of the palliative care team reported minimal or no experience within the context of CF care. Despite relatively high levels of CF end-of-life care understanding among both teams (more than 80% with somewhat to fully understanding), levels of preparedness were low. Only 11% of CF and 19% of palliative care team members reported to feel “fully prepared,” and about one quarter of each team felt minimally or not prepared at all for that type of care. Notably, 58% of CF members had no (21%) or minimal (37%) general palliative care training. Likewise, 69% of the palliative care team had no CF-specific training, and the remaining 31% had minimal training. Importantly, all respondents showed a desire for additional education in CF end-of-life care — CF team members specifically in general end-of-life care, and palliative care members in CF-specific knowledge. Participants also reported that the preferred method for this additional training would be shadowing the other team (CF or palliative care teams), rather than online learning or formal seminars.

<https://tinyurl.com/ycxx8gxs>

4DMedical’s Lung Imaging Technology Receives FDA Clearance As COVID-19 Intensifies Global Focus On Respiratory Health

4DMedical announced that it has received clearance from the U.S. Food & Drug Administration to market its XV Technology™, a patented four-dimensional lung imaging process that rapidly and automatically analyses any functional lung impairment from a single X-ray. It provides critical information about the functional and structural state of a patient’s lungs in the treatment of illnesses. 4DMedical’s XV Technology™ process is a software-as-a-service (SaaS) diagnostic tool, available through secure cloud subscription, and can be implemented immediately, utilizing existing hospital and clinical infrastructure with no capital expenditure or training required. Imaging departments simply elec-

tronically send an X-ray (using existing fluoroscopy equipment) to 4DMedical. 4DMedical software then rapidly and automatically analyzes and applies its proprietary algorithms to identify and quantify any functional impairment. The software generates a ventilation report and sends it to the hospital to enable clinicians to determine the most effective treatment course of action and allocation of finite hospital resources. The end-to-end process can be completed and a report generated within three hours. The unique 4DMedical technology accurately and quickly scans lung function as the patient breathes, to provide sensitive, early diagnosis and to monitor changes over time. The Software-as-a-Service (SaaS) scans deliver much more complete results, showing even subtle variations in lung function down to the finest details, using lower levels of radiation than traditional methods.

<https://tinyurl.com/y77kjqnq>

A Longitudinal Assessment Of Non-invasive Biomarkers To Diagnose And Predict Cystic Fibrosis-associated Liver Disease

In the present study, the researchers sought to assess the efficacy of AST to Platelet Ratio Index (APRI), fibrosis index based on 4 factors (FIB-4), AST/ALT ratio, platelet count, gamma-glutamyl transferase (GGT), and GGT platelet ratio (GPR) in anticipating cystic fibrosis-associated liver disease (CFLD) development. From January 1, 2002 to December 31, 2014, data were obtained from CF Foundation Patient Registry for patients aged 3–21 years at Johns Hopkins. Information collected included demographic characteristics, splenomegaly, hepatomegaly, ascites, and variceal bleeding, AST, ALT, GGT, platelet count, and FEV₁. At the end of the study, 144 “healthy” CF, 12 CFLD, 19 CF-associated pulmonary disease (CFPD), and 4 CFLD with CFPD cases have been identified. According to findings, GPR, GGT, APRI score, and platelet count were potentially useful biomarkers while CFLD development was not

predicted by FIB-4. In order to analyze the utility of these biomarkers in clinical practice, cost-effectiveness studies are required. <https://tinyurl.com/ybozq638>

Semaglutide In Cystic Fibrosis-related Diabetes

Despite evidence that inadequately controlled glycemia is correlated with worse clinical outcomes, in a majority of patients, cystic fibrosis-related diabetes is not well managed, researchers sought to analyze the impact of adding semaglutide, a glucagon-like peptide-1 receptor agonist, to basal insulin to control glycemia in one of those patients. According to results, there has been a significant improvement in glycemic control, drop in glycated hemoglobin A1c from 9.1% to 6.7%, and stable euglycemic trend on continuous glucose monitoring within 3 months of treatment start. There was no rise in concentrations of plasma pancreatic enzyme. Findings suggested that low dose semaglutide in combination with basal insulin was able to replace prandial insulin and control glycemia.

<https://tinyurl.com/y8ln3pfy>

For CF Patients With Pancreatic Insufficiency, Encala Improves Fat Absorption

Investigational treatment Encala (Lym-X-Sorb) is safe, well tolerated, and increased dietary fat absorption in patients with cystic fibrosis (CF) and pancreatic insufficiency. While CF is most commonly associated with the lungs, it can also affect digestion, impairing nutrient absorption, including fats, and leading to poor nutrition. Malnutrition due to chronic malabsorption is an important intervention target in CF care as it affects growth, development, pulmonary function, immune system function, and survival. Studies have found that medications to treat symptoms of CF, including pancreatic enzymes and CFTR modulators, only show limited effectiveness in improving the absorption of fats. To address this,

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Envira Health developed Encala, an oral treatment specifically designed to increase fat absorption in CF patients. The Phase 2 trial (NCT00406536) investigated Encala as a nutritional supplement in patients with CF and pancreatic insufficiency. To evaluate fat intake, the researchers primarily used a measure-

ment called coefficient of fat absorption (CFA), which uses a stool sample to determine how much fat was absorbed in digestion. The CFA value was measured in participants at the start of the trial, as well as after three months of treatment. The researchers reported that subjects with low baseline CFA and more severe

fat malabsorption had a dramatic improvement in CFA with Encala treatment, accompanied by improved plasma fatty acid and growth status.

Laura is 72 and has CF. She is a former director and President of USACFA. She and her husband, Lew, live in Northville, MI. ▲

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IMPORTANT RESOURCES

Medical Assistance Tool (MAT): <https://medicineassistancetool.org/> PhRMA's Medicine Assistance Tool (MAT) is a search engine designed to help patients, caregivers and health care providers learn more about the resources available through the various biopharmaceutical industry programs. MAT is not its own patient assistance program, but rather a search engine for many of the patient assistance resources that the biopharmaceutical industry offers.

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