CF Roundtable®
A NEWSLETTER FOR ADULTS WHO HAVE CYSTIC FIBROSIS
WINTER 2021

Hostage No More

By Angeline Chase

My name is Angeline and I have cystic fibrosis. What a difficult thing to admit to myself even though it's been more than six years since my diagnosis. I still love to live in denial by taking a "healthy day" and pretending I'm not sick by skipping my treatments. Yes, I am aware of the flawed logic. I wasn't diagnosed until I was 31 years old, so doing treatments is not something that I am accustomed to. I just acquired a Hill-Rom Vest a few months ago; before that I just had nebulizer treatments and an Acapella device for airway clearance. Even that seemed to devour too much time. But I am getting ahead of myself. Let's start from the beginning.

Some people who are diagnosed late in life have never heard of CF, but not me! I was hospitalized when I was six months old because I was so little. The doctors were worried because I wouldn't eat and I couldn't gain weight. The doctors thought I had cystic fibrosis. I was later told that children with cystic fibrosis died when they were teenagers, but I was so lucky because it turned out that I didn't have it. Throughout my childhood, I came down with pneumonia frequently and I coughed routinely. I told people I had allergies or asthma, although I had never been diagnosed with either one of those.

ANGELINE CHASE

Well, at 31 I found out that the doctors had done two sweat tests—one was positive and the other was borderline. Rather than trying to figure out why, they sent me home on a high-calorie formula. I suspect the doctors decided not to pursue a diagnosis back then to protect my family from being denied insurance as they were self-insured dairy farmers.

Having expected a long and fulfilling life, I obtained a Bachelor's Degree in Nursing and I got married. Shortly after, we had a daughter. However, we divorced after five years. I later remarried a widower who had four children with his late wife. I grew up with 12 biological siblings, so his four children were just part of the package deal. We decided to have a daughter of our own and then decided one more baby couldn't hurt. We ended up having twin boys. So, at the time of my diagnosis at 31, I was a mother to eight children, two of which were nine-month-old twin boys.

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INSIDE THIS ISSUE

CF ROUNDTABLE
FOUNDED 1990
Vol. XXXI, No. 1

CF Roundtable (ISSN 1057-4220) is published quarterly by the United States Adult Cystic Fibrosis Association, Inc. (USACFA), a totally independent, 501(c)(3) tax exempt, nonprofit corporation whose Board of Directors all have CF. Articles in *CF Roundtable* may be reprinted only with advance written permission from USACFA. All submissions to *CF Roundtable* become the property of USACFA and should include the author's full name, address and phone number. Submissions are subject to editing as needed and we reserve the right to edit any comments that disparage another person either by name or situation. Requests for anonymity will be honored.

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EDITOR'S NOTES

Happy new year, readers! This issue you'll read about our two newest directors, **Daniel Gonzalez** and **Terry Wright**. We're thrilled to have both of them join us. You can read about their stories on pages 50 and 51. Speaking of Daniel, both he and **Ashley Wilson** write about their experiences as students and prior scholarship winners in this issue.

For this issue's focus topic, you'll hear from an array of adults with CF who were diagnosed later in life. **Victoria Greene** chronicles the 9,357 days before her diagnosis while **Lisa Bentley** talks about her CF diagnosis as a blessing amidst the pursuit of her sports career. Our focus topic section also features **Luisa Palazola**, who writes about her mother's background setting the stage for getting the right diagnosis and treatment. **Danielle Lassak** talks about the litany of GI issues leading to her diagnosis and the importance of continually advocating for yourself. **Dr. Jean Hanley** recounts her relentless and long journey throughout medical school to get answers and, ultimately, her CF diagnosis. You'll also read about **Jerry Cahill**'s diagnosis at age 11 and how that impacted his outlook on life. **Megan Felch** writes about her myriad health struggles leading up to diagnosis while **Darryl Collins** describes the positive effect his CF diagnosis has had in his outlook on life.

Kathy Russell talks about the early days of treatments and diagnosis in her "Speeding Past 50" column while **Molly Pam**, for her "Family Matters" column, interviews a couple (who reached out to her after her NACFC talk) about their infertility journey. **Dr. Xan Nowakowski** writes about rare mutations in multiracial and multiethnic lineage in her new column, "Pearls of Wisdom." **Beth Sufian** answers questions about SSI benefits and health insurance coverage in her column. In "Live Out Loud," **Lara Govendo** writes about the unpredictability of life—how to navigate the sudden changes and why it's important to let go of the illusion of control over our lives. **Mark Tremblay** writes about his quirky dog, Zappy, and what Zappy has taught him about CF and family dynamics. In her "Spirit Medicine" column this issue, **Isabel Stenzel Byrnes** writes about acceptance—what you can and cannot control. **Dr. David Gudis** succinctly covers early data on Trikafta's effect on CF sinuses as well as what's new in the treatment of chronic CF-related sinuses in his "Ask the ENT" column this issue.

Laura Tillman expertly collates all the latest CF research and developments from the internet in this issue. For our "In The Spotlight" interview this issue, **Andrea Eisenman** and **Dr. Jean Hanley** talk with one of our directors, Grace Knight, about her college (and now law school) experience. In the "Voices From The Roundtable" section you can read about **Devin Wakefield**'s creative tips for dealing with CF and **Sonya Ostensen**'s experience with a COVID-19 diagnosis and recovery.

I hope you enjoy reading this winter issue as much as I did! In the words of Effie Trinket from *Hunger Games*, may the odds be ever in your favor, Sydna.

Publication of *CF Roundtable* is made possible by donations from our readers and grants from Sustaining Partners - AbbVie, Gilead Sciences, Maxor Specialty Pharmacy, Mylan, The Smith Foundation in memory of Jeannine Ricci, and Two Hawks Foundation in Memory of Dr. Lisa Marino; Pearl Sustaining Partners - Boomer Esiason Foundation, Cystic Fibrosis Foundation, John & Susan McMurry Fund in memory of Peter McMurry, Scholarship for the Arts in memory of Helen M. Eisenman; Diamond Sustaining Partner: Marina Day, Trustee of the McComb Foundation; Endowment Partner - Diamond Level - Nancy Wech (in memory of daughter, Lauren Melissa Kelly & in honor of son, Scott Kelly).

Information From The Internet...

Compiled by Laura Tillman

PRESS RELEASES

In CF, Certain Factors Could Predict More Severe Hemoptysis, Study Finds

Factors such as diabetes, a specific type of fungal infection, and previous mild to moderate episodes of hemoptysis (coughing up blood) may help predict the risk of massive hemoptysis in people with cystic fibrosis (CF). Older age, advanced lung disease, *Pseudomonas aeruginosa* lung infections, CF-related diabetes, portal hypertension (high blood pressure in the liver's portal vein), and liver cirrhosis have all been described as risk factors for hemoptysis in CF. But it is still unclear which factors could predict the development of massive hemoptysis in CF patients who have experienced mild or moderate episodes in the past. To investigate which factors could lead to massive hemoptysis, a group of researchers did a retrospective analysis of the clinical data of all adult CF patients who had a history of at least one mild to moderate hemoptysis episode. Massive hemoptysis was defined as a volume of coughed blood greater than 240 mL in 24 hours, or more than 100 mL over two or more days. Coughed blood leading to airway obstruction or changes in blood flow was also considered a symptom of massive hemoptysis.

LAURA TILLMAN

Mild to moderate hemoptysis was defined as an episode resulting in a volume of coughed blood between 5 and 240 mL. The findings suggest that "BPA, metabolic disorders and recurrent mild-to-moderate haemoptysis may predict MH [massive hemoptysis] occurrence in CF patients with at least one mild-to-moderate haemoptsis episode in the past.

https://tinyurl.com/y38l2lbm

Study: Screening For Hearing Loss In CF Patients Feasible With Tablet-

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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, high-resolution photo of yourself as well as your name and contact information. Email all submissions to: articles@usacfa.org. Or go to our website: www.cfroundtable.com/newsletter.

Winter (February) 2021: Late Diagnosis. (Current issue)

Spring (May) 2021: Sexuality and Sexual Health in CF. How was your experience of puberty and adolescence influenced by having CF? In what ways has your sexual health been affected by CF? Do medications or treatments affect your sexual health? When and how do you bring up CF when dating?

Summer (August) 2021: Alternative Therapies and CF. What alternative therapies do you use in addition to your prescribed treatment plan? What resources do you use? How has the addition of alternative therapies helped your CF? What advice would you give to those seeking other options for their CF care? Was your CF clinic receptive to adding in these alternative therapies?

Autumn (November) 2021: Balancing Caregiving for Self and Others.

ASK THE ATTORNEY

Be Informed

By Beth Sufian, J.D.

CF Roundtable readers have submitted important questions related to the law in the past three months. People with CF who receive Social Security disability benefits have many questions about eligibility. Many people with CF are considering a return to work when COVID-19 risk reduces sometime in the future. Anyone considering a return to work should make sure they understand the Social Security rules related to work and continuing benefits or continuing Medicare or Medicaid before returning to work.

If you have questions about your own situation, please email the CF Legal Information Hotline at CFLegal@sufianpassamano.com or call 1-800-622-0385. Nothing in this article is meant as legal advice and is only meant to be legal information.

Question 1: I have lost my job and cannot afford to elect health insurance coverage through COBRA. How can I purchase a policy on the Affordable Care Act Website?

Answer: If a person is in need of health insurance, the person can go to www.healthcare.gov to see what policies are available in his/her area. The open enrollment period for purchasing a policy ended on December 15, 2020; however, a person can get special enrollment if that person meets one of the special enrollment reasons that allow enrollment with no exclusion for a pre-existing condition. If a person loses their insurance coverage, that person has 60 days to purchase a policy on www.healthcare.gov. That person can also purchase a policy from a specific insurance company, but that person will not have access to possible help with premiums, which is only available to those who enroll in a plan through www.healthcare.gov.

Question 2: Why is my Supplemental Security Income ("SSI") benefit reduced by 30 percent because my parents pay my rent?

Answer: A person who receives SSI benefits must have low income and low assets. SSI also has rules that consider who pays for a person's housing and food. If a person on SSI is unable to pay their share of rent and food in addition to certain other expenses, then Social Security reduces the SSI check by 30 percent each month. This is called the Social Security SSI Household Deduction.

A person must have a written agreement to pay their share of household expenses (rent, food, electricity, utilities, and garbage fees) if the person wants to avoid the SSI Household Deduction. The written agreement is typically submitted to the Social Security Administration ("SSA") during the SSI application process. The written agreement must contain certain language. If the person who receives SSI benefits cannot pay their share of household expenses, then the SSA will use the SSI Household Deduction to reduce the monthly SSI benefit check.

BETH SUFIAN

For example, if the SSI recipient lives with their mother and the rent is $2,000 then their share of rent is $1000. In most states, the maximum SSI benefit is $783 a month. The person in this example would not be able to pay their share of household expenses because their share of rent alone is over the amount the person will receive from SSI. Social Security would reduce the SSI check by 30 percent. California and New York also add state money to the federal SSI amount of $783, so, if a person lives in California or New York and receives SSI benefits, the total amount the person will receive when the state supplement is added is higher.

If a person cannot initially pay their share of household expenses and then moves and is then able to pay their share of household expenses, the person can submit a rental agreement to Social Security to try to have the SSI Household Deduction removed.

The CF Social Security Project will have funding from the CF Foundation to assist with Social Security SSI Household Deduction issues for some individuals with CF starting January 2, 2021. Please contact the CF Social Project for more information at CFLegal@sufianpassamano.com.

Question 3: I receive SSI benefits and I am thinking of taking a part-time job. Will I be jeopardizing my SSI benefits and Medicaid benefits?

Answer: People with CF who have never worked or have not worked

enough to qualify for Social Security Disability benefits are only eligible for SSI benefits. SSI recipients must adhere to all of the asset and income restrictions set out in the Social Security regulations. If a person goes over the low-income asset and income criteria, the person may lose both the cash SSI benefit and the Medicaid coverage that SSI recipients receive.

In the 13 states that have not expanded Medicaid eligibility for low-income adults under the Affordable Care Act, one of the only ways to be eligible for Medicaid as an adult is to be on SSI. Therefore, it is extremely important for people with CF on SSI who live in those 13 states to make sure they maintain eligibility for SSI.

If a person who receives SSI benefits works part time, Social Security will deduct $1 from the monthly benefit check for every $2 earned from work activity. So, if a person earns $400 from work activity in one month, then the person receives a deduction in the SSI amount of $200 for that month. Typically, Social Security finds out about the work income after the SSI benefit has been sent to the person, thus triggering an overpayment in a future month.

The Social Security Plan to Achieve Self-Support ("PASS") program allows a person to work part time and save money for a work goal and the work earnings do not reduce the SSI check.

Generally, Social Security work rules limit work activity to 20 hours or less per week *and* making no more than $1,260 per month from work activity. If the person is self-employed, the monthly work amount limit is $910 per month before taxes are taken out of the work earnings.

Question 4: I receive Social Security Disability Insurance ("SSDI") benefits. I have stopped working part time due to the COVID-19 risk. Does my SSDI check increase?

Answer: An SSDI benefit amount does not increase if a person is no longer working part time. If a person has been receiving SSDI and works part time making under $1,260 a month (before taxes are taken out of the check) or $910 a month if self-employed, the SSDI check remains the same if the person stops working part time.

SSI is the Social Security benefit that reduces the monthly SSI benefit by the amount of any monthly work income. ▲

Beth is 55 and has CF. She is an attorney who focuses her law practice on disability law and is the Treasurer 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.

TILLMAN continued from page 3

Based Tests

Tablet-based hearing tests can accurately screen for hearing loss in people with chronic lung diseases such as cystic fibrosis (CF) without specialist supervision. This finding suggests that tablets may be an inexpensive and practical addition to programs that monitor patients for signs of hearing loss. Life-threatening bacterial infections are common among patients with chronic lung diseases such as CF. When caused by certain microbes known as gram-negative bacteria, patients often are prescribed repeated treatments with aminoglycoside antimicrobial medicines, such as azithromycin, which can lead to hearing loss — a process referred to as ototoxicity (also known as "ear poisoning"). Hearing loss can be minimized through early detection of its symptoms, but developing effective outpatient programs to monitor for these presents challenges. Conducting a hearing test — audiometry — in a sound booth is costly, time-consuming, and requires a trained audiologist. To address this challenge, a team analyzed the accuracy of tablet and web-based hearing tests. These tests could be performed by non-specialists in outpatient settings. Web- and tablet-based results were compared to those from a hearing booth. Results from the tablet-based test correlated well with those from the hearing booth. The tablet test correctly detected most cases of hearing loss (high test sensitivity) and it proved to be especially accurate in detecting instances of no hearing loss (high test specificity). Although the web-based test proved better at accurately identifying patients who truly had hearing loss, it was less sensitive overall and more likely than the tablet-based test to identify a patient as not having hearing loss when, in actuality, they did. Both tests outperformed the Hearing Handicap Inventory for Adults questionnaire — a standard self-reported measure of hearing loss — across nearly all measures. Furthermore, tablet-based tests were associated with a good user experience, with participants reporting a high preference for them, followed by the booth-based and web-based formats. This study highlights the significant [hearing-related] impact of antimicrobial prescribing in adults with CF, and presents data highlighting the applicability of tablet-based audiometry as a practical screening tool with high accuracy that can be used within integrated ototoxicity monitoring programmes in chronic lung disease to identify ototoxicity at an

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SPIRIT MEDICINE

The Spirit Of Acceptance

By Isabel Stenzel Byrnes

We are well into nearly a year since we sheltered in place with COVID-19. Many of us CFers have been extra careful about going out. Many of us have adapted. We have more Zoom events than we know what to do with. We have found ways to occupy our time: cooking, reading, exercising, browsing YouTube, working, or pursuing hobbies.

When I was a child, my parents had an old wooden frame that looked much older than it was hanging in the bathroom. The top line read in calligraphy, "The Serenity Prayer." I don't know where it came from and my parents were never part of the recovery movement, where this prayer is most famous. I never understood what it meant as a kid. But now, as an adult, I have truly appreciated its words: *God grant me the serenity to accept the things I cannot change, courage to change the things I can and wisdom to know the difference.*" Isn't this particularly relevant for living with CF? And living through a pandemic?

This article is about the spirit of acceptance. Acceptance means understanding something is what it is. The pandemic. Having CF. A dysfunctional family. A loss of any kind. Only after we fully accept what *is*, can we move toward doing something about it. This includes grieving and experiencing the emotional and spiritual fallout of what is.

As a grief counselor, I see so many people who protest the death of their loved one. The disbelief and denial are natural early on in loss; people simply don't want what is. They want what they used to have. And it is a long, slow, hard process to accept fully—intellectually and emotionally—that their loved one is not coming back and this is the life they have now.

In Buddhism, there is the story of the two arrows. The first arrow is the thing that hurts when it hits us. That could be a cancer diagnosis, a divorce, the death of a loved one, or an unmet goal. The disappointment is a blow; the unexpected interruption stings. The second arrow is what hits us next. The second arrow is our reaction or response to the first arrow. It can be anger, regret, sorrow, grief, fear, guilt, denial, depression, or self-inflicted numbing or self-blame. The first arrow is pain. Pain is inevitable in life—we cannot avoid it. The second arrow is suffering. Suffering is our attitude toward our pain. Suffering is optional. And so, our life mission is to come to terms with our suffering.

In a recent support group, we talked about COVID-19 and sheltering in place. One woman said, "There's nothing I can do!" Another said, "I have so many choices in what I can do!" It is interesting that the exact same situation brings such different responses. I've had a chance to practice my responses to another health scare that has come up lately. I can freak out, I can get down, I can deny it. Instead, I am practicing to just let it be. I will face what I need to face and deal with it. I'll find the wisdom to know what I can control and surrender what I can't.

> *"Only after we fully accept what* is, *can we move toward doing something about it."*

ISABEL STENZEL BYRNES

Sometimes acceptance implies passivity. There is a Japanese word, *shikataganai*, which means "it can't be helped." Sometimes this has been used to describe Japanese culture as weak or indifferent. Rather, it's a Zen philosophy to save energy by acknowledging what is and using that energy for things that can be changed. When a challenge comes my way, I sure would like to save energy!

Acceptance does not mean resignation, cowering under oppression, or giving up. We all have a survival instinct; accepting we have a problem is the first step toward cultivating motivation for problem solving. This is the American way. To survive with CF, we must possess creativity and ingenuity when it comes to figuring out alternatives or

solutions. The development of modifier drugs is a prime example of not accepting this disease as inevitably fatal; thousands of people have worked really hard over the years to find drugs to treat it.

One of the great strengths of the human spirit is defiance. To me, defiance is determination. It's raising your fists in the air *after accepting what is* and proclaiming your power to persevere. Michelle Compton, a dear friend and beloved member of the CF community, wrote a poem about defiance of end-stage CF: "I am coming to terms with my life...I am coming to terms with my death—and finding beauty in both. I am not trapped. I refuse to be frightened. I will not be conquered." She describes a freedom in acceptance. What an awesome attitude! If I were Michelle, I'd be so proud of myself for arriving at this realization. In some ways, acceptance and defiance can raise confidence and self-love.

And acceptance does not mean we have to *like* what we have to accept. We can hate what is, but soften around it. Accepting that our choices are limited right now during this pandemic or accepting that we have to go on disability or need a transplant invites us to examine what is in front of us. Acceptance leads to awareness. Similarly, awareness also leads to acceptance. When we look at our lives, we realize this may not be the life we want but it's the one life we have. Acceptance can mean less drama and more harmony in your inner life as well as with relationships. Imagine if we accepted people for who they are! Life could be easier. The Bible says, "For God gave us a spirit not of fear but of power and love and self-control." (2 Timothy 1:7) Acceptance will bring solace to our spirits. May you find its power. ▲

Isa Stenzel Byrnes is 49 years old and has CF. She lives in Redwood City, California. She is 17 years post-lung transplant.

SPEEDING PAST 50

Better Late Than Never

By Kathy Russell

What a year 2020 has been. I am so tired of P and P (pandemic and politics!). The pandemic has worn us down for most of this year and there is no end in sight. I am tired of having to stay home, having to wear a mask, having to wear gloves, having to keep six feet between myself and anyone else, and just having the concern about a vicious virus. The politics just kept getting more and more irritating. I think we could do better by using the method that is used in the United Kingdom where campaigning is limited to about six weeks prior to an election. Wouldn't that be enough time to let all of us know all that we need to know about any candidate? Oh well, this is the system by which we live.

At least I am feeling well. Trikafta seems to be working well for me and I am so grateful for the good feelings I am enjoying. I haven't always felt this well. When I was very young, I felt really rotten most of the time. No doctors would believe my mother when she would tell them of my health troubles. If they didn't see it with their own eyes, it just couldn't be so. Since we lived out in the country, and I rarely was able to get to the doctor's office while I was feeling so ill, they just discounted all of my mother's reports.

Despite my frequent bouts of lung infections and pneumonia, no one believed my mother when she talked with them about the status of my health. I missed a lot of school. Every time some "bug" went around the school, I was sure to get it. It seemed as if I always had an ear infection, a cold, or worse. I was teased for missing so much school and my parents were rebuked for allowing me to stay home when I "should have been in school." I remember one classmate's mother told me that it was a good thing I wasn't her daughter because she wouldn't have let me stay home from school for a "runny nose." To myself, I thought it was lucky for me that she wasn't my mother!

By the time I was 12, I was losing ground with my health. I was getting sicker and sicker. When I was in the hospital, some doctors decided that I should be tested for CF, even though I was "too old to have it." They did what were called duodenal drainages. This entailed putting a tube through my nose and down the esophagus, into my stomach and into the duodenum at the other end of the stomach. The only problem was that my nasal passages were too small to allow the tube, with its steel perforated ball, to pass into my esophagus, so it had to go down through my mouth. The goal was to ascertain if I was producing the correct amounts of digestive enzymes. I would have to lie on my right side, with the tube hanging out of my mouth and draining into an emesis basin that was placed on a chair next to the bed. This procedure was started at about 7 a.m. and continued all day until almost dinner time. Of course, I was not allowed to eat or drink anything while the tube was in. Since the result was less than what it should have been, the whole procedure was repeated the next day. This went on for a week.

"Despite my frequent bouts of lung infections and pneumonia, no one believed my mother when she talked with them about the status of my health."

KATHY RUSSELL

They never got the results they wanted, so they decided to try something that was quite new back then—a sweat chloride test. Back then, these were done differently from how they are done now. My back was cleansed with acetone and a piece of gauze about eight inches square was put on my back and covered with a large square of plastic that was taped down completely. The combination was left on for several hours. This also was repeated for a few times. Since I was allergic to the tape that was used, my back had blisters and was rather sore after that.

It turned out that my sweat chloride was extremely high. The doctors couldn't believe that I could have such a high sweat chloride and still be alive at 12 years of age. They didn't think that I could survive for long because I was "so old." They didn't bother with many treatments, since I was "so old." I didn't even have to do nebulizer treatments or sleep in a mist tent. I am so happy that I didn't have to sleep in a tent. I think that, if I had slept in that wet environment, I might have gotten pneumonia more often than I did.

My own private doctors couldn't quite believe that I had CF. What little they knew about it meant that children with CF couldn't live to 12 years of age. They would say that I had a "fibrocystic lung condition." Their denial did not help me with my acceptance of the diagnosis. In their defense, I will say that they had attended medical school in the 1920s and early '30s so cystic fibrosis was not something that they studied in school. After all, CF wasn't identified until 1938 and it was thought to be a disease only in infants.

Having a diagnosis didn't change the attitudes of people outside of our family. They didn't *get it* and were proud of their ignorance. At least my own family did get it and did their best to keep me well and active.

At the start of the school year after my diagnosis, a doctor at the medical school where I was diagnosed sent a letter to my school telling them that I wasn't allowed to participate in any physical activities. That doctor had never even seen me and did not know what would be best for me, but I had to comply with the edict in the letter. At least I was allowed to keep up my normal activity at home. I still participated in ballet and acrobatics, or tumbling as it was called then, and kept very active. I am sure that activity was much better for me than inactivity. With all of my movement I was able to cough up the junk in my lungs and was able to keep breathing.

Fortunately for me, at that time I didn't have too many bad bugs in my lungs. Mostly, I cultured only *Staphylococcus aureus.* It was somewhat sensitive to the antibiotics that were around at that time. There were more antibiotics available in 1956, when I was diagnosed, and they were not quite as expensive as the earlier ones.

It seemed to me that the docs tried each new antibiotic as they came out. Some of them had some long-term effects that were not positive. Some of them caused me gastric symptoms, others caused damage to my teeth, and still others seemed to do very little.

I was able to get through the eighth grade without missing a single day of school. I realize that for many people that would be no big deal. For me, it was a very big deal since I had missed the equivalent of three years of school in my first seven years. My eighth-grade year was the first time that Asian flu had come around. Only two of us in my class failed to contract that flu and both of us were the only two with perfect attendance.

High school was another case all together. I started out okay, without missing any school until January. I was sick for a week or two then. I missed a little time here and there until the end of third term. I was studying for finals and suddenly was very tired. I told my mother that I was going to take a nap and I asked her to wake me in an hour. She tried to wake me after an hour and was unable to rouse me. She kept trying for about three hours. My breathing was okay and I seemed to be sleeping comfortably, so she let me sleep.

The next morning, when she still could not wake me, she called my doctor. He said to let me sleep, as long as I was breathing okay and didn't seem to be getting congested. I slept for about three days. At that point, my parents took me to the doctor. He could find nothing wrong that would be causing me to sleep. He decided that I was exhausted and needed rest. I missed the final 10 weeks of that school year. After a few weeks, the school district sent a tutor to help me with my studies. She tried to help me, but I was so weak that I couldn't do much work. Even though I missed a quarter of that year, I still was able to start my sophomore year with my class.

The rest of my time in high school was much less dramatic and I graduated with my class. I then went on to nursing school and eventually got a job in a hospital. I worked in hospitals until I was in my 30s. Since I kept picking up viruses and infections, my doctors wanted me to stop working. They were fairly certain that I would be exposed to many fewer ills if I stayed out of hospitals. They were correct.

I applied for Social Security Disability Insurance but was denied. I appealed and tried a couple more times, but I was denied each time. I decided that living without the money I was used to earning was much better than dying from some rotten bug. We had to adjust our budget to manage without my salary, but we made it and I have lived long enough to collect on the money that I paid into Social Security.

I am 76 years old and doubt that I would have lived this long if my mother had not been so insistent about getting the doctors to pay attention and treat me for cystic fibrosis. So, even though I was not diagnosed until I was 12 years old, I believe that it was better late than never.

Please stay well and be happy.

Kathy ▲

Kathy is 76 and has CF. She and her husband, Paul, live in Gresham, OR. She has been a Director, Treasurer and President of USACFA, as well as a past Editor of CF Roundtable. *You may contact her at krussell@usacfa.org.*

PEARLS OF WISDOM

Rare Mutations In Context: Multiethnic Biography, Multiracial Lineage, And CF Diagnosis

NEW COLUMN

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

Welcome to the first official entry in my "Pearls of Wisdom" column! If you've been reading *CF Roundtable* regularly, you probably spotted the teaser in our Fall 2020 issue for this column. So, you know that I'll be focusing on lessons learned in my journey as a medical educator and public health program evaluator living with cystic fibrosis. I can't think of a better way to get started than to share with all of you some lessons I've learned about my own CF in the context of my personal and family history! Like many other things about me, such as my sexuality and gender, my experiences of both ethnic identity and CF diagnosis have been complex—and characterized by feeling "in between" different ways of being.

In this issue of CF Roundtable, we're sharing many different stories about the diverse experiences of late diagnosis in our adult community. These include several narratives from patients of color, some of whom have very diverse racial and ethnic backgrounds and some who can trace their families' origins to a few specific places. Across these stories, we can easily see how the experiences of adult patients who are Black, Brown, and/or Indigenous have been shaped by racist stereotypes of who can and cannot have CF. Likewise, we're featuring several narratives from ethnic minority patients within the broader community of white adults with CF. These stories also illustrate complexities in the diagnosis and treatment journey that can wind up reinforcing other forms of discrimination and harm.

Both racial and ethnic inequality—and the intersection of the two—persist in the diagnosis and management of CF. The diverse stories in this issue of *CF Roundtable* demonstrate the urgent need to address inequity and injustice in all aspects of both pediatric and adult care. For people with CF and other progressive diseases, time is often the most valuable resource we have. An early diagnosis and early treatment can make tremendous differences in both the quality of our lives and the quantity of living we can do. So, when people who already experience racial and/or ethnic marginalization struggle even to get diagnostic testing for CF, those existing hardships get exacerbated by lack of access to proper care. In this way, late diagnosis itself can both illustrate systemic inequality within the CF community and deepen the harms done by those disparities.

Like racism and ethnocentrism themselves, disparities in diagnosis and treatment begin to do harm the moment a child comes into the world. The newborn screening that has enabled many white patients born in hospitals to access quality CF care from their earliest days of life only tests for a few dozen of the most common CFTR gene mutations. Because CF can be caused by over 2,300 known variants on the CFTR gene, a lot of us get missed by that early screening! And of course, this screening was not even available when many of us who are now adults were born. I was five years old when the CFTR gene was first discovered and remain the only known CF patient in the US with my specific CFTR variants! Although I am racially white—meaning that I am read as white in most situations and thus experience white privilege—newborn screening would have led my family to believe that something other than CF was causing my health challenges.

> *"Like many patients with multiracial lineage and/or multiethnic backgrounds, I don't have any common CFTR mutations."*

XAN NOWAKOWSKI

Sweat testing did not help much either, largely because my providers could not manage to collect enough sweat for a valid sample! I am not good at sweating; even in very hot environments I will only sweat a little on my brow line. The fluid that comes out of the skin along my hairline is so corrosive it will eventually eat through the band on a hat. But, because the process for collecting sweat sample uses only

the skin on the arms, I never did manage to produce a useful sample—not at age four when I was originally tested for CF and not even at age 33, when the tests were repeated with modern methods for research purposes. This was around the same time I learned what CFTR variants I actually had. Unsurprisingly, I did not have any copies of either the 508Fdel or other common CF-causing mutations.

This is where my heritage and lineage become relevant. A total of three people contributed to my physical and cultural history. There's my mother, whom I am related to both genetically and socially. Her mother was German and Danish; her father was Tuscarora and Scottish. So, I knew I had some Indigenous lineage and heritage from her side of the family. Culturally, I am Polish on my father's side—his family lived in Poland until the early twentieth century. My Slavic ethnicity[1] and heritage have deeply influenced my social and cultural consciousness. However, my father and I are not related genetically because I was conceived using sperm from an anonymous donor.

I've often referred to this third person simply as "the donor" but have increasingly come to understand them as something more. Connecting with members of that part of my genetic tree has made that other part of my history very real and present for me. Over the past few years, thanks to genetic testing and a lot of detective work, I've been able to learn about the other side of my genealogy. My biological father seems to be mostly of Creek descent, from the Natchez region within the Creek nation. The family also traces a lot of their lineage back to Egypt; a few Sicilian traders joined the family later on and settled in what is now central Mississippi. Most of that family—and my half-brother from the same donor—are still living in the area today.

So why does all this matter for my journey with CF? Well, like many patients with multiracial lineage and/or multiethnic backgrounds, I don't have any common CFTR mutations. This means my road to diagnosis was more complex even after the CFTR gene was discovered, and although I had presented pretty textbook CF symptoms from my earliest days of life. I got a tentative diagnosis of CF at age five after my tiny, invalid sweat samples came back borderline. But this was not confirmed until I got my complete CFTR genes sequenced at age 33. The time I lost in the interim will never be given back to me; sometimes I still struggle emotionally with this. These days, though, I don't know if I would want to go back and do anything differently. Too much else in my life—too much that is good and meaningful and fulfilling—has come from my own struggles in trying to get a conclusive diagnosis and appropriate care.

Foremost among these positives is my unique and nuanced ability to advocate for racial and ethnic justice in CF diagnosis and treatment. Patients like me are presently making up for lost time without the benefit of transformational drugs like Trikafta, because our CFTR mutations do not qualify us to take modulators. Some of us who may be modulator eligible have also missed our chance to benefit from these therapies because the damage our CF has already done to major organs like our livers and kidneys is already too extensive to allow us to safely take these drugs.

If you think my story of not getting a conclusive diagnosis until age 33 sounds harrowing, consider all the patients with darker skin or other more strongly racialized features who have had to fight even harder for an even longer time to get diagnosed and access appropriate care. Several of my personal heroes in the CF community were in their 50s and 60s when they were diagnosed. Think about the injustice of that, and also about the tremendous strength and determination it takes to survive that long with CF without any guideline based care services. And think about how all of those hardships intersect with other forms of injustice.

As a multiethnic white adult with CF who has survived a lot—and learned a lot along the way about my own multiracial lineage and how that may have contributed to some of the struggles I faced in getting a conclusive diagnosis—I can use my voice constructively to educate, advocate, and uplift. Really examining how we diagnose CF—and how we intentionally transform diagnosis approaches and services for patients of marginalized racial and ethnic backgrounds—will be a vital priority in this time of so many exciting changes in adult care. Making late diagnosis a thing of the past will not be possible without robust and sustained attention to racial and ethnic oppression. By actively focusing on justice for racial and ethnic minority patients, we can ensure that every person with CF gets the opportunity to become an adult patient and share our wisdom with those who are just starting out on this complex journey. ▲

Dr. Alexandra "Xan" Nowakowski is 37 years old and has CF. Xan is a director of CF Roundtable, *in addition to being 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. You can find their contact information on page 2.*

[1] Ethnicity is multifaceted. It comes from lineage, experience, socialization, acculturation, etc. So, although I am not genealogically Slavic, I am ethnically Slavic because I was raised by a Polish American parent with all of their customs and history.

ASK THE ENT

Your ENT Questions Answered

By David Gudis, M.D.

Q: I have chronic sinusitis because of my CF. What's new in the treatment of CF-related sinus problems?

A: Patients who have cystic fibrosis have a very high incidence of chronic sinusitis—by some measures, over 90%. Historically, CF-related chronic rhinosinusitis ("CRS") has been very difficult to control. Many of our *CF Roundtable* readers and their friends have undergone multiple sinus surgeries and some may still have sinus problems. Moreover, your sinuses can serve as "reservoirs" for the types of bacteria, like *Pseudomonas*, that can then infect the lungs. Typical symptoms of CRS include nasal obstruction or congestion, nasal discharge, sinus pain or pressure, and decreased sense of smell. Interestingly, many patients with CF do not have sinus symptoms, even if their sinuses are chronically infected.

But we have some GOOD NEWS! Over the last several years, rhinologists (ear, nose, and throat doctors who subspecialize in complex sinus surgery) have developed and studied new and better ways to treat sinuses. A combination of the proper surgery and the proper post-operative management can really help patients feel better and keep the sinuses healthy.

DAVID GUDIS

And there's more! Early data shows us that the new CF modulator medications are great for sinus problems, too. As many of our readers know, there are four cystic fibrosis transmembrane conductance regulator ("CFTR") modulators: ivacaftor (Kalydeco), lumacaftor/ivacaftor (Orkambi), tezacaftor/ivacaftor (Symdeko), and elexacaftor/tezacaftor/ivacaftor (Trikafta). Our CF Center at Columbia University published a study in the *Journal of Cystic Fibrosis* demonstrating that patients taking Trikafta have a huge improvement in sinus symptoms. Several other CF Centers have since published similar data. Be sure to speak to your CF doctor about these options.

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

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

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early stage.
https://tinyurl.com/yyxnuten

Self-Management Intervention Supports Treatment Adherence In Adults With Cystic Fibrosis

A self-management intervention aided adults with cystic fibrosis in achieving greater adherence to inhaled medications, habit strength, lower perceived treatment burden and higher BMI over 12 months compared with usual care. However, results demonstrated no significant difference in exacerbations or FEV1 between the intervention and usual care. Since untreated adherence for nebulized therapy in adults is about 30% a Study Team developed a self-management intervention, described as underpinned by behavioral science therapy and with input from patients with cystic fibrosis to improve adherence to inhaled medications. The current study aimed to evaluate effectiveness of the intervention compared with usual care. The primary outcome of the study was exacerbation and secondary outcomes included adherence, habit strength, BMI and FEV1.

At 52 weeks, the level of adherence in the intervention group improved to about 52.9%. However, researchers observed no significant difference in exacerbations between the intervention and the usual-care group or FEV1. BMI slightly increased, habit strength significantly increased, and Cystic Fibrosis Questionnaire Revised (CFQ-R) treatment burden significantly declined.
https://tinyurl.com/y58e9686

S. *Aureus* Found Mostly in Lung Mucus, Not Tissue, Pig Model Shows

Using pig lungs as a model, scientists discovered that *Staphylococcus aureus* preferentially colonizes the mucus circulating there rather than organ tissue itself. According to the researchers, these findings may open new ways of treating such lung infec-

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The CF Roundtable 2020 Award Winners

The *CF Roundtable* has presented two awards to members of the CF community for over 20 years. Readers submit nominations and the *CF Roundtable* board votes on who receives the award.

The Jacoby Angel Award is named in memory of Dr. Jack Jacoby who was both an adult with CF and a CF physician at the St. Vincent's CF Center in New York City for more than 15 years. The Jacoby Angel Award recognizes a person who has followed in the footsteps of Dr. Jacoby by dedicating themselves to helping others.

On October 18, 2020, *CF Roundtable* presented both the Jacoby Angel Award and the Founders Award during the *CF Roundtable* virtual Hope for the Future event.

Terry Wright is the recipient of the 2020 Jacoby Angel Award. Terry Wright is the President and founder of the National Organization of African Americans with Cystic Fibrosis, an organization that works to help engage, educate, and raise awareness of CF in both the African American and the broader community. His efforts have already directly increased the number of African American adults who are now being diagnosed with CF in the U.S.

Terry was misdiagnosed for more than 54 years. He suffered numerous health issues due to an incorrect assumption in the medical field that African Americans do not have CF. After his diagnosis Terry could have focused on himself but instead decided to work to make sure no one else would have to suffer because of a misdiagnosis. Terry's *CF Roundtable* blog post this past spring chronicling his journey to his CF diagnosis has helped educate the CF community on issues African Americans with CF face in the United States.

Terry has spent his life helping others. He has been actively involved in his community, volunteering for many local organizations and working to help others even though he himself was facing serious health issues. He is a dual-certified Master Gardner and Naturalist. He had a 30-year career as a certified personal trainer. In January 2020, Terry was awarded a CFF Impact Grant, becoming the first recipient from Arkansas.

TERRY WRIGHT

MARTHA MARKOVITZ

Previous recipients of the Jacoby Angel Award include Robyn Petras, Susan Burroughs, Michele Compton, Jerry Cahill, Pammie Post, Isabel Stenzel Byrnes, and Dr. Paul Quinton.

The Founders Award recognizes a person, either with or without CF, who has made an outstanding contribution to the adult CF community. The award was named in honor of the group of adults with CF who founded the U.S. Adult CF Association ("USACFA") and created *CF Roundtable*. They worked tirelessly to bring information to the adult CF community at a time when there was no internet and no efforts to connect adults with CF in order to provide information and support.

Martha Markovitz is the recipient of the 2020 Founders Award. Martha has been a social worker at the University of Southern California CF Adult Care Center for 40 years. Before there was a CF Adult Center, she served as a social worker at the CF Pediatric Center. Martha is known as the "Queen of Social Workers." She has been a mentor to hundreds of social workers who work in CF and has imparted her knowledge of how to help people with CF. She has been a leader in advocating for adult care when there were very few speaking out for the needs of adults. Martha has personally led the way in advocating for assistance with insurance issues, access to medical treatments, and implementing standards of care for CF Centers to follow, which has made access to CF adult care better for thousands of adults with CF.

Martha is known for meeting her patients where they are and figuring out ways to help them even if it takes her hundreds of hours. She has a heart of gold and retired at the end of October 2020. Martha has been a pioneer in adult care, and the lives of all adult with CF are better off because of her tireless commitment to making the lives of people with CF better.

Past recipients of the Founders Award have included Lisa McDonough, Dr. Jim Yankaskas, Darlene Hello, Dr. Jerry Nick, James Passamano, Dr. Jennifer Taylor-Cousar, and Cathy Chacon. ▲

When I found out I had an incurable, chronic, life-limiting illness I was devastated. I was overwhelmed, to say the least. The guilt was unbearable. Had I known I had a life-threatening illness would I have lived life differently? Would my husband have ever considered marrying a woman who was, at the very least, living with an illness that required constant vigilance and, at the worst, left him yet again a widower, now with even more children? I also felt vindicated: after all the years of doctor's visits and illnesses, only to be told it was another cold, I finally had answers—cystic fibrosis.

My first visit to a cystic fibrosis clinic did not go how I expected. My husband and I loaded up our twins and drove in to find out how long I had left to live. But when we got there, the visit was much longer than the 20 minutes I initially anticipated. In hindsight, it was poor planning to take the twins to what ended up being a three-hour long visit. Sadly, the clinic answered almost none of my questions. Rather, the doctor asked me nearly every question known to man and then they asked me to cough into a cup. I had spent my whole life avoiding coughing in front of people and not spitting out my mucus—I wasn't about to start now. They took x-rays and then said they would call. So, I went home and waited for what felt like forever. When my doctor finally called with x-ray results, I quickly asked about my life expectancy. She told me the average age and what factors might influence that number, but that ultimately, it was hard to predict. I hung up the phone and began researching on the internet. I joined some Facebook groups to learn more and connect with others. Overall, I felt completely lost. In retrospect, I should have made a list of questions for my first CF clinic visit, but I had no idea what to expect at the time. I thought it would be more like a cancer diagnosis—you're told you have cancer, how long to live, and what treatment options are available. That is what I was ready for and I thought it would happen in 20 minutes, maybe 30 minutes, tops. And I am a nurse! I am not sure what other people go in thinking, but I was not ready for this.

Let's fast forward to my first hospitalization: it was at Thanksgiving and it didn't go well. I still had very young children at home who were having a hard time with mom being gone from home. In my denial, I mistakenly assumed I would never be hospitalized for cystic fibrosis. I had mentioned it during one clinic visit and it was waived off as a future possibility only if things got bad. And then, out of the blue (OK, maybe not that out of the blue), I got super sick. I waited until I was really, really sick. My doctor had no clue what bug was making me so ill. I was hospitalized for 10 days and now, in hindsight, I can tell you that I should have remained in the hospital for at least 14 days. I was discharged early because a doctor came to see me and found me hiding in the bathroom, on the floor, crying my eyes out. I wasn't eating or sleeping, and my anxiety level was through the roof. It was such a shock to me being in the hospital for such a long period of time. Prior to this hospitalization, I had only been hospitalized to deliver my babies. But the length of time for this stay and not knowing ahead of time what to expect was very hard for me to accept. And there is the obvious—being in the hospital completely sucks. You have no control, no privacy; people continually come and go from your room, effectively disrupting everything; you feel terrible physically; and you're away from your usual comfort items. On top of all that, I felt guilty for being away from my responsibilities.

> "I thought it would be more like a cancer diagnosis—you're told you have cancer, how long to live, and what treatment options are available."

The next hospitalization was still hard, but better overall. This time, I cried out in the open rather than in the bathroom. At this point, I had three years of a CF diagnosis under my belt and I knew a little something about how to deal with it but still very little about being hospitalized for CF. Most patients leave after a couple of days in the hospital—why did I need to be there for two weeks? I was not as sick as the first time, so why did they need to keep me for so long now that I could breathe *and* talk at the same time? During this admission, I found the nurse case manager and asked why I needed to stay for so long. I told her that I had been diagnosed three years prior and that even though I myself was a nurse, I had no clue what was going on with my care. As a nurse, I had worked in the operating room, in home-health settings, and in case management. I had no experience working with other CF patients. Despite having researched cystic fibrosis, it still made no sense to me. It's one thing to know something based on research and an entirely different thing to live it. Everyone who walked into the hospital room assumed this wasn't my first rodeo. The nurse case manager was very understanding and passed my concerns along to my care team. She brought me a book that explained what being hospitalized with cystic fibrosis really looks like. She also asked the social worker to come talk to me. It still wasn't perfect, but, for the first time in a long time, I took a deep breath.

Not unsurprisingly, I met my catastrophic out-of-pocket maximum that year. That's what my insurance calls it, anyway. $5,500 per year. That only includes the things that the insurance counts. Somehow, I still met the criteria for having had a catastrophic year. It's not hard to see why. Copays for this, a percentage of that, hospitalizations here, and daily medications everywhere. I thought catastrophes were supposed to be a once-in-a-lifetime event, not a yearly expense that we have to plan for and pay. When do the so-called catastrophes stop? They don't. I keep getting hospitalized. I keep getting sick. I keep needing medication, antibiotics, doctor's visits, and lab work. So, as I merrily count down the days until Christmas, I also feverishly schedule everything I can into the end of the year while there are no copays and no out-of-pockets costs since I've already hit the maximum. I continually hope that I can have a small break from the "catastrophe" that is my new normal.

It's nice to tell my story and reflect on some of the things that have happened to me. I would have loved a "What to expect now that you know you have CF" book and I would still love a "if you have this happen, it means this" book or a "these are all the organizations that could help you and how" book. When I got a sweat test, my husband thought I was crazy. He was sure there was no way I could be diagnosed with cystic fibrosis as an adult. But here I am. I no longer fit into my old world. That world has changed out of necessity. Some of my friends seem to understand what a big change this diagnosis has been, and they have continued to travel this road with me, but others have fallen by the wayside. I am not a great fit in the cystic fibrosis world, either. Some people, who have been diagnosed for longer, find it hard to be friends with people who are new to the world of CF and are still trying to understand all of it. I know I am still trying to figure it out. I am the new kid in a class that has been together since they were in kindergarten and it is hard to join the club. Especially since it's a club with such strong bonds. I am much more comfortable with my cystic fibrosis diagnosis now. In the end I was never told when I was dying, but with each bug I have grown, with each PFT I take, and with each antibody I react poorly to, I calculate what "we'll see" is turning into and I worry that I won't be able to fulfill the commitments I made to be a mother to these eight children and to grow old with a man who has already been a widower once. I no longer ask the question "how long until cystic fibrosis kills me?" Instead, I try to focus on living in the moment and refuse to allow the future to hold me hostage any longer. ▲

Angeline Chase is 37 years old and has CF. She lives in Kaysville, Utah. She likes to listen to audiobooks, watch Asian melodramas on Netflix, garden, hike, play with animals (especially her little dog Millie), tickle babies, and has taken up playing a Nintendo switch game (adventure fit ring) for exercise.

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tions, including treatment-resistant infections, enabling a lower amount of prescribed antibiotics against *S. aureus* than is now normally given. Antibiotics often fail to fully eliminate lung disease and its symptom, leading some to question whether antibiotic use is best possible treatment for *S. aureus.* The bacteria appear to localize in mucus plugs in the lumens of bronchioles. Bronchioles, one of the smallest lung airways, are connected to the alveolar ducts that house the alveoli. Researchers found that when *S. aureus* colonizes pig lungs it tends to form aggregates in mucus, rather than invading the organs' tissue and forming abscesses. Unlike previous reports in mice, *S. aureus* in pig lungs were not seen to invade the animals' lung tissue. Instead, these bacteria seemed to concentrate and form large aggregates in the artificial CF mucus that had been added to the organs. No abscesses were observed. According to the team, these findings may provide valuable insights into the effects *S. aureus* has on the lungs of CF patients, which could lead to new and more effective treatments for infections caused by these bacteria.

https://tinyurl.com/y3xqudmq

AND

https://tinyurl.com/y2azt24n

Twincer May Be Better Alternative To Nebulizers For CF Colistin Treatment

Twincer, a dry powder inhaler (DPI), was shown to be a more appealing approach for the inhalation of colistin when compared with a nebulizer. Using a nebulizer, a small device that turns liquid medicine into a mist, is the most common method of administering inhaled antibiotics as a treatment for CF patients infected with the bacteria *Pseudomonas aeruginosa.* DPIs are a more patient-friendly option because they have a much shorter administration time, are more stable, do not require an external power source or refrigeration, and are three to six times more efficient. Notably, it takes about 1–2 minutes per dose to administer colistin with Twincer, compared with 10-20 minutes with a nebulizer. Twincer is a disposable inhaler for a single use that allows the delivery of relatively high doses of antibiotics. Researchers did a real-life evaluation study, with a small number of patients, which showed that colistin dry powder inhalation using the Twincer is a patient friendly alternative to nebuli-

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Building Antibodies With Balance

By Sonya Ostensen

The information provided below is only one individual's personal experience and is not meant to be medical advice. Please contact your CF care center if you have any concerns about COVID-19 or other CF-related illnesses.

SONYA OSTENSEN

While I downplayed the symptoms in my head, reality crept in. A 102-degree fever plus body aches and fatigue equaled a sickening dread in the pit of my stomach. My husband tested positive for COVID-19 and, as he came back into the house with his mask on to grab clothes in order to self-quarantine in a different location, I couldn't help but wonder if this would be the last time we would see each other. It was an emotional departure as our six-year-old daughter and I stood sobbing on the front porch. Turns out, we were able to reunite two days later when I came down with similar symptoms and tested positive for COVID-19 as well. It hit us so fast. Suddenly, our anxiety and worries if we would survive overshadowed the chore of what to fix for dinner. Would this tear apart our family? Would my daughter grow up without her mom or dad or both? I needed to snap out of it and had a pep talk with myself, vowing to focus on the positive and draw from that energy. Listen up self! There will be no panic!

My daughter was not tested per the doctor's recommendations as she didn't have symptoms; thus, we quarantined and monitored her condition. Thankfully, my daughter remained mostly asymptomatic except for a dry cough. So, on the first day after my diagnosis, I climbed a tree with her. Why? Because she asked me to—because it was my way of taking control and because it's what a parent does for their child. It could have been my last chance to ever climb a tree with my daughter, and I was not going to miss the opportunity. I will tell you that it took every ounce of energy to climb the tree, and all of my joints and muscles hurt beyond belief—worse than any flu aches and pains. However, I did it and it felt good to breathe in the fresh air, listen to the birds, and watch the butterflies. As we talked and played make believe, I savored every moment. We were in the present enjoying life.

It has been over a month since the first day of symptoms. I am pleasantly surprised at how well both my husband and I have recovered. Neither one of us had to go to the hospital and our daughter has remained unaffected. Although I am not back to my baseline and do not know whether my lung functions will suffer any long-term repercussions, I do feel strong and energetic. So, I wanted to share a list of things I did to heal and promote recovery from COVID-19.

1. Water—I drank water like a camel preparing for a trip to Timbuktu.
2. Sleep—the fatigue is real. Sleeping was key the first few days. I've heard that the fatigue tends to linger off and on for months afterwards.
3. Upon the advice of my CF team, I ordered a pulse oximeter online and monitored my peripheral capillary oxygen level (SpO_2) regularly. Any reading below 90 is not normal. An SpO_2 reading below 88 can be dangerous and might warrant a trip to the hospital.
4. I continued taking 50 mg of zinc every day. Zinc is a mineral with antiviral properties that helps strengthen the immune system. Zinc is a preventative and essential nutrient—your body does not produce or store zinc on its own, yet it is needed for many metabolic processes.
5. Vitamin C to help my immune system.
6. Whole food dietary supplements that contain concentrated fruit and veggie extracts. This is a normal regimen for our family. When I run out, there is a noticeable energy deficiency. Ideally, we should have doubled our doses; however, we were running low and I wanted to make it last so all our bodies would benefit during Covid-19 infection.
7. ADEK vitamins—continued normal regimen of one vitamin per day.
8. Buried Treasure ACF Liquid Rapid Immune Support—this is an over-the-counter, immune-boosting supplement. I discovered this a few years back and always take it when I first feel the onset of a cold. There are over 15 different natural ingredients, so, if you are considering this product, make sure to discuss with your doctor.
9. Per doctor's orders, I doubled my

existing nebulizer treatments—albuterol, 3% hypertonic saline, and Pulmozyme. I also continued my normal course of inhaled antibiotics, which happened to be Tobi that month.

10. Increased airway clearance.

11. Continued use of 500mg Azithromycin three times a week

12. Tylenol for my throbbing headaches, which were compounded by any screen time whatsoever.

13. Movement—after two days of lying around and sleeping, I realized it only made the body aches worse and my lungs felt congested. Even though I had no energy, walking for short periods of time gave some relief. There is a delicate balance between activity and resting during illness—both are needed.

14. Miso soup and sweets were the only things that tasted good. I was able to maintain a small amount of taste until the eighth day. Smell was completely gone by the sixth day. My appetite was zero from the onset of symptoms.

15. Sinus rinses with the addition of Alkalol every other day. Alkalol is an over-the-counter saline solution containing essential oils and natural extracts (eucalyptus, spearmint, and menthol, to name a few) that act as a mucous solvent helping to clear nasal passages for sinusitis, post-nasal drip, and allergies.

16. Trikafta—the drug that without a doubt gave my body the extra boost to recover and most likely prevented hospitalization. The heartfelt gratitude I feel for the timeliness of this modulator is indescribable. I remain hopeful for the 10% of our CF community to have a modulator medication available as soon as possible.

While taste and energy have returned, sadly my sense of smell is not back. It was diminished before COVID-19 due to chronic sinusitis and multiple sinus surgeries; however, now it is worse. I did not completely realize this until my daughter and I did a turtle nest dig with the UCF turtle guide well after recovery from COVID-19. After turtle nests hatch, they are dug up to count the number of eggs that have hatched, among other things. As you can imagine, this is a stinky process. While the guide and my daughter were turning their heads away from the repugnant smell, I could not smell anything. COVID-19 often causes anosmia, a temporary loss of smell. So far, my olfactory senses haven't returned but *c'est la vie*; if that is the extent of long-term damage, I accept this small loss with grace and humility.

> "Looking back, past CF exacerbations have left my body more debilitated than COVID-19."

Looking back, past CF exacerbations have left my body more debilitated than COVID-19. Unfortunately, not everyone has been so lucky, those with and without CF. This time, there were no feelings of trepidation that my time on earth was ending. However, as I write, my good friend who does not have CF, has been diagnosed with COVID-19 and is fighting for her life in the ICU. With a heavy heart this leads me to search science for cause and effect of this strange virus. It has affected so many lives in an odd variety of ways.

All of this leads back to balance. A popular theory based on genetic analysis reveals horseshoe bats as the origin of this novel coronavirus. Wildlife trade, markets, urbanization, and farming threaten biodiversity around the world. History has shown us that all of these foster opportunities for viruses to become zoonotic diseases. Mass clearing of forests, tremendous amounts of fertilizer and pesticides continually feeding into aquifers, harmful farming practices, and construction on wetlands are throwing ecosystems out of balance causing an alarming rate of species extinction. We, too, are becoming imbalanced, resulting in an unhealthy society, both physically and mentally. This is a wake-up call to look within and rediscover personal equilibrium in the hope of spreading a more cohesive, sustainable way of life in order to heal each other and our home—planet earth. It all seems so daunting, much like cystic fibrosis can be; however, not impossible if we continue to persevere, making one small change at a time. As Francis J. Braceland said, "[w]e can be sure that the greatest hope for maintaining equilibrium in the face of any situation rests within ourselves."

Author update: As of day 41, Sonya received a negative COVID-19 test. Her antibody tests came back positive on day 50. Her pulmonary lung function tests only dropped by three percent. She still has not recovered her sense of smell. Most importantly, her great friend has walked out of the hospital and is on the road to recovery! ▲

Sonya Ostensen is 45 years old and has CF. She lives in Melbourne, Florida, with her husband and daughter. She received her Bachelor's in Science in Environmental Sustainable Resource Management from Ohio State University. She loves to travel with her family and experience new cultures, and she has a passion for wildlife rehabilitation. Her favorite activities include gardening, baking, walking the beach, and climbing trees with her beautiful daughter. Sonya is a CF Roundtable *director. Her contact information is on page 2. You may also message her through Facebook at Facebook.com/Sonya.Ostensen*

FOCUS TOPIC

LATE DIAGNOSIS

Becoming An Open Book

By Megan Felch

I found myself so angry—not angry because I found out that I have cystic fibrosis at 33 years old, amidst an unprecedented pandemic, but angry because I always had a suspicion that I have it. None of the doctors I had seen in my 33 years had figured it out. Maybe I'd still have a gall bladder? Maybe my kidneys might be in better condition? Maybe I would have less damage in my lungs? Oops, I'm being pessimistic again—it's hard not to get lost in the "what if" cycle.

I had never met a doctor who could solve my mucus problem. Every primary care doctor from birth to 33 blamed it on asthma, allergies, sinus infections, bronchitis, and/or pneumonia. All could be solved with new rounds of inhalers and antibiotics every few weeks and, if I was lucky, every few months or even a whole year. At 23, in between what had become normal lung infections, I was physically sicker than I had ever been. Every time I ate, I broke out in a sweat, I screamed in pain, and I couldn't keep anything down. My doctor said there's no way you're pregnant and you're too young to need your gallbladder removed. This particular doctor we saw made me take a pregnancy test on the spot. When he didn't believe the result, he ordered a blood test. Ironically, the doctor's nurse practitioner had prescribed me birth control a year prior because I had lost too much weight and couldn't gain it back. Not unsurprisingly, he was wrong about the pregnancy speculation and my gallbladder came out shortly thereafter.

Countless times I heard, "You have indigestion. Don't eat these foods and your mucus will go away." It didn't. I also heard, "Have this surgery and your sinuses will be fixed, then your mucus will go away" or "Take these allergy shots every day for 30 days and you'll have relief for up to six months." My primary doctors said, "Come in as soon as you have any symptoms so it won't turn into pneumonia again." They were all wrong and I had lost hope. I stopped telling doctors how I felt and, for a brief period, stopped going to the doctor entirely (I don't recommend this). I remember when someone told me that my cough sounded like I had a yeast overgrowth from having too much sugar in my diet. That was a new one! Everyone was willing to offer their unsolicited advice: "don't drink milk," "only drink room temperature water," "only drink ice cold water," "clean up your diet," "chew gum," and "don't chew gum." I stopped telling my family and friends how I was feeling. I was so tired of people asking if I was sick that I made a sarcastic joke about it: "I don't get sick." No one I knew coughed like me. No one I knew was sick like me. No one I knew had this much struggle, took this much time off work, and was this tired every day.

> "I was scared because I had already researched CF and had read the doomsday articles on the internet that told me I only had five years left to live. Cue the emotional breakdown."

MEGAN FELCH

Shutting down came easy to me. I have never been one to divulge my emotions, my personal life, or my health. I have treated doctor's appointments like business transactions ("I'm good, how are you?"), skipping right over the long list of symptoms I've had over the last three months to take the spotlight off me. I've even spent sessions trying to get my therapists to talk about themselves so that I wouldn't have to talk about me. Money not well spent.

I hid my cough as best as I could, but anyone who knows this cough knows that it's impossible to hide. Eventually, my cough was too much to bear. Another few years on and off antibiotics every three months and I was researching all of the time. What

could be making me this sick? For years I had wondered if I had cystic fibrosis. Based on my research it had to be CF or primary ciliary dyskinesia. Or possibly early-stage heart failure according to a *Washington Post* story I had read. I finally asked my doctor if I could be tested for cystic fibrosis. He told me that I would be sicker, that I would have been diagnosed as a baby, and that I wouldn't have made it this far. Maybe he's right, I thought, maybe it is just seasonal allergies.

Finally, in the spring of 2020, I was tested with a genetic testing kit. The person reading the test asked, "Has anyone told you that you have cystic fibrosis before?" She explained that I had two mutations that were not common and that I should go to the cystic fibrosis clinic as soon as possible to confirm. The first few minutes of my first clinic appointment confirmed my suspicions but having the suspicion still couldn't prevent the shock from my new diagnosis. I was scared because I had already researched CF and had read the doomsday articles on the internet that told me I only had five years left to live. Cue the emotional breakdown.

It's now been about nine months since my diagnosis. My clinic has been so incredible. This community has accepted me and my "CF light" with open arms. Despite my rare mutations, I qualified for Kalydeco and started doing two breathing treatments per day. Finally, my cough is (mostly) gone. I still can't believe it. I made a promise to myself that I will not dismiss my symptoms anymore. I take notes, I ask questions, I go over every single symptom with my clinic. I was so afraid of finding out that my only condition was hypochondria that I didn't fight for the best care. Some of the best advice I've been given from the cystic fibrosis community is to put myself first, advocate for myself, to be the annoyingly detailed and thorough patient because no one else is going to do it for you. And to stay off the life expectancy side of the internet.

I wish I had more to say, I wish I had all the answers. The truth is that I don't. I'm still finding out all that's "wrong" with me. I know I'm not the only person to be diagnosed during COVID-19 because unfortunately many of us weren't tested as babies. To be honest, the pandemic might have made the adjustment easier for me. It's hard to feel insecure in a mask when everyone else is wearing one, too. My hope is that anyone else who has been recently diagnosed uses the resources available: connect with a peer, attend the conferences, talk with your clinic, talk to your social worker, and talk to your dietician. It's hard to not shut everyone out, but I know that's what I need to do in order to accept it myself. ▲

Megan Felch is 33 years old and has CF. She lives in Dallas, TX. Megan was raised in Pennsylvania, studied economics at Penn State, and works in commercial real estate. Megan likes to spend her free time drawing portraits in charcoal, exercising, playing with her rescued bulldog, Winston, and torturing her boyfriend, Nathan. She can be reached by email at mcfelch@gmail.com.

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zation, with no significant differences found in the clinical outcome regarding lung function decline and exacerbation rates. The researchers acknowledged several limitations to their study, especially its small sample size. Thus, further studies are needed to confirm these findings.
https://tinyurl.com/yy5aascm

Intravenous Vs Oral Antibiotics For Eradication Of *Pseudomonas Aeruginosa* In Cystic Fibrosis (TORPEDO-CF): A Randomised Controlled Trial

Researchers undertook this multicentre, parallel group, open-label, randomised controlled trial to compare intravenous ceftazidime and tobramycin vs oral ciprofloxacin in terms of effectiveness and safety in the eradication of *Pseudomonas aeruginosa* in patients with cystic fibrosis. Findings showed that intravenous antibiotics, in comparison with oral therapy, failed to provide sustained eradication of *P. aeruginosa* in a greater proportion of patients with cystic fibrosis and was more costly. The intravenous group had fewer hospitalisations during follow-up compared with the oral group, but this conferred no benefit over oral treatment since intravenous eradication often needs hospitalisation. In the light of these findings, the use of intravenous antibiotics to eradicate *P. aeruginosa* in cystic fibrosis was not supported.
https://tinyurl.com/y4clx4pm
AND
https://tinyurl.com/y33f9n3w
AND
https://tinyurl.com/yyv4tu4l

Delafloxacin—A Novel Fluoroquinolone For The Treatment Of Ciprofloxacin-Resistant *Pseudomonas Aeruginosa* In Patients With Cystic Fibrosis

This investigation was undertaken to test in vitro susceptibility of delafloxacin against a population of *P. aeruginosa* isolated from adult cystic fibrosis (CF) patients. In addition, delafloxacin and ciprofloxacin in vitro susceptibilities against CF *P. aeruginosa* were compared. Experts also evaluated where delafloxacin may add advantage in treating CF *P. aeruginosa*. Findings revealed

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Late Cystic Fibrosis Diagnosis: Growing Up In The Dark Ages

By Jerry Cahill

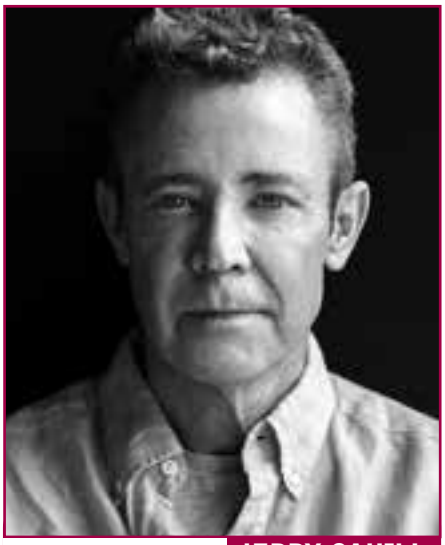
> "I chose to never be defined by my disease, but by my character and resilience."

According to the doctors who diagnosed me in 1956, I shouldn't be here today; I shouldn't have lived past the age of 16.

I was born on a typical day in June—the fourth boy, in what would eventually be a family of six children. As an 8-pound 9-ounce baby, I was seemingly the picture of health. It wouldn't be long before my parents realized that I was anything but that.

My childhood was not like that of others. I was chronically ill and regularly diagnosed with everything from allergies to colds to asthma and bronchitis to pneumonia. Finally, after years of misdiagnoses and struggling with health issues, I was diagnosed with cystic fibrosis via a sweat test at the age of 11.

I started regular breathing treatments, but it wasn't an easy process since I began at such a late age. My siblings and friends would be outside playing on sunny, beautiful spring days in our Brooklyn neighborhood (where I used to be pre-diagnosis) while I followed a regimented, daily routine of nebulizers and digestive enzymes. Long ago, the enzymes were in powder form that I had to mix with juice and hold my nose just to get them down. And don't get me started on the postural draining regimen—my mother and brothers had to pound on my back and chest twice daily to loosen chest congestion. Some days it was painful to the point that I felt more like a punching bag than a little boy.



JERRY CAHILL

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greater activity of delafloxacin vs ciprofloxacin, given similar breakpoints of these fluoroquinolones. With sensitive isolates, equal effectiveness was displayed by both delafloxacin and ciprofloxacin, however, the value of delafloxacin was observed with more resistant isolates to ciprofloxacin. While ciprofloxacin was suggested to be employed as the first line fluoroquinolone for treating CF *P. aeruginosa*, experts also concluded that delafloxacin has potential in treating ciprofloxacin-resistant *P. aeruginosa*.
https://tinyurl.com/yytnoehc

Inhaled Tobramycin Effectively Treats Initial *P. Aeruginosa* Infections In CF

Inhaled tobramycin alone works as well at eradicating an initial infection by *Pseudomonas aeruginosa* bacteria in people with cystic fibrosis (CF), and its use in combination with an oral fluoroquinolone, such as ciprofloxacin or levofloxacin, appears to be redundant. The study results also suggest that inhaled tobramycin is as effective in a real-life setting as in clinical studies, validating current trial-based treatment guidelines.
https://tinyurl.com/y4evgysq

Bronchitol Inhalation Powder Approved In US For Adult CF Patients

The U.S. Food and Drug Administration (FDA) has approved the dry inhalation powder Bronchitol (mannitol) as a maintenance treatment to be used along with other therapies for improving lung function in adults with cystic fibrosis (CF). The therapy is expected to be launched in the U.S. market in March 2021. Bronchitol's active ingredient, mannitol, is a sugar alcohol that is able to attract water. When directed to the lungs, using a small dry powder inhaler device, mannitol draws water into the airways and moisturizes the mucus, making it easier for patients to expel the mucus. By increasing mucus clearance, Bronchitol can help improve lung function and quality of life for people with CF. It's the only inhaled dry powder indicated to be used along with other treatments

Despite all of this—and probably because of it—I became a fighter. I chose to never be defined by my disease, but by my character and resilience. I stayed as active as possible, integrating exercise into daily routine, which led to my becoming a student athlete through college.

I excelled in college, graduating *cum laude*, and moved on to a successful career in the fashion industry, all while no one knew I had CF. Once the disease inevitably caught up with me, I moved on to volunteer at the Boomer Esiason Foundation, where I am able to place my drive and focus on helping others with cystic fibrosis.

In 2012, I tackled getting a double lung transplant like I do anything else—I brought a positive attitude and the determination to get back to living my life the way I want to live it as quickly as possible.

Looking back, being diagnosed later in my childhood may have been a blessing. I lived 11 years without cystic fibrosis being a constant part of my life and I believe that ultimately led to the way I dealt with it. I knew it was a part of me; I knew I had to be vigilant, aggressive, and always compliant with my treatments; but I didn't have to let it be the defining factor in my life.

A cystic fibrosis diagnosis, even a late one, is not a death sentence. It is a license to live a life you never imagined through perseverance. You never give the disease control; you take control over it. If you're struggling with it, remember my story, the struggles I faced all those years ago, and become a survivor. For me, the dreaded, predicted age of 16 came and went—and I was still here, thriving. Don't get me wrong, it was never easy and still isn't. But the life I have been able to live is worth every moment of the fight. ▲

Jerry Cahill is 64 years old and has CF. He lives in Brooklyn, NY. He loves biking, jogging, weight training, and coaching high school pole vaulting. You can watch his documentary, Up For Air, here: https://youtu.be/p35qG-Bjwig.

to improve lung function in adults with CF. Bronchitol offers a portable and discreet option for CF management, with no routine cleaning or maintenance of the inhaler device required. Participants in three Phase 3 studies (NCT00630812, NCT00446680, NCT02134353) were randomly assigned to receive either Bronchitol or a placebo for 26 weeks, and entered an open-label treatment portion for an additional 26 weeks. The main goal of these trials was to determine improvements in lung function, measured by change from baseline in forced expiratory volume in one second (FEV1), over 26 weeks. Other measures of lung function and pulmonary exacerbations, or respiratory infections leading to worsening of lung function, were also examined over time. Overall, results showed a significant improvement in lung function in treated patients when compared with a placebo. The rates of pulmonary exacerbations also tended to be lower after Bronchitol treatment — though not significantly lower in all trials. While one of these trials examined Bronchitol in patients 6 and older, the safety and efficacy of this treatment in children and adolescents is still not clear. Thus, the current indication for Bronchitol is for clinically stable adults only. The most common

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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

Don't Stop Believin'—Journey To CF

By Jeanie Hanley, M.D.

I grew up in a large family with five brothers and three sisters and was the eighth child in the pack. To top off the enormity of my parents' caring for such a large household, my sister Theresa, who was a year older than me, was always sick with bronchitis, sinusitis, and pneumonia. She was one of the primary reasons that, at an early age, I wanted to become a doctor—to find answers to this mysterious respiratory disease, one that doctors couldn't adequately treat or sufficiently diagnose. In my preadolescence, I started having the very same symptoms she had. My hope to someday figure this out took on a new dimension and became even more urgent. But, I was very young. How much could I do except study hard, so I could eventually get into medical school.

Our care alternated between our family doctor, pulmonologists, and specialists at academic centers. Theresa, who was the sickest, was tested once for cystic fibrosis. The doctor told my mom that the sweat chloride result was not high enough and added that testing was really an academic exercise only, not a realistic consideration, since CF was not found in the Hispanic population (circa early 1970s).

Despite two more siblings (making four of us in total) developing recurrent bronchitis, pneumonia, and GI problems, the possibility of cystic fibrosis was not revisited in my youth. Over the years, my siblings and I had been worked up for many diseases; none panned out. Instead, we were given a generic diagnosis of an unusual form of asthma and allergies because my brother had tested positive for allergies; yet none of us improved much on the suggested asthma therapies.

During the first year of medical school, my fellow students and I were often guinea pigs for our professors' research. For one protocol, we all had to cough onto agar plates that, on mine, grew *Pseudomonas*. The professor used its sweet-smelling odor as a class-

JEANIE HANLEY

learning experience. I looked healthy enough on the outside so he brushed it off as a contaminant. If only I knew then what I know now.

When I brought up the possibility of my family members having CF to other professors, they quickly dismissed it and attributed it to "medical student syndrome" where students think they are experiencing the symptoms of the disease being studied. A few also blatantly stated I was the wrong skin color (olive)/ethnicity (Mexican-American) and/or too old to have CF at 24 years old. By the time of my med school graduation, despite the contrary responses from my professors, the seed was firmly planted and sprouting that cystic fibrosis could explain my siblings' and my symptoms.

As our symptoms progressed to include hemoptysis, Theresa and I were then diagnosed with bronchiectasis and allergic bronchopulmonary aspergillosis ("ABPA"). While the diagnosis fit better, it didn't explain everything (e.g., Theresa's GI symptoms, my brother's infertility, and the fact that neither my brother nor oldest sister suffered from the symptoms of ABPA, but did have recurrent pancreatitis and recurrent respiratory infections).

During my pediatric residency, I had treated a handful of kids with cystic fibrosis. One was Black while the others were non-Hispanic white. The possibility of CF grew as I realized that not all cases followed the textbook description, but could affect different races and cultures. The gene had recently been discovered (1989) and CF genetic testing was in its nascent stages. There was no way to prove we had CF since our sweat chloride tests were in the normal range. Although my suspicions were strong, there was also a seed of doubt. Even so, I believed that I was getting closer to the answers to our diagnostic dilemma.

I decided to further my medical training by subspecializing in allergy, asthma, and immunology and this helped me on my quest to further define our illness. During my two years in the subspecialty, I had access to many specialists—not only allergists, but geneticists, pulmonologists, infectious disease doctors, and others. Many noted my chronic cough and assumed I was the "typical doctor" and not compliant with my asthma meds. If I only had a nickel for every time someone said "doctor, heal thyself," I'd have pockets overflowing with coins.

The good news is that my time in the allergy subspecialty would ultimate-

ly pay off. It began when I attended a national medical conference. As I perused the program, a presentation about cystic fibrosis caught my eye. It was starting soon, so I ran over to participate. Excited to make it (and out of breath), I quickly entered the room. Immediately, I realized this was not meant for allergists-in-training like me. It was a small group discussion of 15 prominent CF Center directors, like Drs. Bonnie Ramsey and Richard Moss, who were running the meeting. A waiter was serving breakfast and gestured for me to take the one seat available. Hesitantly, I took it. It was a relief that the group didn't kick me out, but, in fact, welcomed me after introductions, even though I was not a CF director. This meeting was the turning point and led to the answers I craved.

Besides getting a delicious breakfast, I learned about the different classes of CF mutations, which was groundbreaking information at the time. The presenters also explained how each mutation class affected different organs in severity and conferred age-related differences. After the discussion ended, I presented my family to Dr. Moss from Stanford, who listened intently, asked a few questions, then encouraged me to get the family tested with the six CFTR mutation analysis that was available at that time. I'm forever grateful for his thoughtfulness and encouragement, which was the first I'd ever received.

Upon my return, a geneticist I had worked with agreed to test my family, if only to prove to me to give up this quest for the possibility of CF. At Christmas time, I took the buccal (inner cheek) scrapings of my parents and all siblings. The results showed that my three siblings and I who had chronic respiratory and GI symptoms had one 508Fdel mutation. Another sibling, with no symptoms, carried the same mutation as did my mother. Like so many past doctors, the geneticist brushed it off as part of the CFTR mutation carrier frequency in the general population. But his ulterior motive of trying to convince me of my folly fell through, as it only convinced me to forge on. I knew the low sweat chloride threw everyone off and even I had a trickle of a doubt. I looked too healthy on the outside even with my chronic, junky cough and many other symptoms. But, like the Stanford professor, I needed someone who would truly give weight to my words and think outside the box.

During my second year of allergy training, I became even sicker, taking massive doses of oral steroids and receiving intramuscular shots of tobramycin and oral ciprofloxacin to control the bacterial *Pseudomonas*. My allergist colleague treating me had confirmed ABPA, but also diagnosed "mucoviscidosis" a catch-all term meaning an illness with thick mucus. Mornings were the worst for symptoms, yet I always went to work, inhaling albuterol and Mucomyst in the car, no matter how fatigued I was. One gloomy morning, there was a Grand Rounds talk on rare diseases, where a scientific paper was discussed describing two sisters in their twenties who were newly diagnosed with CF. They both had normal sweat chloride tests and were diagnosed by a research lab that was conducting rare CFTR mutation analyses. I felt this was the ticket! As soon as I had a breather (pun intended) that day, I contacted the principal investigator and postdoctoral fellow who wrote the paper. They readily agreed to analyze my family's genetics. The day turned bright and filled with hope.

Thank goodness for the holidays during which I could take advantage of having all my family together (yet again!). I drew their blood and sent it off for testing. Two months later, the postdoc called me to say they had found the other mutation, D1152H (a class 4 mutation) and that, indeed, we did have CF. As an aside, he asked if I had Ashkenazi roots since D1152H had been found exclusively in this population. I didn't know it at the time, but, years later, an ancestry analysis would confirm that we did have these roots.

The postdoc thought I'd be sad, but I was ecstatic. I finally had the name of the disease that plagued my family. I had proof and a diagnosis that fit. My sister Theresa and I were so excited and had to celebrate that night. We couldn't have been more eager to start receiving the proper treatments and therapies.

Officially I was diagnosed at a CF center by an excellent USC CF pulmonologist who was surprised, but knew about adult diagnoses in CF. I was 33 and his first adult-diagnosed patient. My oldest sister and brother would be treated at USC too, but not until ten years later. For them, the diagnosis was more difficult to accept and it prevented them from receiving proper care.

At my first CF evaluation, the pulmonologist immediately hospitalized me. This is when my practical introduction into CF began. No medical textbook could prepare me for it and I initially felt like a deer in headlights. Even though I was very familiar with hospital routines, it hadn't been as a patient, only as a doctor, and I wasn't used to feeling helpless or dependent on anyone. Learning good hospitalization care and routines from the perspective of a patient took several stays and lots of help from my CF care team, hospital staff, and other patients hospitalized with CF, whom I could easily socialize with back then. With the proper diagnosis and treatments of regular tune-ups, daily therapies and, very recently, CFTR modulators like Trikafta, my health markedly improved.

I believe that had I and my siblings

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The Story Behind An Unlikely Champion

Lisa Bentley

LISA BENTLEY

I was diagnosed with CF when I was 20 years old. Yes, I was a sick child; but I was child with allergic reactions, so most of my ailments were attributed to allergies. When I coughed up blood, it was blamed on the feather pillows in the hotel room, wearing a wool sweater, or having dust-collecting stuffed animals on my bed. It was normal for me to be on antibiotics for most of the winter. In March, my parents would throw me and my three siblings in the car and we would drive from Toronto, Ontario, Canada, where we lived, to the saltwater beaches of Florida. Hypertonic saline was not an official therapy at that time, but it usually did the trick and we all returned healthy, tanned, and happy!

Ironically, it was my sister's illness that led to the CF diagnosis. She was always healthy but started to cough up blood when she was 24 years old. Doctors tested her for every possible illness, ranging from cancer to tuberculosis. They eventually did a sweat chloride test, confirming the CF diagnosis. Testing for all of us immediately followed. My brother; however, was not tested as he was in the process of adopting children and did not want CF on his records. Testing confirmed my CF diagnosis. Later, after my brother adopted his two children, he too was diagnosed at age 35. This explained his infertility.

My CF diagnosis came while I was a student at the University of Waterloo. I was so happy to have a diagnosis—now I could get the proper antibiotics rather than throwing broad-spectrum antibiotics at my CF bacteria. I carried on as usual—studying hard, running track and cross country, and experiencing all that university had to offer. I was very healthy when I was healthy. My main challenge was returning to baseline when I was ill, but a solid dose of antibiotics always worked.

I believe that had I been diagnosed as an infant I would have been sheltered. Instead, I had a very normal upbringing as a child and young adult. I loved sports and gradually transformed from a varsity runner into a competitive triathlete while at university. When I graduated, I became a math and computer science teacher. I balanced teaching with my passion to swim, bike, and run competitively. Seven years into my teaching career, I resigned and pursued a career in sports. CF created challenges, but managing my infections wasn't an obstacle. I often raced with an active CF bacterial infection, but my love of triathlon overshadowed my day-to-day dealings with CF.

The St. Mike's Adult CF Clinic was instrumental in managing my CF and, specifically, my lung health. Often, I would call from abroad while racing in Hawaii, New Zealand, or South America, getting advice as to whether to start a course of antibiotics, increase my inhalers, resume Tobramycin or the like. But my doctors never told me that I could not race or train. They knew that exercise made my lungs stronger and helped me clear mucus. Ultimately, they knew that my body and lungs would dictate my pace and my effort, and so I was not in any danger. Ironically, it was my primary CF physician, Dr. Tullis, who suggested I resign from teaching to pursue sports full time. I was frequently sick trying to manage a full-time teaching career with training and racing on the world stage. She reminded me that I could always come back and be a teacher—now is the time to do sport. I would bet a million dollars that she has never had that conversation with another patient!

The CF team was also realistic. While they did deal the hard realities when needed, they never discouraged my training. One January, while away at a training camp in Florida, I was advised to return home when *Pseudomonas* showed up in my sputum. Dr. Tullis wanted to eradicate the bacteria quickly and I needed to get on a Tobramycin routine as soon as possible. Meanwhile, during another exacerbation, the doctors offered to bring an exercise bike to my hospital room to increase my sputum production since physiotherapy alone wasn't working. At that time, my lung function had steadily dropped from 100% to 55% over the course of six months. I desperately wanted to avoid IV antibiotics, but I relented and spent the next five days in hospital followed by five weeks at home on IVs. I was 40 years old and that was my first and last hospitalization. That is how good my CF clinicians are at keeping me healthy.

I was blessed with an amazing career in sports. I raced for 20 years—10 of those as a full-time professional athlete. I raced 33 Ironman races (3.8 km swim, 180 km cycling, and a 42.2 km run) and won 11 of them. But my real gift is CF.

> "Regardless of wellness, we must embrace who we are and love our whole self, including loving our CF."

I don't think I would have had that success in sports without it. And that is a bold statement coming from an endurance athlete whose lungs are her engine. But each time I raced, I had purpose to bring hope to families with children with CF. As a parent, all you need to know is that there is one person thriving in spite of CF and that is hope! And I could be that hope. So, during the difficult and discouraging moments of my nine-hour races, I always had a reason to finish and do my best. I made a promise to myself to be the best person on that start with CF. With a mission statement like that, I could not fail!

All any of us can do is our best with the deck of cards we are dealt. Regardless of wellness, we must embrace who we are and love our whole self, including loving our CF. When you are compassionate with yourself, you can lead with your whole heart and reach your full potential, whether in life, business, school or sports.

Yes, CF has become my superpower despite a collapsed lung, IV antibiotics, inhaled therapies, and not being able to have children of my own. I have had more complications from CF in the last 10 years than in my first 40 years. I am grateful that, at 52 years old, I am very much alive, well, and maintaining a lung function around 88-90%

Let's all make CF our superpower and open up our lives and hearts to the incredible possibilities. ▲

Lisa Bentley is 52 years old and has CF. She originates from Toronto, Canada. She is an 11-time Ironman Champion, speaker, and coach. Lisa is also the author of An Unlikely Champion. You can find out more about her and order her book at www.lisabentley.com. You can follow her on Instagram at lisa_bentley123.

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been born today, that either CF or CF-related metabolic syndrome ("CRMS") would have been diagnosed at birth through newborn screening. A definitive diagnosis of CF would have then been given in early childhood when our symptoms began, rather than as 30-something adults. There still will be adults in their late 30s and older who, like me, will be diagnosed late in life because they missed out on newborn screening which began 30 to 40 years ago (depending on the U.S. state). I'm hopeful that late-diagnosis journeys like mine will be a thing of the past.

To this day, I feel fortunate to have had access to physician specialists and researchers who readily offered their expertise and helped me to achieve the correct diagnosis years in advance of the comprehensive CFTR mutation analysis that would take many more years. The physicians who dismissed my suspicions also figure prominently in my mind and taught me an important lesson: to not fall into the trap of only thinking "inside the box."

Dealing with my late-diagnosis journey, hospitalizations, and CF therapies have given me a greater understanding and perspective of the health struggles of others and ramped up my advocacy as a doctor. Reading insightful books on personal CF journeys like *Power of Two* were life changing as I realized I shared many experiences with those who were diagnosed at birth. This led to my becoming involved with national CF organizations like USACFA (that produces this *CF Roundtable*) and CFRI, where I was able to forge deeper connections with those in the CF community. Together these connections have eased the journey through CF, and for this, I'm very grateful. ▲

Jeanie is 58 years old and has CF. She is a director and former president of USACFA. She lives in Los Angeles and welcomes your comments. Her contact information is listed on page 2.

9,357 Days: A Late-Diagnosis Journey

By Victoria Greene

9,357 days. That's how many days I lived my life not knowing what was wrong with me. So many times, everyone said, "Why do you stay sick so much?" So many of my teachers believed I was playing hooky: There's no way one person can be sick this often." All I could say in reply was, "I have a low immune system."

Having a diagnosis to put with my illness is like finally finding out the gender of your baby. You know you have something in there. You feel it, you know it's something, but that confirmation just makes everything real. Most people would be devastated to hear those two words: cystic fibrosis. But for me, it was a sigh of relief. I *finally* had a name to put on my symptoms. I finally had a reason why I stayed so sick. I wanted to go back to all of my teachers and say, "I told you I wasn't faking!"

My life has changed drastically since that day. I now have breathing treatments, antibiotics, peripherally inserted central catheter ("PICC") lines, hospital stays, and tons of doctor appointments. Before, when I would get a cold, it was just that, a cold. Now, the moment I get a small sniffle, I have to report to the doctor for antibiotics immediately. There is no cold too small, no sniffle too short. I have to be careful where I go, whom I'm around, and what I do.

I wonder often what my life would have been like had I been diagnosed early. What would be different? Would I have ever had my child? Would I have ever gone to college, met my husband, and got married? It's hard not to wonder what if. My life is a constant battle of pacing myself. Some days I feel like I could run a marathon (although there aren't many of those days). Some days I feel like getting out of bed is going to put me into the grave. It's like walking a tightrope between what I can do and what I want to do. Will what I do today put me in the hospital tomorrow? Is that runny nose just a cold or the start of a *Pseudomonas* infection?

VICTORIA GREENE

How many people have questioned me about whether I'm really as sick as I say? How many people have said I could not have cystic fibrosis because I'm not underweight? How many times have I felt like the world is against me and no one cares?

The answer is too many to count. I feel like a prisoner in my own body and mind. It's crazy to say, but I almost wish I could look sick just so people would believe that I really am. It's crazy to even speak those words, but it's true. We live in such a superficial society where if you cannot see it, it cannot possibly be true. If someone has a broken arm, they get people to sign their cast. But with an invisible illness, there's no cast. There's no sympathy. There's only judgement and heartache because our pain is on the inside and not the outside.

Invisible illnesses are real. And the emotional toll they take is far worse than any of the pain we feel. So, when someone tells you they are sick, believe them. Ask them more about their illness. Study it yourself. Saying "you don't look sick!" seems like a compliment, but it isn't. It is a slap in the face and essentially, you are calling them a liar. You may mean well, you may not realize you are passing judgment, but you are. And the difference in "I didn't know that, how are you doing?" and "you don't look sick; I would have never known" could be the difference in a bad day and a good day for that person.

It's okay to cut off toxic family members or friends. It's okay to cry. It's okay to not be okay. But just know, late diagnosis doesn't make you any less sick than the people who have known from the very beginning. If I've learned anything, it's that my story can help others who are teenagers and struggling with no name to their illness. That alone is enough for me to believe that everything happens for a reason. I am so thankful that, even though it was late, I now know that I wasn't just "being weak" or it wasn't "all in my head." Keep searching, keep fighting—you're worth it. And if you're a parent of a child who is sick and you know deep down something is wrong, keep fighting. Never give up. ▲

Victoria Greene is 28 years old and has CF. She lives in Milton, Florida. She is an aspiring grant writer and mother of a child with autism. She loves concerts and photography. You can reach her via email at toriw8674@yahoo.com.

PHOTO BY ALEJANDRA CHAVERRI

For My Granddaughter

Your world has never
    Been on hold
You dive into the deepest
    Water like the
Swimmer you were born
    to be
Your grace under pressure
    And all you must endure
Keeps me in awe
Your sparkling self dances
    Into my heart with
      Love,
Grannie

-E. Dougherty, 2002

FROM OUR FAMILY PHOTO ALBUM...

OSTENSEN FAMILY: (KURT, SONYA AND DAUGHTER MAISLYN) VOLUNTEERING TO PREPARE FOOD BASKETS TO DELIVER TO FAMILIES FOR THANKSGIVING MEALS, BASKET BRIGADE, IN MELBOURNE, FL.

ASHLEY WILSON

SYDNA MARSHALL WITH HER DOGS, HUSKER ON THE LEFT AND CUTTY ON THE RIGHT, OUT FOR A SHORT WALK ON A RARE SNOW DAY IN AUSTIN, TEXAS.

MEGAN FELCH DOES YOGA IN ACROPOLIS, GREECE, IN DECEMBER 2019.

DANIELLE AND DAN LASSAK ON THEIR WEDDING DAY ON SANIBEL ISLAND, FLORIDA, JANUARY 19, 2019.

LISA BENTLEY, DOING WHAT SHE DOES BEST: COMPETE AND WIN.

VICTORIA GREENE WITH SON, BENTLEY, AND DAUGHTER, AUBREE.

JOHN AND JEANIE HANLEY AT ZION NATIONAL PARK, UTAH.

SARA KOMINSKY RACING OVER 26 MILES WITH A 20-POUND RUCKSACK ON HER BACK. HERE SHE IS AT THE TOUGH RUCK FINISH LINE.

LUISA PALAZOLA

JERRY CAHILL CYCLING.

GRACE WITH HER BROTHERS, KEN AND WILL KNIGHT, IN AUSTIN, TX.

FAMILY MATTERS

When Infertility Leads To A CF Diagnosis: An Interview With Tom And Mary Helmers

By Molly Pam

A few months ago, I gave a talk at the North American Cystic Fibrosis Conference ("NACFC") about the importance of integrating sexual and reproductive health care into CF clinics. After my talk, I received an email from my pediatric CF nurse, Mary Helmers. In her more than 36 years treating CF patients at Stanford Children's Hospital (first as an inpatient nurse, then as nurse coordinator of the adult and now pediatric clinic), she watched CF transition from a fatal, pediatric disease to a more manageable chronic condition with many different manifestations that led to more adult and late-childhood diagnoses.

Mary wanted my thoughts on discussing CF-related infertility and future family goals with her adolescent patients. As we talked, she divulged that she has a personal reason for wanting to make sure her male patients understand they will likely need assisted reproductive technology ("ART") to have children before they find out the hard way—her husband was diagnosed with CF subsequent to their infertility diagnosis. I have known Mary since my own delayed diagnosis, in 1998, when I was a very sick nine-year-old girl, but I never knew late diagnosis was also close to her personally. I asked if I could interview her and her husband, Tom, and they readily agreed.

Before I present the interview, I want to give a short background on CF and infertility. Because 95% of people with CF born with male organs are born without the vas deferens (the two tubes that transport sperm out to the penis during ejaculation), they are clinically deemed "infertile." This condition is called congenital bilateral absence of the vas deferens ("CBAVD"). This is distinct from being diagnosed as "sterile" because most still produce sperm, they just need science to help them get it out to create an embryo. There are many different ARTs that can help people with CBAVD have biological children. Testicular sperm aspiration ("TESA") and microscopic epididymal sperm extraction ("MESA"), referenced below, are two methods of extracting sperm. Intracytoplasmic sperm injection ("ISCI") is a method of injecting a single sperm directly into a mature fertilized egg to create an embryo. During the early 1990s, around the time Tom was diagnosed, many adults were being diagnosed with CF due to CBAVD when they were unable to have biological children.

TOM AND MARY HELMERS

When were you diagnosed with CF and how did you find out?

TH: I was 34 years old when diagnosed through UC Irvine Fertility Clinic when my wife and I were going through fertility treatment to have children. I had already been diagnosed with CBAVD, and, at the time of the diagnosis, I did not know it correlated to CF. A physician from Italy was doing his fellowship in reproductive medicine and was involved in a research study looking at males with CBAVD and the correlation to CF. I had lab work drawn for genetic analysis and I was found to be homozygous for R117H.

Were there any signs you might have CF prior to your diagnosis?

TH: Yes, I recall getting bronchitis starting in my teens and throughout adulthood, as well as having very salty sweat. My baseball caps were always white from my sweat. I had a productive cough with illnesses and tended to cough more in the early morning upon waking, especially when taking a shower (the steam made me cough). I also noticed my mucus was pasty and thicker.

How did you feel about the diagnosis?

TH: I was surprised. Then, I started thinking about it more and realized that I did have symptoms, and I knew that because I was around people with CF through Mary's work. I realized I was salty and had a cough, and those correlated with CF.

Can you describe your experience with infertility testing?

TH: Mary and I were attempting to have children and did not have success in getting pregnant despite not using protection for six months. We waited another six months and went to see Mary's ObGyn. She ordered a semen analysis for me and lab work only for Mary. Her doctor decided that doing a semen analysis first was the most cost-effective and least invasive option. The results came back—azoospermia, meaning no sperm was detected in the semen sample.

I was then referred to a local urologist who performed a transrectal ultrasound, which confirmed my CBAVD. He referred us to a Stanford University urologist who told us that we had two options: adopt or use donor sperm. Our local urologist gave Mary the number of a physician at UCLA who encouraged us to see two fertility doctors, Drs. Sherman Silber and Ricardo Asch, at UC Irvine who had some success with pregnancies in couples where the men had been diagnosed with CBAVD.

Can you describe your experience and the process you took to conceive biological children?

TH: Our experience was definitely one no one would probably believe. At UC Irvine, after two rounds of IVF, each of which required a MESA surgery for me and a round of infertility drugs, injections, etc., for Mary, we had no success. Shortly after our second attempt at UC Irvine, we learned that Dr. Asch had used eggs and embryos from his patients without their consent and gave them to other women. It was a nightmare for the fertility world.

Dr. Silber then took a group of patients to Brussels, Belgium, to access ICSI, a newer fertility technology not available in the U.S. After another MESA surgery for me, many rounds of injections and medications for Mary, and ICSI to fertilize the egg, we had a successful pregnancy. We had Matthew, our oldest son (who is now 26), in September 1994. We later went back to Belgium for a frozen embryo transfer ("FET") with no success. We then headed back to California where I had another MESA surgery and Mary had more medications and injections. John, our youngest child (who is now 23), was born in July 1997 as a result of a FET.

We went through nine rounds of fertility treatments and ultimately, we were lucky to have had Matt on our fourth attempt and John on our seventh attempt. Mary did all of the research and never gave up hope. She would not take "no" for an answer and, were it not for her persistence, we would not have our two healthy, biological children, Matt and John.

What do you wish reproductive health care personnel knew about CF?

TH: That *all* men with CBAVD should have genetic testing for cystic fibrosis and, if they test positive, additional testing for any mutations. Their partner must also get genetic testing prior to proceeding with fertility treatment. Healthcare personnel should also be aware that some men may have atypical CF but should still be tested, even if they may not appear to have CF or experience symptoms indicative of CF.

How does CF affect your everyday life right now?

TH: I do not feel it impacts my everyday life. I am aware of it, but I don't do treatments unless I get sick and, even then, I may just take an antibiotic.

Are you eligible for modulators?

TH: Yes, I was in the Vertex 770-110 trial back in 2013 and then took Kalydeco for a bit, but I ultimately decided there was no benefit in me taking it, so I chose to stop.

Were there any signs Tom had CF prior to diagnosis?

MH: To be honest, it was a running joke for us at first. I used to say, "If I didn't know better, I'd think you have CF...in fact, I would say you have CF" since I was a nurse coordinator in a CF clinic. Tom would laugh it off and insist he didn't have CF. His sweat tasted very salty and he had a pronounced cough, especially in the morning upon awakening. He shared with me that he had frequent bronchitis as a teen and into adulthood.

How did you feel about your husband being diagnosed with the disease you treat?

MH: It was definitely weird, but I think a part of me always knew he did have CF. I just could not figure out how he did, since he did not present like all my other patients who, at that time in the late 80s, were very sick.

When we went to UC Irvine for fertility treatments, we were asked to participate in a research study. The fellow explained, "We are doing a study to see the correlation between CBAVD and men with CF. I am not sure you know anything about this disease but we would like to test your blood to see if you are a carrier of a gene for CF." I said, "Yes, I know about CF. I am the nurse coordinator for a CF center!"

Immediately following this, I called my colleague, Dr. Rick Moss, and asked him if he was familiar with the correlation of CF and CBAVD and whether he could evaluate Tom. Needless to say, Tom was worked up at Stanford since this was all new to them as well. Dr. Jeff Wine, a researcher at Stanford, had a field day with this in his research lab—being a homozygote for R117H is 1:1,000,000 odds!

Continued on page 32

PAM continued from page 31

How did the diagnosis change the way you treat your CF patients?

MH: I think it was beneficial to the adult CF men under my care at the clinic. I was aware of the fertility issues which CF men exhibited. I knew the questions to ask the fertility specialist, I knew what they could expect, and I knew the cost and potential outcomes of the treatment options. It enabled me to give the adolescent and adult CF men whom I cared for better information on male infertility. I know I did counsel several adult males, a handful of whom had their own children because of my advice. I felt like it was meant to be—that I had this experience so I could help my patients. At the very least, they would know that if they wanted to have children they could and if not, that was OK, too. That, in itself, was priceless. I know it was for us.

How has the process of having biological children changed for CF men, from the time your children were conceived to now?

MH: All treatments are now more TESA/ICSI (non-surgical procedure) rather than MESA/ICSI (surgical procedure), and there is no more IVF—all embryos are developed though the ICSI procedure. The fertility clinics know about atypical males with CF and much more about CBAVD and its correlation to CF. Advances in fertility treatments are also better and more fertility clinics pre-screen for CF mutations now, either the basic or extended panel. Not all 2,000 plus mutations are checked without an insurance fight, but it is a start!

What do you think is important for male CF patients to know about CF-related male infertility (CBAVD)?

MH: Most importantly, it's important they know that they are *not* sterile and they can have their own child if that is something they want. Unfortunately, I still come across articles stating CF men are sterile, when really, they are infertile.

I would reiterate the importance of knowing the right questions to ask so no one feels taken advantage of by fertility clinics. It is also important to make sure their partner gets a full sequencing/mutation analysis to ensure they do not have a child with CF. If their partner is a carrier, then I would recommend pre-screening embryos (a/k/a PGD) to check for CF prior to implantation of the embryos into the partner. If this is something they can afford or chose to do, it gives them the options but does not mean they need to follow through with it. We were not given all the options and I had to fight to get answers. Options are always good.

On a more personal note, when I order a semen analysis for a patient, I teach both adolescent and adult males that they might only have a small amount of liquid in the container. When Tom gave his sample, the lab tech came out, pointed to a full container, and said "Excuse me, sir, we need this much." It was another person's sample that filled the container. Tom's barely filled the bottom circumference of his container. It's unfortunate that the lab tech embarrassed him.

What are some of the questions you suggest asking at fertility clinics?

MH:

Questions to make sure it is a reputable fertility clinic

How long have you been in existence? How long have you been doing male infertility treatments?

Have you had any complaints or violations at the clinic?

Genetic Testing

Will you test my partner for CF?

Which panel do you use? Can I get the whole panel?

Does my insurance cover the full panel? Some insurance companies may only cover the basic panel; however, you can fight that with a prior authorization since your partner has CF. It may require a letter from your physician to get the full panel approved.

Procedures

What are your success rates for men with CBAVD?

Which treatment has the most success: MESA/ICSI or TESA/ICSI

As a cautionary note, ensure they are using ICSI rather than IVF as that is now the protocol for CBAVD ▲

Molly Pam lives in San Francisco, CA, with her husband Adam and betta fish Sally. They have been learning bridge during COVID-19 lockdowns. If you want to play bridge with them online, email Molly at mpam@usacfa.org.

Cussing Your CF Away

By Devin Wakefield

This article is not intended to be medical advice or curative. Please always talk to your doctor before starting new treatments.

Cussing has helped me with so many CF problems. Seriously, just yelling "$&*(!!" at the top of my lungs to my backed-up bowels has literally blasted my way out of a sticky situation many a time. With such splashy results, I wanted to be sure I shared my trick with the CF community. You would be surprised just how many problems get solved by insulting your mucus plugs' ancestry. Why, just yesterday, I was sitting on the couch, and I insinuated that a mucus plug's mother was unworthy of love, and—*pop!*–I coughed it up! Another time, when my liver enzyme levels were high while I was a teenager on IV antibiotics, I expressed my desire for my CF-related liver disease to put various suggestive objects into its inflamed bile ducts. The next thing I knew, my laboratory results were credited as a medical mystery.

The most memorable time cussing has helped me fight CF was an episode of coughing up blood. I could not stop for the life of me. I had gone to the hospital because of an earlier bout of hemoptysis after an increasingly frequent series of gory episodes had concerned me and my medical team. As night loomed outside, I lay myself down to sleep and saw all the lights from various medical equipment reflected in the window. Just as I set myself horizontal, I could feel bubbles gurgling in my chest, and I immediately sat upright again, coughing. I grabbed a pink, kidney-shaped bowl and spat blood. The thing with coughing up blood is how *easy* it is to keep coughing up more, however much you know it's not good, it's not

DEVIN WAKEFIELD

right, and desperately want it to stop. As I spat again and again into the bowl, I thought to myself "this blood couldn't get laid in a textile-free bar! #$%@ you! Your mother urinates like a fire hydrant." Suddenly, the blood stopped. My FEV1 skyrocketed. I have no idea why this works, but it just does.

My first experience with this has been with bowel obstructions. They have also presented me with many an opportunity to practice this trick. When I was 12, I had such an intense series of obstructions, I had been going back and forth from the hospital back to my home almost every week. Each time I went back, the doctors put their exciting enemas to work. I often felt such pain that I would cry out. It was only when I begun using language best known for peeling bananas that I noticed my stays at the hospital would be shortened. I began to experiment. I found "*#&$!!!" to be *far* more effective than "#&$*!!" while "@#&$ $#@$ ^@#%!!!" could be used with great results. My poor mother required quite the explanation, but, once she understood, she joined me in my quest for health. Even my nurses, upon understanding what we were doing, would vigorously chant "#&$*!!" with my mother while I strained on the toilet. The resounding echoes of our cussing in the bathroom would soon be joined by a productive splash and evolve into cheers of joy.

I must warn you, sometimes this works *unexpectedly* well. I caution you of a time I was riding my bike. Another rider rudely cut in front of me without warning, and I had to swerve mightily to keep myself balanced. I assumed only someone with a bowel obstruction could behave so rudely in their rush to get home. With that in mind, I kindly informed this other rider in quite colorful language that his mother has the wit of a "@#&$ing" flea. He turned his face to me (I presume to thank me) when his eyes suddenly widened in astonishment and his eyebrows vanished under his helmet. I looked at him in confusion until I noticed the pants clasped tight around his respectable bottom had suddenly turned a much darker shade. Soon, something frightening began to drip through. The poor fellow—I had not waited for him to at *least* hop onto a toilet! I could only console myself with the fact that he no longer suffered from intestinal distress. What a lesson we both learned that day.

With this newfound knowledge, I hope you, dear reader, have a new tool to face CF. Remember that practice makes perfect, and, when your friends join in, you can accomplish so much. Your health, I hope, will reach new heights, and you will soon feel an unending peace. #&$* you, #&$* your health, and have a wonderful day! ▲

Devin Wakefield is 29 years old and has CF. He lives in Seattle, WA. In his free time, he likes to run, hike, and backpack. His contact information is on page 2.

Sicker Than I Look: Stronger Than You Know

By Danielle Lassak

Ever since I can remember, I have been dealing with gastrointestinal issues. In elementary school, I complained of stomachaches to my parents. I had recently transitioned to a new school at that time, so my parents, understandably, wrote it off as nerves. Only, the stomachaches never stopped.

Fast forward to transitioning to a new middle school. I began to notice bloating that accompanied the stomachaches. Aside from the abdominal discomfort, I was seemingly healthy. It was easy to blame the bloating on hormones and adolescent body changes, but I started to feel like something might seriously be wrong.

In high school, it was progressively worse. The stomach discomfort and bloating were increasingly painful. I began to feel nauseated on occasion. I struggled with diarrhea and constipation constantly. I was never overweight but, in search of an answer, I blamed it on my diet. I was making a conscious effort to eat healthy foods (most of the time). The diet did not help.

The day of my senior prom I was so excited to wear the light blue prom dress I had somehow convinced my mom to buy me. I was maybe in third period when I started feeling dizzy, hot, and nauseated. I knew I couldn't make it through the school day, so I begged my principal to let me go home early and still allow me to go to prom that night. He was so kind and understanding. He reluctantly agreed, but I could see the genuine concern on his face.

I looked at myself in the mirror as soon as I got home. I looked pale and something was off about the whites of my eyes. I didn't pay close attention to it because I was so tired. When I woke up from my nap, I began the process of getting ready for my senior prom. I did my makeup, my mom did my hair, I threw on my dress, and I was off!

Unfortunately, the magic was short-lived. Not even two hours into the night I began to overheat, feel nauseated, and so incredibly tired again. I thought I was going to faint in front of my entire graduating class. I apologized to my date and called my dad to pick me up. I was devastated.

DANIELLE LASSAK

I seemed to be getting worse every day, so my mom scheduled a doctor's appointment that following Monday. The doctor confirmed my skin and eyes were jaundiced. Not surprisingly, the lab tests confirmed my liver enzymes were elevated. They scheduled more tests to look at my gallbladder, but ultimately ruled it out as the culprit. During the two weeks of missed school and in between doctor's appointments, my entire body itched. It was so bad that I actually scratched myself raw. I had no appetite and an unquenchable thirst. When I wasn't drinking water, I was sleeping. Thankfully, I started to feel better around week three. I was so eager to go back to school—I felt like I had missed so much. My doctor wrote it off as an unidentified virus that attacked my liver and that was it.

Except, it wasn't. I continued having GI issues. I continued feeling sick. It was a part of my daily life except some days were worse than others. Year after year, it got worse.

Fast forward to age 23. I worked at my dream job at a great company with lots of opportunities. I had worked so hard to get there. I tried to avoid allowing my sickness to get in the way, but at times it was unavoidable. Again, I was attempting to heal myself with diet, with exercise, and by reducing stress (as much as possible given I was working 10-12-hour days). On this particular morning, I could barely get myself into work. I had skipped dinner the night before and breakfast the morning of. When lunchtime rolled around, I ordered a healthy sandwich on whole-grain bread. It sent me over the edge. I began feeling dizzy and nauseated. Sound familiar? I aggressively swung open the glass doors, ran by our office security, and shut myself in the sick room where I hovered over a trash can and hoped for the sickness to pass. When I realized it wasn't going away, I walked to my desk, grabbed my purse, and quickly exited the building without telling a soul. I drove myself home and called my mom who knew this was out of character for me and urged me to go to urgent care. On my way out the door, I was hallucinating. I blamed it on my contacts shifting, got in my car, and drove myself to urgent care.

I busted through the doors hoping the staff could see me right away. That

wasn't the case. I took a seat where I patiently waited to see a doctor. By this point, I was drenched in sweat and hallucinating yet again. The windows appeared to be waving like blacktop does on a hot summer day, but it was the dead of winter in Ohio.

They called my name and took me back to see the doctor who gave me anti-nausea tablets and asked if I felt better. I didn't. He took my temperature and immediately called an ambulance. I spent three days in the hospital. I was diagnosed with IBS and gastroparesis. I imagine this sounds familiar to many late-diagnosed CFers.

Life went on. I continued to focus on my career. Yet, every year I grew sicker and sicker. Five years ago, when I met my now husband, I told him I knew something was wrong with my health. He believed me and he stood by me when even my doctor of over 12 years refused to believe anything was wrong. I begged for more tests. I begged for help.

Fast forward to age 28. It's the morning of our wedding and I felt sick. I think I was running on adrenaline, because it was, nevertheless, one of the best days of my life. The morning after returning home from our honeymoon in St. Lucia, I woke up and I knew something was seriously wrong. There was so much pain in my upper abdomen. I was bloated beyond belief. I snuck out of bed so I wouldn't wake my husband. I began the process of scheduling a virtual doctor's appointment for that day, hoping it was as simple as a prescription they could send to the pharmacy. The pain worsened and I decided it was best to go to my normal doctor in person. When my husband woke up I told him something was wrong, and I was going to the doctor. He offered to drive me there and, as the exit approached, I told him to keep driving because I thought I was dying. He sped to the emergency room. It felt like my insides were tearing apart. I couldn't feel my extremities and apologized to him and told him I loved him because I genuinely thought that was my last day on earth.

The emergency room doctors confirmed I was suffering from pancreatitis. They explained that drinking is typically the cause and told me I probably had too much to drink on our honeymoon. I was treated like an alcoholic or a typical junkie with pancreatitis even though my toxicology test showed nothing was in my system. I was treated this way even after explaining, in chronological order, my past medical episodes to the medical staff. No one documented what I said. No one believed me.

Over the next year and a half, I continued to experience bouts of pancreatitis. It became a dark cloud that hovered over me. I was in a constant state of fear wondering when I would be balled up in the fetal position and crying in excruciating pain for days on end. I was treated like an addict when I asked for pain medication. I truly cannot describe the pain and agony that accompanies pancreatitis. It is, by far, the worst pain I have ever endured.

I knew there was an underlying cause. I knew it was related to my past medical episodes. All I needed was for a doctor to listen to and believe me. I sat across from my doctor during my current bout of pancreatitis, which lasted over three weeks. I went over my previous test results from prior medical episodes, insisting they were all related and begging him to help me get to the bottom of it. He told me all of it was "normal" for a 20-something-year old and asked me if I had ever considered anxiety meds.

That was our last appointment. I mustered the courage to challenge my doctor's opinion. I found the confidence to leave him in search of a doctor who would help me. And, that is exactly who I found and exactly how I was diagnosed. By this point, I had permanent sinus damage and scar tissue covered 10 percent of my pancreas. Because of this new doctor, I didn't suffer further damage. Because of this doctor, I was able to take Trikafta, which I like to refer to as a miracle medicine. This medication has turned my life around. Thankfully, I haven't experienced a bout of pancreatitis since I started taking it. I have more energy and I feel so much better. I laugh more, smile more, and every day is a gift.

Cystic fibrosis, for me, was hearing "you don't look sick" so many times, I lost count. It was sleeping with a heating pad velcroed around my waist for three years straight, even though the outside world didn't see that. A large portion of my paychecks went toward medical bills I could barely afford in my early 20s, even though no one knew that was why I was broke. CF meant countless doctor's appointments and a lot of normal test results that very few saw. It meant canceling plans at the last minute because I felt sicker than on the day I had agreed to go out with friends and family. CF meant calling off work at the last minute because I knew I couldn't mask my symptoms that day. It was working an eight-hour day during an active bout of pancreatitis, despite no one having a clue.

Cystic fibrosis for me was also being my own best advocate. It was never giving up even when I felt hopeless. It was me finding my voice, believing in myself, learning my strength, and being the captain of my own ship. Cystic fibrosis has many different faces and I am one of them. I'm relieved to have a diagnosis and I'm so incredibly grateful there was a medication available to me that truly gave me a second lease on life. I will never take it for granted. ▲

Danielle Lassak is 30 years old and has CF. She lives in OH. She's a wife, a dog mom, a cat mom, and an IT project coordinator. She enjoys decorating, cooking, and traveling! She can be reached via email at Danielle.Lassak@gmail.com.

A Diagnosis That Came Full Circle

By Luisa Palazola

I can't imagine a life without cystic fibrosis. Sometimes, it boggles my mind to think that, for nine years, I was a *relatively* healthy kid. That is, until I developed relentless walking pneumonia at eight years old. My pediatrician couldn't figure out what was going on and I was getting sicker.

I remember the first time I heard the words cystic fibrosis. I was sitting in an examination room with both of my parents, and a doctor we had never met walked in holding a picture of my lungs, indicating the right one had collapsed. He told my parents, "This to me looks like cystic fibrosis." I later learned things must be pretty serious if the radiologist walks into your room. Within a few weeks, I was being tested at my local children's hospital, Le Bonheur, for *sis-tick fy-bro-sis*—wondering why I had two diapers wrapped around my now sweaty arms. It was my first sweat test in a hospital that would later become sacred to my healing and where I would undergo many more sweat tests. I was never admitted to the hospital throughout this process, a decision that I now think was probably a poor one. But I recovered well at home with oral antibiotics and the new medications and treatments to heal my lungs.

I remember the day we found out I have CF. I remember sitting in the car with my mom, asking her if I was a survivor, like in Destiny Child's new song, knowing with some granule of intuition that my life was on the cusp of a massive transition. I know my diagnosis came as a shock to my dad and was followed by a long period of grief and mourning. But my mom has a different story. My mom is an immigrant from Venezuela, the daughter of a former head nurse of the tuberculous unit in her city. My mother grew up surrounded by and immersed in the world of not only medicine, but pulmonology. Sputum cultures, x-rays, bronchiectasis, and lung inflammation were all familiar terms, reminiscent of a childhood 4,000 miles away. I think this background helped empower my mom to advocate for her daughter—a skill set she homed in on the first weeks after my diagnosis. Like I said, I was never admitted to the hospital when diagnosed, despite a collapsed lung and pneumonia. In fact, the pulmonologist I first saw made a poor impression on my mom when they remarked we were unlikely to be able to afford the Vest. From there, my parents found a new doctor to treat me.

My new doctor wasn't trained in pulmonology, rather in allergy and immunology. However, his father was one of the pioneering doctors in CF medicine. We had met him serendipitously and I believe it was because of him that I fared incredibly well for the next five years. But, as we all know, cystic fibrosis is unpredictable and shameless. I started having massive hemoptysis and soon surpassed the scope of his practice. It was terrifying not knowing what to do next—the memories from my diagnosis haunted my mom and me. However, my pediatrician (another Venezuelan mom) pushed for an admission at the children's hospital.

And so, I was back in the place where I was diagnosed, even sicker than when I first went with a collapsed lung and double lung pneumonia. This time, I was sitting at the edge of life and death multiple times a week. And, as luck would have it, the hospital had just hired a new chief of pulmonology—a well renowned man from Kentucky ("Dr. S.").

LUISA PALAZOLA

Without a doubt, Dr. S. saved my life. We quickly became friends, despite the fact that I was the sickest I'd ever been. This relationship has endured past my pediatric days. The three of us—Dr. S., my mom, and I—learned to advocate for my body. Between Dr. S. and my mom, I learned how to use my voice, to not fear asking questions, and to foster a love for anatomy and experimental medicine.

It's funny, I had always hated my children's hospital because of how I was treated at diagnosis. Life, however, has a funny way of working out. Now, Le Bonheur has become a place that is, to this day, sacred and critical to my healing.

Remember how I said that this hospital would become a place for many, many sweat tests? I still go there, at 27 years old, because it is the site for many drug trials. I think in the last two years, I have had over 15 sweat tests, this time without diapers!

I am amazed at how my story has taken different turns and twists. I think of myself as that skinny, sick nine-year-old child with a collapsed lung, wondering if I am a survivor. I also think of myself today: many sweat tests later, with curves and hips for days, and two powerfully functioning lungs. Who would have thought? I think of the hospital that diagnosed me and healed me, many times over the last 17 years. And, I think, yes, baby Luisa! You are a survivor and, more importantly, you have persevered with some of the kindest and most resilient folks at your side. ▲

Luisa is 27 years old and has CF. She lives in Memphis, Tennessee. Through her writing and rambling, she injects her perspective as a Hispanic woman and as a lover of life and stories in all her interactions. Luisa is an avid coffee lover and worldwide adventurer. In her downtime, she practices Portuguese and takes selfies with raccoons. Follow her on Instagram at @ladilla93 or email her at luisapalazola@gmail.com.

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adverse reactions, occurring in 3% or more of patients, include cough, hemoptysis, oropharyngeal pain, vomiting, bacteria sputum identified, pyrexia, and arthralgia. Before treatment initiation, however, patients are required to undergo a Bronchitol tolerance test. Here, healthcare providers give patients a first dose to evaluate if the treatment will cause bronchospasm (a sudden tightening in the walls of the airways) or a decrease in FEV1 and in oxygen blood levels. If any of these events occur, the treatment will not be prescribed.
https://tinyurl.com/y6dbkofv
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Translate Bio Resumes Enrollment And Dosing In Phase 1/2 Clinical Trial Of MRT5005 In Cystic Fibrosis

Translate Bio announced that enrollment and dosing in its Phase 1/2 clinical trial for MRT5005 in cystic fibrosis (CF) has resumed. In March 2020, the Company had announced a pause to enrollment and dosing in the clinical trial in response to the COVID-19 pandemic. MRT5005 is the first clinical-stage mRNA product candidate designed to address the underlying cause of CF by delivering mRNA encoding fully functional cystic fibrosis transmembrane conductance regulator (CFTR) protein to the lung epithelial cells through nebulization. MRT5005 is being developed to treat all patients with CF, regardless of the underlying genetic mutation, including those with limited or no CFTR protein. The FDA has granted MRT5005 Orphan Drug, Fast Track and Rare Pediatric Disease designation.
https://tinyurl.com/yxgk6a7t
AND
https://tinyurl.com/yy98s5kh

A. *Fumigatus* Lung Infections Linked To Age, Long-Term Antibiotic Use

Among fungal infections in CF, *A. fumigatus* is one of the most common. This fungus is known to cause allergic bronchopulmonary aspergillosis (ABPA) and acute lung infections. Co-infection of *A. fumigatus* with *P. aeruginosa* is also associated with more hospitalization and poorer lung function. Researchers analyzed data and found records of fungi in CF patients of all ages, with the infection rate of *A. fumigatus* rising until about age 49, after which it declined. Candida albicans and related species were the most common fungal species in patients' lungs *fumigatus* and related species were the next most common species. They found that patients with chronic *P. aeruginosa* infection had significantly poorer lung function, as measured by forced expiratory volume in one second (FEV1), than those without. Patients without chronic *P. aeruginosa* infection but with at least one positive *A. fumigatus* infection also had significantly lower FEV1, implying that *A. fumigatus* may directly impair lung function. Those with at least two *A. fumigatus*-positive cultures also experienced significantly more pulmonary exacerbations requiring antibiotic treatment. These results show a decrease in *A. fumigatus* prevalence in CF patients older than 50 years. This might reflect that getting older with CF presupposes a milder CF disease or a better lung health status, which prevents *A. fumigatus* colonisation. Compared to patients without *A. fumigatus*-positive cultures, those with at least two had significantly lower body mass indices and significantly

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As Normal As Possible

By Darryl Collins

I was diagnosed with cystic fibrosis in 1992 at 24 years old. While I was growing up, doctors had always treated me for bronchitis. I started having more severe respiratory symptoms when I was 19. In 1988, I had pleurisy and pneumonia. It wasn't until years later that my illness reached a dire state.

I had gotten to the point where I wanted to find out what I had so that I could be treated appropriately. My mom and dad took me to some good doctors to try and figure out what was wrong with me. I went to an allergy specialist and a lung specialist—both doctors told me to get out and live my life because they thought I was going to die young. This was a very frustrating and depressing time for me—I just kept getting sicker, until I found a doctor who really cared about me. This doctor had me do a sweat chloride test, which turned out to be negative. He sent my case before a group of doctors who determined that if I did have CF, it was only a mild case. I tried to check into Vanderbilt University Medical Center, but they didn't have an adult CF clinic at the time. When I went to Memphis, I saw a doctor who had been treating people with CF. They ordered genetic testing—CF showed up in the results. I'm glad I didn't learn that I had a terminal illness until I was an adult. I think it would have made it more difficult growing up if I had known that I had a serious disease. The only resources I had were just a few people with CF whom I had met while going back and forth to Memphis.

Eventually, in 1995, I went back to Vanderbilt where they had just started an adult CF clinic. I met some friends who, unlike me, had gotten diagnosed early in life. Vanderbilt had what they called "CF days." They had families get together twice a year. Dr. Preston Campbell was one of the doctors there; the whole team was awesome. He had invited the CEO of the CF foundation to attend. The foundation gave various educational sessions that were very helpful.

DARRYL COLLINS WITH HIS WIFE ALLI, AND SON HAYDEN, AT THE TRANSPLANT GAMES IN HOUSTON, TEXAS (2014).

I'm fortunate to have a mild case and to not have been as sick for as long as most CFers. I didn't let CF dictate who I was. I stayed above it by working my butt off. I could not go to bed wheezing. I would get up and do my Vest, hypertonic-saline, and flutter as much as it took. However, CF took a toll. Around January 2012, I became very sick and I was eventually listed for transplant. I was on the transplant list for 18 days, receiving a double lung transplant on May 30, 2012, at 44 years old.

I feel very grateful to have met my donor's family. He was 22 when he passed away. My donor was married with two beautiful girls. We were blessed to have spent Christmas with his family in 2018.

I have a different philosophy than what I've heard other people with CF say. For some, it makes them cherish life more. Personally, I hate CF! I would not choose to have it. I would hope that I could have a zest and care for life without it and still have a close relationship with God.

> "My advice for newly diagnosed patients is to keep regular appointments, commit yourself to staying on top of CF, and don't worry about your PFTs—it's just a number."

Making new friends has been the biggest resource for me. I enjoy attending the virtual conferences. I love talking to people who have CF and have had a double lung transplant. It seems like we have a deeper connection than before I got transplanted. My advice for newly diagnosed patients is to keep

regular appointments, commit yourself to staying on top of CF, and don't worry about your PFTs—it's just a number. Make as many CF friends as possible. Love, laugh, and be your own normal. Become a child of Jesus, putting your faith in Him; I wouldn't be here without Him. Be your own advocate. Keep the doctors in check, even if you have to be a pain in the butt! I had one doctor out of at least 20 who understood my fight for life. She happened to be the only CF doctor who visited me after my transplant.

CF can be lonely. It's hard to find someone to love you along with the demands of CF. I went through a bunch of heartaches before I finally met my wonderful wife—beautiful Alli, who is a nurse. God couldn't have put me with anyone better. We met in 2008 and got married on May 30, 2013, exactly one year after my transplant. Now we get to celebrate my new life and our new life together on the same anniversary. I also adopted her son Hayden, whom I am very close to and love with all my heart.

I hope my story helps give a different insight into how CF affects me and connects us to each other. Having CF has changed my life. I cherish life more after having had my transplant. When you're faced with death, it should change your perspective on life. It has given me more of an affinity for people who suffer and are sick. Life is precious. Live it one day at a time! ▲

Darryl Collins is 52 years old and has CF. He lives in McMinnville, Tennessee, with his wife, Alli, and son Hayden. Prior to his double lung transplant in 2012, he taught physical education. Currently, Darryl is an adjunct professor at MTSU and has coached girls' and boys' basketball for 17 years. He enjoys being active and has been an enthusiastic participant in the Transplant Games of America. You can reach him by email at collinsd1968@gmail.com.

My Experience As A Student With CF

By Daniel Gonzalez

For most of us with CF, it has become a part of our daily lives and responsibilities. We have to do our treatments, go to doctor appointments, and sometimes even be hospitalized. So, we have to learn to make our treatments a part of our daily routine. The life of a student who has CF is very different from that of the average student. However, we must learn to adapt and overcome to progress and continue our lives.

DANIEL GONZALEZ

Around my third year of high school, I started having major issues with my liver and, after about two months in the hospital, I was placed on the transplant list. I had some issues throughout my last two years of high school and missed some days here and there but got through without major issues. However, when I was applying for college, I realized I needed to choose my college carefully as it could mean needing to change doctors, having to find a new liver transplant center, and a new pulmonary doctor, along with all my other specialists. This could potentially mean complications in my care as my doctors wouldn't know my case as well as my current team.

So, I had a big decision to make when choosing my college while considering the added pressures on my health. For this reason, I applied to colleges within four hours of my transplant center (four hours being the time they give you to get to the hospital after you get the call that they have an organ). Toward the end of my senior year, I had more health issues and realized that I needed to stay right where I was currently living. For this reason, I applied to my local community college and took classes online. I felt sick at random times, and this allowed me the flexibility I needed for study and work.

The next year or two, I continued studying at a community college while taking care of my health. I did well but, quite frankly, I did not perform up to my standards. I tried putting more time in and worked harder, but it only helped a little. About this time, my transplant clinic called to let me know they had a liver ready for me. I arrived at the clinic and, about five to ten minutes later, I was being rolled into the operating room. After my transplant, I contacted the school and withdrew from the semester in the full expectation that I would be back the following semester to continue my classes.

Unfortunately, this was not the case. I had various complications post-transplant and was in and out of the hospital every few days. These stays sometimes lasted a day or two and other times a few months. In the end, I was in and out of the hospital for about two years, first for my liver and the issues that arose after my transplant, then later for issues subsequent to both my hospital stays and my resultant condition. My lungs and kidneys took a hit in part from all the medications I had been given. It took a while to regain my health and stay out of the hospital for more than a month or two.

After I was able to stay out of the hospital for a bit, I started thinking about how I would be able to continue my studies. It became clear that I needed a way to continue studying without having to leave every time I was admitted to the hospital. For this reason, I decided to continue studying in an online learning format. However, the community college I was in before my transplant required in-person testing. With my health being very volatile and requiring often lengthy hospital visits every few months, I needed to find a way to continue school remotely. That's when I found Penn State World Campus, which allowed me to work toward my degree through a fully online experience. I could do my work

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and take my tests online even when I was in the hospital.

For a while, I did OK and did not have many hospital admissions. However, I was eventually admitted and had to do my schoolwork while in the hospital. At first, it was difficult to find the time to do my work without being disturbed by the parade of doctors and other healthcare providers coming in and out my room. So, I started looking at the times during which they visited and managed to find the time to study. At first it was a bit difficult, but I got used to my schedule and soon I was able to do everything without much trouble. The lack of energy we all feel at times is even more likely to happen with chronic illness. It is important to push through without overdoing it. When I was sick, I often rested while doing my schoolwork.

> "It is important to push through without overdoing it. When I was sick, I often rested while doing my schoolwork."

Because I was able to step back and examine my responsibilities—health, studies, and daily life—I was able to make a schedule that worked for me. My enhanced organizational abilities have helped me to be very close now to successfully completing my studies. In the end, you could say that my health complications have had a positive impact on my studies because I learned to develop tools that will help me in the future. Indeed, I have kept up good grades no matter what difficult situations have come my way. In the end, my experience with health complications and school has helped. I have learned how to be better organized and adjust to difficult schedules that change often. It has been overwhelming at times, but I was also provided with tools that can help me in my future. I have learned that it's possible to continue working on my goals without sacrificing my health. Since being hospitalized, I have experienced different situations that have affected my studies, yet I have managed to maintain good grades and have been able to learn from the experience.

I think that it is important for students like me to know that while your CF can have an impact on your student life, there are ways to work around it and meet your needs. Each person's situation is different and will present different challenges. However, if you put your mind to it, you will be able to find a way to overcome any difficulty. Many of us may feel overwhelmed by our treatment needs, medications, and feeling sick. This may lead to us forget things in any of these areas, such as taking medicine. Having a good outlook about your needs is a good place to start so as not to neglect anything.

Students with CF have lots of pressures and responsibilities; however, we also have experience with facing difficult situations. It is a good idea to channel this experience in your schoolwork and daily lives. ▲

Daniel Gonzalez is 24 years old and has CF. He lives in Cedar Park, TX, and is currently working toward his Bachelor's in Business Administration through Penn State World Campus. He is a previous winner of the Higher Education Scholarship (f/k/a The Lauren Melissa Kelly Scholarship) offered through CF Roundtable. *Daniel is a volunteer for* CF Roundtable. *He enjoys spending time with friends, playing video games, and relaxing. His contact information is on page 2.*

more ABPA, CF-related diabetes, and arthritis or other joint disorders.
https://tinyurl.com/y2ncttcd

FDA Clears AP-PA02 Phage Therapy Trial For CF *P. Aeruginosa* Infections

The U.S. Food and Drug Administration (FDA) cleared Armata Pharmaceuticals' investigational new drug (IND) application for a Phase 1b/2a clinical trial of AP-PA02 for the treatment of the *Pseudomonas aeruginosa* bacterial infections that are a hallmark of cystic fibrosis (CF). AP-PAo2, conversely, utilizes a cocktail of viruses called bacteriophages to attack the infecting bacteria. Because phages are specific to bacteria — they cannot infect the cells of the patients' organs — they cause fewer side effects than antibiotics and can target even antibiotic-resistant microbes. AP-PA02 delivers phages specific to *P. aeruginosa* directly to the lungs via inhalation.

The new trial — called SWARM-P.a. — is a multi-center, double-blind, randomized, placebo-controlled, single ascending dose (SAD) and multiple ascending dose trial that will assess the safety and tolerability of AP-PA02 in CF patients with chronic pulmonary *P. aeruginosa* infections. Results from this study - SWARM-P.a. - to reflect the manner in which phage attack dangerous pathogens, will be the first clinical trial to evaluate a phage-based therapy as a potential treatment for *Pseudomonas aeruginosa* airway infections. This clinical trial will contribute to the evaluation of the potential of phage to combat multi-drug resistant infections, and potentially usher in a new era in the fight to develop alternatives to antibiotics. While the study will initially evaluate AP-PA02 in combination with standard antibiotics, the ultimate goal with this product candidate is to replace antibiotics as a front-line therapy. AP-PA02 is a second-generation phage product candidate for *P. aerugi-*

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IN THE SPOTLIGHT

With Grace Knight

By Jeanie Hanley and Andrea Eisenman

Grace recently joined the *CF Roundtable* board with plans to interview other adults with CF, to learn their (your) stories, and to share them through our blogs. It made perfect sense to interview her first! Throughout her young life (she's 22 years old), Grace has demonstrated such perseverance and determination, especially during her college years when she battled severe infections and yet kept up with her courses. Be prepared to be energized by her strength and conviction. As a bonus, read about her somewhat humorous experience during one of her hospitalizations and how it ended up being one of her higher moments. She's now enrolled in law school to enhance her advocacy for others. Please welcome Grace Knight, our newest star. Spotlight please!

GRACE KNIGHT

Where do you live?

I am from Tyler, TX, but am attending law school at the University of Texas in Austin where I will be living for three years.

When were you diagnosed?

I am homozygous for 508Fdel and was diagnosed when I was a year and a half. When I was born, I was a normal size, but over the next few months I didn't gain any more weight. My mom had a baby book for parents and in it she read about CF. She was convinced that I had it, but the doctors refused to test me. She was told by many people that I was fine, and she needed to calm down, but she kept pushing. Eventually, I was tested and my parents found out that I had CF.

What was your major in college?

My major in college was English with a concentration in creative writing. I always knew I wanted to major in English, but I added the creative writing later. During my freshman year I took a creative writing class with a professor who inspired me. I ended up having to go into the hospital for six weeks and he worked around the syllabus to allow me to still participate in the class from my hospital room. I was incredibly challenged by the class and decided I wanted to focus on that throughout my college career.

What challenges have you had during college?

In my freshman year of college, I was infected with *Mycobacteria abscessus*. This bacterial infection was terrifying and it took us a very long time to figure out what was causing the dramatic downfall in my health. When I started college, my lung function was 80% and, by January, it had dropped to 39%. I was exhausted by the time I was hospitalized in February. I had just transitioned to adult care, so my doctor was fairly new. He spent a lot of time blaming me for my own illness, even though there was no way I could have prevented this bacterial infection. I am very diligent with all of my medications, and there were many times when I would contact my care team with concerns about my health and would never get a response. I was in the hospital in Philadelphia for two weeks with pneumonia. When they finally discharged me, I had recovered a little bit of my lung function, but I was still really sick. When I had a follow-up appointment the next week, my doctor told me there was nothing else he was going to do to help me. He gave up. I was incredibly angry and frustrated because I was willing to do anything to get better and he just walked away. At that point, my mom and I decided that it was best for me to go to another doctor, one that another CF patient had recommended. This new doctor was in Denver and the next week we flew out there. I was still going to school during this time, and I was able to complete my classes online through Skype while I was in the hospital. Once I got to Denver, the new doctor knew right away that I had mycobacteria from the CT scan. The culture grew straight off the bacterial slide! He said it was one of the most aggressive forms of M. *abscessus* he had ever seen. The treatment for this bacteria would be four weeks in the hospital, three months of IV antibiotics after that, followed by nine months of oral antibiotics. It was an intense cocktail, and, when August eventually came, I ended up taking a medical leave from school because I was still so weak. I eventually returned to Penn a semester later, but my lung function would not recover until two years later after starting Trikafta. This entire experience shaped most of my college years. I spent a lot of time catch-

ing up from the semester I had lost. I joined clubs as a junior, when everyone else joined as freshmen. I went to summer school the following summers, when everyone else did internships. I was scared, angry, and frustrated by the limitations my disease held over me, and the worries I carried around were very different from those of my classmates. This is not to say that I didn't have a good time in college, because I definitely did. This is to say that going to college with CF is incredibly tough, and that graduating from college with CF is a feat worthy of being celebrated.

How did the college clinic experience affect you?

School has always been my happy place, so, when I had to leave it because of my health, I really struggled. Because I came so close to not surviving, I was convinced that the next cold I got would take me out completely. I was really scared and it took me a long time to heal from that trauma. I improved my outlook by staying busy with things I was interested in and loved doing. This reminded me of all the good things in life. About a year after M. *abscessus* treatment, my friends and I ran a half-marathon. Completing that race gave me a lot of control over my disease. I didn't feel so hopeless anymore and I ran another one a few months later. Pushing my body to the extreme reminded me that even if CF took me down a little, it certainly would not win.

How have CFTR modulators worked for you?

Because of my mutation, I have been on Orkambi, Symdeko, and Trikafta. I even did the clinical trial for Orkambi and I gained so much weight that I am certain I had the drug. However, after being on Orkambi for a while, I started experiencing chest tightness and major hair loss. I ended up stopping the medication because there were more side effects than benefits. Once Symdeko came out I started on that one, too. I didn't see much improvement, but it's possible I got over colds a little quicker. I didn't have any side effects so I just stayed on it until Trikafta came out and then I switched. I have been on Trikafta since November, 2019, and it has been a dream. I feel incredible and it is almost like I don't even have CF anymore. At my doctor's recommendation I still do all my breathing treatments, etc., but my quality of life has greatly improved. I have gained about five to seven pounds, rarely cough, and am able to breathe so much easier. It has changed my life.

What are your current health issues?

I still have digestive issues and am taking the same amount of enzymes as I did before Trikafta. I tried taking less enzymes once and it was not good (you know what I mean). However, I don't have as much discomfort in my stomach and don't have to eat as much throughout the day. About two years before starting Trikafta, I had my fifth sinus surgery, so my sinuses were still doing well when I started the drug and have continued to stay clear.

How does your joining *CF Roundtable* as a director fit into your passion for writing?

I can't remember the first time I encountered *CF Roundtable*, but it has always been a huge source of information and support for me. Growing up, I only knew a few people with CF so being able to read an entire newsletter from CF patients was incredibly valuable. *CF Roundtable* has also helped me financially as I am a past recipient of the Lauren Melissa Kelly scholarship. I studied English and creative writing in college, so becoming a *CF Roundtable* director aligns perfectly with my passion for writing.

What pearls can you share regarding the demands of CF and classes?

In college I found that knowing my priorities was really helpful. My health always came first because without that you can't do much of anything. After that came my classes and friends. When I was sick, I made a lot of sacrifices with regards to my social life and I made it a point to have strong relationships with my professors so they would be accommodating to my needs. I have found that if professors know your name and know you truly care about the material, they will be much more open to helping you if you have to go into the hospital, etc. For me this meant going to office hours as often as I could, being present in every class unless I was sick, and being incredibly open with them about my disease. When I did eventually get sick, I always went to them with a plan so all they had to do was say yes or no, instead of asking them to think of a solution. I always loved school and going to class so, even when I was sick, I found my joy in academics.

What inspired you to go into law?

Not to be cliché, but it was definitely my experience with CF. Advocacy is an important part of the patient experience. You have to stand up for your own health, and, if you can't, then you need someone who can do it for you. When I was sick my freshman year, my mom stood up for me when the doctors didn't listen. Being a lawyer means being a professional advocate. It is your job to have your client's best interest in mind and get them the best outcome. After years of advocating for my own life, I decided I wanted to become a lawyer to help others advocate for theirs.

How do your friends deal with your having CF?

My friends are incredibly understanding of my disease. They came to visit me when I was in the hospital during freshman and junior year and they

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LIVE OUT LOUD

The Journey To Contentment: What To Do When Life Doesn't Go As Planned

By Lara Govendo

According to societal standards, I should be married by 25, complete with two kids, a white picket fence, and a successful career. As if this were the picturesque life to strive for and anything outside of that wasn't considered "good." However, my life with CF has been anything but predictable—or plannable, for that matter. There has never been a real sense of certainty. I've learned that fighting *what is* robs us of joy and the peace we seek. Accepting the life we've been given and finding the good in it is the best that we can and should do.

Society has sold the lie that we are in control of our lives. Consequently, if our lives don't go as planned, we freak out. We are used to order, schedules, and routines. When that doesn't happen, we are unable to deviate and adapt. This, in turn, leads to increased emotional outbursts, irrational thought processes, and complete internal shutdowns.

On the flipside, those of us with CF have learned that we can't truly envisage what may happen next. We are seasoned professionals in the world of unpredictability. We hit every curveball out of the park. It's what we do. We can roll with the punches. We're adaptable. And we're better for it.

We don't have the luxury of being set in our schedules, unable to deviate from the norm. If we're not flexible, we spend our whole lives wishing for something that is out of reach: perfection. In reality, though, perfection is an unattainable goal—for everyone. We hold the illusion of control with a white-knuckled grip for fear of what will happen if we're not in charge. Tuning into what is *underneath* the pain of not knowing what will happen next is necessary for growth and healing.

The stages between sitting in the unknown and acceptance of the present is where the tension resides. We all go through different seasons of life that bring us through various struggles or celebrations. This ebb and flow between the seasons is a constant throughout our entire lives. We never have that moment of arrival where everything is perfectly in alignment. We wish away seasons, that turn into years, that turn into the course of our lives. Presence in each season is pertinent. We can't wait until *(fill in the blank)* happens before we start living. Life is being lived right now. We either partake or remain in that place of longing for something different.

> *"Tuning into what is* underneath *the pain of not knowing what will happen next is necessary for growth and healing."*

That's not to say it's not OK to dream, have goals, and tentatively plan the future. The difference is being content in the season that we are in no matter the circumstances. It's not easy. It hurts. It's hard. And our hearts long for things to be different. But we have to find ways to be here, right now, in this space wholeheartedly.

LARA GOVENDO

No, I didn't get the life I had envisioned so long ago. And I'm so thankful to God that I didn't. I would have never chosen to have my chest sawed in half, say goodbye to my lungs, and hello to new ones; but, oh, how I wouldn't change one thing if given the choice. The greatest gift is contentment. The ability to find ways to cope and be OK with however things turn out brings peace.

I've had to come to terms with death more times than I care to count throughout my journey in life. I'm sure that you have, too. I've had to grieve the life I had envisioned and heal those deep wounds. I've learned that being

bitter about circumstances doesn't change them; rather, it makes them so much harder to muscle through. Adapting the mindset of becoming better from what we're going through has afforded me the opportunity to learn from any season that I'm in and to be grateful that I am *still* here to experience it.

We have to learn how to accept what *is* and be all right with it, somehow, someway. Yes, we grieve the life we thought we would have. We have to feel those feelings and sit with the magnitude of letting go of what we had pictured for our lives. Next, we need to heal that part of us. It's important to give attention to our process, whatever that looks like for each of us. Then, we can step into wholehearted living. Loving the life we have been granted is essential for cultivating a good life.

Together we can provide the path that leads to contentment. We all have the ability within us. If we give ourselves the tools, support, and encouragement, we can allow ourselves to live differently; moreover, we can encourage others to do the same. Let us become people who embrace the life we've been granted. In doing this, we will pave the way for others in their journey toward true joy in the face of any circumstance. ▲

Lara Govendo is 34 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 contact information is on page 2.

KNIGHT continued from page 43

are always encouraging me to keep fighting. When I can't go out with them because I am sick, they completely understand and have never made me feel different because of my disease.

Favorite things to do?

I enjoy playing the violin, writing, running, going to museums, and listening to music.

Do you have an amusing CF story?

Last summer when I was in the hospital, I was on IV colistin among other drugs and it made me feel terrible. I didn't want to eat and was struggling to keep my energy up. The nurse asked me if I wanted some medicine to help me eat and I said, yes, please. He didn't tell me what it was, just that I could take it before each meal. I started taking it and I think it helped me eat, but then I would be so tired and lazy afterward. The next day, about an hour or so after taking the medicine, I started feeling really weird and started hallucinating. My heart rate skyrocketed and I felt super anxious and paranoid. The doctor rushed in and was like WHAT IS HAPPENING? We had to do an EKG and then I had to wear a heart monitor for 24 hours to make sure I was OK. Turns out, the medicine they gave me to help me eat was actually just straight THC, and I'm pretty sure I was tripping from it. I had never done marijuana before so the whole thing was traumatizing but thinking about how I was and the whole hospital freaking out really makes me laugh.

Who or what inspires you?

My family—their patience, strength, and support—are so inspiring in the face of such a debilitating disease. It is because of them that I am still alive today. ▲

Jeanie Hanley is 58 and has CF. She is a Director and the Past President of USACFA. Andrea Eisenman is 56 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.*
If you would like to be interviewed for "In The Spotlight," please contact either Andrea or Jeanie.

CF: THE MIND GAME

What My Yorkie Has Taught Me About CF And Family

By Mark Tremblay

I've spent more hours than I'm willing to admit in front of my laptop thinking about ways to discuss CF family dynamics while my 19-year-old, half-blind, and deaf Yorkie snores next to me. To be honest, I reckon my relationship with Zappy, in many ways, mirrors the relationship between me and my family.

Before I get into that, however, let me tell you a little about Zappy, formally known as Zaphod Beeblebrox, otherwise known as, the President of Life, the Universe, and Everything in *The Hitchhiker's Guide to the Galaxy* (by Douglas Adams). I thought he'd live up to the name by rising above his circumstances. Fortune never did shine on that ole boy from birth. He spent his formative years in an abandoned trailer in Apalachin, NY, with a cruel owner who ran an illegal puppy mill and cooked meth to make ends meet. It's worth noting that the town he and I both grew up in gained national infamy for three things: it was the site of the largest mafia bust in history (November 15, 1957); it is located in a region known for being the meth capital of the country; and is the last stop on the heroin highway (on the southern bank of the Susquehanna River). With such notoriety, it may come as no surprise that Apalachin is located in the poorest, most rural, county with the highest poverty rate in New York State.

In the midst of my divorce and bankruptcy in my late 30s, I was trying unsuccessfully to adopt a friendly shelter dog when I came across an article in the *Sun Bulletin* about some very sick Yorkies at a Tioga County shelter in need of emergency adoption. My heart broke when I read that Zappy (f/k/a Reilly at the shelter) and three other dogs were abandoned when their owner was arrested and failed to tell the authorities that she was living in an abandoned trailer with 24 Yorkies in southern Apalachin. By the time a dirt biker came across the trailer in an overgrown field two weeks later, only four of them remained.

In order to make it, Zappy developed incredible survival skills—breaking into cabinets, escaping his holding pen, leaping onto the counter to drink from the clogged kitchen sink, as well as cannibalizing his fellow captives, all of which made him a terrible dog by most standards. In fact, he was so traumatized by the experience that it took weeks before he'd get out from behind my couch, and it took months before he'd let me pick him up without trying to bite me. But he survived and, 15 years later, he'll occasionally lean against my leg while he naps on the couch. However, the trauma left some terrible scars: the constant desire to be alone in the dark, the urge to fight any dog in sight, consistent barking at any and everything, excessive thirst, regular vomiting, and pooping/peeing everywhere.

> *"Fortunately, when I was diagnosed, my family fought hard and initially was very CF-focused."*

MARK TREMBLAY BRUSHING ZAPPY AFTER HIS BATH.

Unlike my family, who had no idea what they were getting into before I was diagnosed with CF, I knew exactly what I was getting into with Zappy and chose him, anyway. As hard as it is to hear, especially for CF patients, a recent study found that 90% of children who tested positive, in utero, for CF were aborted—families would, given the choice, choose not to have children with CF if they knew what they were getting into ahead of time.

Nevertheless, once a CF sweat test comes back positive, families generally respond to the devastating news either by becoming CF-focused such that CF becomes the *raison d'être* for a family's existence or by hunkering down to try to survive the disease while doing as many normal family things as possible amidst juggling CF emergencies and treatment.

Like Zappy, I was, by most standards, a terrible human (infant) from the moment my mom brought me home. Like most infants, I refused to take naps or sleep at night; I was covered in my own diarrhea most of the time, which I liked to smear all over everything; I cried all the time; I had a peculiar habit of climbing out of my crib and balancing on the handrail; and, to top all that off, I was sick—I required hours of chest PT, mist tent cleaning, and room cleaning every day. Most likely, upon my diagnosis, my mom had to grieve the healthy baby she had wished for. Fortunately, when I was diagnosed, my family fought hard and initially was very CF-focused. I was featured in CF promotions, spoke at CF fundraisers, and made appearances on local TV and/or radio shows to raise awareness about the then little-known disease that was the number one genetic killer in the U.S. However, like many families, they eventually burned out, perhaps when they realized all their efforts wouldn't add a day to my life or they couldn't afford to keep neglecting their other children, so they gradually shifted to survival mode. It's important to note that many families, in fact, don't survive intact. Parental divorce rates are 20% higher in CF families, and CF patient's siblings often cut themselves off from their family as they grow older due to jealousy for parental attention; feeling neglected, especially in childhood; or lack of attention, particularly during critical milestones or personal achievements (e.g., birthdays, sporting events, and other extracurricular activities).

In my case, as I got older, my drinking escalated, run ins with the law increased, my dad got sick with cancer and eventually died, and my brothers went off to college and had their own families. Thus, CF became even less of a priority for my family. To illustrate, in the last 20 years, I've had 32 hospitalizations. My mom visited just once, while on her way to my brother's house.

Similarly, when I first met Zappy, I tried everything I could to correct his learned behaviors but after too many failed attempts, I shifted into more of a Zappy-survival mode: the constant battle to overlook his overwhelmingly annoying qualities to meet my responsibilities as his owner until he passes. Also, I'm somewhat ashamed to admit that since I picked up my Siberian Husky puppy, Patton, four years ago, I've become way more attached to him, rather than to Zappy, because he loves to play and always wants to snuggle.

Having these two dogs has given me the perspective to better understand, empathize, and accept my own family dynamics living in a health-centric world with healthy family members. Thus, I'm not surprised that, when my family does get together, certain family members try to hide impending colds because they're loath to change their plans, particularly if it means disappointing their children or preventing them from seeing grandma. On the flip side, particularly in the eyes of older family members, I have to be okay with mom preferring to see her grandchildren rather than her son, just as I would rather wrestle with Patton than blot Zappy's pee.

Normally, I have a rule against giving advice. As the leader of the CF Fighters for Recovery and Freedom group, I'm often asked for advice and demur because I strive to empower group members to find their own solutions and follow their own paths. However, I will make this rare exception on how to deal with family-of-origin issues:

Do not "should" on yourself to go home or visit your family. Only you can determine if you are socially distancing out of self-preservation (e.g., fear of COVID-19 or flu) or socially isolating out of your own dysfunction.

If you have to, keep your visits home short to mitigate risk of exposure and/or stress that may result from being around certain family members. It's better to make a brief appearance and leave on a positive note than wait until things have soured and leave feeling down.

Similarly, if home is far away and you have to stay a couple of days, carve out plenty of time away from the family dynamics to relax, breathe, and destress.

Many CF patients benefit from individual or group counseling to process family and life issues, particularly in the winter months when frigid weather, fear of illness, and family issues abound.

Above all else, if it's emotional wellness that you desire, your objective lies in finding peace and acceptance with your family of origin, whether through direct communication or emotional processing in the form of counseling or self-help. This is true no matter whether your role in the family requires a lot of patience (like Zappy), or very little patience (like Patton), or whether your family is CF-centric or in CF survival mode. Remember, it's never too late to begin emotional healing, so there's nothing preventing you from starting or restarting your journey today. Good luck, Godspeed, and *bon voyage*! ▲

Mark is 51 years old and lives in Albany, NY, with his wife, MaryGrace, and stepson. He holds a Master of Arts in Psychology from Marywood University and a Master of Public Administration from Syracuse University. Mark has worked for six years in the New York Governor's Division of Budget and currently works full time at the Department of Health. He is the President of CF Vests 4 Life Foundation. He and his wife love cycling, church ministry, and riding their motorcycles. You can follow them on their YouTube channel, "Breathing Grace,"and/or join him at the CF Fighters for Recovery and Freedom group (Wednesday nights at 7:00 p.m. ET).

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, 9450 SW Gemini Drive, PMB43881, Beaverton, OR 97008-7105. Or email to: cfroundtable@usacfa.org**

ANNIVERSARIES

Birthday

Sara Kominsky
Riner, Virginia
49 on September 18, 2020

Sydna Marshall
Austin, Texas
40 on September 19, 2020

Andrea Eisenman
New York, New York
56 on November 28, 2020

Wedding

Sara & Dan Kominsky
Riner, Virginia
24 years on September 21, 2020

Sydna Marshall & Adam Keys
Austin, Texas
7 years on October 26, 2013

Transplant

Sara Kominsky, 49
Riner, Virginia
Bilateral lung transplant
3 years on October 12, 2020
(re-transplant on March 12, 2020)

Julie B. Ice, 49
Crestwood, Kentucky
Bilateral lung transplant
9 years on December 18, 2020

Joanne Schum, 56
Webster, New York
Bilateral lung transplant
23 years on September 12, 2020

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nosa that follows AP-PA01, the first phage product developed by Armata. AP-PA01 was shown to successfully treat a CF patient with resistant *P. aeruginosa* infection when used in combination with antibiotics. According to Armata, AP-PA02 offers significantly improved therapeutic features compared with AP-PA01.
https://tinyurl.com/y5s7tsf8
AND
https://tinyurl.com/y5s7tsf8

BiomX Testing Potential Therapy For CF Lung Infections

BiomX has unveiled a platform designed for more rapid and efficient development of phage therapy, which the company is using to test potential treatments, including one for *Pseudomonas aeruginosa* infections in people with cystic fibrosis (CF). BiomX focuses on bacteria-targeting therapies that use phages — viruses that can infect and kill bacteria cells. The company's BOLT (BacteriOphage Lead to Treatment) platform is designed to allow for rapid development of phages that can target particular bacteria and move quickly into human studies. According to BiomX, the upcoming CF program aims to test the investigational therapy in 20 CF patients. Each individual will be treated for 10 days, and researchers will assess the treatment's safety and efficacy in terms of bacterial counts, lung function, and quality of life.
https://tinyurl.com/yy462rb5

Patients With Cystic Fibrosis And Advanced Lung Disease Benefit From Lumacaftor/Ivacaftor Treatment

Several studies have assessed safety and efficacy outcomes for lumacaftor/ivacaftor therapy. This study reports on lumacaftor/ivacaftor's impact on lung function, physical performance, and health-related quality of life (HRQOL) in a subpopulation of people with Cystic Fibrosis with advanced pulmonary disease who would not fulfill inclusion criteria for these other studies. This follow-up review examined lumacaftor/ivacaftor's effect in a highly selected CF population. Inclusion criteria included low percent predicted forced expiratory volume in one second (ppFEV1), fast deteriorating ppFEV1, low body mass index (BMI), and difficult-to-treat infections. Primary endpoints included change in ppFEV1 slope, cardiopulmonary exercise testing (CPET), and all domains of the Cystic Fibrosis Questionnaire-Revised (CFQ-R). Secondary outcomes included change in ppFEV1, BMI Z-score, and sweat chloride concentration. The findings suggest that lumacaftor/ivacaftor reduces lung function decline, improves lung function, physical performance, and HRQOL to a greater extent in PWCF with severe lung disease than previously

Overcoming The Odds

By Ashley Wilson

"What do you want to be when you grow up?" With twinkling eyes, little Ashley would answer, "I want to be on Broadway." Looking back, I get a chuckle at how big I could dream. Many things have changed over the years, and I have grown up a bit since that time, but the one thing that hasn't changed is how big I dream. There have and will continue to be different obstacles to face as I continue to work toward my dream.

When I started my first semester of college in 2016, I was told by a doctor that 85% of cystic fibrosis college students will decline in health, drop out of college, or even take more than four years to graduate. I remember this doctor distinctly asking me, "Will you be the 85% that fails or the 15% that succeeds?" I looked him in the eye and said, "15%, of course." I wanted to prove to that doctor and to others that I can be successful despite living with cystic fibrosis. After finishing two years of community college, I transferred to my dream university. I was excited for all that the school would have to offer and was eager to begin preparing for my career. Unfortunately, due to the air quality, my lung function went

ASHLEY WILSON

downhill, and I was only able to stay at that university for one year. I felt like I had let everyone down—I had become that 85%. Since that time, I wish I could have given my younger self this advice: "No, you did not meet that 15% standard of that past doctor, but you have succeeded in your own 15%. Being a college student with cystic fibrosis is a *huge* accomplishment. It does not matter if you finish school in four years or seven. It is okay to go at your own pace—15% is continuing to go for your dream through all the obstacles you have had to face. You should be so proud of yourself!"

I am currently finishing my general education at Monterey Peninsula College and will be transferring to California State University of Monterey Bay in spring 2021. I will be studying to obtain my Bachelor's Degree in Cinematic Arts. I would like to have the opportunity to work for film companies that inspire and make a difference in the community. I would also like to create a company in the future assisting businesses with the resources to grow through marketing and media. I am incredibly thankful to have received the Higher Education Scholarship (f/k/a The Lauren Melissa Kelly Scholarship), offered through CF *Roundtable*, as it will assist in allowing me to continue my education as a cinematic arts major. ▲

Ashley Wilson is 23 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 and her podcast at Blooming Roses.

recognized.
https://tinyurl.com/y3538tsc

Alternative And Experimental Therapies Of *Mycobacterium Abscessus* Infections

Mycobacterium abscessus is a nontuberculous mycobacterium notoriously known for causing severe, chronic infections. Treatment of these infections is challenging due to either intrinsic or acquired resistance of M. *abscessus* to multiple antibiotics. Despite prolonged poly-antimicrobial therapy, treatment of M. *abscessus* infections often fails, leading to progressive morbidity and eventual mortality. Clofazimine and rifabutin are known anti-mycobacterial antibiotics, repurposed for use against M. abscessus. Novel antimicrobials active against M. *abscessus* include delamanid, pretomanid and PIPD1 and the recently approved beta-lactamase inhibitors avibactam, relebactam and vaborbactam. Previously unused antimicrobial combinations, e.g. vancomycin–clarithromycin and dual beta-lactam therapy, have been shown to have synergistic effect against M. *abscessus* in experimental models, suggesting their possible use in multiple-drug regimens. Finally, engineered phage therapy has been reported

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Meet A New Director—Daniel Gonzalez

Hi! My name is Daniel Gonzalez. I am 24 years old and was diagnosed with CF when I was two years old, before I can even remember. I was born in Saltillo, Coahuila, in Mexico and moved to Austin, Texas, when I was about eight years old to receive better care for my CF. I currently have my Associate's Degree in Business Administration from Penn State and am currently working toward my bachelor's degree.

Over the past few years, I have learned more about the difficulties that come with CF and how varied these can be. It has given me a new perspective into how CF is different for every single person. Hearing about how each person deals with their own challenges has made me realize that we all share some similar experiences despite our differences. This means that we can learn from each other's experiences.

The realization that we can learn from one another led me to look for new ways to learn about others' experiences and the different organizations that help those with CF. This became my reason for choosing to work with USACFA for a school project. As I discovered more about the organization and its purpose, I realized that I wanted to get more involved. I applied for and was awarded the LMK scholarship in the fall of 2020. After volunteering with treasurer duties, I decided to officially join the board.

DANIEL GONZALEZ

I enjoy spending time with my friends. We usually pass the time by playing billiards (although I am not very good) and video games. I like to play all types of video games that allow me to experience different adventures and stories. I also enjoy watching a wide variety of TV shows and listening to music. ▲

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to be clinically successful in a severe case of disseminated *M. abscessus* infection. While many of these experimental therapeutics have shown activity against *M. abscessus* in vitro, as well as in intracellular and/or animal models, most have little if any evidence of effect in human infections. Clinical studies of *M. abscesssus* treatments are needed to reliably determine the value of their incorporation in therapeutic regimens.
https://tinyurl.com/y52d3yvy
AND
https://tinyurl.com/yydbmxgp

Polyphor Receives Award Of Up To USD 3.3 Million From Cystic Fibrosis Foundation To Support Clinical Development Of Inhaled Antibiotic Murepavadin

Polyphor AG announced a funding agreement with the Cystic Fibrosis Foundation to advance clinical development of its novel class antibiotic, inhaled murepavadin, in cystic fibrosis (CF). Inhaled murepavadin is a highly potent and selective antibiotic against *Pseudomonas aeruginosa,* including multidrug resistant strains. The award will fund a Phase Ib/IIa clinical trial of inhaled murepavadin. The Phase Ib/IIa trial in adults with CF, assessing safety and tolerability (both overall and local) of ascending doses of inhaled murepavadin, is planned to be initiated in Q4 2021. Polyphor's inhaled murepavadin is currently being developed as a precision antibiotic specifically targeting chronic *Pseudomonas aeruginosa.* An inhaled antibiotic could make it easier for someone with a *Pseudomonas* infection to take the drug from home. In addition, the drug, which targets the outer membrane of bacteria, specifically focuses on *Pseudomonas,* which might have certain advantages over broad-spectrum antibiotics. It is the first member of the Outer Membrane Protein Targeting Antibiotics (OMPTA), a novel class of antibiotics. If approved for commercial use, inhaled murepavadin would be the first new class of antibiotics for Gram-negative pathogens in the last 50 years. It would also be potentially the first agent to target specifically *Pseudomonas aeruginosa* bacteria.
https://tinyurl.com/y52sfjsn
AND
https://tinyurl.com/y4x7nuj5
AND
https://tinyurl.com/y6sbjo2u
AND
https://tinyurl.com/y3caprgr

Matinas Biopharma Awarded Up To $3.75 Million From The Cystic Fibrosis

Meet A New Director—Terry G. Wright

Terry G. Wright is the president and cofounder of the National Organization of African Americans with Cystic Fibrosis ("NOAACF"), a 501(c)(3) organization with a mission to connect individuals with CF, help build diverse communities, and, through its national platform, raise CF awareness within the African American community and beyond.

At the age of 54, Terry was unexpectedly diagnosed with cystic fibrosis. He has also experienced extensive bacterial infections and recurrent fungal infections including sinusitis, bronchitis, pneumonia, aspergillus, and *Burkholderia multivorans*. Terry has also endured chronic pancreatitis as a side effect of his CF diagnosis.

Terry is the recipient of a 2020 Impact Grant from the Cystic Fibrosis Foundation as well as being the first person from Arkansas to receive this honor. He is also a recipient of the 2020 Jacoby Angel Award, the highest award presented by the U.S. Adult Cystic Fibrosis Foundation ("USACFA"), publishers of *CF Roundtable*.

Terry likes to plant seeds of life, love, and hope to help bring change for the better for cystic fibrosis patients, their families, and caretakers. He is an Arkansas dual-certified Master Gardener and Master Naturalist. He was elected the 2016 Pulaski County Master Gardner of the Year award winner and was a finalist for the 2017 Arkansas Master Gardener of the Year. He completed the standard curriculum in Permaculture Design in 2018. He has also been honored to formerly serve on the North Little Rock Green Committee and Commission on Environmental Efficiency. He has an impressive 38-plus-year career as a certified personal fitness trainer.

TERRY G. WRIGHT

Today, he wholeheartedly utilizes his deep-rooted passion for gardening, nature, agriculture, horticulture, fitness, nutrition, and health to help individuals from all walks of life to achieve the best in health! As a consequence of his own life-altering health challenges, coupled with the numerous challenging health issues he has witnessed firsthand in the lives of others, Terry is fully committed to utilizing his expertise in gardening, agriculture, horticulture, nutrition, and fitness to help others combat medical issues and achieve a better quality of health and life. Terry lives in North Little Rock, Arkansas, with his wife and Butterbean, Dr. Michele R. Wright. ▲

Foundation To Support Development Of Oral Amikacin (MAT2501) For The Treatment Of NTM Infections In Cystic Fibrosis Patients

Matinas BioPharma Holdings, Inc. announced that it has been awarded up to $3.75 million from the Cystic Fibrosis Foundation (CFF). The award will support preclinical development of MAT2501, Matinas' lipid nano-crystal (LNC) oral formulation of the broad-spectrum aminoglycoside amikacin, toward an indication to treat nontuberculous mycobacterial (NTM) lung disease. The CFF award will allow Matinas to rapidly advance the development of MAT2501 and will support preclinical in vitro and in vivo studies, along with several of the toxicology studies necessary to progress MAT2501 into Phase 2. MAT2501 is an oral version of the broad spectrum aminoglycoside antibiotic amikacin (encochleated amikacin) used to treat bacterial infections, including NTM and various drug-resistant, gram-negative bacteria. MAT2501 utilizes the Company's proprietary LNC platform to achieve oral bioavailability, limit toxicity and enable targeted delivery to sites of infection. MAT2501 is designed to deliver high levels amikacin directly to the lungs, based on a lipid nano-crystal delivery technology that allows for a more targeted treatment, while improving the antibiotics' safety and tolerability. Currently, amikacin can only be delivered by injection or through inhalation. IV and inhaled amikacin, however, are associated with major side effects including toxicity to the kidneys and ototoxicity or permanent hearing loss with long-term use. Matinas believes that MAT2501's ability to orally deliver high levels of amikacin directly to the lung and without use-limiting toxicity, distinguishes it from all available therapies and could provide

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Announcing Our New Scholarship For The Arts

USACFA is proud to announce their newest scholarship, the Scholarship for the Arts, established by Andrea Eisenman to honor her mother, Helen Eisenman. Helen valued education and had a great appreciation for the arts; she found immense joy in music, opera, photography, and fine arts. She would be delighted knowing this scholarship would benefit other adults in the CF community interested in pursuing a degree in the arts.

This scholarship will award two deserving students $5,000 each toward their tuition in their respective field of the arts: fine arts, computer graphics, design, music, choral, photography, filmmaking, creative writing, and poetry, to name a few. It is open to anyone seeking a degree, from an associate to a doctoral degree in the creative arts.

Helen was a single mother devoted to her daughter, Andrea, who has cystic fibrosis. She made many sacrifices in order to help Andrea live a long and healthy life. Helen also fit in her passion for the arts. She was a talented photographer, writer, and editor (she used to proofread *CF Roundtable*, too).

Helen was born in 1928, near Vienna, Austria, and was a Holocaust survivor. She and her parents were admirers of the arts in Vienna and later in New York City, where they resettled after 1940. Helen was proficient in several languages—German, French, Portuguese, and English—which would later serve her well in her career writing subtitles for foreign films. She majored in English at Queens College and later landed her first job writing for radio. Helen later joined the film industry as an editor and, eventually, was known as the "Doyenne of Subtitles"—the go-to person for subtitling foreign feature films.

In her 50s, Helen combined her love of learning and photography by pursuing a master's degree at the New School of Social Research in NYC where she studied new media studies. This led to a few photo exhibits in Long Island, NY. For her thesis, she taught kindergarten children this medium, creating a photography program and building a darkroom in a local public school. It was a way for young children to communicate through images about their lives and the world around them.

Helen eventually added Spanish to her language proficiency repertoire in order to broaden her freelance subtitle writing. Having her own company allowed her to care for her daughter, Andrea, who received a double-lung transplant in 2000. Helen never let CF get her down and never stopped fighting for her daughter. She instilled in her daughter the appreciation of life and the arts. Andrea went to college and became a graphic designer with a minor in silkscreen printing. Currently she volunteers her time to USACFA and *CF Roundtable*.

To apply for this scholarship, please specify what type of creative degree you are pursuing (e.g., fine arts, graphics, music, singing, photography, filmmaking, writing, poetry, dance, theatre, and other performance arts). Candidates should have a minimum GPA of 3.0. Please submit essay answers, electronic samples of your work for consideration, transcripts, and a letter from your physician confirming CF diagnosis.
https://www.cfroundtable.com/arts-scholarship ▲

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Edward Gulewitz (in honor of Betsy Rush)
Patty Hardesty
Cynthia Hays (in honor of Zach Hays)
Elizabeth Hissing
Henry Hofflich (in honor of Jonathan Miller)
Barbara Jenkins
Douglas Johnson
Abigail Huntington Kadesch
Linda Keathley-Stamey
Adam Keys
Raymond Keenan
Glen and Rhonda Keysor
Janet King
Edgar Laumbacher
Debra Maloney-Evans (in honor of Erin Evans)
Cathleen Marceau
Tom and Maureen Marlow
Loretta Mulatz
James and Ann Nash
Carol Navidonski
Wanda Olson
Wynne Proctor (in memory of Laird Proctor)
Karen Quinn
Sydney Rabinowitz
Nancy Riecker (in honor of Kevin Reicker)
Michelle Ruckersfeldt
Paige Russell (in honor of Kathy Russell)
Joanne Schum
Iris Shaw
Connie Smith
William and Jennifer Staasheim (in memory of Cynthia Adams Ellis)
Susan Stein
Laura Tillman (in memory of all those we have lost)
Jeanette Trecek (in memory of James Trecek)
Laurie Worth

SILVER

Susie Baldwin (in honor of USACFA directors)
Jean Hanley (in memory of Liz Marshall)
Ozmatic (in honor of CF community)
Karen Scott (in honor of Nicholas)

GOLD

Colleen Adamson
Phyllis Kossoff (in memory of Stephanie Lynn Kossoff)
Dick Marshall

PLATINUM

Ozmatic (in honor of Erin Fenney and USACFA Board of Directors)
Beth Sufian (in support of Hope for the Future awards ceremony)
Aileen Vizel
Devin Wakefield

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DIAMOND SUSTAINING PARTNERS

Marina Day, Trustee of the McComb Foundation

an important solution for patients and physicians.
https://tinyurl.com/y6ecmy7q
AND
https://tinyurl.com/y5fzqsyr

FDA Support Given To Advancing Potential Gene Therapy For CF

Spirovant Sciences' lead gene therapy candidate for select cystic fibrosis (CF) patients, SPIRO-2101, was given rare pediatric disease and orphan drug designations by the U.S. Food and Drug Administration (FDA) to support its development. SPIRO-2101 is an inhaled gene therapy, designed to replace the faulty cystic fibrosis transmembrane conductance regulator (CFTR) gene with a working copy. The therapy uses a harmless virus called adeno-associated virus, or AAV, to carry the genetic sequence of the healthy CFTR into patient lung cells, which can then produce a functional CFTR protein. SPIRO-2101 aims to treat CF patients with class 1 mutations or who cannot tolerate treatment with an existing CFTR modulator. The company estimates this covers about 10% of the total patient population. Class 1 mutations are those in which either no messenger RNA (mRNA) — the intermediate molecule between DNA and a protein — is made (class 1A), or in which the cell produces damaged mRNA that it cannot turn into a protein (class 1B). Current CFTR modulator therapies do not treat these mutations.

SPIRO-2101 has been engineered to target the cells of the outer layer of the airways, called epithelial cells, with high efficiency, allowing the therapy to be delivered where it is needed most.
https://tinyurl.com/yy4pm5r4
AND
https://tinyurl.com/y3jotuxf

Potential Breakthrough For Cystic Fibrosis As Moscow Scientists Use Gene-Editing To Correct Part Of Mutation That Causes Disease

Scientists at Moscow's Bochkov Medical Genetic Research Center have successfully corrected a mutation of the CFTR gene. Using genetic editing, they replaced the parts of the DNA that cause mutations. The researchers focused their efforts on the most common mutation, F508del. The scientists removed specific parts of the DNA of the mutated cells using the CRISPR/Cas9 gene-editing tool, replacing them with a healthy copy of the desired segment. The editing took place on a cell line known as CFTE29o-. The specialists managed to correct around three percent of the cell line, and about five percent of the stem cell culture. To cure a person of the disease, scientists believe around 10-15 percent needs to be edited. Despite the progress, the scientists note that the relatively low efficiency of genomic editing means that it cannot yet be used inside a patient's body.
https://tinyurl.com/y4a23rrf
AND
https://tinyurl.com/yy865xoc

Calithera Biosciences' CB-280 Arginase Inhibitor Trial In Progress Poster Presented At The North American Cystic Fibrosis 2020 Virtual Conference

The randomized, double-blind, placebo-controlled, multiple ascending dose-escalation study (NCT04279769) is exploring CB-280 versus placebo in adults with cystic fibrosis and chronic infection with *Pseudomonas aeruginosa* who are stable on cystic fibrosis medications, including cystic fibrosis transmembrane conductance regulator (CFTR) modulators. The study is evaluating the safety, pharmacokinetics, pharmacodynamics and biological activity of four dose cohorts versus placebo. Enrollment in this study is ongoing and Calithera expects to share interim data in 2021. Preclinical study results suggest CB-280 significantly improved lung function and reduced *Pseudomonas aeruginosa* colony-forming units. Arginase inhibition with CB-280 resulted in improved central airway resistance in CFTR knockout mice, and decreased lung infection in wild type and DeltaF508-CFTR-expressing mice infected with *Pseudomonas aeruginosa.*
https://tinyurl.com/yyu4pk62

Ionis' Inhaled Antisense Medicine Demonstrates Potential As A Novel Treatment For Cystic Fibrosis

Ionis Pharmaceuticals, Inc. announced that data from a clinical trial of IONIS-ENAC-2.5Rx demonstrated a significant decrease in the expression of epithelial sodium channel (ENaC) in healthy volunteers. The study represents the first time an antisense medicine delivered directly to the lung via a nebulizer has shown a significant reduction in ENaC messenger RNA levels. IONIS-ENAC-2.5Rx is an investigational antisense medicine designed to reduce the expression of ENaC in the lung. ENaC is believed to be hyperactive in cystic fibrosis. The data demonstrate attractive tolerability and safety for IONIS-ENAC-2.5Rx with substantial target reduction and the convenience of once weekly administration.
https://tinyurl.com/yy3uunfz

Envara Health Launches Medical Food To Address Fat Malabsorption In Cystic Fibrosis Patients

Envara Health has launched a clinically supported medical food called Encala. The new product is formulated to address fat malabsorption in patients with cystic fibrosis and other exocrine pancreatic diseases by providing both easily absorbable fat calories and improving the absorption of fat and nutrients in accompanying meals and beverages. A study found that children with cystic fibrosis and pancreatic insufficiency that had low dietary fat absorption saw significant improvement in fat absorption after three months of treatment with Encala, as well as increase plasma linoleic acid, a-linoleic acid, and total fatty acids. Compared to placebo, these subjects also saw improvements in height, weight, and body mass index.

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According to Envara, the structured lipid in Encala does not require PERT or digestion to be absorbed. Encala is neutral in taste, gluten-free, non-GMO, and plant-based. It is a source of highly absorbable long-chain fats, including fatty acids, and can be an alternative to medium-chain triglycerides derived from palm, coconut, or vegetable oils. Encala is recommended for use under medical supervision in adults and children over one year of age.
https://tinyurl.com/y2va6h2k
AND
https://tinyurl.com/y4ppw43r ▲

Laura is 73 and has CF. She is a former director and President of USACFA. She and her husband, Lew, live in Northville, MI.

REMINDERS

- Please notify us immediately of any address changes. Returned mail wastes money and delays mailings.
- We would like to act as a referral source for active adult support groups. Please send us your group name, leader's name and phone number, number and age range of your members and geographical area covered, and we will add you to our referral list.
- Please let us know of the major occurrences in your life (e.g., marriages, births, completion of educational degrees or training, career advancement, transplants, anniversaries, birthdays), and we will print your information in **Milestones**.
- Share your ideas for **Focus Topics**, feature articles or any suggestions for improvements you may have to help make *CF Roundtable* more relevant and interesting to you.
- You can reach **USACFA** and ***CF Roundtable*** at any time by email at **cfroundtable@usacfa.org**
- Send your questions of a general nature regarding legal issues that relate to CF to our legal advisor: **Beth Sufian, Esq., call: 1-800-622-0385 Email: CFLegal@sufianpassamano.com**
- You may subscribe at www.cfroundtable.com

Published by the United States Adult Cystic Fibrosis Association, Inc.
CF Roundtable is printed on recycled paper.

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 healthcare 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.

United Network for Organ Sharing (UNOS): Phone: 1-888-894-6361 http://www.unos.org/
Call for information on transplant centers, access for all patients needing organ transplants, and general transplant information.

Transplant Recipients International Organization, Inc. (TRIO): Phone: 1-800-TRIO-386 http://www.trioweb.org/index.shtml
An independent, nonprofit, international organization committed to improving the quality of life of transplant recipients and their families and the families of organ and tissue donors. For information, write to: TRIO, 7055 Heritage Hunt Dr, #307, Gainesville, VA 20155 or email them at: info@trioweb.org

American Organ Transplant Association (AOTA): Phone: 1-832-930-AOTA (2682) http://www.aotaonline.org/
Helps defray out-of-pocket travel expenses for transplant recipients. Helps to set up trust funds. For more information, write to: Administrative Service Center, American Organ Transplant Association, P. O. Box 418, Stilwell, KS 66085. Preferred method of contact is email: aotaonline@gmail.com

ADA: To learn how the Americans with Disabilities Act (ADA) applies to you, contact the Disability Rights Education and Defense Fund (DREDF): Phone: 1-510-644-2555 or email at info@dredf.org