CF Roundtable®
A PUBLICATION FOR ADULTS WHO HAVE CYSTIC FIBROSIS
SPRING 2023

Our Life As A CF Couple And As Parents

By Amanda Boone

My husband Ryan and I have been together for 11 years and married for almost 10 of those years. Unlike a lot of CF couples, we did not meet online as one might think. In 2011, Ryan was working full time at his family's physical therapy clinic as an office manager and I worked full time as an apartment manager in Austin, Texas. I had been through quite a rough year, physically and mentally. I was divorced at 28 years old and my health had started to really decline, mostly because I struggled with compliance. I lost a lot of weight and my lung function started to dip. Right before Christmas in 2011, I was hospitalized for three weeks because I was extremely sick—I was coughing up blood and my lung functions tanked.

PHOTO BY MARIJO SCHUBERT

RYAN AND AMANDA BOONE.

I was a part of a patient advocacy group in 2011 at our CF clinic's hospital. It consisted of adult CF patients and parents of children with CF, in addition to people from our care team. At this time, the cross-infection guidelines were not as strict. Our clinic formed this group and recruited us. I became close friends with Tabitha (Tabby), a mother of a child with CF, Noah. She told me that she had another good friend, Ryan, whom she met at other CF events. He was going through a divorce and needed a friend. He was dealing with some of the issues I had dealt with the prior year and his health was starting to take a hit. She asked me to give him a call. I knew of Ryan but did not know him personally at the time—Austin was still a fairly small CF community back then. We became friends in the fall of 2011 and mostly talked on the phone and on Facebook as meeting in person was frowned upon by clinic.

A mutual friend of ours, Amberlyn, passed away in November, 2011, after Ryan and I became friends. It hit us both hard. Amberlyn was extremely loved in our community and generally. She started a non-profit called Amber's Angels. Volunteers would deliver gifts to kids in the hospital at Christmas and other times. We cried with each other on the phone. Debbie, Amberlyn's mom and a good friend of ours, held a memorial

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EDITOR'S NOTES

As we usher in spring, a season of rebirth and regrowth, it feels fitting that our main focus topic this issue centers around the many pathways to becoming a parent and dovetailing that, we'd like to welcome **Jacob Greene** to our growing board of directors. You can read about Jacob on page 29. Additionally, **Dr. Xan Nowakowski** has stepped up as Secretary for the USACFA board of directors. Xan already serves as our scholarship committee chair. They are such an important part of the work we do here and they're continually striving for our organization to reach further and better reflect the diversity of our CF community. If you're interested in joining our board or starting your own column in our publication, we'd love to connect with you. Send us an email at cfroundtable@usacfa.org so we can set up a time to chat about our mission.

For the focus topic, **Katy Monte** writes about the long process of how their daughter, Riley, came to be—having her sister as a gestational carrier. In their "Pearls of Wisdom" column, **Dr. Nowakowski** writes about the relatively new (at the time) technology of artificial insemination to help histoincompatible couples conceive, a key part of their own birth story and CF diagnosis. We're also sharing **Nicole Kowal**'s journey with pregnancy, all thanks to Trikafta. **Katherine Lockwood** chronicles her struggles with infertility in her article for the focus topic. For our second focus topic, we're following up on how people are doing on modulators three years later. **Suzanne Joyce** shares her experience with increased anxiety while taking Trikafta in conjunction with her transplant medications.

Beth Sufian answers questions about both SSDI work limits for those who want to work part time and asset and income limits for those on a low-income Medicare plan in her "Ask The Attorney" column. **Isabel Stenzel Byrnes** notes the importance of loving your body as it is, even when the going gets rough and medical issues are taking over your quality of life. For our "Pet's Perspective" column this issue, **Colleen Adamson**, on behalf of Penny, relays how getting old isn't for the faint of canine heart—there are lots of vet visits and acupuncture, to say the least. For our "Transplant Talk" column, **Andrea Eisenman** writes a cautionary tale about the proliferation of skin cancer after transplant, including her recent bout with cancer on her eyelid. **Maggie Williamson** shares her recipe for Creamy Lemon Orzo with Asparagus and Peas in her "Culinary Corner" column this issue. I'm definitely adding asparagus to a future Farmhouse Delivery order! **Laura Tillman** expertly collates all the current CF research in her "From the Internet" column. If you read part one of **Rachel Johnston**'s two-part story about climbing to Everest Base Camp in the prior issue, then you're in for a treat with the rest of her story! This issue, **Dr. Nowakowski** interviews Jes Davis about their career as a filmmaker and professional actor as well as their ongoing work with Black Lives Matter for our In the Spotlight feature!

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

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Information From The Internet...

Compiled by Laura Tillman

Cystic Fibrosis Clinical Trials | A Drug Pipeline Analysis Report 2023

Over 80+ Cystic Fibrosis pipeline therapies are in various stages of development. Leading Cystic Fibrosis companies developing novel drug candidates to improve the Cystic Fibrosis treatment landscape include Algi Pharma, Verona Pharma, Translate Bio, Calithera Biosciences, Krystal Biotech, SpliSense, and others. Promising Cystic Fibrosis pipeline therapies in various stages of development include OligoG, Ensifentrine, MRT5005, CB280, KB407, SPL84231, and others. Discover more about the emerging Cystic Fibrosis drugs @ Cystic Fibrosis Treatment Drugs. Find out more about the Cystic Fibrosis treatment options in development @ Cystic Fibrosis Clinical Trials.
https://tinyurl.com/mw34dxhc
AND
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LAURA TILLMAN

Cystic Fibrosis Life Expectancy: What To Know

The life expectancy for people with cystic fibrosis (CF) has greatly improved over time. Early diagnosis and treatment can improve both life expectancy and quality of life for people with the condition. According to the Cystic Fibrosis Foundation's 2021 annual report, the median predicted survival

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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/publication.

Spring (May) 2023: CF and the Pathway to Parenting (Current issue)

Summer (August) 2023: Returning To Work or School (Deadline: June 15, 2023)
Have you been able to return to work or school since starting a modulator and what role did modulators play in that decision to return to work/school? How did you go about returning? Have you been able to return full time or part time? What advice do you have for others looking into returning to work or school? How has going back to school affected your family and/or intimate relationships? How are you navigating changes in those connections? What propelled you to return to work/school? Was there a financial need or desire to get a degree?

Autumn (November) 2023: The 10% Left Behind (Deadline: September 15, 2023)
In general, what are your feelings about being ineligible for modulators? What are you most hopeful for in terms of future treatments while not being able to take modulators? What other therapies on the horizon give you hope? What do you feel while watching others benefit from modulators? If you are worried you will be left behind, what feelings does this elicit?

Winter (February) 2024: Organ Transplants (Deadline: December 15, 2023)

ASK THE ATTORNEY

SSDI Work Limits And Medicare Low-Income Plans

By Beth Sufian, J.D.

If you have questions about laws related to Social Security benefits, Medicaid, Medicare, health insurance, employment, and education rights, you can contact the CF Legal Information Hotline at CFLegal@sufianpassamano.com or 1-800-622-0385 to set up a time to speak to an attorney. All calls are confidential and there is no cost to the caller. The CF Legal Information Hotline (CFLIH) is funded by the CF Foundation, but CFLIH employees are not employed by the CF Foundation. The CFLIH is now in its 25th year.

The information provided in this article is only meant to be general information. A person should verify the type of benefit they receive and review the Social Security rules to make sure that Social Security work income and asset criteria are followed. Nothing in this article is meant to be a guarantee that a person will be eligible for SSDI, SSI, or any other government program.

In the past three months, *CF Roundtable* readers have sent many questions related to the ability to work part time and still receive Social Security Disability benefits. There have also been questions about Medicare and requirements for low-income Medicare plans that provide coverage at a reduced premium with assistance for some Medicare copays.

Question: Does a person who receives Social Security Disability (SSDI) benefits have to work less than 20 hours a week and make less than a certain amount per month in work earnings, or do you only have to meet one of those requirements?

Answer: In order to maintain eligibility for SSDI benefits, a person cannot work more than 20 hours a week *and* cannot make more than a certain amount per month from work activity. In 2023, the maximum amount a person on SSDI can make per month is $1,470 before taxes are taken out of the work check.

There is a possibility that Social Security will provide up to nine trial work months to go over the allowable monthly income amount. However, Social Security uses a lower number to trigger use of a trial work month. Once the trial work months are used and a person goes over the maximum monthly amount of $1,470, the person can lose SSDI benefits. This is a very confusing rule and a person should make sure they understand when a trial work month is used.

If a person is self-employed, the work earnings amount per month is lower and there are a variety of ways Social Security determines work income if a person is self-employed. In addition, a person's medical condition must still meet or equal the medical criteria listed in the Social Security Listing.

BETH SUFIAN

Question: Does the value of a car owned by a person who receives SSDI benefits count in terms of eligibility for SSDI benefits?

Answer: SSDI eligibility is not affected by assets, so the value of a car is not counted when determining eligibility for SSDI. It is important to know which type of Social Security benefit you receive as the asset rules are different for SSDI and SSI.

Question: Does the value of a car owned by a person who receives Supplemental Security Income (SSI) benefits count in terms of eligibility for SSI benefits?

Answer: If a person is receiving SSI benefits, the person must have low assets and low income. This is different than the eligibility rules for SSDI. A single person who receives SSI benefits has a maximum asset limit at all times of $2,000.

The value of one car is not counted toward the SSI asset limit. The car is owned by a person if the title of the car is in their name. If the title of the car is in a parent's name, then the person on SSI is not considered the owner. However, the value of a second car is counted toward the SSI maximum asset limit for those receiving SSI. For example, if a person who receives SSI owns a car that is worth $5,000 and owns a second car that is worth $2,000, then the $5,000 car will not count toward the $2,000 SSI asset limit. The second car will be counted and its value of $2,000 will mean the person cannot have any other assets except the person can own one house.

Question: Does a spouse's income and assets affect eligibility for SSI?

Answer: Yes. If a person, who

only receives SSI benefits then gets married, then the spouse's income and assets will affect eligibility for SSI benefits. A family of two can only have $3,000 total in assets at any given time. In addition, the spouse will have a limit on how much income the spouse can make from work activity.

Understanding the effect of marriage on eligibility for SSI benefits is important. In 11 states in the U.S. there is no access to Medicaid for adults unless the adult receives SSI benefits.

If a person lives in a state that has not expanded Medicaid to low-income adults, then it is very important to make sure marriage will not result in the loss of Medicaid if the person with CF gets married. Sometimes marriage to a spouse who has access to health insurance means that a loss of SSI or Medicaid will not be a problem. But if the spouse does not have access to health insurance that will provide good coverage, the person with CF should assess how they will access coverage for care and treatment.

Question: If I have a low-income Medicare plan do I need to keep income and assets below a certain amount?

Answer: If a person has Medicare and is on a low-income Medicare plan, then assets, work income, benefit amount and spouse's assets and work income may be considered when determining eligibility for the plan. Different Medicaid low-income plans have different rules that can be found at www.Medicare.gov.

Other government programs may count assets differently than Social Security when determining eligibility such as food assistance or housing assistance programs. A person should check the asset criteria for any federal or state program that is available for people who have low income and low assets. ▲

Beth Sufian is 57 years old and has CF. She is an attorney who focuses her law practice on disability law and is the Vice President of USACFA. Her contact information is on page 2. You may contact her with your legal questions about CF-related issues at CFLegal@sufianpassamano.com.

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SPIRIT MEDICINE

Loving The Body As Is

By Isabel Stenzel Byrnes, L.C.S.W., M.P.H.

I have struggled with body loathing all my life. This morning, in a flash of awareness, it occurred to me that I love my body. Really? Am I lying? Well, in this journey toward spiritual wholeness, I do. I love my body. It is my truth. I commit to the conviction that I must love my body. How do we love a body that doesn't always work?

Much of my body loathing has had to do with CF-induced appearance changes, discomfort and pain, or not being able to do what I wanted to do. My body is so darn high maintenance; I'd rather be doing most anything else than taking care of my body's endless demands.

I am 19 years post-lung transplant and my body has responded the right way to the immunosuppressive drugs all these years, which has spared me from rejection. In suppressing my immune system, I am now living with metastatic lung cancer. In exchange for being a long-term survivor, my lungs are slowly diminishing, both by cancer and rejection caused by reducing immunosuppression. In exchange for reaching the age of 50, my joints and muscles are tight and achy. I have vision, gut, sinus, bone issues, brain fog, and, my nemesis, diabetes. The list goes on. It is just my body; it is a lot. But everyone's got something, right?

I know many people with cystic fibrosis naturally feel frustrated by the work it takes to care for the demands of a CF body. I try to take care of my body with all I've got, but, to be honest, at this stage of my life I triage my needs all the time—I exchange time and energy for quality of life. My CF body fits into all the other parts of my life that make demands to live a rich, fulfilled, engaging life.

And yet, there is so much in this body still deserving of love. This body is my vessel that allows my spirit and soul to be housed so I can experience the world, be of service to others, and be in relationship with so many other loving bodies out there. It may only allow me to perform at around 60% capacity, not 100%, to fulfill my spiritual and mental desires, but it's still functional. This body is still working well enough. I am still able to move, speak, think, plan, laugh, and do things I enjoy with less intensity and speed. Wow. There is vitality.

I recently applied for disability and submitted pages upon pages of my medical records. What a humbling experience. On paper, I look like a royal medical trainwreck. The health history and medication list are literally multiple pages long. Someone sitting at a social security office must've been very compassionate—I was approved in two weeks' time. I was shocked!

And then I felt an overwhelming sense of gratitude. I was rewarded with disability benefits because this body was ill enough to qualify. I am grateful that I am mostly functioning relatively well; I am living more than dying. And, before disability, I was managing a stressful job and a time-consuming medical regimen. I was exercising hard. This body climbed mountains, took me around the world, and helped me race in the pool at the Transplant Games. What a glorious miracle this was! But I cannot love my body only when it is well. That's conditional love. Our spiritual charge from God is to engage in loving kindness with all beings, which includes unconditional love. It is hard to do! But our bodies deserve unconditional love, too. So even with chemotherapy, I love my body because it can still function more than I thought it could. It is taking a toll and my weakness and fatigue are getting worse, but I'm still here. My fierce and life-loving willpower and discipline, cultivated by a lifetime with CF, help to fuel this body still, with tenderness and patience. I'm loving my body

> *"So even with chemotherapy, I love my body because it can still function more than I thought it could."*

ISABEL STENZEL BYRNES

for how well it is tolerating the treatment, how it keeps going despite the assault of poison on top of everything else. I love this body for responding to the medications that I'm on and for switching to whole fat dairy and actually gaining weight! It is working. I'm blessed to have started this cancer journey with a reservoir of stamina and strength.

I look around at the waiting room of my hospital clinics and am amazed to see so many people struggling with their bodies: some are old; some are morbidly obese; some are barely walking; some are bald, cancer patients; some are lung transplant patients on oxygen; and some barely stand up from their wheelchairs. I know everyone has a relationship with their own bodies. If we despise and curse our bodies when they aren't working well, we are spending a lot of emotional energy that could otherwise be used to love what we can, whom we can, for as long as we can.

So how do we give love to our bodies? We listen to it. When we are tired, we rest; hungry, we eat; thirsty, we drink. We put our hand on our hearts and can talk to it, encourage it to recover. We can touch and caress the parts that hurt or, even better, get a massage! We can stretch and do yoga and breathe out the tension. We can watch the IV drip into our veins and have faith and trust the body will respond. We can nourish it with wholesome foods and water. We can do as much movement as is comfortable for the body we are housed in, so we circulate our blood and all its healing chemicals. We move slower. We can laugh at it and with it, lovingly. We can accept this is the body we've got. And we abstain from comparing this body to the perceived healthy, ideal body out there. All perfect bodies will, at some point, change and fail.

I am so inspired by my dear friend who recently died of complications from CF and her lung transplants. This remarkable author and patient advocate, Tiffany Christensen, wrote about the importance of loving her body by being patient and giving it space and time for healing. She wrote, "One week and a day after the placement of my PD catheter, an amazing and wondrous thing happened: I felt better. I was not in so much pain and I felt like 'myself.' My body had done what it always has done, healed from the medical wounds, but not on the timeline I had demanded. The body is funny like that. It doesn't listen to reason, bargaining, or commands. It heals in its own time and in its own way. I know this after 49 years of chronic illness and, yet, once again, I betrayed my body with my need to meet an intellectual expectation of healing rather than the magical, peaceful, beautiful process that it is." Amen, Tiffany. Her wisdom outlives her body.

This body deserves love. It is a living creature. I believe it houses a spirit of its own, that is in connection with my larger spirit. Both spirits have to engage in collaborative magic. Much like plants and animals are happier when we talk to them with loving words, so, too, are our bodies. There is much spiritual healing happening when we love our broken bodies deeply and fully.

I love my body. I hope you, too, can say that to yours. Whether your body is stronger with modulator drugs and you can push harder or do more with your body or whether you continue to endure the spectrum of symptoms of CF, may you find your own ways to give it love and talk to it lovingly. Body, mind, and spirit are beautifully woven together and self-care to one of that triad is self-care to all three.

To end, I share with you an inspirational quote that has uplifted me for many years.

Birthright

"Despite illness of body or mind, in spite of blinding despair or habitual belief, who you are is whole. Let nothing keep you separate from the truth. The soul, illumined from within, longs to be known for what it is. Undying, untouched by fire or the storms of life, there is a place inside where stillness and abiding peace reside. You can ride the breath to go there. Despite doubt or hopeless turns of mind, you are not broken. Spirit surrounds, embraces, fills you from the inside out. Release everything that isn't your true nature. What's left, the fullness, light and shadow, claim all that as your birthright."

—Danna Faulds ▲

Isa Stenzel Byrnes is 50 years old and has CF. She lives in Redwood City, California, with her husband, Andrew. She is 19 years post-lung transplant.

PEARLS OF WISDOM

Pathways To Parenting: The Other Side Of The Table

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

When we announced our Focus Topic on CF and the Path to Parenting, I spent some time thinking about what I would write as my column for this issue. I don't have children and have known for my entire life that I do not want to become a parent by any means. But after giving it some thought, I realized that my life as a childfree person with CF still affords plenty of insight about family planning and parenthood in the context of our shared health experiences. After all, my own family's unique path to parenting may well explain why I have CF.

Let's rewind a bit. I was born in 1983 as the result of a science experiment at the University of Mississippi Medical Center. Too perfect, right—how many scientific faculty members at medical schools can claim *that* as our origin story? I was made in a lab before ever growing up in one. And yes, show and tell at school was always interesting. But before I was busily spending my free time dissecting brains and dipping slides to develop images of how cells were growing and moving around, I was a little clump of rapidly growing tissue myself. My parents were able to make me because they had some help from an anonymous sperm donor who contributed samples to the research study they joined.

My parents, who have no history of CF in either of their families, were both fertile but could not conceive a baby together. This phenomenon was becoming more widely recognized by scientists in the early 1980s. Reproductive health scholars took interest in exploring the relatively new technology of artificial insemination to see if it would reliably benefit histoincompatible couples like my parents. One of these studies just happened to be located at the same medical school where they held faculty positions. Three trials later, they got me—and an eventual crash course in a disease they'd only known about previously from their work.

What we still don't know—and may never find out—is whether I would have had CF regardless of whether my parents had done the artificial insemination study or pursued in vitro fertilization as an alternate option. CF is frequently hereditary, but not always. Every genetic mutation, whether CF-related or not, started out as a spontaneous one. Sometimes these mutations happen during a parent's life, making it possible to pass the altered gene to offspring. Other times they happen while a child is developing in the womb.

XAN NOWAKOWSKI

We don't know whether my CFTR mutations came from my mom and biological father. But because I have double expression of the same extremely rare mutation, it's likely that neither of them passed it to me and that something that happened during my mother's pregnancy would explain how my own CFTR genes wound up coded differently. Before I was born, my parents didn't know I had CFTR gene mutations or that I would have CF clinically. I joined the world in December of 1983—nearly six years before the CFTR gene was even discovered, let alone when any genetic testing to detect potential CF in developing babies became available.

Being born via donor conception in the early 1980s posed some drawbacks. Laws about mandatory information sharing for families of children conceived via anonymous sperm donation missed my birth cohort by two years. People born after 1985 get access automatically to details about their background and origins that I spent decades seeking out. For me—and for my half-brother who does not have CF—there are always more questions than answers.

At the same time, I feel glad that my parents didn't know before I was born that I would have CF. If they had known, I might never have gotten to live at all. We've spoken frankly about the fact that my mother and father would have considered aborting the pregnancy had they known I would have this disease. I'm not sure what choice they would ultimately have made and neither are they. And I don't think my own opinions about the inherently eugenic elements of

choosing to terminate a pregnancy because of chronic disease should supersede the essential human right of people to self-determine about gestation and childbirth. I may not agree with the specifics of someone's reasoning when choosing abortion, but I will defend their right to get one no matter what.

And although it would be easy for me as a childfree person to say "if people don't want to deal with the possibility of a sick child, they should just adopt" as if the word "just" belonged in that sentence, I cannot do so. I've known for most of my life that there is no "just" when it comes to expanding a family. Indeed, my parents tried for years to adopt before finally giving up because even getting an interview would have taken another 10 years or more. I can't pretend to understand firsthand the heartbreak of that experience—or of learning once they had me that I would live with a life-threatening disease that forced all of us to reckon constantly with my mortality.

So what have I learned about pathways to parenting from being a childfree person with CF whose life began with donor conception? Lots—and it all comes back to the vital importance of supporting people in their individual family planning journeys. Many people with CF now find themselves having options they'd never anticipated for expanding their families.

Apart from just living longer thanks to better overall care, plenty of people in the adult community are thriving on CFTR protein modulators. Drugs like Trikafta can't replace a missing vas deferens—the tube that allows sperm to get into the seminal fluid before someone ejaculates—in people born without one. Folks with testicles who have two copies of the DF508 mutation often don't have a vas deferens. But in people with vaginas, often taking a modulator will thin out the cervical mucus—the wet substance that lubricates the canal—enough for sperm to pass through into the uterus and fertilize an egg. Producing children through sexual reproduction is now much easier for many folks with CF thanks to modulators!

Options for assisted reproduction have also expanded since I was born. I've seen many folks with CF use either artificial insemination or in vitro fertilization to have children. Accessing these services has become easier and more affordable than in times past, although we still have miles to go in achieving true justice on that front. IVF also offers the option of intentionally selecting eggs that do not carry any CFTR gene mutations, and testing in advance for other genetic diseases. Whether parents use this information to plan intentionally for how they will meet their kids' medical needs or select the specific sample they want to use to make an embryo based on this information, it's a huge advantage.

Many folks with CF are also sharing joyfully these days about their adoption and fostering journeys. With visibility increasing all the time for our adult community—and better information circulating about what people with CF can do with our lives as we continue to live longer and thrive more—it's becoming easier for prospective parents who want to open their hearts to children who are already in the world to welcome kids into their lives. I have to say, of all the stories I've seen where people wanted to be parents and then actually got to do so, these make me smile the biggest. I still think the kid who didn't get to grow up with my parents because they had me instead lost out. Then again, I'm terribly biased.

Finally, it's important as we think about all the ways people with CF can now have kids that we remember to support members of our community who don't want that life. Some of us realize we're childfree as adults; some of us have known since we were still young children ourselves. Either way, it's vital for our loved ones to affirm our desire *not* to raise children. As my own parents always told me, it's better to regret not having children than to regret having them. I've never regretted being childfree for a moment—I always knew becoming a parent wasn't my journey in life.

Likewise, I always appreciated the insight I gained from my lack of interest in having children. People who would pressure me to change my mind usually changed their tune quickly when they learned I had a fatal genetic disease. Ableism remains as deeply ingrained in our society as ever. Yet I *always* had the full support of my parents in following the path that would best for me—whether it led me toward parenting or brought me nowhere near it. And being a little older and wiser these days, I deeply appreciate what being childfree allows me to give with the time and energy I would otherwise spend on my own children. Supporting my fellow adult CF community members on their parenting journeys remains a joyful part of that always. ▲

Dr. Alexandra "Xan" Nowakowski is 39 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 Associate 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.*

PET'S PERSPECTIVE

Sweet 16

By Penny, translated by *Colleen Adamson*

Hi, there! My name is Penny, and I am a 16-year-old miniature schnauzer. I am named after my parents' home states (PA [Penn] and NY). Pretty creative, I guess. There is another miniature schnauzer in my neighborhood, also named Penny, because she came from PA. People get us confused all the time, which makes no sense to me since I am obviously cooler and better looking.

I came from my grandparent's dog, Nellie. Mama picked me out after sitting on our puppy blanket for a LONG time. I don't know why it took so long; I sat there staring at her the entire time while she played with my sister, who proceeded to poop right next to Mama! I thought that was funny. My sister was trying to tell her not to pick her in no uncertain terms! The rest, as they say, is history.

I had a hard time adjusting from rural life to city life. I didn't like all the cars going by on the street, since my grandparents live in a quiet, rural area, where I did not see many cars. We three started hanging out on the back patio and grassy common area, and slowly moved from there to walking on the sidewalks. No sweat! I met a lot of friends (especially boyfriends) that way, too! Now Daddy's whole family calls me "The City Dog". BOL (Bark Out Loud)!

When I was younger, our next-door neighbors took care of me during the day. I loved that because I got to play with their two kids AND I got to visit with my boyfriend Rocky! We grew up together, and it was so sad when they moved. I missed him for a while, and I didn't understand why he wasn't around anymore. After that, two really nice women (Kate and Mary) with their own dog-walking business took care of me. They walked me for a half hour some days, and other days I was with them all day (something called doggie day care) and went on all their walks with them. There were days I walked five or six miles. Talk about being fit! I met more friends and boyfriends! Not that I'm boy crazy, but I kind of am. Then something called COVID-19 came, and they stopped walking me and my friends. But the good news was I had Mama and Daddy home with me all day every day! Daddy is usually upstairs working, but Mama is downstairs with me. I see her doing something called "breathing treatments" in the morning and at night. I usually sleep through those since they are boring to watch,

PENNY WITH HER PEANUT BUTTER.

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rate for people born in 2017–2021 is around 53 years. This means that 50% of people born in those years are expected to reach more than 53 years of age. It is important to remember that these figures are estimates and are based on previous medical studies. Even with improved treatments, there are factors that can affect the life expectancy of people with CF. New medications are opening doors to better long-term lung function. Effective treatments include:

Bronchodilators: These are medications that help open airways by relaxing the lungs' muscles.

Airway clearance techniques (ACTs): ACTs are techniques that include clapping or huffing to help clear mucus buildup. In addition, nebulizers, which are devices that turn liquid medications into an inhalable mist, can be beneficial. Exercise machinery and vests that vibrate the chest may be used as well.

Antibiotics: These medications can treat recurrent lung infections.

CFTR modulators: These are medications that can help prevent mucus buildup tied to the genetic mutations of CF.

Additional treatment avenues that can improve life expectancy for people with CF are lung and liver transplants in the event of severe organ damage.

https://tinyurl.com/55fncevh

Cystic Fibrosis Chronicle: Why Has The Often-Deadly CF Gene Not Passed Out Of The Human Genome? And What New Treatments Are Being Developed?

The mutation that causes cystic fibrosis arose in the early Bronze Age and spread across Europe during ancient migrations. Cystic fibrosis is the most common fatal genetic disease in the US, and yet has genetic characteristics that should hinder it's spread or remove the mutation from the gene pool altogether. We would expect natural selection to eliminate alleles with

but it seems like she feels better after doing them. Go Mama! Mama also sometimes has a nurse come in to do an infusion. I really don't understand what's going on, so I sit on my bed right next to her and watch the whole time, just in case.

Getting old is not for the faint of heart, am I right? I lost my hearing about two years ago; I am not sure why. I do not mind thunderstorms or fireworks as much now, so that's good. I also have weakness in my back legs, and I don't have very good balance anymore. I started slipping on the hardwood floors on our main level, so I now wear socks that have slip proof bottoms. They work great, but I do like to take them off sometimes. I like to keep my parents on their toes! I also wear a diaper because I have some bladder control issues, but they are very pretty with colorful flowers on them (so they tell me, anyway, since I don't see colors). I got a new harness that covers my whole body, which helps Mama and Daddy keep me balanced when we go out for walks. The new harness also has a handle on it that helps Mama guide me or carry me up the stairs.

COLLEEN ADAMSON AND PENNY "SIT."

Daddy just carries me up the stairs, but Mama likes to hold on to the banister since she has balance issues, too. I also have dry eyes, so I see a special veterinarian for that.

I just started acupuncture therapy. I really like it. I get treats during the sessions! I LIVE for treats! I will do ANYTHING for a treat, even a bath! Not that I'm food motivated or anything. Can we just talk about peanut butter for a second? Delicious, am I right? My parents put my pills in peanut butter so it's easy for me to take them. Do I care? No! Because peanut butter IS THE BEST! Sorry I got a little distracted there for a second. What was I writing about? Oh, yeah, acupuncture. I do feel like I am walking better even after just a couple of sessions. The vet said it usually takes around four or five treatments to notice a difference.

My parents really do take very good care of me, I must say. I love them very much, and I know they love me. There is nothing better than that, not even treats! ▲

Colleen Adamson is 54 and has CF. She and her husband Scott got Penny in 2007 when she was only 10 months old. She is the love of their lives.

negative effects from a population, and yet many populations include individuals carrying such alleles. So why are these deleterious alleles still around? What might keep natural selection from getting rid of them? First the longer projected life expectancy means that survivors have more of an opportunity to pass along these killer mutations. Second, CF is a recessive genetic disorder; this means both parents must carry one copy of the gene for a child to have a chance of being born with the disorder and, even then, the risk is one in 4. In most cases, these two characteristics would have limited the spread of the deadly gene. However, in the case of CF, they created an evolutionary niche that has allowed the gene to grow in the population to the point where approximately 10 million Americans carry it. This is partly due to the gene's recessive nature: you can carry one copy without exhibiting any symptoms of the disease. Only a genetic screening will alert you to the presence of the GF gene in your DNA. This has allowed the gene to 'silently' propagate though the population. Lastly, it's been proposed that carrying one copy of the gene for CF actually conferred an evolutionary benefit to prevent people from dying of tuberculosis and cholera. In other words, the negative effects of the genes involved were counterbalanced by their positive evolutionary contributions. This hypothesis is now regarded by many as the best explanation for why such a lethal genetic disease became so common.

https://tinyurl.com/2kc7no84

CFTR Mutations Impair SARS-CoV-2 Virus, Study Finds

CFTR gene mutations significantly reduce the cell entry and replication of SARS-CoV-2, the virus responsible for COVID-19. The research team noted that several studies have suggested that incidence of severe COVID-19 in CF

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FOCUS TOPIC

CF AND THE PATHWAY TO PARENTING

CF And My Pathway To Parenting

By Katherine Lockwood

KATHERINE LOCKWOOD HOLDING HER DEBUT BOOK "WHY ME, MAMA?" AT THE MOEBIUS SYNDROME CONFERENCE IN ATLANTA, GA, THIS PAST JULY.

My husband and I got married, bought a fixer upper and made it ours. One holiday, I presented a baby card to both sets of our parents. "Pregnant?" they asked in excitement. "No," I replied. I blurted out that we had genetic testing and Arden isn't a carrier, so we are able to have biological children together. The testing cost us four thousand dollars.

A couple years later, we decided it was time to grow our family. We took one last trip—a two-week journey through France: Paris, Troyes, and Champagne. Fitting for a last hurrah—exploring the wine and champagne houses and toasting to our future. We arrived home and I stopped my birth control. I switched from oral meds to insulin and changed a few other meds per my doctor's advice. I had spoken to my doctors and they were on board. At the time I was given the ok to start trying, my lung functions were at 83%.

Not pregnant.

After a few months of trying, I tried figuring out my ovulation, got a tracking app and a few more months passed.

Not pregnant.

I spoke to my doctor and they recommended making an appointment with a specialist. It can take a while; you can always cancel.

I got blood work, a sperm sample from Arden, and a hysterosalpingography (HSG) test. The HSG test is an x-ray procedure used to look at your uterus and fallopian tubes (yes, this hurts but only for 30 seconds). I read somewhere that many get pregnant following an HSG as the dye somehow cleans out your fallopian tubes. That was hopeful.

Unexplained infertility.

The doctor said we were lucky—we came in early, we were young, and we lived in Massachusetts, where fertility coverage is mandated. The process would be two IUIs, which are required by our insurance, and then, if that doesn't work, we would move onto IVF.

We decided to keep trying on our own. Another year passed. We explored foster care and took the training but didn't go any further. My husband's family had three trees come down through their home so they moved in with us for a year. We went to therapy. Another degree was completed. We traveled more, especially when I was ovulating. I tore my knee in three places from an unfortunate jump in a bouncy house. I could not drive and, because I worked an hour away, I was out of work for six months. Eventually, we returned to the doctor.

We had to redo all the testing per insurance. We did three IUI's. My husband was praised for attending all my appointments as I was on my back having a procedure done while my leg was broken.

Not pregnant.

I returned to work. We had our own gynecological egg hunt on Easter: 11 eggs; 10 mature and seven fertilize. We got four embryos. We opted for preimplantation genetic testing, which tests the chromosomes and supposedly reduces the risk of miscarriage. We paid $3,900 for this extra testing. A few weeks went by and I got the call that three of the four embryos were chromosomally normal. Three girls. Visions of three twin cribs side by side in a warm nursery entered my mind.

A couple of months later, we did

our first transfer. The call came two weeks later.

Pregnant...wow, ok!

Our beta numbers were excellent. They doubled and continued to rise until, at 11 weeks and three days, our baby no longer had a heartbeat.

I applied for jobs while bleeding in the bathtub as I was sick of staying at a job an hour away just because I'd worked there long enough to feel comfortable taking maternity leave. We spent the next couple of months in despair. I went to therapy. I talked about getting an Australian merle puppy. I read books about pregnancy loss and childlessness not by choice. I spent multiple hours at appointments.

We decided to try again. This time there was no excitement, only anxiety. The transfer happened. Our beta number was low. Only 41, while our first transfer was over 200. I cried in the bathtub, thinking, "it didn't work." But it did, and the number kept rising. We passed the twelve-week appointment. We finally let our guard down after the anatomy scan but, the next day, the world shut down due to COVID-19.

I wish I could say that our path became easier after that. Unfortunately, I can't. However, we now have two wonderful girls: Rose, who is two and a half years old, and Magnolia, who is one. And we will not be getting a merle puppy anytime soon.

My advice is to prioritize your relationship with your partner—let them into your world and find ways to continue to have fun together. Plan things to look forward to. Therapy can be a helpful tool.

The path to parenthood for us started years before we were ready to physically start trying. I never considered that we could have difficulty getting pregnant. Both my medical team and I were always more concerned about my health and how my body would experience pregnancy (my body did great).

Infertility is really common. In general, one in seven couples have difficulty conceiving and one in four experience pregnancy loss. Unfortunately, these things aren't talked about, which leads to people feeling like they are more alone in their experience. We told everyone about our first pregnancy because we thought we were in the safe zone. I recommend only telling people who can grieve with you as much as they can celebrate in the early weeks. A benefit to people knowing is that they shared their own experiences; ones I would never have known otherwise. ▲

Katherine Lockwood is 35 years old and has CF. She lives on Cape Cod with her family. She recently published a children's book, Why Me, Mama?, *after her daughter was born with an even rarer condition than her own, so that all little ones with differences could see themselves represented in children's literature and know that they matter. Katherine works with individuals and couples as a therapist and specializes in infertility and disability issues. You can reach her at KatherineML22@gmail.com or acorncottagepress.com.*

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patients is lower than what was expected. Because of this, the team set out to uncover the cellular mechanisms that could explain this situation. The results suggest that CF may hamper the cytokine release syndrome triggered by SARS-CoV-2 S protein stimulation in airway epithelia, thus strengthening the hypothesis that CF may constitute a biological advantage by decreasing the risk of developing unfavorable COVID-19 outcomes.

https://tinyurl.com/4j2x36ys

AND

https://tinyurl.com/2ehwxt9t

AI Model Can Help Detect Collapsed Lung Using Chest X-rays

A new study shows that an artificial intelligence (AI) model can accurately detect simple and tension pneumothorax on chest radiographs. A pneumothorax is a collapsed lung that occurs when air leaks into the space between the lung and chest wall. This air pushes on the outside of the lung and makes it collapse either wholly or partially. The study noted that early detection of pneumothorax is crucial, as the severity of the condition will determine the need for emergency intervention. A pneumothorax is typically detected and diagnosed using a chest X-ray and radiologist interpretation, but the authors hypothesized that AI could help improve this process. The researchers explained that first, AI could assist in triaging chest radiographs for sooner interpretation by a radiologist based on the suspected presence of a pneumothorax. Second, it could provide a second 'set of eyes' to support identification of a pneumothorax. The findings suggest that the ability to accurately detect and rapidly triage pneumothorax with an AI model could assist with earlier identification and improve patient care if integrated into the clinical workflow.

https://tinyurl.com/4x42cx66

High-Dose Vitamin D Therapy Successful For Patients With Cystic Fibrosis

Vitamin D levels in patients with a deficiency and cystic fibrosis increased with a single high dose, or stoss therapy. Vitamin D deficiency can lead to poor bone health (osteomalacia) and has been associated with worsening lung

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A Long And Twisty Road To Becoming Mom

By Katy Monte

I was born in 1984 and diagnosed with CF when I was 13 months old. Growing up, I knew that one day, if I was lucky enough to still be alive, I would love to become a mother. Starting at an early age, I spent a large portion of my time in the hospital for CF exacerbations and, because of this, I also knew about a lot of older CF women who got pregnant and became moms, but, ultimately, lost their lives because the pregnancy was very hard on their bodies. That scared me a lot. As I got older, I also learned that there were many different paths one could take to become a mother.

I started dating my now husband, Joey, in 2008. We met in 2001 at a restaurant where we both eventually worked. We became really great friends at first because our age difference was an issue at the time. He is three years younger than me and, while that is not an issue now, it was when we worked together at the restaurant as he was 14 and I was 17. I knew that, in a few years, he was going to make someone really happy. I ended up moving away to go to college and then nursing school. While I was in nursing school, I lived at home again and worked a few nights a week at the restaurant. Joey was now 20, almost 21. We went out a few times and ended up dating. I knew he was the person I was going to marry someday. We had talks about the future and we both wanted kids. We had conversations about how it might not be the best for me to carry a baby, if I could even get pregnant, and that we might have to look into different avenues to becoming parents. From the beginning this was never an issue with Joey. He wanted me around for the long haul, so if getting pregnant would somehow jeopardize that, he was willing to try something else.

KATY AND RILEY MONTE.

In 2011, I was told that I needed to be evaluated for a double lung transplant. I decided that for me, personally, a chance at transplant and extra years with my family and Joey were worth all the risks that came along with it. I was on the transplant list for six and half months and was transplanted in November of 2011. The transplant was a success—I felt amazing, like I had a brand-new lease on life. In 2012, Joey proposed. Our wedding was set for October 2013. At the end of 2012, I was admitted to hospital with a virus and ended up on life support, ECMO, and dialysis. The doctors didn't know if I would make it, but I did. I came off all life support and slowly recovered.

I had started seeing a reproductive endocrinologist a few years prior to this because I always had really painful menstrual cycles, and I usually got sick every month right before my period came. Our wedding was fast approaching and I made it clear that I had always wanted to be a mom, so my reproductive endocrinologist suggested I freeze my eggs because of all my medical issues. I discussed this with Joey and we were excited to proceed. I scheduled the date, ordered my medicine, and, just as I was about to pay for the procedure, the head doctor at the fertility office, without even seeing or speaking to me, called me to say it was cancelled because I was "too risky" a patient. I was crushed and didn't know where to go from here.

Right before our wedding, I started getting short of breath just walking small distances. When we got back from our honeymoon, after having many different tests and ruling other causes out, I was told I was in chronic rejection. I started treatment to try and stabilize my lung function but nothing worked. My only option was a second double lung transplant. My kidney function, however, prevented me from getting listed. After my life support episode, my kidneys never recovered completely. I was told I needed a kidney transplant before I was able to be relisted for a second double lung transplant. Two of my sisters got tested and were both a match. Christine was a slightly better match, so she donated one of her kidneys to me in July 2014, and I was actively listed for lungs again in August. In December, I got sick and was admitted to the hospital and was told that I was too sick to continue to wait for lungs at home. I knew that I was either going to be lucky and blessed with a second double lung transplant or I was going to die waiting in the hospital. After two dry runs, I was finally transplanted with my second set of donor lungs in February 2015. I went home in March and started to really dream about my future.

In 2016, I found a different fertility center apart from my reproductive endocrinologist. They knew all about my complicated medical history and had several other CF patients and were willing to let me try and freeze my eggs. The hope was to freeze embryos and have my sister be our surrogate. I did two rounds of hormone shots and retrievals, but the embryos that were harvested never made it to the freezing stage. My husband and I didn't know what to do next, but I wasn't ready to give up on my dream of becoming a mom.

> "Being a mom and seeing Joey as a dad is the highlight of my life."

In 2017, two years after my second lung transplant, I was healthier than I had been in a very long time, and I got the approval from my doctors to try and carry a baby myself. My husband and I tried for six months naturally and nothing happened. We moved on to IUIs. We did five and they all failed. Each month, the anticipation of wondering if this could work, followed by the devastation when it didn't, was a lot. It was like a rollercoaster of emotions, from hope and excitement to defeat. After the failed IUIs, we decided we were going to take a quick break and start IVF in 2018. Unfortunately, in 2018 I had some kidney complications and had to restart a medication that was not safe to be taken while pregnant.

At this point, we still had my sister willing to be a surrogate for us, but we didn't have any embryos. We decided to purchase a batch of donor eggs. We went through a ton of egg donor profiles online and found one that looked like me and had a similar background and characteristics. Six eggs came in the batch. We fertilized them with my husband's sperm, and two of them made it to the freezing stage. We now had two embryos ready to go. We were going through the process of drawing up legal contracts and getting screened by the fertility center and ready to transfer an embryo into my sister, when I found out I was unexpectedly pregnant. We did not move ahead with the transfer into our surrogate. I couldn't believe that after all the time trying to get pregnant naturally, and then the failed IUIs, that I just randomly conceived. I called Joey right after I took a test and we were both freaking out; we couldn't believe it. I called my doctors right away and was taken off the medicine that was not safe for pregnancy. I was in utter shock that I was pregnant but also really excited and, if I'm being honest, a little scared. Unfortunately I miscarried around nine weeks. I was completely devastated, but I also had hope that one of the two donor egg embryos we had frozen was going to work for my sister. We transferred one in January 2019 and it took. At the eight-week scan, there was no growth or heartbeat. We transferred the last embryo in July 2019 and it never took; it was a failed transfer. We were back to square one.

Deep down I kept thinking and asking myself why I was able to get pregnant after all of that time just to end in a miscarriage. I had this gut feeling deep down that maybe even though I was three years older, that it was a sign saying my eggs were healthier. I felt like it was someone saying, try one more time. I decided to try and harvest my eggs one last time at the fertility center where I was doing the IUIs. The doctor had a different protocol of medications and, on New Year's Eve, 2019, they harvested 11 eggs. Six embryos fertilized successfully and two healthy embryos made it to the freezing stage! We were ready to try one last time with my sister as our gestational carrier and then COVID-19 happened. By the time procedures like an embryo transfer were allowed again and the timing was right for my sister, we had a scheduled FET (frozen embryo transfer) set for September 2, 2020. On September 11 we got the phone call that said she was pregnant and her numbers were looking amazing. We were cautiously optimistic at this point because we had both been here before. I lost the baby at nine weeks, and she lost one at eight weeks. We didn't get to go to any doctor's appointments with her because of the rules they were following for COVID-19, but, after every appointment, we were reassured that things were looking great. We started to actually get excited that our dream of becoming parents was finally happening. We did a gender reveal in November and found out we were having a baby girl. On May 11, 2021, my sister delivered our baby girl, Riley Kathryn Monte.

Being a mom and seeing Joey as a dad is the highlight of my life. Joey was ready to give up at a certain point but now thanks me for being so stubborn and never losing heart. I often catch myself driving and looking in the rearview mirror in utter disbelief that she's real and all mine! She's almost two years old and I can't believe how fast the time goes. I am so thankful for modern medicine, awesome doctors, and my sister for helping our dreams come true. It took so many years for us to get here from the time we just started thinking about freezing my eggs, to my failed retrievals in 2016, and all that followed after. I hope one day to share this all with Riley, so that she can see just how badly she was wanted and how much she is loved. ▲

Katy Monte is 38 years old and has CF. She lives in Bayside, NY. Katy is a registered nurse but currently a stay-at-home mom. She loves reading, exercising, and spending quality time with her family.

concert for her at a local honky-tonk. Country music artists who knew and loved her sang in her memory. The event raised money for Amber's Angels. Tabby and Debbie were best friends and the four of us made plans to go. I told Ryan I would see him there. I didn't think too much of it, really. Ryan walked through the door and we saw each other for the first time. We immediately had chemistry. He asked me to dance, but he asked other girls, too. We danced three or so feet apart and moved our heads away from each other; however, we were holding hands, too, so it is sort of comical now to think we were trying to stay safe. From that night on, we kept making up excuses to see each other, staying three feet apart. After a couple weeks, it was impossible not to admit that we were in love. I was in love with someone I had never even kissed! It was strange and refreshing. We talked about it at length and what we were going to do. We were both the happiest we had ever been, especially after everything we had gone through with our previous relationships.

We looked at each other's clinic reports and sputum cultures and discussed with what bacteria our lungs were colonized. We both mainly grew *Staphylococcus aureus* and we both had only grown *Pseudomonas* a handful of times each in our lives. We both told each other that if we had anything that was detrimental, such as *B. cepacia* or MRSA, we would not be able to continue with our growing relationship. We both felt that life was too short and we decided to dive in! We married on September 21, 2013, on a beach in Port Aransas, Texas.

Our doctors and nurses were surprisingly very supportive. Ryan and I were active in the CF community and my health improved a lot being with him and becoming 100% compliant. His health did, too. We were both still working and enjoying married life when I dropped the B word on him. Oh, boy, did I want a baby badly! Ryan is five years younger than I and he was not quite ready for a baby. In fact, I'm sure he thought I was a tad crazy at this point. I have always just thought I could do it all. I've always loved children and being a mom was my dream. Ryan finally came around to it and we saw a fertility doctor. We knew we didn't want to pass on CF. We spoke with our CF team and I was cleared to have a baby. After a little while, I realized my body could not handle a pregnancy Instead, we found a surrogate. I went through IVF and had my eggs retrieved in June of 2014. Thankfully,

PHOTO BY MARJO SCHUBERT

AMANDA BOONE AND HER SON, RYDER.

I was very fertile at the age of 31 and we got seven embryos. The first and only embryo transfer into our surrogate took place in October of 2014! Ryder Boone was born on June 11, 2015.

My health had begun deteriorating between 2014-2019. Some would say motherhood and its responsibilities took a toll on my health and others would say it's just part of getting older with CF. I would argue both of these reasons contributed to the decline in my health. To be perfectly honest, it started even before Ryder was born. I came down with the flu in December of 2014. It was the first year that I realized the flu shot did not always work. I was hospitalized and very afraid. Our surrogate was 12 weeks into her pregnancy, and I was truly thankful I was not. My health never completely recovered after that hospitalization.

I required more IV antibiotics and hospital trips. I was not too happy about the whole situation, as I had planned to continue working after he was born. Instead, my doctor and I came to the realization that I should not go back to work so I medically retired. I had two months to wallow and then I had a newborn baby to take care of. Ryan had just started a new job as an IT tech and was very busy. I was worn out but blissfully exhausted in my new role of both CF patient and mom.

By the time Ryder was two years old, I was in and out of the hospital routinely and it was taking a toll on my health. I was at a point in my health that if I didn't do something I was afraid I would not live long. My allergies and asthma were horrible in Austin and it just exacerbated the issues in my lungs. I told Ryan that we should move to Colorado Springs as I had googled best places to live with allergies and asthma and then cross-referenced those cities with accredited CF care centers.

I had heard many wonderful things about the National Jewish adult CF clinic in Denver, Colorado, and I was ready for a change. So off we went! We lived in a B&B for a while to see if it was going to work out. During that time, I saw the CF clinic in Denver and my body was in worse shape than I thought. They immediately admitted me to the hospital. It was the week of Thanksgiving and our short-term lease was about to end. After some consideration and talks with the doctors and our family, it was decided we needed to stay in Colorado. I was stuck in the hospital alone for Thanksgiving and the three weeks following while Ryan and Ryder went back to Texas to get our house packed up and sold. I came home for

five days at the beginning of December, only to have to be rushed back to Denver because I was so ill. I spent Christmas and my birthday two days later in the hospital away from my family and everyone I knew. It was one of the darker periods in my life. Thankfully, my mom flew in to be with me. My Dad and the rest of our family helped Ryan get moved. We reunited as a family on December 28, 2017, and moved into another short-term rental in Manitou Springs. I was on IV antibiotics at home and still pretty sick.

2018 was another rough year for me health wise. I was in the hospital six times that year. In 2019 my health saw some stability, but my lungs were still severely damaged. I was being hospitalized every two to three months. The visits were not as critical as previous times—my CF team had found a decent antibiotic cocktail that worked for me. The National Jewish CF Care Team were my saving grace. In October 2019, Ryan and I went to see our doctor for our quarterly CF clinic visit. I always pack a bag because I never know when I will be admitted, and we live a couple hours away from the hospital. The visit was uneventful for both of us, which, for me, is rare. While we were at the appointment, Ryan was on Facebook and saw that the newest genetic modulator, Trikafta, had been approved by the FDA. It was not projected to come out until March of 2020. We were all in shock and so happy. Ryan had been on the drug through a clinical trial and had already seen wonderful results. He gained weight and his health was very stable. He recovered from viruses and a bad sinus surgery quicker than usual. Unfortunately, he did not have a huge increase in lung function.

By November 2019, my lung function was down to 44%, and I couldn't get it over 50% unless I had just been on a round of IV antibiotics. I felt unwell most of the time and I slept a lot. I used oxygen at night and during light exercise. It was hard to play with Ryder and keep up with simple house chores.

> "Some would say motherhood and its responsibilities took a toll on my health and others would say it's just part of getting older with CF."

My husband and family helped us out a lot during this time. My doctor referred me to a transplant clinic.

I felt defeated at first. The Cystic Fibrosis Foundation changed the way they do transplant evaluations and they wanted us to see transplant clinics earlier, if possible, so that we could be on their radar and get information. My doctor explained that since my health is so up and down it would not be a bad idea. I had anxiety about this and expressed to her that I was worried about what would happen to me if I was really sick and we did not have a plan. I think she thought this visit would help ease some of my worries.

Ryan and I each have only one copy of the most common CF mutation, DF508. My second copy is a rare mutation, E60X, which is a nonsense mutation and rather severe. Because neither one of us has two copies of DF508, we have never been eligible for any of the prior modulators. After finally getting insurance approval and copay assistance, I was able to start Trikafta on Friday, November 8, 2019. I was excited, but not expecting too much.

My health and life changed on day five of Trikafta. I started coughing up tons of mucus and then I started to breathe again. I honestly could not remember the last time I could take a deep breath like that. It woke me up in the middle of the night. I was in shock and could not go back to sleep. I decided to do my PFTs with my home spirometry to check my lung function. I blew a 56% FEV1, which was a 12% jump in four days! I started crying. My PFTs had been hovering around 41-50% over the last year, and rarely at 50%. My numbers had dropped to the lower 30s with lung exacerbations. I was in disbelief but felt elated. I posted this on my Facebook page: "It is such an exciting time in CF and I'm so happy for my CF friends that are eligible! I cannot wait until children can benefit from this and I pray that their lung function stays high or stabilizes so they will never have to endure the things we have. I have hope for my future." I did recognize the grief I felt for my friends who remained ineligible and hoped that CFF had plans for them as well.

I attended my transplant clinic appointment the day after this health breakthrough. I was turned down for a transplant because everything looked stable. My oxygen saturation was good and my lungs actually sounded fairly clear. I was told to go home and continue the new miracle drug I had begun. The physician said she hoped I would never need a transplant; however, I would still be monitored to make sure.

I just turned 40 last December! I am living the dream. I have far exceeded the age my parents were told that children with CF lived to. When I was diagnosed at 10 years old, children were expected to only live until they were twelve.

There were some scary times following the Trikafta boom, as we all endured the COVID-19 pandemic in 2020. We were fortunate to have just received Trikafta prior to this new virus. We were deathly afraid, but somehow managed to stay COVID-19 free for two years. When we did get it, thankfully it was very mild for us. We both got the flu and rhinovirus and fared better than we would have prior to Trikafta.

Our son Ryder is now seven years

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old and in the second grade. He is in his second year of Scouts. Watching him grow up is painful and joyous at the same time. It's just going by too quickly! We enjoy hanging out with him and talking about life. We moved into our little farmhouse on acreage in April of 2020. We can see beautiful mountain ranges (Pikes Peak and Sangre de Cristos) from our front porch. All of the years we saved and saved have paid off. We are both on disability now, and it can be hard at times, but we make it work and live rather frugally. We have one horse, one labradoodle, Stormy, and a new poodle puppy named Trooper. We had goats for a bit, and that was crazy but fun! Our "first born" Moogie, our 11-year-old husky, passed away in September of 2022. He is missed every day, and this year has been difficult, especially for my husband.

Ryan stays busy with ranch chores, fixing our vehicles that forever seem to have problems, and solving all of our IT issues. He is still very active and hasn't gained a whole lot of weight on Trikafta because of it. I, on the other hand, have gained 30 pounds! It is uncomfortable to me and I am working on body image issues. However, I am still incredibly grateful. I would rather be a little fluffy but alive! I have become a pretty decent cook and baker. I enjoy entertaining our friends and family and filling their bellies.

I am rarely hospitalized now; usually just once a year, and almost always after a virus, not just a CF exacerbation. I decided to go back to "school" last summer to obtain my paralegal certificate. The Claire's Place Work Proudly program sponsored me in an online program with Boston University. It was a 14-week long program. I loved it and hope to get a part-time remote job in the near future. I am working out the kinks with how to keep my medical benefits and go back to work without risking my health.

We are sort of lost on what to do next due to this drastic change in our lives. We may actually have to plan for living a long time now! It is not something we are set up for. We still have to do breathing treatments, and CF is still a big part of our lives. However, we get much more time to live "normally" now than ever before.

Becoming CF grandparents is a reality that could very well happen now, even if those grandbabies are fur babies. Ryder says he's going to have lots of dogs and no kids, so we shall see!

This article is for Noah and Tabby. Noah passed away the summer of 2018 waiting for a double lung and liver transplant. He was only 10 years old. I will forever be thankful to you for inspiring your mother to get involved, as I would not have my little boy or my husband without you. I hope my faith will someday be as strong as yours. ▲

Amanda Boone is 40 years old and has CF. She lives in Colorado Springs, CO, with her husband, Ryan, who also has CF. They have a son, Ryder, and two pups, Stormy and Trooper.

TILLMAN continued from page 13

disease in persons with cystic fibrosis (CF). Despite this knowledge and prescription of daily vitamin D supplementation for persons with CF, vitamin D deficiency has remained common in adult and pediatric cystic fibrosis clinics. Investigators found that stoss vitamin D dosing can be successfully implemented in routine cystic fibrosis care and increases vitamin D levels in and vitamin D deficiency.
https://tinyurl.com/3ekwrt9b

Bisphosphonates For Osteoporosis In People With Cystic Fibrosis

Around 23.5% of people with CF experience reduced bone mineral density (BMD), commonly known as osteoporosis, which increases the likelihood of bone fractures. The short-term and long-term effects of fractures (e.g. ribs and in the spine) may make lung disease worse, and hospitalization more frequent. Bisphosphonates are drugs that increase BMD by slowing down how fast bone is resorbed. This current review of the literature found that bisphosphonates consistently increased BMD in adults at the lumbar spine and hip regions. Treatment with bisphosphonates did not appear to reduce the rates of fractures (either in the spine or elsewhere) or deaths in adults. Severe bone pain and flu-like symptoms were commonly linked to intravenous bisphosphonates, especially in people not using corticosteroids. The reviewers concluded that additional trials are needed to determine if bone pain is more common or severe (or both) with the stronger drug zoledronate and if corticosteroids lessen or prevent these adverse events. Additional trials should also assess gastrointestinal adverse effects in the stomach and digestive tract which are linked to oral bisphosphonates. Trials with larger numbers of participants with longer follow-up are needed to show how bisphosphonates affect fracture rate and survival.
https://tinyurl.com/ybbwh8pc

Position Paper: Models Of Post-Transplant Care For Individuals With Cystic Fibrosis

There is no consensus on the best model of care for individuals with CF to manage the non-pulmonary complications that persist after lung transplant. The CF Foundation virtually convened a group of international experts in CF and lung-transplant care. The commit-

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Surrounded by Angels

Angels illuminate my soul with faith and my heart with hope.
My life has been filled with Angels.
It seems they have always been here.

I've seen invisible Angels and heard their gentle whispers guiding me
along my way. I've felt their nurturing presence in times of joy and in times of despair,
when I needed a friend and no one was there.

Surrounded by Angels, I am humbled.

Angels are beings in the known and the unknown.
I have encountered and known them in many forms:
family, friends, teachers, healers, and through the kindness of strangers.

Throughout my life, my Guardian Angels have been an integral part of my journey.
They encouraged me to attend a party where I met my future husband, Rahim.
They led me to China where we adopted our daughter, Maleka Rose Mau-my favorite "Angel on Earth."

Surrounded by Angels, I am blessed.

My Angels have had a full-time job watching over me and protecting me.
During the twenty-six months anticipating a double lung transplant, they consoled me with the confidence that I would survive.

November 5, 2000: I felt an abundance of Angels—
From the hospital åcall, the drive to Stanford,
during the hours of waiting, and into the surgery.
They were with the surgeons in the operating room, helping us all.
Angels were stationed at my bedside in the Intensive Care Unit
Two months of constantly holding me in their prayers, strengthening my will to live.

PHOTO BY RAHIM MAU

Surrounded by Angels, I rejoice.

An Angel resides both in Heaven and within me.
He lives on in my body with every breath I take.
I am eternally grateful to him and his generous family, for giving me a precious gift, and for teaching me the true meaning of "The Gift of Life."

Surrounded by Angels, I am alive.
Thank you God, for surrounding me with your Angels.

— N. Mau, 2003

FROM OUR FAMILY PHOTO ALBUM...

PHOTO BY AMANDA ROWE

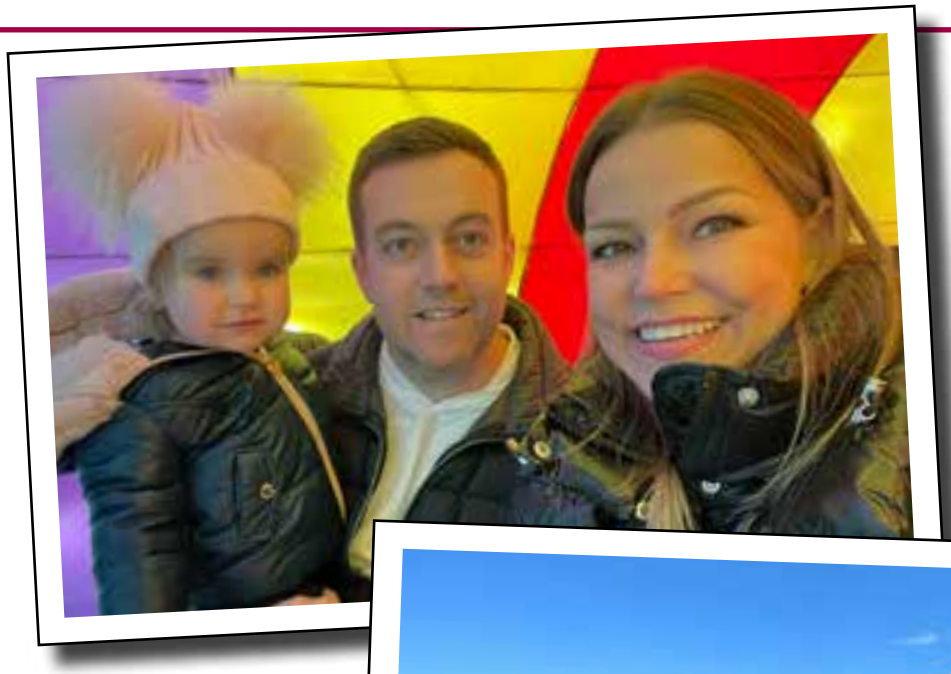
THE KOWAL FAMILY: NICOLE, MICHAEL, AND THEIR SON, ERNIE.

JES DAVIS

SUZANNE JOYCE PARTICIPATING IN BEACH YOGA, REDINGTON SHORES, FLORIDA.

PHOTO BY MARIJO SCHUBERT

THE BOONE FAMILY: RYAN, AMANDA, AND THEIR SON, RYDER, WITH THEIR DOGS, MOOGIE AND STORMY, AT THEIR RANCH NAMED ROCKIN' RLB RANCH, COLORADO SPRINGS.



FROM LEFT: RILEY, JOEY, AND KATY MONTE.

JACOB GREENE IS STANDING ON THE GOLDEN GATE BRIDGE WITH HIS GIRLFRIEND, JACLYN HODGSON, AS THEY TOUR AROUND THE CITY WITH FAMILY THE AFTERNOON BEFORE JACOB'S MEDICAL SCHOOL WHITE COAT CEREMONY.

RACHEL JOHNSTON HIKING TOWARD MOUNT EVEREST BASE CAMP.

ROSE, ARDEN, KATHERINE, AND MAGGIE LOCKWOOD.

Part II: Saving A Life On Everest

By Rachel Johnston

Editor's note: The story below is part two of Rachel's two-part story depicting her climb to Everest Base Camp four years ago. Part one was featured in the Winter 2023 issue of *CF Roundtable*.

Rachel worked in an ICU for two years and, in 2017, quit her job, sold her car, put all her things in storage, and left to travel the world. She joined a missions organization called Adventures in Missions and went on a mission trip called "the world race." This took her to 11 countries in 11 months (El Salvador, Guatemala, Honduras, Nicaragua, Côte d'Ivoire, Ghana, Nepal, India, Thailand, Malaysia, and Indonesia). One of those countries was Nepal—where this adventure took place—at the time she was 26. That year was life-altering for her. She got sick several times and really questioned God's goodness and plans. But she had no idea He would take her to the heights He did. She continued traveling for another nine months with a different organization called G42, with whom she spent six months in Spain and three months in Iraq, working in a refugee camp and underground church.

Day 6:

As soon as I saw him, I knew. "Can you come in here please? He's making this weird, scary noise that I've never heard before." Before I heard it, I already knew. In critical care, it's called, "*the death rattle*." The all-too-familiar sound pierced my heart. So many times before I had heard this sound and not let it permeate me. This time, though—this time I felt it. Deep. In my very being. This man was going to die.

***

I woke up that morning after a hellish night. The enemy left me absolutely and utterly defeated. I woke up anticipating having to announce to my teammates that I could no longer go on and to go ahead without me. I was preparing myself to spend three to four freezing nights here in this small, abandoned tea lodge at 4,400 meters completely alone.

Today was another acclimation day—a shorter hike than our first, but

RACHEL JOHNSTON AT MOUNT EVEREST BASE CAMP.

grueling, nonetheless, or so I had heard. I stayed behind to rest. I lay in my bed most of the morning, just staring at the wall and trying to use my human comprehension to decipher just exactly what God's plan was in having me come on this trip, only to fail halfway. Perhaps I had made the wrong choice coming in the first place. Maybe my expectations for this adventure with God were too high. I was lost—lost in a sea of confusion and condemnation, drifting farther and farther away from God.

I was still lost in my head when my teammates came back, and we all sat down together in the lodge for lunch that afternoon. I was trying to gather my words to tell them I would not be able to continue with them, when one of the trekkers we had met on the trail came into the small, sunlit dining room and interrupted us. "Hey, Rachel, you're a nurse, right?" Oh, gosh, I've heard this question before. Usually it's preceded by a "Could you look at this weird spot on my (insert awkward body part here)?" or "So I've had this weird rash." Reluctantly I said, "Yes...?" "Well, there's this guy downstairs in the next lodge who isn't doing so well. I thought you might be able to take a look at him? He's not really responsive." Any form of the word unresponsive is an immediate red flag to a critical care nurse. I left the remains of my cup of hot chocolate and moved quickly toward the door, leaving my jacket. As I followed him down the stairs, out the front door, and toward the next lodge, I immediately started praying.

February is the very beginning of the season on Everest. Even though it is "technically" in season, the villages are still ghost towns, the weather is cold and unpredictable, and any form of medical treatment is nonexistent.

Working in the ICU in Oregon, I have never dealt with altitude sickness or been exposed to it. The little research I did about it before I left, I knew that, at its very worst end stage, it evolved into cerebral and/or pulmonary edema, both of which I am very familiar with in the ICU.

I walked toward the sick man's open room door. There were several people gathered outside. He was lying flat on his bed, in his many layers. I smiled and met his gaze through his half-opened eyelids.

I immediately snapped back into my nurse mode, which had been sitting dusty on the shelf for the past seven

months. I sat next to him and grabbed his hands. They were cold. As I asked him, "How are you feeling?" I reached for his pulse. It was fast and irregular. He responded with a few simple words in English but it was clear that he didn't speak much English. A woman we had been trekking with spoke English and knew Chinese as well, which just so happened to be what this man spoke. Through translation and looking through this man's bag and wallet, we discovered that he was Chinese, travelling all by himself, without a guide, porter, or insurance. He had arrived in Lukla a day after us and had arrived here in Dingboche three days ago. A trek of 1,760 meters or almost 6,000 vertical feet that had taken us five days he had done in just *two*. The only medicinal treatment for altitude sickness is Diamox, which I was able to get him to swallow before he became extremely lethargic. But the only *real* treatment for mountain sickness is to descend in altitude and fast.

I walked outside his room to talk with the people who had gathered, including the owner of the lodge. They were discussing what to do. Most were under the impression that because he didn't have any insurance or money, there was nothing that could be done. That's when the woman came out of the room and said, "Can you come in here, please? He's making this weird, scary noise that I've never heard before." My heart began to beat faster as I quickly went back into the room and got the man into a sitting position, maintaining his airway. He had developed flash pulmonary edema. He was drowning in his own lungs, gurgling evident with every breath.

My take-charge critical care nurse hat had to be put on and I knew it. I walked out of the room back to the group arguing outside. I interrupted them and, as politely as I could, let them know that this man was going to die, and soon, if he didn't get down in elevation. He didn't have time to be carried down the mountain; it would take hours to get to the next village and, even then, there were still no medical facilities. He needed a helicopter evac stat.

Thankfully, while I was in the room, another man fought very passionately to get the owner of the lodge to finally call for the helicopter. It would come from Lukla and would be here in 30 minutes. There was a slight glimmer of hope. *Okay*, I thought, *30 minutes, that might be just enough time.* My heart plummeted when, five minutes later, the lodge owner came in and said there were no helicopters in Lukla and rescue had to come from Kathmandu, a two-hour flight. There was only one option: prayer. A sense of urgency rushed over me as I showed the woman who had been translating for us how to keep the Chinese man in an upright position in order to maintain his airway. I hurried out the door and started toward our lodge. I was on a mission.

> "Maybe my expectations for this adventure with God were too high."

"The wonder and mystery of prayer is God waits until someone asks."—Billy Graham

"Prayer moves the arm of God."—Doris Johnson

It was time to gather the troops. As my team filed into the tiny room, I was anxious about what God might do. I simultaneously felt anxious, nervous, excited, and scared As we laid our hands on his limp body and began to pray, a feeling of peace superseded all of those emotions. The next two hours were the longest of my life. I was counting the minutes, giving oxygen via a spray can-looking device that someone had found buried away in a room, trying to keep this man conscious and breathing, and constantly had two fingers on his pulse, waiting. Just waiting for it to stop.

As a nurse in general you must be a multitasker, but as a critical care nurse, those skills must be even more fine-tuned. My nurse brain was on overload. There was nothing left that I could do to help this man. All I could do was keep him upright to prevent him from drowning in his own lungs. I knew it would be horrendous if his heart stopped and I had to perform CPR on this man, here, in the mountains all while having a crowd of onlookers. I also knew that CPR would have no effect if I couldn't get an airway, which was impossible to do since his lungs were filled with fluid.

After what felt like an eternity, the lodge owner finally walked in and announced that the helicopter would be here within minutes. Before I even knew what was happening an old, tiny Sherpa had picked up the man and put him on his back and began carrying him to the "landing pad," which is essentially a tiny pile of flat rocks on the edge of a cliff.

My anxiety went into overdrive when I saw that the man was now lying flat and face first on the man's back. I knew that we had to get this man to the helicopter, but I also knew that now all the fluid in his lungs would obstruct his airway.

We reached a large boulder next to the landing area and got the man into a seated position again, with the help of several people, as he was now dead

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The Miracle That Was Unexpected And Unplanned

By Nicole Kowal

On Saturday March 5, 2016, I married the man of my dreams, Michael Kowal. And from that point on, we tried for a baby. After months of nothing happening, we looked into infertility treatments and other possibilities to conceive our own little human. At the time, surrogacy was not allowed in the state of New York and adoption was insanely expensive. Some agencies I spoke with had a hard time adopting out to someone who had a "terminal illness." Michael got tested to see if he carried a faulty CF gene. Luckily he does not, but, either way, we were going to keep trying. On December 7, 2017, I had a hysteroscopy, dilation and curettage (D and C), polyp removal, and endometrial ablation. The doctors opined that if we were going to become pregnant on our own it would be shortly after this surgery. Within three days after this surgery, I was diagnosed with cystic fibrosis-related diabetes. During all this, my sister and her daughter (our niece) lived with us. Gabby is our niece whom we treated as our own; she is also our God Child. We did everything with our Gabby—she was our child in any work function and life event to which we were allowed to bring kids. We would pretend she was ours at all times and snuggle her and love her more than she would ever know.

I was feeling defeated after all these months and issues of not being able to create life with the love of my life. We had a massive four-bedroom house that we had this feeling we would never fill. As much as we didn't want to give up about expanding our family and carrying on my husband's family's last name,

NICOLE AND ERNIE KOWAL.

PHOTO BY AMANDA ROWE

it just did not seem to be happening. We listed the massive house in search for a smaller home, and no longer cared about the amount of bedrooms because we gave up on becoming parents. In August 2019, we bought our forever home in a very small community known for its corn, cows, and tractor pulls. We love where we live and our new one-bedroom home with its two and a half bathrooms (have to have enough bathrooms with CF tummy).

Along came Trikafta in November 2019—that medication made me feel absolutely amazing. "The Purge" certainly happened and with EVERY SINGLE ORGAN of mine. It was amazing how outstanding I felt, how much life became better! But, here came COVID during March of 2020. And I was mandated to stay home until further notice, but the story did not end there: It became very interesting.

On April 10, 2020, I knew something was off. I knew somewhere in the mass of old bathroom items I had an expired pregnancy test. I took that test while my husband was working out, and it came out with an answer I never thought I would see: "PREGNANT." I was shaking, excited, terrified, and shocked. I walked into where my husband was working out and told him to get up now, and then I showed him. We both didn't believe it, and every day for four days he brought home different pregnancy test brands for me to try; all of them turned out to be positive. After I called CF clinic, they suggested this would be happening as Trikafta cleans out EVERYTHING, making people more prone to becoming pregnant. So here we were in our newly purchased one-bedroom home: After mentally being okay with never having children and just being amazing aunts and uncles, we were now being upgraded to parents.

All because of Trikafta, our little miracle happened, when we least expected it and weren't even thinking of it. We were pregnant during a very stressful time with COVID. My husband was not allowed to come to any of my doctor appointments or FaceTime me while I was there. It was very lonely at some times and at others so exciting. During my pregnancy I had preeclampsia and my pregnant belly was always showing more than what's considered normal at each week during my pregnancy, probably due to the diabetes. On Saturday November 14, 2020 my doctor advised that my blood pressure was too high and to drive to the hospital, so I

drove myself and my husband to the hospital. We got there at about 11a.m. and I began getting treated for stroke. Our miracle man Ernest ("Ernie") was born at 2:53 p.m. that very day, perfectly healthy and to the song "Don't Stop Believing" by Journey (our doctor asked what music we wanted on during the C Section). We were able to both add a room to our home and add this amazing gift with no change in my health. And Gabby was under the impression Ernie was now her little brother.

Of course, the thoughts of my CF interfering with being a good parent crosses my mind all the time, but I also have Ernie "help" Mommy do things to make sure I stay on top of my own health. He helps with scanning my CGM and holds my nebulizer when I do it. I fully believe in talking about Mommy's cough, potty breaks with him, and how being "sick" means we need to rest. Being open has helped me be the person I am today and the parent I have become. Although right now things are becoming different, as toddler years are a whole new ball game, I am forever grateful for our miracle man and all of life's precious moments.

To anyone out there looking to adventure down this road, be yourself and own your issues. Let your doctors know your stance on life and your hopeful adventures and desires. Tell your loved ones your plans and goals. And truthfully just be prepared for whatever happens, because life will most likely not go the way you expect but turn out much better than you ever imagined. I wouldn't change a thing that happened to us. I love every aspect of our journey and I would love to talk to anyone about what theirs could be. ▲

Nicole Kowal is 35 years old and has CF. She lives in Buffalo, NY, with her husband. Her email is: abnormalnicole@gmail.com.

FOCUS TOPIC

CF AND USING MODULATORS

Trikafta Letdown

By Suzanne Joyce

I've been anxious most of my life. I'm anxious right now while writing this piece. I had no idea that this anxiety would skyrocket as a result of taking the drug I'd been waiting to have for 57 years.

I started Trikafta in 2020 in the hopes of seeking the miraculous experience of clear sinuses. I was one and a half years post-transplant at the time. If my pancreas or GI system also improved, that would be gravy. My sinuses were a mess and sharing *Pseudomonas* with my new lungs. To begin Trikafta I had to first switch from one anti-fungal to another, which, in turn, required that I increase my daily dose of tacrolimus, a nonnegotiable immunosuppressant, one of many post-transplant drugs.

Within days, my sinuses were wide open, my sense of smell increased, and I was happily somewhat stunned. During the course of the first three months, I also noticed a change in my GI system: Some of my symptoms were gone but replaced with different ones. My need for insulin had begun to decrease a bit.

SUZANNE JOYCE

After several weeks on Trikafta, my anxiety slowly intensified. Situational anxiety became unbearable. I'd experience panic attacks—having that end-of-the world feeling. I increased my exercise and mindfulness practices in order to get through the day. It appeared that the combination of Trikafta with my transplant meds was making my anxiety escalate significantly. After three months it was obvious I had to stop taking Trikafta. Clear sinuses and using a bit less insulin could not outweigh living in a crazy wound-up mind.

While my clinic has left it up to me to decide to try a modified dose, I am afraid to try it. I know others have had success with a modified dose but I'm hesitant. I'm afraid I won't get enough of the benefits and that my existing anxiety will just increase. If anyone else has dealt with anxiety issues and tried the modified dose, I'd like to connect with you. ▲

> "It appeared that the combination of Trikafta with my transplant meds was making my anxiety escalate significantly."

Suzanne Joyce is 60 years old and has CF. She lives in Clearwater, FL. She has been enjoying blues and rock music at outdoor venues and exploring nature in Florida. You can email her at exesq1@gmail.com.

Speakers Bureau Updates

Speakers Bureau members Isabel Stenzel Byrnes, L.C.S.W., M.P.H., and Xan Nowakowski, Ph.D., M.P.H., gave a team Zoom talk for the Delta Phi Epsilon sorority's cystic fibrosis service group at St. Norbert College focusing on diverse experiences of CF as people live and age. Isa and Xan shared stories from their own lives with CF, highlighting how even though both are multiethnic people from multiracial backgrounds, they have had many differences in their journeys as well as similarities. Both speakers then answered questions from participating students about health disparities in the U.S. CF community.

To inquire about our speakers for your events, go to: www.cfroundtable.com/speakers-bureau.

weight. I quickly pushed away the blanket that was covering his face and reached for his pulse again. Still fast and irregular. But I could no longer hear his gurgling because he was barely breathing—yellow fluid from his lungs was leaking from his mouth.

My mind went back to the thought of having to perform CPR on this man. He was barely holding on. I couldn't believe he had made it this far, still alive. As I gave him the rest of the oxygen that was left in the can, my eyes were glued to his chest, watching it sporadically and faintly rise and fall, waiting for it to stop at any second.

I have never been happier to hear the sound of helicopter propellers in my entire life. As we watched it fly toward us, my heart could start beating again. It stopped for a brief second as the helicopter flew next to us and then continued to fly on past us. We looked on in disbelief. Someone quietly uttered the words we were all thinking: "Is that one not for us?" In that instant, all my hope vanished. This man's fate was sealed. You know that saying that goes something like, "You wouldn't be able to fully appreciate joy if you didn't know sorrow"? I felt the utter despair of sorrow and the overwhelming joy of hope all in the same 10-second time period, as the helicopter began to make a wide right turn, back toward us. Instant. Tears. That's never happened to me before. I didn't know where they had come from or how they had gotten there that fast, but I couldn't stop them. This man was going to *live*.

The helicopter was on the ground for no longer than two minutes. We picked the man up and carried him the last 20 feet to the helicopter. I tried not to trip over the huge, sharp rocks as I cradled his heavy, dead-weight leg. The wind from the mountains combined with the helicopter was overpowering as we approached the door. As soon as it opened, my heart instantly sank. There was nothing inside, only a bench seat.

> "God stepped in, and I didn't even have to ask Him to. A beautiful assurance came over me as God let me know that the man was going to be okay."

There was no medical equipment of any kind, no IV bags, no AED, nothing. Not even a heart monitor. I doubt there was even a first aid kit on this helicopter. In under 60 seconds he was up in the helicopter and the doors were closed. The winds were even more fierce and we were forced to turn our heads away from the force. I turned just in time to see the helicopter disappear from my sight as it dove down in elevation as quickly as it could, vanishing behind the cliff on which it had just precariously landed.

I hadn't even taken two steps before the enemy attacked me again. *You could have done more. He won't make it another two hours without medical treatment; it was all in vain. Maybe if you had prayed a prayer with more faith, God would have healed him.*

I shared my disappointment about the lack of medical equipment on the chopper with the man with whom we had been trekking. And as matter-of-factly as any medical professional should have known, he said, "Well, now he's going down in elevation and that's the most important thing."

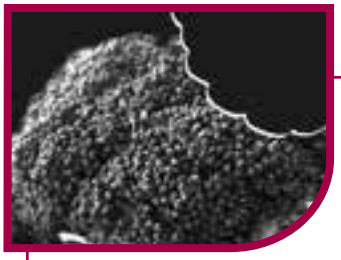
I locked myself in my tiny, cold lodge room. As I sat there on my single bed, staring at the wall, trying to process all that had just taken place, God stepped in, and I didn't even have to ask Him to. A beautiful assurance came over me as God let me know that the man was going to be okay. He then allowed me to see the situation through His eyes, instead of through the lens of the lies of the enemy. He saved this man's life. Just because it wasn't through an instantaneous miracle, it didn't make it any less miraculous. All the pieces fell into perfect alignment as every single person on that mountain that day played a role. An English-speaking Chinese translator, an ICU nurse, and a team of prayer warriors, all together in an almost abandoned village on the doorstep of Mount Everest—God chose to use us to save this man's life.

"Christ has no body now but yours. No hands, no feet on earth but yours.
Yours are the eyes through which he looks compassion on this world.
Yours are the feet with which he walks to do good.
Yours are the hands through which he blesses all the world.
Yours are the hands, yours are the feet, yours are the eyes, you are his body.
Christ has no body now on earth but yours."—Teresa of Ávila

What a grand and marvelous mystery that the God of the universe not only allows, but deeply desires to include us in all His ways, in all His *adventures*. His love truly is extravagant. I got an even bigger picture of this grace-filled, awe-inspiring, just downright outrageous love two days later when I finally stepped foot on Everest Base Camp.

We also found out on our way down the mountain, that the Chinese man had made it to a Kathmandu hospital alive and he was expected to make a full recovery. ▲

Rachel Johnston is 32 years old and has CF. She currently lives in Oregon with her husband. She has traveled all over the world as part of various mission trips. She came back to the United States in June 2019 and got married in November 2021.



CULINARY CORNER

Creamy Lemon Orzo With Asparagus And Peas

By Maggie Williamson

Spring is here and, even though the days are longer, we may still have that transition time between cold and warmer days. There is a bit of comfort here with the orzo and cream, but the lemon, asparagus, and peas bring in some fresh elements. This is a vegetarian dish, but you can add roasted chicken breast or thighs for protein. I call this dish the quick version of risotto. You do not have to stand over your stove stirring for 30 minutes but you get a similar result to risotto with the use of orzo. This one-pot dish comes together pretty quickly, which is great when you aren't feeling your best, but still want a healthier option besides takeout or a frozen meal. The key to this recipe is to prep everything before you start cooking. If you do that, all of it comes together quite quickly in the end. I have put all prep work in the ingredient list to give you a head start.

Creamy lemon orzo with asparagus and peas

Yield: 4 servings
Prep time: 10 minutes
Total time: 30 minutes

Ingredients:

2 tbsp olive oil
1 medium to large onion, finely chopped
2-3 garlic cloves, minced
1 cup of orzo
2-3 cups of vegetable or chicken stock
1 lemon, zested and juiced
½ cup frozen peas
7-8 asparagus spears, ends chopped off and cut into half-inch pieces
½ cup grated parmesan cheese
¼ cup heavy cream
1 tbsp chopped fresh herbs (dill, parsley, thyme, or basil all work)
salt and pepper to taste

Preparation:

Step 1:

On medium heat add olive oil in a medium pot. Add onion and a pinch of salt. Sauté for 5 minutes or until the onion is soft. Add chopped garlic and cook for another minute.

Step 2:

Add orzo and stir until all orzo is coated in oil. Add 2 cups of stock and cook for the number of minutes it says on your orzo package. I recommend cooking until al dente.

Step 3:

As the orzo nears the last 3-4 minutes of cooking, add peas and ½ inch asparagus pieces. Add zest of 1 lemon and its juice. Add the parmesan cheese and stir. Test to make sure your asparagus is cooked through after four minutes or continue cooking until you can pierce a fork through it. If you notice the liquid evaporating too quickly before the orzo is done cooking, add a bit more stock to ensure the orzo cooks and the dish doesn't become dry.

Step 4:

Add salt and pepper to taste and pour in the heavy cream. Stir to combine and cook for another minute to let the orzo soak in the cream just a bit. Top with fresh herbs of your choice. ▲

Maggie Williamson is 35 years old and has cystic fibrosis. She received a double lung transplant in 2014. She now lives in the U.K. with her British husband, Tom, and their Bengal cat, Charlie. You can find her and all of her cooking delights on Instagram @justasprig

MAGGIE WILLIAMSON

Meet A New Director: Jacob Greene

Jacob Greene is a 24-year-old medical student with cystic fibrosis and is our newest member of the USACFA Board! Born in Tigard, Oregon, Jacob was diagnosed with cystic fibrosis at birth when he was treated for meconium ileus. He grew up in Seattle, Washington, Seattle Children's Hospital served as his home CF center throughout his childhood. In junior high and high school, Jacob was involved with robotics, Boy Scouts, his school's track and field and wrestling teams, and did medical research at Fred Hutchinson Cancer Research Center. These experiences, primarily his lived experience with CF and work at Fred Hutch, piqued Jacob's interest in pursuing a career in medicine. In 2017, Jacob moved to California to start college at Stanford University. At Stanford, Jacob did neurology research in the Monje and Gibson Labs. This work led to Jacob being published in the journals *Cell* and *Neuron*. Jacob also volunteered with Stanford Health Care's Cardinal Free Clinics and was a Residential Assistant in Roble Hall. In 2021, Jacob graduated from Stanford with a Bachelor of Science with Honors in Biology. Following graduation, Jacob worked at Genentech in the Research and Early Development group working to develop drugs for neurological diseases like Alzheimer's and multiple sclerosis. This work led to a publication in *Glia*. In July of 2022, Jacob left Genentech to begin medical school at the University of California San Francisco (UCSF) School of Medicine. At UCSF, Jacob is a co-coordinator of HealthLink—a program for high school students interested in medical careers. He also serves as the UCSF School of Medicine delegate to the Medical Student Section of the American Medical Association, sits on the Student Medical Education Council, and does research in the Jonathan Pan Lab. Jacob joins the USACFA Board of Directors after having written for the *CF Roundtable* publication and participating as a panelist in our scholarship application webinar in December. We're thrilled to have Jacob on our board! ▲

JACOB GREENE

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tee reviewed literature and shared the post-lung transplant model of care practiced by their programs. The committee then developed a survey that was distributed internationally to both the clinical and individual with CF/family audiences to determine the strengths, weaknesses, and preferences for various models of transplant care. Discussion generated two models to accomplish optimal CF care after transplant. The first model incorporates the CF team into care and proposes delineation of responsibilities for the CF and transplant teams. This model is reliant on outstanding communication between the teams, while leveraging the expertise of the CF team for management of the non-pulmonary manifestations of CF. The transplant team manages all aspects of the transplant, including pulmonary concerns and management of immunosuppression. The second model consolidates care in one center and may be more practical for transplant programs that have expertise managing CF and have access to CF multidisciplinary care team members (e.g., located in the same institution). The best model for each program is influenced by several factors and model selection needs to be decided between the transplant and the CF center and may vary from center to center. In either model, CF lung transplant recipients require a clear delineation of the roles and responsibilities of their providers and mechanisms for effective communication.
https://tinyurl.com/ywk4cad8

Living Near Composting Sites May Increase Risk Of Worse CF For Adults

Living closer to composting sites, where solid urban waste is collected and stored, may increase the risk of more severe disease for adults with cystic fibrosis (CF). Close proximity to such sites was linked with worse lung function and more fungal infections among adults. The study also noted a higher risk of pulmonary exacerbations, or periods of sudden symptom worsening, in this patient population. Notably, no

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TRANSPLANT TALK

Call Me A Skin Cancer Incubator—A Cautionary Tale

By Andrea Eisenman

Every time I turn around, I have a new skin cancer. Just a side-effect of post-transplant life, which is mainly due to being immunosuppressed in order to keep my non-native lungs from being attacked by my body's roving "security" system. And because my immune system is not as active as non-transplanted people's, I run the risk of infections, fungus, and cancers—mainly skin cancer (although, I have had lymphoma, but that is for another time).

Alright, full disclosure: I ran around as a child on the beach with no hat on—I hated them. And I was raised by sun-worshipping Austrians who had light eyes, fair skin, and constellations of freckles encompassing their exposed skin. Nobody knew the harmful damage that sun exposure could wreak on people later in life. I was the first generation in my family to use sunscreen instead of baby oil and mercurochrome while out in the sun.

Not surprisingly, my family had a history of skin cancers for the obvious reasons. I sometimes went with my mom and grandmother for their skin check appointments. This was not foreign to me. I saw them get skin cancers removed almost monthly. They wore the white bandages from lesion removal by scalpel or freezing off by liquid nitrogen. Then Mohs surgery was developed and widely put to use. This new technique was developed by Frederic E. Mohs, M.D. Mohs surgery improves patients' outcomes by ridding them of all remaining cancer in one surgical visit instead of having to repeatedly come back to get more tissue excised. The Mohs procedure focuses on just the cancer cells—instead of taking wide and deep swathes of skin, only cancerous growth with a clear margin around it is carefully removed. The area is stitched up rather than leaving a gaping hole to slowly heal. Before Mohs, the sites where I had many skin cancers removed would resemble a gunshot wound because excess skin was removed as well as the cancer. I have one of these gems on my shoulder from a squamous cell carcinoma removal.

ANDREA EISENMAN THREE MONTHS AFTER SURGERY.

From my early sun exposure and my immune status, I had four Mohs surgeries on my scalp about ten years post-transplant, one on my ear, and others removed without Mohs. Thankfully I was introduced early on to a talented Mohs surgeon who now does all my skin cancer surgeries. She or my regular dermatologist can biopsy and confirm the skin lesion is either basal, squamous, or melanoma. However, Mohs is not used for removing melanoma—doctors still use the old "dig deep and gouge it out" technique. I only had one of these procedures, thankfully. Otherwise, I see a regular dermatologist who is aware of my immune-compromised situation and she knows I need a skin check every three to four months to get things in their nascent stage. She will biopsy suspicious skin lesions or moles that look like they are changing shape or color. Once the report returns as positive for cancer, I see my dear Mohs surgeon.

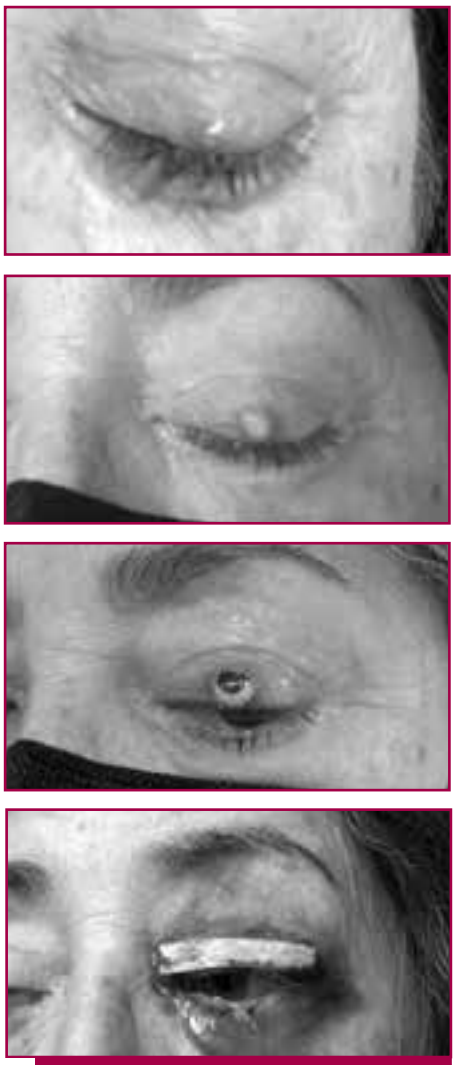
The next part of this story might be upsetting to some. It still freaks me out and I went through it. I am usually the one who notices a bump or an elevated area of flaky skin and I go to get it biopsied. I saw a dreaded bump on my eyelid right near the lash line. I tried to get in to see my regular dermatologist but, because she changed hospitals, I was considered a new patient and could not get in quickly. I did get in quickly to my Mohs surgeon. I showed her the small bump and she said it is probably nothing, a benign growth, but because it is you, we should be thorough and biopsy it. The worst part was the biopsy process. She had to inject my eyelid with lidocaine to numb it to slice off the smallest area to send to the lab. I apologized and told her that I am fine with other parts of my body being cut, jabbed or sliced, but get near my eye and it is going to be a fight. So, I was quite squirmy as she drew near with the dreaded lidocaine syringe. She assured me she is way worse when it comes to her eyes, and her nurse nodded in agreement. It was nice to be understood. I tried to breathe deeply through it all.

I learned with relief it was a benign

growth. However, after I finally saw my regular dermatologist, she looked at it and the bump was still slightly there and flaky so she sprayed it with liquid nitrogen, which was not pleasant to say the least. About five days later, the scab that formed from the nitrogen freezing fell off. It was finally smooth. Unfortunately, about two months later, the bump was back as if nothing had been removed, frozen, or biopsied. I thought, no way, not cancer. I just could not deal with the thought of having to get another injection on my eyelid so I let it go for a week. Then, it started getting bigger. A white head formed on it and it kept getting bigger. I was asking people what they thought it was the entire time. Many thought it was a cyst. I thought, ok, so this can be lanced and I will be fine. I emailed my dermatologist and was eager to see her because in 15 days, the "cyst" had quadrupled in size.

Part of me dreaded investigating this as I feared it would require another eyelid injection or I'd develop an infection from the cyst bursting. Way far back in the depths of my brain, I feared cancer. "THIS IS CANCER!" it screamed. I could not see my dermatologist for two weeks and when I finally got there, she said, "I cannot touch that. You need to see an ophthalmologic surgeon or a Mohs surgeon." She said I should go see the surgeon who does my Mohs surgeries. I called her office and sent photos of my eyelid via text. They saw me an hour later and biopsied it. She said it was not a cyst and probably a basal or squamous cell carcinoma and that I would not only need Mohs but also an ocular plastic surgeon to repair the eyelid after she removed the growth.

She recommended three different ocular surgeons. Even though she removed the majority of the growth for biopsy purposes, the cancer was growing back rapidly. It was very upsetting



FROM TOP TO BOTTOM: ANDREA EISENMAN'S LEFT EYE FROM BEGINNING TO THE DAY OF EXCISION.

and made me very afraid of what lay ahead. Once I had the biopsy report five days later, I started calling the surgeons. With some cajoling and telling the surgeon's office manager that the cancer is on my eyelid, that it was growing rapidly, and that I am immune suppressed, I was able to see him quickly. I had to go into serious self-advocator mode, meaning, aggressively pushy. The first doctor I met with was my Mohs surgeon's favorite. When I met him and heard his plan of action, I was certain he was the right surgeon. I got a good feeling about him when he declared, "This has to be removed ASAP." He put me on his surgery schedule for three days later.

It was then arranged with my Mohs surgeon's office. They fit me in the same day. I was to get Mohs on Monday morning at 8 a.m. and, once the margins were clear, run uptown to get the plastic surgery in the operating room. I reported early to my Mohs appointment. I knew it would be several hours and passes of the scalpel to get this area cancer free. Because I was not going to be sedated for this process, I took an Ativan to calm myself. My face was draped. A giant opaque contact lens was placed in my eye. She numbed me and then she began cutting. What she cut was then sent to her lab on the premises to check for cancer-free margins. This happened two more times and the third time was the charm. Even though I was numb, on the last round, I heard the scissors clip and knew it was a section of my lashes. It was 10:30 a.m. and I was bandaged up with sight only in one eye.

I could not eat anything that morning because I was going to be sedated for my second surgery—thank goodness! I was brought in to the operating room rather quickly, saw the ocular plastic surgeon and his associate, who explained they may need to do a skin graft, which they would take from behind my ear, most likely. The Mohs surgeon had sent him a photo of my eyelid so he knew what he was dealing with before I got there. My surgery was going to take two or three hours due to the skin graft.

As luck would have it, since I am 58 and never had an eye lift, the ocular surgeon was able to take my extra (sagging) skin near my brow bone and move it to cover the area on my eyelid. No skin graft was needed. Because the Mohs surgeon had left a hole in my lid, the ocular surgeon removed a slice or

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IN THE SPOTLIGHT

With Jes Davis

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

Age: 33
Home: Oakland, CA

Readers, I'm thrilled to introduce my friend and fellow community advocate Jes Davis! I first met Jes through Andy Lipman's initial *CF Warrior Project* book and the social media groups he created for participants to connect with each other. Since then, we've stayed connected and worked together on several advocacy initiatives focusing on racial and intersectional justice within and beyond the CF community. When they're not organizing for Black Lives Matter, Jes continues to build their career as a professional actor and filmmaker. And they still find time to do an occasional modeling shoot, making still photos come alive with shining confidence. They maintain a lively and thoughtful social media presence across multiple platforms offering sneak peeks at their latest projects. Jes is a courageous advocate, a master storyteller, a generous friend, and a fashion icon. I jumped at the chance to celebrate their contributions to the adult CF community with an interview!

What should our readers know about you?

CF is like my partner in crime. I spent years demonizing it because that's how the world perceives disease. But if I die, the CF dies; it needs me to sustain itself. So it's kind of a working relationship. I don't know if other patients feel connected to that sentiment; to me it's stronger than thinking you have to destroy part of yourself. Society sees disease as something to conquer and defeat. For me the "defeating" aspect is seeing illness as a partner.

JES DAVIS SEEN AT A MARCH FOR ABORTION RIGHTS IN NEW YORK CITY, JULY 1, 2022.

That's how you stop it having such agency over your life.

How did your CF first get diagnosed?

I was three years old at diagnosis, even though I showed symptoms at birth. My CF wasn't diagnosed right away because of racism. My mother works in the pharmaceutical industry; she has a wealth of medical science knowledge. She recognized certain symptoms like fat intolerance and tailored meals for me and my brother accordingly. But most doctors we saw were white, cisgender, heterosexual men who didn't take kindly to being challenged by a Black woman. When my mom changed my brother's diaper in the clinic, the nurse recognized immediately that the smell wasn't normal. She'd been trying to tell them exactly this and getting dismissed! Obviously, it shouldn't be that hard to be heard so getting diagnosed was not a great experience. We were still met with racism afterwards, even from our CF specialist. Someone called Child Protective Services on my mom because my brother was struggling to gain weight—nobody had diagnosed his diabetes. The medical industry wants to blame Black patients rather than understand disease in each person. This kind of arrogance is rampant.

Can you tell us about your racial justice work?

My community organizing work hit a stride during the George Floyd protests. During 2020 I also connected with some CF patients who are also Black and generally of color, like some Indigenous and Latin folks who don't have Black heritage specifically. I heard their stories about being in doctors' offices, their relationship to the CF community, and having to explain the disease. There's still a lot of dismissal—not just from doctors, but from people in general—for Black people with diseases that aren't associated with our race. My activism emerged from the racism I endured in clinic and school. That emboldened me in taking more of a stance. People want to stifle movements because advocacy makes them uncomfortable. They're afraid of change, and of you as an activist. It's both a sword and shield when we are advocating for change. I'm always confronting people who are complacent—because complacency is complicity.

What are you working on right now?

The film that I'm producing right now is called *POV*. It's told from the perspective of a feminine person on a date with a masculine person. This is going to be purely from the feminine perspective because we always get the masculine perspective on how people relate to the dating world and those spaces. We're going to highlight how objectifying these perspectives can become. People don't realize how much porn influences the objectification of a lot of bodies, particularly more feminine ones. These same perspectives often appear in movies outside of porn itself, with things like shot angles and composition, because people respond to them. We wanted to do something that flips this dynamic and shows how a masculine person can also be objectified by a feminine view. I like watching porn and find empowerment in saying that openly. A core goal of the film is empowering femmes in a sexual space while also seeking accountability from mascs.

How did your acting and filmmaking career get started?

Parents fear sickness from other kids—even if someone's cough isn't contagious. Having that isolation forced on me, I embraced being alone so it became enriching. My brother also has CF; I had some camaraderie. But I've always loved watching movies because I didn't need friends to do that. Movies became a coping mechanism for CF exacerbations or depressive symptoms or anything else. I moved into acting and writing as well; when I was a kid I did a Little Caesar's pizza commercial. My love of performing came from these early experiences where I could "take off" my CF and dive into a different reality. Over time I became passionate about using my body to create. This helped me see it wasn't broken or damaged just because I had a chronic illness.

What drew you to working in the arts?

I acted out the death scene from *Bambi* as a kid. Having CF means confronting your mortality at a very early age. When you have a chronic illness you get told you're not invincible—and that you could die, possibly soon. This drew me to the tragic elements in performing. So what I've learned from having CF helps me in some ways, but I still have to fight for my roles. The industry remains racist and I don't audition as much as my white peers do. I feel like we're on the precipice of changing that with more inclusion and diversity. But we still have to fight virtue signaling and performative inclusion. People need fully fledged characters instead of flat ones who only exist as "check marks" for diversity. As an actor of color, I often have to create my own work. The upside is that I now have a platform to share my experience my way.

> "Someone called Child Protective Services on my mom because my brother was struggling to gain weight—nobody had diagnosed his diabetes."

How do you prepare for a new acting role?

It changes from role to role. As a society we have very black and white thinking, which has marginalized so many people. Gray areas are often uncomfortable for people to acknowledge. So when I approach a character, I always like to see the gray—I feel like there's more information in those areas. Humanity is gray; it's not all good. I look at emotional, mental, and physical aspects and how those all bleed into one another. And I focus on creating different sensory inputs for the audience rather than relying entirely on my body or voice. I want to be inclusive with my performance. In the process, I'm dismantling this white cishet male ideal of the method actor, which is very predatory. There's no "right" way to pursue a character—only how you feel and how you're making the audience feel.

Have you ever declined a role because of health challenges?

I don't think so, but on the other hand I've done lots of stubborn things to avoid being hospitalized. For example, in high school I was force-feeding myself so that I could stay on the track team. My doctors at the time didn't even see this as something negative. They just praised me for "being so compliant" and "working so hard" instead of thinking about what I needed. I spent so much time and energy appeasing my white male doctors. Now I do things that make me comfortable and promote others doing the same. I've pushed my physical limits for certain roles, though. I've taken roles where I have to smoke a cigarette or stay up all night. I didn't want CF to limit my choices—and I wanted to prove to myself that I could do it. I've turned down roles before, but usually for other reasons; maybe I didn't like the story or the character.

What do you feel proudest of in your activism work?

People rallied when I got passionate about feeding the community and started a community fridge. They came

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together and protected that effort. I saw people get inspired and passionate; I met folks who were excited to help. I organized a clothing drive. I organize school drives and resource drives for communities that are underserved and neglected—predominantly ones that are home to Black and Indigenous people of color. Feeling guilty about not making it to every action remains a struggle—but I go back to the idea of gray areas. You can't be everywhere all the time or expect that of yourself as an activist. I try to give myself as much grace as possible so that I can work smarter and not harder.

Tell us more about your own journey with CF.

I've always had a very tiny physique. I got picked on in school and people thought I had an eating disorder. Being small was ridiculed because it was also coveted, especially in the performing arts. I think we all should be able to exist in our bodies without societal judgment. CF can contribute to that, depending on where you fall physically. Everyone in my family is tall except me and my brother. All those nutritional issues probably stunted our growth. So I have a height insecurity; I'm always in heels. People think I'm taller but I'm just in stilts all the time! I feel like the odd one out.

Do you have a dream role you'd like to play?

I used to want to play Josephine Baker. Her struggle was like mine; she was a black sheep who never fit in. She had to travel abroad to find a community that really enjoyed her performance and expression. Being a Black femme in the South who was lighter-skinned, and seeing the hypocrisy in colorism, was also relatable for me. But nowadays I'm drawn less to biopics and more to roles that are physically demanding. I've always wanted to do an action role! Lately I've been submitted for a couple of Marvel Cinematic Universe productions.

What helps you cope with your CF?

The biggest thing helping me cope is knowing I've survived thus far—meaning I've outlasted the idea that CF was all I'd ever be or do. When I feel more defeated or depressive, I always appreciate not being defined just as a CF patient. At the same time CF can be invisible, which makes people question whether it's really a fatal disease. I think of it as a badge of honor and an important part of me rather than something to destroy. But I do have survivor's guilt and sometimes I'm distant from the CF community because of this. It's painful when you form bonds and then people die. I'm working on that in therapy. How can I be involved and still protect myself emotionally? How do I not feel like I'm abandoning my community while also not forcing community? Right now that means advocating when I have capacity and resting when I don't.

What advice would you give your younger self?

I was a different person before 2020. I've always been very rebellious, anti-establishment, and anarchist. But before that year, part of me was still flirting with the idea of the status quo and trying to fit in. I see this in the people I dated and befriended, and in my working relationships. These days I really protect and value my comfort, integrity, and values above everything else. So I would tell my younger self "Don't look back. Keep going forward." As a society, our projections of our past hold us back—especially when we try to make the past our present.

Do you have a funny CF story?

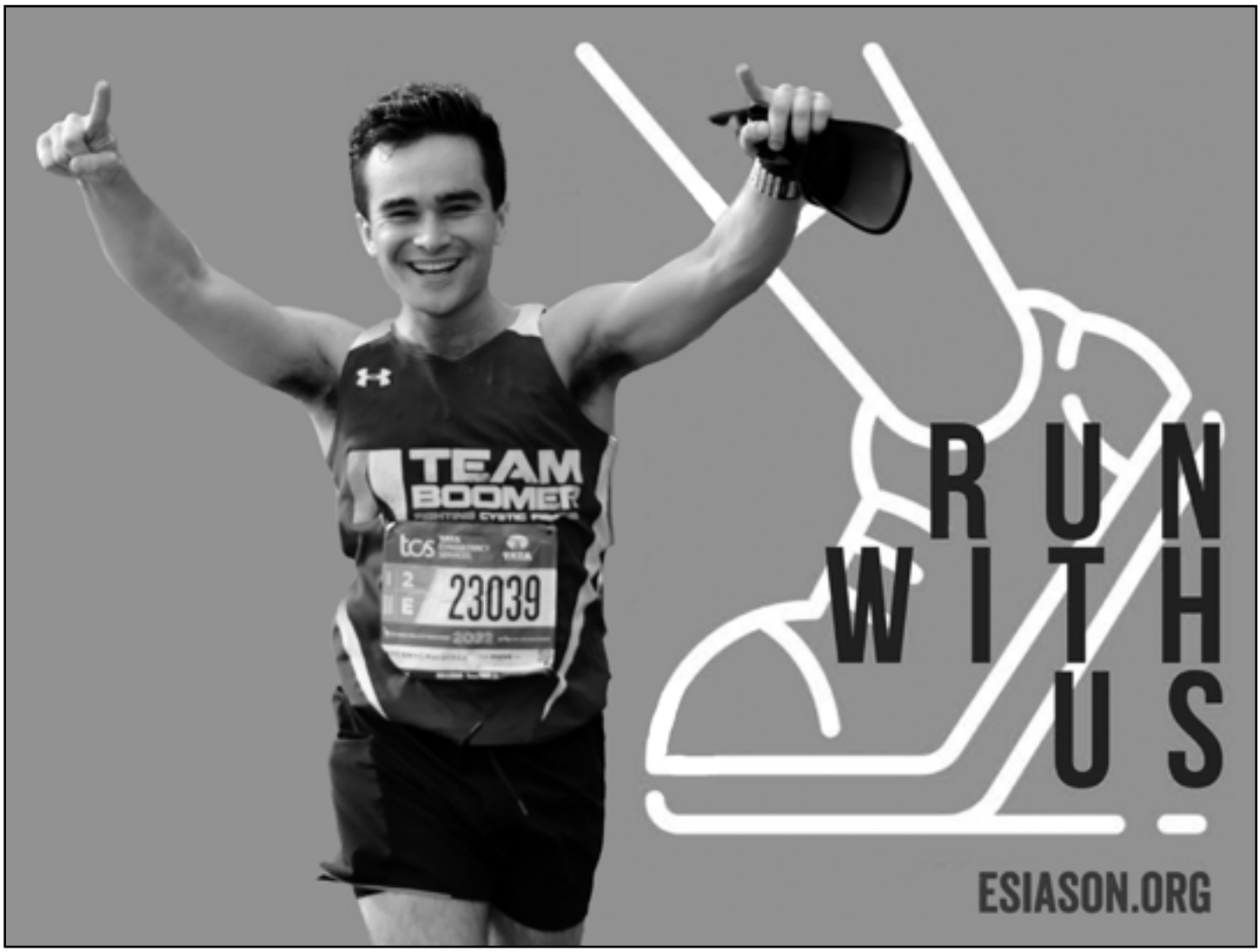
My mom had this bag of "nasty capsules"—enzymes that she would find on the floor. She'd keep them in her purse; whenever my brother and I didn't want to do treatments she'd threaten us with those! She's a science person, pragmatic and good at problem solving. That running joke about the nasty capsules was genuinely helpful. My mom and dad used "tough love" and dark humor to show me and my brother we could be self-sustaining. To this day I usually do well at remembering to take my meds—including Trikafta which has reduced my treatment burden. I have my own little tricks, like keeping enzyme capsules in an Altoids tin so they're easy to carry.

What would a perfect day look like for you?

Getting up, taking my meds, minimal coughing all day. Maybe doing some rigorous exercise. Spending time with my partner. And doing what I love to do, which is creating. Whether that be my fashion designs, my visual art, or my film work—just being able to create in an open and comfortable space. Somewhere in there, I'll go to the beach because I'm a Californian. Much as I love New York, the beach will always be my home. ▲

Dr. Alexandra "Xan" Nowakowski is 39 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.*

If you would like to be interviewed for "In The Spotlight," please contact Xan Nowakowski or Andrea Eisenman. Their contact information is on page 2.



EISENMAN continued from page 31

section of my lid where the hole was and then stitched the two sections together. The affected left eye opening is now slightly smaller than my unaffected right eye. The left is also perkier, instead of sagging closed like my right. He did a fabulous job. Unless up close, you cannot see the carnage. That is not to say I did not suffer depression for several weeks after from the trauma of those two surgeries. I felt defeated. But eventually I knew I was fortunate to have the care of two great surgeons who made it their mission to cure me.

I have gone back three times since surgery to check in with the ocular plastic surgeon. He is pleased with how it is healing. I am missing a section of lashes in the middle. Thankfully my vision is not impaired and I get to live to tell this tale. I was told by a nurse that this kind of cancer can spread by traveling up through the nasal passages to the brain. They had actually seen this in another patient.

My dermatologist, the Mohs surgeon, and then the ocular plastic surgeon all basically said the same thing to me: "Thank goodness you got this addressed quickly!" I wish I had not been so afraid of an eyelid injection and moved faster. Had I to do it all over again, I would not have hesitated as I did. This is a cautionary tale to those who are immunosuppressed. Do not put off any suspicious bumps or lesions. Mountains quickly form out of mole hills! You will be happier if you act swiftly. ▲

Andrea Eisenman is 58 and has CF. She lives in New York, NY, with her husband Steve and dogs, Willie, Roscoe, and new girl, Trixie. Andrea is the Executive Editor for USACFA. She enjoys cooking new recipes, playing pickle ball, biking, tennis when possible, and staying active as her health allows. Her contact information is on page 2.

impact on lung function was seen for children living within or beyond about 2.5 miles of composting sites. If further research confirms the present study's results, the associations identified could have important implications on i) the living environments of pwCF, ii) clinical advice to pwCF; iii) public health advice provided to vulnerable populations living near permitted composting sites (PCS); and iv) how PCS are regulated and permitted.
https://tinyurl.com/2q5bwbhs

The Clinical Association Between Aspergillus Fumigatus And Respiratory Outcomes In Adolescents And Adults With Cystic Fibrosis

Positive *Aspergillus fumigatus* (Af) culture was not associated with lower patient-reported respiratory-related quality of life (QOL). Yet, positive Af culture was associated with both lower FEV1 percent predicted and increased frequency of severe pulmonary exacerbations (PEx) warranting intravenous antibiotics in adolescents and adults with CF. Future studies are required to better understand the direct role of Af in lung disease progression in CF.
https://tinyurl.com/2eyj3knd

Airway Bacterial Community Composition In Persons With Advanced Cystic Fibrosis Lung Disease

The progression of lung disease in people with cystic fibrosis (pwCF) has been associated with a decrease in the diversity of airway bacterial communities. The absence of a dominant genus, presence of methicillin-susceptible Staphylococcus aureus, and greater bacterial richness positively correlated with lung function. Higher relative abundance of the dominant genus and greater antimicrobial use negatively correlated with lung function. PwCF with a low diversity community and dominant genus had reduced lung transplant-free survival compared to those without. In summary, a considerable proportion of pwCF with advanced lung disease do not have airway bacterial communities characterized by low diversity and a dominant genus and these individuals had better survival. An understanding of the antecedents of low diversity airway communities– and the impact these may have on lung disease trajectory - may provide avenues for improved management strategies.
https://tinyurl.com/yn5kkmts
AND
https://tinyurl.com/2ol9wfwt

Diabetes Is Associated With Increased Burden Of Gastrointestinal Symptoms In Adults With Cystic Fibrosis

Individuals with CFRD overall, have a higher gastrointestinal (GI) symptom burden, according to CF-specific GI symptom questionnaire CFAbd-Scores. The CFAbd-Score total score (0–100pts), its 5 domains, alongside nine specific GI symptoms associated with diabetes mellitus (DM), were compared between CFRD and non-CFRD groups. Total CFAbd-Score and the two domains: gastroesophageal reflux disease and disorders of appetite were significantly higher in the CFRD group compared to the non-CFRD group. Among the nine GI symptoms commonly reported as elevated in DM, bloating and nausea were significantly more common in individuals with CFRD compared to those without.
https://tinyurl.com/34upuzer
AND
https://tinyurl.com/2va6j58h
AND
https://tinyurl.com/2kkkta8n

Giving CGM Access To All People With Type 2 And Other Diabetes

People with cystic fibrosis-related diabetes (CFRD) require careful management of blood sugar levels to avoid complications such as heart disease, kidney disease, and nerve damage. There is evidence that lack of glucose control worsens lung function and that fast blood glucose tests are not effective in identifying glucose variability. One of the greatest challenges when addressing CFRD is initially screening for diabetes. The CF Foundation currently recommends an oral glucose tolerance test (OGTT) for people with cystic fibrosis over 10 years old. However, rates of annual OGTT testing in adults are very low – only 30% of adults with CF were screened for diabetes. Continuous glucose monitors (CGM) may potentially be an effective tool for expanded screening. CGM may also be helpful in the ongoing management of CFRD, both with and without AID (automated insulin delivery) technology. The limited data that exists suggest that use of a CGM can make the diagnosis of diabetes earlier, improve outcomes and motivate people to make healthier choices in diet and lifestyle, no matter if they are taking insulin or not.
https://tinyurl.com/mry8veb6

Drug-Drug Interactions With CFTR Modulator Therapy In Cystic Fibrosis: Focus On Trikafta®/Kaftrio®

The combination of CFTR modulators ivacaftor, tezacaftor and elexacaftor (Trikafta®, Kaftrio®) significantly improve outcomes, including survival in a broad range of cystic fibrosis patients. These drugs have complicated metabolic profiles that make the potential for drug interactions an important consideration for prescribers, care providers and patients. Prolonged survival also increases risk of age-related disease and their associated pharmacotherapy, further increasing the risk of drug interactions and the need for increased vigilance amongst care providers. The authors systematically searched the literature for studies identifying and evaluating pharmacokinetic and pharmacodynamic drug interactions involving the components of Trikafta®/Kaftrio®. They also searched electronic databases of drugs for possible drug interactions based on metabolic profiles. 86 potential drug interactions were identified, of which 13 were supported by 14 studies. There is a significant need for research to describe the likelihood, magnitude and clinical impact of drug interactions.
https://tinyurl.com/4hszfb7p

The Higher Education (Formerly The Lauren Melissa Kelly) Scholarship

The application deadline for the Higher Education (formerly the Lauren Melissa Kelly) Scholarship is June 30, 2023. Any student seeking a degree in higher education, from Associate to Ph.D., is welcome to apply. We look for students who demonstrate tremendous academic achievement, community involvement, and a powerful understanding of how their CF—matched with these achievements—places them in a unique situation to gain leadership roles within the community. We believe that any higher education is a strong foundation for advocacy and involvement in the CF community.

Nancy Wech established this scholarship in honor of her daughter, Lauren Melissa Kelly. This semester's winners

IN MEMORY OF
LAUREN
MELISSA KELLY

demonstrated outstanding potential, just like Lauren years ago. Lauren was an inspiration to all who knew her. An incredible leader and scholar, her drive and success are the foundation of her memory. She was transformative in every aspect of her life. She had distinguished herself as a member of the Golden Key Honor Society, Mortar Board, Phi Upsilon Omicron, Gamma Beta Phi, Delta Gamma sorority, and was chosen as one of ten Senior Leads at the University of Georgia. She acted as one of the re-founding members of the Phi Kappa Literary Society and was significant in the metamorphosis of the Z Club into the William Tate Society. Although Lauren lost her battle with cystic fibrosis late in her senior year, her hard work and memory continue to live on through her inspiring involvement.

Two scholarships are awarded each application cycle. More information, including the application and relevant deadlines, can be found on our website. For questions about future scholarships or anything related to the application process, please contact us at scholarships@usacfa.org. ▲

Effect Of Elexacaftor/Tezacaftor/Ivacaftor On Annual Rate Of Lung Function Decline In People With Cystic Fibrosis

Elexacaftor/tezacaftor/ivacaftor (ELX/TEZ/IVA) was shown to be safe and efficacious in people with cystic fibrosis (CF) with ≥ 1 F508del-CFTR allele in Phase 3 clinical trials. ELX/TEZ/IVA treatment led to improved lung function, with increases in percent predicted forced expiratory volume in 1 second (ppFEV1) and Cystic Fibrosis Questionnaire-Revised respiratory domain score. Here, the impact of ELX/TEZ/IVA on the rate of lung function decline over time was evaluated by comparing changes in ppFEV1 in participants from the Phase 3 trials with a matched group of people with CF from the US Cystic Fibrosis Foundation Patient Registry not eligible for cystic fibrosis transmembrane conductance regulator (CFTR) modulator therapy. Participants treated with ELX/TEZ/IVA had on average no loss of pulmonary function over a 2-year period. ELX/TEZ/IVA is the first CFTR modulator therapy shown to halt lung function decline over an extended time period.
https://tinyurl.com/27rszwuf

Lived Experiences Of People With Cystic Fibrosis That Were Not Eligible For Elexacaftor-Tezacaftor-Ivacaftor (ETI): A Qualitative Study

Elexacaftor-tezacaftor-ivacaftor (ETI) represents a significant step forward in cystic fibrosis (CF) care and could change the course of CF lung disease and quality of life for many people with CF (PwCF). However, several PwCF cannot benefit from these modulators because their rare mutations are not eligible for treatment. This study aimed to investigate the lived experiences of PwCF who are not eligible for ETI. Data were collected through semi-structured interviews. The investi-

Continued on page 38

In Memory

Cynthia Denice Lazenby, 50
Oxnard, CA
Died on May 9, 2022

Immediate family members may send in the names of CF adults who have died within the previous year for inclusion in "In Memory." Please send: name, age, address and date of death.
Send to: *CF Roundtable*,
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TILLMAN continued from page 37

gators found that PwCF who are not eligible for ETI experience intense disappointment and conflicting emotions that can influence their decision-making linked to diminishing/renewal hope. Integrated care, including mental health monitoring programs, should be provided to these patients to aid them in overcoming their disappointment and to improve their coping.
https://tinyurl.com/2jfdadjs

Elexacaftor/Tezacaftor/Ivacaftor Projected Survival And Long-Term Health Outcomes In People With Cystic Fibrosis Homozygous For F508del

A series of phase 3 clinical trials have demonstrated that elexacaftor plus tezacaftor plus ivacaftor (ELX/TEZ/IVA) is safe and efficacious in people with cystic fibrosis (pwCF). The impact of this treatment on lifetime clinical outcomes and survival, however, has yet to be assessed. The researchers used a person-level microsimulation model to estimate the survival and lifetime clinical benefits of ELX/TEZ/IVA treatment versus other CFTR modulator combinations (tezacaftor plus ivacaftor [TEZ/IVA] or lumacaftor plus ivacaftor [LUM/IVA]) or best supportive care (BSC) alone in pwCF. The median projected survival for pwCF homozygous for F508del-CFTR treated with ELX/TEZ/IVA was 71.6 years. This was an increase of 23.2 years versus TEZ/IVA, 26.2 years versus LUM/IVA, and 33.5 years versus BSC alone. Treatment with ELX/TEZ/IVA also reduced disease severity as well as the number of pulmonary exacerbations and lung transplants.
https://tinyurl.com/ye2x4nf3

Long-Term Tezacaftor/Ivacaftor Safety And Efficacy In People With Cystic Fibrosis And An F508del-CFTR Mutation: 96-Week, Open-Label Extension Of The EXTEND Trial

Study 661-110 (EXTEND) is a phase 3, open-label, three-part rollover study designed to assess the long-term safety and efficacy of tezacaftor/ivacaftor (TEZ/IVA) in participants aged ≥12 years homozygous for F508del (F/F) or heterozygous for F508del and a residual function mutation (F/RF). TEZ/IVA was shown to be safe and efficacious for up to 120 weeks in Part A. Results from Part B, which evaluated safety and efficacy for an additional 96 weeks found that TEZ/IVA was generally safe and well tolerated over a further 96 weeks; safety data were consistent with Part A. Improvements in ppFEV1 and pulmonary exacerbation rates were maintained for an additional 96 weeks in Part B. The most common adverse events, which were generally consistent with common manifestations of CF, included infective pulmonary exacerbation of CF, cough, nasopharyngitis, hemoptysis, and headache. Lung function was maintained over 96 weeks in both genotype groups.
https://tinyurl.com/bdznxdbw

CFTR Modulators Safe While Pregnant, Breastfeeding: Case Series

Data on the safety profile of CFTR modulators to guide patients during pregnancy is still scarce. The Phase 3 trials that supported their approval have excluded pregnant women with CF. Also, recent studies suggest the therapies can cross the placenta and reach the fetus' blood circulation. However, treatment with CFTR modulators was safe during pregnancy and breastfeeding in two women with cystic fibrosis (CF). Despite being advised to stop treatment, both women continued on their CFTR modulator during pregnancy and breastfeeding without any safety concerns. Continuation of CFTR modulators during pregnancy and lactation requires careful multidisciplinary considerations and patient discussion regarding risk and benefit.
https://tinyurl.com/3rxk55bj

CFTR Modulators Found To Improve Antibiotic Efficacy In CF: Study

Despite their established clinical benefits, response to CFTR modulators can vary from patient to patient, suggesting other factors may influence the effectiveness of modulators. Because bacterial infections commonly occur in CF lungs, the impact of CFTR modulators on lung bacteria also may impact their efficacy. To find out, a team of scientists tested the effects of CFTR modulators, either alone or combined with antibiotics, on bacteria samples isolated from CF lungs. The CFTR modulators tested included ivacaftor (sold as Kalydeco), lumacaftor, approved in combination with ivacaftor as Orkambi, and tezacaftor, sold in combination with ivacaftor as Symdeko. Elexacaftor, combined with tezacaftor and ivacaftor as the triple combination therapy Trikafta, and Trikafta itself, also were assessed. Samples of two bacteria commonly found in CF airways—*Staphylococcus aureus* and *Pseudomonas aeruginosa*—were collected from CF patients at the early stages of colonization, and again at later stages when the bacteria had adapted to the CF lung environment. The bacteria were then exposed to increasing levels of CFTR modulators. Antibacterial activity was measured by the lowest concentration of medicine that effectively suppressed bacterial growth, called the minimum inhibitory concentration, or MIC. Lower MIC values indicated more potent antibacterial activity. Results revealed that ivacaftor had the most potent antibacterial activity for all the *S. aureus* samples. This was followed by elexacaftor. Lumacaftor and tezacaftor showed little or no signs of antibacterial activity. Triple combination therapy elexacaftor/tezacaftor/ivacaftor (ETI) also showed strong antibacterial

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activity. By contrast, all CFTR modulators showed little or no activity against *P. aeruginosa* samples. Next, the researchers mixed CFTR modulators with antibiotics to measure potential additive or synergistic effects. Ivacaftor enhanced the activity of the antibiotic linezolid in most *S. aureus* samples while enhancing amoxicillin, vancomycin, and teicoplanin on a more limited scope. Lumacaftor and ivacaftor both enhanced vancomycin and teicoplanin against *S. aureus*. Even though CFTR modulators showed no activity against *P. aeruginosa* on their own, ivacaftor strongly enhanced the activities of colistin and polymyxin B, against more than 95% of the *P. aeruginosa* samples. Triple combination ETI showed an additive effect on the activities of colistin and polymyxin B in several *P. aeruginosa* samples.
https://tinyurl.com/yc4xp3v9 ▲

Laura Tillman is 75 years old and has CF. She is a former director and President of USACFA. She and her husband, Lew, live in Northville, MI.

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- 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.
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- You may subscribe at www.cfroundtable.com

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IMPORTANT RESOURCES

Medical Assistance Tool (MAT): https://medicineassistancetool.org/ PhRMA's Medicine Assistance Tool (MAT) is a search engine designed to help patients, caregivers, and 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

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