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

Thirty Years, 35 Miles: A Story Of Triumph And Hope

By Marcus Miller

Three years ago, at the age of 27, I found myself hospitalized for a week in the Progressive Care Unit in Wilmington, North Carolina. I was battling the most severe bout of pneumonia I'd ever contracted with a total lung function around 43% at the time. All of this happened during the height of the COVID-19 pandemic and made for a very anxious and uncertain time. Thankfully, in the weeks that followed, I was able to meet my new CF team for the first time since my move to NC. During that first visit, I was able to learn more about Trikafta and the incredible results that were being reported from this miracle drug. It seemed too good to be true; could I really get my health and life back fully? I was excited and nervous when that first little box of pills arrived. I recall thinking, what if this doesn't work for me? Or perhaps the bigger question, what if it does? What will I do then?

After a long journey of ups, downs, and building back my body and mind, I wanted to push my limits and see what was possible with my newfound health. Where is that prohibitive line at now for my body? After much training and reaching several other lofty goals, I decided to attempt my first ultramarathon run. An ultramarathon, for those who don't know, is any run longer than the standard 26.2-mile marathon. Currently only about .03% of the US population has ever run one. My story isn't written to glorify or bring recognition to myself, but rather to bring awareness to and highlight the endless possibilities for folks with CF when they have access to the proper care and life-changing modulators. Hopefully, the day comes soon when CF will indeed stand for *Cure Found* for all of us.

MARCUS MILLER CROSSING THE FINISH LINE OF HIS FIRST ULTRAMARATHON CALLED THE MT. MITCHELL HEARTBREAKER, MARCH 2023.

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"We can't afford the time to sit here and wait for a chainsaw. We'll have to turn around and find a different route," I stated as I hopped back

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INSIDE THIS ISSUE

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EDITOR'S NOTES

Summer is in full swing, but it also means the fall semester is just around the corner. This issue we're focusing on the return to school and/or work. For the focus topic, **Dr. Jeanie Hanley** writes about returning to her medical practice after 17 long years away and what that looks like as far as balancing work and treatments. **Suzanne Joyce** writes about her own experiment with returning to work as an attorney.

In their "Pearls of Wisdom" column, **Dr. Xan Nowakowski** writes about the importance of having a mentor, especially someone with CF, as you navigate both higher education and employment. **Beth Sufian** answers questions about eligibility criteria for both SSD and SSI benefits in her "Ask The Attorney" column. **Isabel Stenzel Byrnes**, in her final Spirit column, writes about the legacy we leave. For our Pet's Perspective column this issue, **Ashley Coleman**, on behalf of Orbit and Axil, relays the importance of mental health support and regular exercise. **Maggie Williamson** shares her recipe for Chopped Broccoli Salad in her "Culinary Corner" column this issue. **Laura Tillman**, our longtime researcher and columnist, expertly collates all the current CF research in her final "From the Internet" column. We're deeply grateful for the countless hours spent each quarter to bring us the latest scientific news each issue. Don't worry, we'll have a new researcher starting this fall! This summer we're excited to introduce our newest column, "Chaptered Lives," by **Andrew Corcoran**. In his inaugural column, Andrew writes about getting the call for new lungs and leaving his old life behind. In our "Voices From The Roundtable" section **Matison Deaton** writes about her struggle with *Mycobacterium abscessus* and the long journey to transplant because of that diagnosis. We love hearing from our readers! Reach out to us if you're interested in sharing your story in an upcoming issue. You can also find a list of focus topics both on our website and on page 3 of this issue.

We're deeply saddened to share the passing of one of the founders of USACFA, Kathy Russell, in May. You can read about her life and legacy on page 21. I hope that we continue to grow as an organization, embody her grace and wisdom, and evolve in a way that honors her commitment and dedication to USACFA throughout her tenure on the board.

We now offer three scholarships, including the newest William Coon, Jr. Scholarship! You can read about each of our scholarships on page 15 or, for even more information, head to our website where you'll also find the application, requirements, and deadline for each. Additionally, we're always looking for people to interview for our "In The Spotlight" column as well as new directors. If you're interested, 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.

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

CF Patients Using Kaftrio/Trikafta May Enjoy 'Near Normal' Lifespans

Use of Kaftrio (elexacaftor/tezacaftor/ivacaftor - Trikafta) may help people with cystic fibrosis (CF) and the F508del mutation in both CFTR gene copies live to a median age of 71.6 and possibly longer if treatment is started in adolescence. In a scenario where treatment with Kaftrio is initiated between 12 and 17 years old, a patient's median estimated lifespan is 82.5 years. That's 45.4 years longer than estimates for CF patients on best supportive care alone. To determine estimates of such benefits with Kaftrio as opposed to other treatment options, scientists used computers to run a microsimulation, a model that mimics the effects of treatment on individual members of the CF population. They drew on data of disease progression from articles published in scientific journals. Data on how well each treatment worked came from Phase 3 clinical trials and extrapolations of clinical information. In addition, it was estimated that people on Kaftrio would experience better lung function across greater spans of life than those on best supportive care and, subsequently, less time with severe lung disease. Life-years were defined as the estimated years of life assigned with a given treatment in these simulations. While living longer,

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LAURA TILLMAN

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.

Summer (August) 2023: Returning To Work or School (Current issue)

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, 2024) Have you had a transplant? If so, for how long and can you tell us about your journey? Tell us the good, the bad, and the ugly. What can you share to offer hope to those who are waiting? Were you prepared for all of it? What do you wish you knew before embarking on this endeavor? Are you waiting for one or deciding whether to get one? What are your hopes or fears for receiving a transplant? Are you in need of a second organ transplant? If so, why? What issues (e.g., diabetes, high blood pressure, anxiety, etc.) have you encountered after receiving a transplant?

Spring (May) 2024: CF and Cancer (Deadline March 15, 2024)

ASK THE ATTORNEY

Back To Basics

By Beth Sufian, J.D.

In the past three months, *CF Roundtable* readers have sent many questions related to eligibility criteria for Social Security Disability or SSI benefits.

The information provided in this article is only meant to be general information. Nothing in this article is meant to be a guarantee that a person will be eligible for SSDI, SSI, or any other government program.

Question 1:

Once Social Security approves my application for disability benefits, will I remain eligible for the rest of my life?

Answer:

No. Social Security rules now require that a person be reviewed every three to five years if the person receives Social Security Disability Insurance (SSDI), SSI benefits, or Social Security benefits based on the work record of a parent who is deceased, retired, or receiving SSDI themselves. Social Security sends Continuing Disability Review (CDR) paperwork in the mail. Also, if Social Security has evidence of work activity that is more than the allowable amount, they may send a CDR. CDR paperwork is *very* important. The paperwork may look like it is easy to fill out, but answers given to the questions often lead to termination of benefits. Often the person completing the forms does not understand the questions and gives answers that are vague or unclear.

Many people with CF are losing benefits because they do not think the CDR is important and mistakenly think they will have benefits for life. A person can contact the CF Legal Information Hotline if they have questions about Social Security paperwork or a CDR.

Question 2:

I have had Social Security disability benefits since I was three years old. Does Social Security review eligibility once I turn 18 years old?

Answer:

Yes. Social Security must review anyone who has been receiving SSI benefits once the person reaches 18 years of age even if a person has received SSI benefits throughout their childhood. Social Security may stop benefits after an 18-year-old review if the person no longer meets the medical criteria or there is no evidence to support a finding that the person cannot work more than 20 hours a week **and** make more than $1,470 a month (before taxes are taken out of the work earnings) from work activity or $1,050 a month from self-employment. The length of time a person has been eligible for Social Security benefits in the past is not a reason to have benefits continued.

BETH SUFIAN

Question 3:

Will Social Security consider me to be disabled as long as I have a diagnosis of cystic fibrosis?

Answer:

No. "Diagnosis" and "disability" mean different things. Diagnosis is a medical determination that a condition exists or is the cause of symptoms. Disability for adults under Social Security rules means the individual is incapable of substantial gainful employment as a result of one or more medical conditions that limit the person's ability to work. For children the standard relates to the child's ability to engage in age-appropriate activities on a daily basis. Social Security considers a person disabled if the signs and symptoms of the condition meet the degree of severity listed in its regulations or is equivalent in severity. The regulations set out specific symptoms and the degree of severity that the medical evidence must show. This is called the Social Security Listing and can be found in the Social Security Blue Book at www.ssa.gov. In addition to showing a person meets one of the listed medical criteria, a person must show daily limitations that prevent the person from working more than the allowable amount. A person can also be eligible for benefits if the person's medical condition equals or is as severe as the Social Security Medical Listing criteria.

Question 4:

Social Security stopped my SSI benefits because it said my household income is over the limit, but my monthly income has not changed. The only change is that I recently got married.

Answer:

SSI has eligibility criteria based on

household income and resources. If a person is over 18 years of age, the parents' income and assets will no longer be counted when determining SSI eligibility. However, if the person who receives SSI benefits gets married, then the new spouse's work income and assets will be counted by Social Security.

If the spouse's assets put the person with the disability over the SSI asset limit, then SSI benefits stop. If the spouse works, then the spouse's monthly income has to be below a certain amount; otherwise the SSI stops.

SSDI eligibility standards do not consider a spouse's assets or work income to determine eligibility for SSDI.

Question 5:

Social Security says my work income exceeds the limits, but my take-home pay is less than the limit listed on the Social Security website. What amount is counted to determine income?

Answer:

When Social Security refers to income, it typically means gross income before the subtracting of taxes, withholdings, or other deductions.

There are many other questions about Social Security, both basic and advanced. However, these few questions should help people with CF who review Social Security befits understand some fundamental principles regarding eligibility.

If you have questions about laws related to Social Security benefits, Medicaid, Medicare, health insurance, employment, or 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 generously funded by the CF Foundation, but CFLIH employees are not employed by the CF Foundation. The CFLIH is now in its 25th year. ▲

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.

MILESTONES

Please share the milestones in your life with our readers. Your successes and achievements may serve as a source of motivation for others in need of an infusion of "positive mental attitude" in the pursuit of their goals. Send us a note specifying your "milestone." Include your name, age, address and phone number. Mail to: **CF Roundtable, 9450 SW Gemini Drive, PMB43881, Beaverton, OR 97008-7105.** Or email to: **cfroundtable@usacfa.org**

ANNIVERSARIES

Birthday

Susie Baldwin
Los Angeles, CA
56 years old on April 10, 2023

Jeanie Hanley, M.D.
Los Angeles, CA
61 years old on June 10, 2023

Employment

Alexandra "Xan" Nowakowski, Ph.D., M.P.H.
Lakeland, FL
Work anniversary
13 years with Florida State University

Wedding

Susie Baldwin and Adam Levy
Los Angeles, CA
Wedding anniversary
26 years on October 13, 2022

Transplant

Paul Albert, 63
Catasauqua, PA
Bilateral lung transplant
30 years on February 10, 2023

Andrea Eisenman, 58
New York, NY
Bilateral lung transplant
23 years on April 25, 2023

Susie Baldwin, 56
Los Angeles, CA
Bilateral lung transplant
10 years on December 21, 2022

Mike Darrar, 56
Post Falls, Idaho
Bilateral lung transplant
12 years on May 3, 2023

Zach Hays, 34
Vernon, Connecticut
Bilateral lung transplant
6 years on February 3, 2023

SPIRIT MEDICINE

Living Out Our Legacy

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

In 2009, I wrote an extra Spirit Medicine article during a flurry of inspiration. I was 37 years old and wrote a list of the ways we leave a legacy. It felt premature to publish, and alas, that was about five laptops ago so I cannot find that document. Now, at 51, I'd like to rehash this idea of legacy.

A legacy is often brought up at the end of life. But, of course, it is not something we necessarily complete at the end of our lives. Every day, we live out our legacies. It is the containment of our mark on the world. Some are divinely ordained; callings that arise within us to pursue our purpose. Some are actions and choices that add up to create a tapestry of our lives. This is usually a tapestry of contentment, satisfaction, and fulfillment—making us feel good about the life we've lived. Legacy is how we make our imprint on the world. It is the ripple effect of our deeds that is everlasting and outlives our short lives. Some can say our legacies are promises to God to live the best life we can live in the time we have.

If we look closely, there are legacies all around us. The library is filled with stories, which are the legacies of peoples' lives or imaginations. The clay granaries hidden in the cliffs of the Southwest are the legacies of the ancient people who survived in such harsh conditions. The statues, plaques, museum exhibits, films, works of music, art, and athletic prowess are also types of legacies.

I thought I'd simply list a few types of legacies, so perhaps you can recognize the ways you are manifesting yours.

- A love story of mythic proportions is one profound legacy. One that comes to mind is the long marriage of Paul and Kathy Russell, leaders of the USACFA organization for decades. I still remember them being glued at the hip at CFRI conferences, laughing in synchronicity and telling endless stories of life with CF back in the day. Theirs was a love story that I believe helped Kathy live for as long as she did. Godspeed, dear Kathy. Thank you for making a difference for our community. The legacy of love always has a ripple effect.

> *People with CF have a unique form of legacy by paying it forward for the next generation through DEI awareness, clinical trials, or organ donation awareness.*

ISABEL STENZEL BYRNES

- Becoming a caregiver is another way to leave a legacy. Many of us make life better for a four-legged animal or other vulnerable beings. For some, this means impacting the life of a child. In my 20s my impact was through being a mentor for my CF camp kid. Now I get to be an aunt to three beautiful nieces. Many of my CF friends are raising their own children and some are even grandparents. Parents get to pass on their ethics, values, and teachings about what's important to live a meaningful life. Maybe becoming a parent is too much of a commitment given health demands. You can still become a mentor to a child who needs a guide in life. Boys and Girls clubs or the Scouts are eager for teachers who show they care.

- Becoming a specialist or expert in something you are passionate about offers a rich legacy: an academic expertise, fixing motorcycles, running marathons, being in a rock band, coaching your daughter's basketball game, or just following your bliss like playing the bagpipes. Isn't

this why we are here? Just to take life and play with it? It sure balances out all the messiness.

- A professional legacy is the chance to contribute your creative energy to something bigger than yourself—an entrepreneurial business, a health care company, an environmental cause, a political or social cause, etc. You can make a difference for good in this large world of ours. And we have a choice: Our passion may have nothing to do with CF; just something we love! And sometimes this legacy contributes to the security of your financial legacy as well.

- People with CF have a unique form of legacy by paying it forward for the next generation through DEI awareness, clinical trials, or organ donation awareness. Our voices raise awareness of these causes to directly impact our community's longevity. It can mean being bold and putting your CF story out there; knowing that stories have the power to invite connection, empathy, open-mindedness, self-discovery, and courage. Our creative sharing has potential to be helpful to others, as evidenced by a recent CFRI Writing to Heal group where writers told their stories with such authenticity and insight. Just an aside, I have been writing Spirit Medicine since 2007, following the footsteps of the late Lisa McDonnell and Catherine Martinet. I remember Rich DeNagel making fun of me, saying, "You know, whoever writes that column dies!" And I replied, "Yes, whoever writes any column dies, Rich. And besides, this will be part of my legacy."

- A spiritual legacy is another form of legacy. As a writer of this column I've tried to write generally about spirituality to make the themes as relevant and non-dogmatic as possible. Truthfully, my faith is defined by God as love and my charge to be the loving being Jesus and God want me to be. To me, a spiritual imprint is what you leave behind in terms of your spiritual principles and beliefs for others to learn from. Perhaps you've learned them from your family or cultural or ethnic heritage. Have you incorporated spiritual habits and practices into your life and your family's? Do you have a lasting, trusting, relationship with a higher power? Perhaps you have close friends or community who have helped you develop your spiritual legacy as well. Are you at peace with the path you've chosen for your spiritual maturity? Perhaps I will be rewarded by my faith; perhaps not. Still, there can be comfort knowing that a spiritual legacy is the treasure chest we invest in which gives us security when we reach the end of this life and enter the "next place."

> *"Our energy can leave an imprint in the lives of those we love."*

- Finally, there are emotional and social legacies we leave behind. For all my life in the CF community, I've been touched by observing the emotional and social legacies of my peers during their adventures with CF. When I think of social and emotional legacies, I think of these questions: Over the course of our relationships with others, how have we made people feel during our intersections with them? What did we bring into our friends' presence by walking into the room? Were we capable of forgiveness, communicating conflicts, listening, and giving and receiving support? Our energy can leave an imprint in the lives of those we love. Our social circles can be enriched by knowing and loving us. Just by living the best life we can live with a serious illness, we convey virtues and principles about the sacredness and preciousness of loving, and emotionally deep relationships with others. For some, it takes a lifetime to learn of the sustaining force of connection. Instead, CF has been the urgent teacher to get us to love each other!

Truthfully, the end of my life is where legacy becomes most relevant. I'd like to believe my spiritual legacy is a reflection from so many CF mentors: *We fought the good fight. We did our very best. We did not become embittered. We chose gratitude. We cherished appreciation. We learned to love ourselves. Our spirits did not break with the hardships of this disease.*

A major legacy in the CF community has been the capacity to face fear head on and still maintain an optimistic attitude towards life. I've done my best to parry the blows of the CF drama without being battered by them. Through suffering and pain we can still hold on to values like kindness, compassion, or non-harm. An ideal legacy is a life lived with no regrets. I could ask for no better community, no more life-affirming and evolved group of friends than those I've found among you. How lucky I've been to be able to be one reflection on your legacy's mirror. Thank you for being part of my legacy. ▲

Isa Stenzel Byrnes is 51 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

Who's In Your Corner? Mentor Support For Adults With CF Returning To Higher Education

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

Now is an exciting time to be an adult with CF working in higher education. In my work with our scholarships committee for USACFA over the past few years, I've seen an explosion of fellow adult community members pursuing new opportunities in education. Adult community members are dreaming bigger and thinking bolder about what may be realistic for them.

A variety of therapeutic advancements in my lifetime have helped to make these welcome changes possible. These include CFTR protein modulators, which I've never been eligible for due to rare genetic mutations. They also include off-label uses of therapies like inhaled corticosteroids, which quite literally saved my life about a decade ago. Likewise, the Americans with Disabilities Act and related legislation have made higher education more accessible for people with CF across all of the spectrum of disease presentation and progression.

In this transitional time, many adult community members also continue to struggle with serious exacerbations and lasting consequences. So it's never been more important for adult learners to have mentors with CF who can speak to "resume gaps" and other unique facets of education and career journeys when writing recommendation letters or providing verbal feedback about a prospective student or employee. Established professionals with CF can also speak to the particular strengths people with the disease bring to our learning programs and practice applications. And working with mentors in our target fields can help us envision and plan our own pathways.

These days our education and career timelines are often long and complex, with plenty of twists and turns. I think about my own college dean, who spoke of learning as a journey of constant "zigs and zags" that mentors help us navigate thoughtfully.

> *"I think about my own college dean, who spoke of learning as a journey of constant "zigs and zags" that mentors help us navigate thoughtfully."*

XAN NOWAKOWSKI

This general wisdom rings true now more than ever for adults with CF and our highly specific experiences. Regardless of whether we're on modulators, many of us are living much longer than we expected to and facing new horizons for our lives.

There's a personal angle here too, as always. I've worked full time—rather an understatement, but true in essentials—for my entire adult life. At one point, I had had three jobs. I've always been the primary source of income for my households. I never stopped working to do my graduate degrees. Being able to work in jobs that intersected with my education and provided valuable learning opportunities of their own was certainly a privilege and a boon, but it didn't help my lung function or my exocrine insufficiency in the slightest.

I'm well aware these days that I survived my journey through two master's degrees and a Ph.D. partly through sheer luck. I powered through a lot of things that should have landed me in the hospital. Sometimes I refused hospitalization outright because I knew well from my studies that the "biographical disruption" of time away would hurt me mentally and socially in ways I feared more than physical impairment or even death.

Absolutely none of this is a flex. Frankly, it's more of a horror story. These choices had terrible costs that I'm still reckoning with today. Some of these are fairly obvious: prosthetic teeth, rebuilt gums, destroyed joints, painful breathing, muscle tremors, impaired circulation, kidney damage, heart problems, and perpetual exhaustion. It

hasn't all been bad news, certainly. I've never had a major organ transplant or been listed for one. Because I live at sea level, I haven't needed supplemental oxygen for any sustained amount of time. But I could do several things physically in my early 20s that I cannot do anymore. Like many adults with CF born in the early 1980s, I have lost many things I'll never get back.

Perhaps most of all though, I internalized a toxic culture of literally working myself to death—something I've dedicated my career to fighting against at a structural level. I've spent the last couple of years taking a hard look at this and making intentional changes to ensure that what I model for my students and junior colleagues reflects what I actually want them to learn. As a sociologist, I've understood well for some time that "do as I say, not as I do" scarcely proves effective as a teaching strategy. We should be able to see diverse examples of healthy behavior in our mentors to help us consider what thriving looks like for each of us individually.

Throughout all this, I never had another adult with CF I could look to for guidance or even just support. That's made for a lonely journey, if one that has taught me a tremendous amount about the importance of mentors who can understand our lived experiences firsthand. It has also taught me a lot about the importance of responsive mentorship in general, something I strive to practice consistently in my own work. Lately this includes not merely embracing but also actively *centering* how I model healthy time management myself for my mentees to assimilate and emulate.

Although I never had a mentor with CF, I absolutely had great mentors who understood how to work with someone with a serious chronic illness—and to learn new things from me in that process. I don't think I would have been able to do a lot of things that I've done in my career without their support. And I really have had great mentorship for my entire life, starting with my parents who raised me in their neuroscience research lab at Rutgers Robert Wood Johnson Medical School. They never for a moment thought that I should temper my aspirations for the impact I wanted to make in the world. My parents never exposed me to the idea that being sick meant I shouldn't set big goals or plan for the future.

> *"We should be able to see diverse examples of healthy behavior in our mentors to help us consider what thriving looks like for each of us individually."*

This may have been easier because although I got a tentative clinical diagnosis of CF at age 5, it would be another 27 years before I got the genetic testing that confirmed I had the disease. By that point though, my lungs were going downhill badly and I'd become much more disabled physically. I'm honestly not sure how I continued doing as much as I did work wise during that time. Fear of running out of time certainly factored in, though. People have asked me sometimes how I completed a Ph.D. in two years while working full time as a research project manager. "I was dying" is the only answer that has ever seemed honest.

Here's the thing, though: I probably *would* have died had my doctoral advisor not urged me to seek medical care and get extra rest when I was dealing with an exacerbation. She didn't have CF, but she understood well how to tell me to slow down in ways I'd actually heed. I still hear her voice in my head and replicate it for my own students. Moreover, I try my best to tailor the messaging to the unique circumstances and needs of each learner.

It's gotten easier to do this as my work with USACFA has introduced me to other adult professionals navigating career transitions and negotiating balance between work and health. It's also become more fun. We're stronger together than we are individually. I've seen this strength in action by teaming up with fellow community members to offer mentorship sessions hosted virtually by students with CF and their campus organizations. I remember vividly how staying on the videoconference for one of these sessions to talk with the student organizer led to a candid and heartfelt conversation about how isolating it feels to have no mentors who are like us—no one who understands the battles we fight daily. Providing that support means the world to me and highlights the power of listening and affirming.

With each new advancement in therapeutic options and access to them, adults with CF are surviving and thriving more than ever. We are moving towards a world where many in the adult community will not be going "back" to school for their first experience in higher education because they will never have left school for long periods of time. Mentorship needs for adults with CF will certainly keep evolving as these transformations in our survival and horizons continue.

Organizations that serve our adult

Continued on page 11

into the pickup truck, water dripping off my jacket as I sat down. It was 6:00 a.m. on a rainy Saturday morning in the mountains of North Carolina, just a bit east of Asheville. My two lifelong and faithful friends and comrades and I were navigating the winding mountain roads as we headed to our first ultramarathon event. We had carefully planned out our time for this morning to ensure early arrival at the starting line that would allow ample time to stretch and mentally prepare ourselves for the task at hand.

As luck would have it, though, here we were halfway to our destination and there was a large tree sprawled across the entire roadway. Apparently, it had been uprooted during the early morning hours by a quick, but fierce, storm. My mind began to fill with negative thoughts and questions as we drove back up the mountain, scouring Apple Maps for an alternate route. What if I'd done all these months of hard training just to arrive late and get disqualified before even leaving the starting line? Or perhaps, I thought, what if this is a good thing and the universe is just saving me from failing miserably and making a fool of myself? After all, the longest race I'd run prior to this was only a half marathon and today's scheduled run was 35 miles.

"Turn right here," I said, as our GPS guided us along this new route. "Time of arrival is 7:30 am, so we'll only be a half-hour late and fingers crossed the director will be understanding and still allow us to run." I laid my head back against the headrest and dozed off for several minutes. At exactly 7:30 a.m., we arrived at the now mostly deserted race headquarters and sought out the director. After a brief explanation of our events of the morning and expressing our eagerness to race, the director graciously agreed. He laid down some quick ground rules and directions and we hit the trail. Soon we were dodging puddles and settling into our pace. What proceeded will forever be one of the most memorable days of my life.

The first five miles of the run passed by quickly and easily. I was soaked to the bone from the continued rainfall but I didn't mind a bit. I increased the volume to my music and got lost in the rhythm as I matched my pace to the upbeat tempo vibrating in my ears. I smiled at the beauty surrounding me and thought to myself, "I must be the luckiest person alive to be able to be here in this moment and be healthy enough to even attempt such a thing." The enormous oak trees, moss-covered boulders, and peaceful river grabbed my attention for a moment. I reveled in the tranquility and then my mind wandered off again.

> "For me, though, the anguish that comes from knowing I have been given the gift of health and not having done more than living in mere mediocrity, becomes a much greater burden to bear."

"This view is incredible," I said out loud, as I stopped for a moment to catch my breath and stretch my calves, which were burning intensely by this point. I had just ascended about 2,800 feet in the last three miles and my body was letting me know. When I gazed out across the vast expanse below me, I could see turkey vultures gliding in the wind. Fog rose out of the valleys and created a giant curtain, mysteriously hiding the peaks from view as if waiting for a stage to be perfectly set and everyone in their places before revealing all its beauty. After some H_2O and energy gel, I continued the journey. The next 12 miles were filled with more amazement at the endless beauty, a stop to shed some clothing as the sun made an appearance and several stops at aid stations to fill my water bladder and refuel on food (candy, pickles, and pancakes).

"19.6 miles completed, now it gets real," I huffed to my friend Mike, as I checked the tracker on my phone. I'd finally caught up with him and we were beginning another long ascent. We would gain roughly 3,000 vertical feet over the next four miles. It was at this point that my body and mind screamed out to quit and simply did not want to go on. But I pushed forward—up and up. The sun kissed my skin and crystals of salt built up on my face and arms.

Around mile 29, I left the last aid station refreshed after being baptized with a bucket full of cold creek water and some Oreo cookies in my stomach. The combination of sugar and adrenaline gave me a second burst of energy and my body switched into cruise control for those last six miles. When I approached the end of the run, a tsunami of mixed emotions washed over me. On the one hand, I was completely ecstatic because I realized I'd come this far and was actually going to complete this monumental run. All the months of discipline and training had indeed paid off. The overwhelming sense of accomplishment made me beam with pride and smile like a kid as I did when I had just caught my first large-mouth bass. My whole life and journey with CF flashed before me like spools of footage from an old projector: all the clinic visits, the hospital stays, and the walk-a-thons. I then pictured each and

everyone's face—others with CF—that I'd known throughout my life who had already passed. My eyes welled up with tears causing my vision to blur. Why me? Why was I allowed to live long enough to receive the new miracle drug? Why was I lucky enough to have the most common CF mutation, meaning I was one of the first to have access to these drugs? Why me instead of Eva, or Jacob, or Danny—or the many other souls that also deserved a chance at a good life? I dwelled on these heavy thoughts for that last hour, which inevitably made that part of the trail zip by quickly, even though my pace had slowed significantly from the first half of the day.

The beginning and end of the race was at a camp situated in a "holler," as they'd say here in the South. There were mountains on both the east and west side, and the long narrow draw ran north and south. As I approached the finish line, I saw the sun start slipping behind the ridge and I could hear cheers and clapping echoing through the valley. I crossed that line with an exhausted body and mind, but a heart that was overflowing with gratefulness—for life, for health, for family and friends. I was grateful for my two best friends who were by my side that day, for the countless messages of encouragement that flashed on my phone screen while I ran. I was grateful for my amazing CF team at Duke University and for the miracle drug that had given me my life back along with the ability to live a much more fulfilled physical life than I ever could have imagined.

***

I feel all the gratitude in the world to be able to have lived this experience and to write about it to all of you. However, I also feel so terribly unworthy to share this, while there are still so many in the CF community without access to modulators or who have debilitating effects from the same drugs that are meant to improve our lives. For all of us who have been fortunate enough to have success with modulators, survivor's guilt can be a truly complex web of emotion to untangle. For me, though, the anguish that comes from knowing I have been given the gift of health and not having done more than living in mere mediocrity, becomes a much greater burden to bear.

So, as I continue to challenge myself and push my body to new heights and greater successes, I will do so with righteous vigor, intensity, and purpose in part to prove what can be accomplished, yes, but also as a tribute to honor those who never could so that in some small way their lives and memory will live on and never be forgotten. ▲

Marcus Miller is 31 years old and has CF. He lives outside Wilmington, North Carolina, about 30 miles from the Atlantic Coast. He has the best pup in the world, a Siberian Husky, named Emma and she accompanies him on most of his adventures. His true passions in life are hunting, archery, running/fitness, hiking and camping, and basically anything that gets him out in nature. If you'd like to follow his adventures or reach out to him, you can find him on IG @marcusrmiller or send him an email at extrasaltyrunner@gmail.com.

NOWAKOWSKI continued from page 9

community are innovating in all sorts of ways to meet dynamic needs within and beyond education. This includes embracing the breadth and diversity of higher education options. For example, some organizations are leaning into support for trade school students and learners seeking additional non-degree credentials. We're also seeing expanded support for college and university learning opportunities, from disciplinary focus to type of enrollment.

I feel more fortunate than ever to work with our scholarships committee at USACFA as our adult community lives longer and thrives more. This year we're working on some exciting new opportunities to better support all of you in your educational goals, and grateful for the support of our donors who are making that possible. In the process, we're embracing opportunities to champion adult community members navigating intersecting forms of oppression in education and in life.

As we grow our engagement in supporting adult learners, we're also embracing new ways of providing mentorship and support for everyone in our community who's pursuing additional education. Our Scholarships webinar from December 2022 (https://www.youtube.com/watch?v=7LbbU0i2ARY) offers a brief overview of what we do to support adults with CF in degree and credentialing programs. Our directors and contributing writers are also eager to connect with you and help you connect with impactful mentoring opportunities for your unique educational journey. Reach out and let us know how we can help! ▲

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.*

FOCUS TOPIC

RETURNING TO WORK OR SCHOOL

Believe It!

By Jeanie Hanley, M.D.

I couldn't believe it—I was a practicing doctor again after 17 years of being away! I had already tried twice in the last 10 years to return to direct patient care and, when those attempts didn't pan out, I started to give up altogether. Fortunately, I had forgotten to unsubscribe from an allergy and asthma physician job board, even after coming to terms that I would absolutely never return to work again. One week after making this final decision, a dream job popped up and fell into my lap for employment as an allergy physician in a private practice. I chose to work part-time hours three days per week. I guess the third time's the charm! Having worked there for one year already, I'm thrilled to say that it's been a fabulous ride so far.

Prior to starting the new job, I was fairly nervous—I wondered if I'd fall ill the moment I started as I had the first time I tried to go back to work 10 years prior. At that time, I spent one day treating allergy patients, went home and suffered a heavy episode of hemoptysis. That was it for that! The biggest difference between then and now was that in the interim I was able to start the CF protein modulator Kalydeco, which changed the course of my life. I distinctly remember after one week on Kalydeco wondering if the changes I was experiencing were real and sustainable. I thought, "Someone pinch me!" My health continued to improve year after year. The regular hospitalizations, productive cough, wheezing, GI issues pre-Kalydeco transformed into days of minimal respiratory and GI dysfunction, improved sleep, and more energy. These changes again motivated me to consider going back to work and I subscribed to an allergy doctor job board to peruse potential opportunities.

JEANIE HANLEY

In December 2019, Kalydeco was replaced with the latest modulator Trikafta, which seemed for me to keep back-to-back colds from progressing to full blown CF flares. This additional benefit prompted me to work in January 2020 helping a pediatrician friend with her patients. I'd barely gotten my feet wet when two months later the pandemic befell the world and I didn't want to risk my health treating patients in-person.

As I considered my third attempt to return to work, there were a multitude of other worries leading up to the first day on the job. So much had changed about medicine during the 17 years that I was on disability. I had only worked in a hospital environment with students, residents, fellows, and other allergy specialists supporting the clinic, which was a totally different structure than a private practice where you're it and you do just about everything for the patient. I thought my head was going to explode trying to remember all I had relearned and newly learned in the months leading up to the first day. I had maintained my allergy and asthma board certification all these years, but I wanted to have all the latest information at my fingertips as much as possible. All my knowledge was tied up into what felt like a tight ball of yarn, which I was worried wouldn't effectively unravel during a patient visit.

There was also Practice Fusion, the electronic medical record system that I had never used before. The whole system was like learning a foreign language. I also met a new set of coworkers—nurses, assistants, billing, and other staff—all of whose names I was trying to remember, along with figuring out who did what.

Nothing could prepare me for the first day, though. I was fairly slow and ended up spending 45 minutes on the first "follow-up" patient. The appointment length was slotted for 15 minutes. The patient requested a test that is only done in allergy research. It was a bizarre request and I tried to explain that only medical research environments conducted this test. It was very frustrating, but eventually the visit ended with what felt like half my hair pulled out. I thought, "Oh, my god, is this how patients are now?" Fortunately, the remainder of the day went smoothly as have the great majority of clinic days since. The patient population has overall been delightful this past year, and I've never come across anyone quite like that first patient so far. Whew!

Three months into practicing, I thought my biggest fear had come true. I contracted the flu, thank you to a

visitor from Las Vegas. Evidently what happens in Vegas does *not* stay in Vegas. My health went south very quickly and I ended up in the hospital on oxygen. Fortunately, the allergist whom I work with was very supportive and, with the help of a physician assistant, covered for me for three weeks.

During those three weeks, I started to feel strongly that returning to work would be too difficult and that with each passing day, the probability was slipping away. After two weeks I still needed oxygen and was fairly breathless with any activity, even showering. I could've used two or three more weeks off, but my colleagues had planned travel soon and wouldn't be able to cover. By the third week, I was off oxygen and, despite my fatigue and weakness, I returned to work (again!) sensing that this would be a day where I'd make it or break it and decide to stop working. I was seriously uncertain if I'd survive the day. As I entered the office, I almost broke down crying but kept it together holding back tears in my eyes. I couldn't believe I made it back. The day ended up being a welcome distraction from my health and showed me that I'd be okay; I was stronger than I thought. Fortunately, slowly but surely, over the next four to five weeks, my baseline lung function and physical and emotional strength returned.

> "The big chunk of 17 years on disability were tumultuous health years and it took every single one of those years to enable me to return to direct patient care."

The big chunk of 17 years on disability were tumultuous health years and it took every single one of those years to enable me to return to direct patient care. I knew others with CF who had tried to return to work full time and ended up leaving the job due to severe exacerbations primarily from not having enough time to take care of themselves. Their experiences pushed me to consider part-time work only, which has been essential to success this past year. Part time allows me to fully take care of myself, get plenty of rest, exercise, keep up my daily breathing treatments, and appreciate time off! Also, I had a feeling that setbacks would occur but hoped they wouldn't be so severe as to stop me from working altogether. I believe anyone going back to work can realistically expect that tweaking of your routines will be needed and yes, setbacks will occur. In a supportive work environment, setbacks don't signal the end of employment but can be overcome. ▲

Jeanie is 61 years old and has CF. She is a physician who lives in Los Angeles, CA, with her husband John. Jeanie has three amazing grown children and two beautiful granddaughters. Her allergy practice is in Glendale, CA. She welcomes your comments and asks that you send them to cfroundtable@usacfa.org.

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people on Kaftrio also were projected to have 13 fewer pulmonary exacerbations over a lifetime than those on best supportive care. The greatest survival benefit relative to those on best supportive care was observed when patients started on Kaftrio at a mean age of 14.9, the study reported. Relative to best supported care alone, the median projected survival was 82.5 years for those who started treatment at ages 12 to 17. Among simulation patients started with Kaftrio between 18 and 24 years old, their anticipated lifespan was 77.9 years; it was 62.2 years for those who beginning treatment at age 25 or older.

https://tinyurl.com/2n32c7fz
AND
https://tinyurl.com/2jmspbwe

Cost Burden Among The CF Population In The United States: A Focus On Debt, Food Insecurity, Housing And Health Services

Advancements in the cystic fibrosis (CF) field have resulted in longer lifespans for individuals with CF. This has led to more responsibility for complex care regimens, frequent health care, and prescription medication utilization that are costly and may not be fully covered by health insurance. There are outstanding questions about unmet medical needs among the U.S. population with CF and how the financial burden of CF is associated with debt, housing instability, and food insecurity. Researchers developed the CF Health Insurance Survey (CF HIS) to survey a sample of people living with CF in the U.S. A total of 1,856 CF patients in the U.S. were included in the study. Of these, 64% faced a financial burden: 55% of respondents faced debt issues, 26% housing issues, and 33% food insecurity issues. A third reported at least one unmet medical need: 24% faced

Continued on page 20

The Return To Work Experiment

By Suzanne Joyce

Practicing law was my second career. I went to law school at age 33. I worked full time after graduation for two and a half years then changed firms and switched to part-time work. Four years later, I stopped working because I was in that "seesaw" place—if I kept up with my work, my health suffered; and if I was fully compliant with my daily health demands, then I couldn't keep up with work. At 43 I wasn't bouncing back after exacerbations like I used to. I hated not being able to fully perform my work responsibilities, so I had to let work go. At age 59, three years post-transplant, I went back to work part time as a lawyer at my old firm for one year. I worked remotely from Florida with my firm in New York City. One benefit of COVID-19 was that it proved to employers that people can work from home successfully. I wanted to return to work as I felt so much better physically and I was desperately seeking intellectual and social stimulation.

I thoroughly enjoyed using my brain again. I did legal research, drafted litigation papers, and discussed cases with partners and associates. I also enjoyed the sense of purpose and accomplishment I regained. I had a job to do. It felt good to make my own money again.

After so many years living without a schedule, I struggled to establish a schedule for working, eating, staying hydrated, and exercising. I had a tendency before to get tunneled in to my work and ignore a lot of these fundamental things. I was surprised how quickly I fell back into that pattern. Additionally, litigation is demanding and has deadlines so there really is no way to just work four to five hours per day. Some days the job demands that you keep going until the papers are done. Sometimes I would hit a wall mentally. It took a while for my brain to build up endurance and hone the ability to focus for longer periods of time. I didn't see that as a bad thing; it seemed normal that I would experience this after not working for so long.

SUZANNE JOYCE

> "I struggled to establish a schedule for working, eating, staying hydrated, and exercising."

I expected that I would have more social interaction with others in the firm. That was something I missed after I stopped working the first time. This did not happen and it was mostly because I was working solely from home—I never went into the office nor did I participate in social gatherings.

Surprisingly to me, I found that it was still a physical challenge at times to even work part time after my transplant. I developed GI problems beyond my normal malabsorption/constipation issues. Randomly, yet not infrequently, I experienced weakness and had to lie down for an hour or so. This of course was very disruptive to my work and frustrated me. I had more specialist doctor appointments, tests, and procedures than before transplant. It took me at least an hour each way to get to these appointments.

My contract ended this past February and was not renewed because of a new nepotism policy established at the firm (my husband works there). I chose not to look for another firm because what I had at my old firm were people who understood my situation and it was highly unlikely that I would find a new law firm that would permit me to have such a flexible work schedule. The legal field is by and large not supportive of people with health issues. At 60 years old, I did not feel like being put in the position of explaining my health situation, having the anxiety of interviewing, the additional anxiety over a potential "new job," or of having the feeling of disappointing someone because I can't always work the hours an employer needs.

Overall, I am glad I did this work for a year. At first, I was very angry and disappointed in the firm for not keeping me on. But now I am ok with not working and the absence of work pressure is actually something I appreciate. ▲

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.

Scholarships Offered By USACFA

USACFA proudly offers three different scholarships! Both the Scholarship for the Arts and the Higher Education Scholarship were set up in memory of a loved one. Our newest scholarship kicks off this fall with a one-time deadline of 10/30/2023. You may apply for more than one scholarship each year, but you may only be awarded one per academic year. If you do not win, your application can be moved to the pool of applicants for another relevant scholarship in the same cycle. For questions about future scholarships or anything related to the application process, please contact us at scholarships@usacfa.org.

Scholarship for the Arts (05/30/24):

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

The Scholarship for the Arts was established by Andrea Eisenman to honor her mother, Helen Eisenman. Helen was a single mother devoted to her daughter, Andrea, who has cystic fibrosis.

https://www.cfroundtable.com/arts-scholarship

IN MEMORY OF
LAUREN
MELISSA KELLY

Higher Education Scholarship (06/30/24):

The Higher Education Scholarship was set up by Nancy Wech, in memory of her daughter, Lauren Melissa Kelly. The academic scholarships of up to $2,500 are awarded to two adults with cystic fibrosis who are pursuing career certifications, associate's, bachelor's, and graduate degrees.

Any student seeking a degree in higher education, from associate's to doctorate, 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.

https://www.cfroundtable.com/highereducationscholarship

The William Coon, Jr.
SCHOLARSHIP

William Coon, Jr. Scholarship (10/30/23):

Any student seeking a degree in any of the following is welcome to apply: business, economics, communications, political science, information, project management, finance, accounting, public administration, or marketing. We believe that any higher education is a strong foundation for advocacy and involvement in the CF community.

William J. Coon, Jr. established $20,000.00 in scholarship funds to be awarded in $2,500.00 scholarships for four students each year over a period of five years, totaling 20 scholarships. Mr. Coon was both a cystic fibrosis patient and a businessman who valued the importance of education and "paying it forward."

https://www.cfroundtable.com/williamcoonjrscholarship ▲

Are you interested in establishing a memorial scholarship honoring a loved one from the CF community who has passed away? Please reach out to us at scholarships@usacfa.org to learn more. A member of our Scholarships Committee will follow up with you promptly!

Encourage Family and Friends to Sign Donor Cards

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To receive donor cards, call:

United Network For Organ Sharing 888-844-6361

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Elizabeth W. Hissing (in memory of Larry Miller)
Dorrie Hulsey
Robert & Susan Kelly
Carol Keenan
James Kuhlman
Johanna Libbert
Robert McLaughlin
Lori Morris-Hughes (in memory of Kathy Russell)
Nancy Moseley
Loretta Mulatz (in honor of her daughter Debra Radler)
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Wanda Olson
Paypal Giving Fund
Richard and Karen Quinn
Cheryl Reynolds
Judith Riley
Maureen Riordan
Gina da Rosso
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Sharon Thompson (in memory of Melissa Thompson)

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GOLD BENEFACTORS
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Cystic Fibrosis Foundation
Helen M. Eisenman Fund Scholarship for the Arts

DIAMOND SUSTAINING PARTNERS
($20,000 AND UP)

William Coon, Jr.
Marina Day – Trustee of the McComb Foundation

DIAMOND ENDOWMENT PARTNER
($20,000 AND UP)

In Memory of Lauren Melissa Kelly

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.

PHOTO BY ROBERT WRIGHT

Hidden Agenda

The invisible motivator

I'm standing in front of fifty architects and engineers
I'm telling the way I want it to be
So we help sustain mankind and our modern lifestyle.
So we'll stop blowing up mountains to find coal.
So we'll prevent climate change progressing.
But it all started with air quality
Me discovering the giant tailpipes in the sky,
spewing coal ash in my air so we can feel cozy indoors.
My audience can't see my core motivation,
my lungs struggling against the bacterial cesspool destroying them from inside.
These lungs crave fresh air, a deep unrestricted breath, healing.
Something these lungs will never experience again.
But their malfunction is your gain, they push me to fight for cleaner air.
The hope our successors will read about our short-sighted lifestyles in history books.
This is my fantastical vision, and these crippled diseased lungs that you don't see,
are really what motivated me, to want to feel like part of the solution.

-E. Hyman, 2010

FROM OUR FAMILY PHOTO ALBUM...

ASHLEY COLEMAN AND HER DOG, AXIL.

LAURA MENTCH CELEBRATES TURNING 70 IN STYLE.

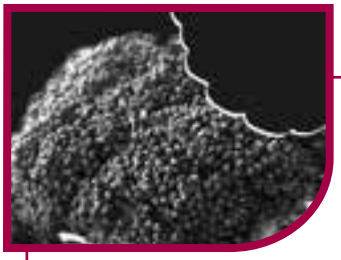
MATISON DEATON WALKED ACROSS THE GOLDEN GATE BRIDGE AND BACK. A GOAL MATISON HAD SET ORIGINALLY FOR ONE YEAR POST-TRANSPLANT BUT DID ONLY TWO MONTHS AFTER HER TRANSPLANTS.

ANDREW CORCORAN IN THE MIDDLE OF HIS PARENTS, MYRA AND JERRY CORCORAN.

MARCUS MILLER WITH THE FIRST TROUT HE EVER CAUGHT IN THE MOUNTAINS OF COLORADO, SUMMER 2023.

SUZANNE JOYCE WITH HER DOG, JADE.

JEANIE AND JOHN HANLEY IN NORTH CAROLINA WITH THEIR AMAZING AND CUDDLY GRANDDAUGHTERS, AMELIA WHO IS 2 AND ALANNA WHO IS 3 MONTHS OLD.



CULINARY CORNER

Chopped Broccoli Salad

By Maggie Williamson

We all know I have a thing for broccoli. The first recipe I shared with all of you was my Cream of Broccoli Soup. Well, here I am again with a family favorite and a perfect dish for summer! Chopped Broccoli Salad has been at every summer holiday get together since I was a kid. Summer to me is about fresh, bright, and easy food to eat and share with others. I love how crunchy this salad is and it looks great at any summer picnic or potluck! This salad can easily be made vegetarian or vegan by leaving out the bacon and using vegan mayo. It is also a super easy side dish to scale up for a large party. Happy summer, everyone!

MAGGIE WILLIAMSON

Chopped Broccoli Salad

Yield: 6 servings

Ingredients:

1 bunch of broccoli
½ pound of bacon, cooked until crispy and then chopped up (optional)
1 red onion
2 tbsp sunflower seeds
3 tbsp dried cranberries or raisins
3 tbsp olive oil
3 tbsp mayo
1 ½ tbsp cider vinegar
½ tbsp maple syrup or honey
½ tbsp Dijon mustard
½ tsp salt
¼ tsp pepper

Preparation:

Step 1:

In a mixing bowl chop the broccoli into quarter-inch pieces, including the stalks.

Step 2:

Dice the red onion finely. Add the dried cranberries, sunflower seeds, and bacon (optional).

Step 3:

In a separate bowl, add the olive oil, mayo, cider vinegar, maple syrup, Dijon mustard, and salt and pepper. Whisk together. Add the dressing to the broccoli mix and stir until fully coated.

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

TILLMAN continued from page 13

unmet prescription needs, 12% delayed or shortened a hospitalization, and 10% delayed or skipped a care center visit as a result of the cost of care. Income is the biggest risk factor for financial burden for people with CF, with people dually covered by Medicare and Medicaid particularly at risk.
https://tinyurl.com/3997v8v4

The Risk Of Colorectal Cancer In Individuals With Mutations Of The Cystic Fibrosis Transmembrane Conductance Regulator (CFTR) Gene: An English Population-Based Study

Studies have demonstrated a higher risk of developing colorectal cancer (CRC) in individuals with Cystic Fibrosis (CF). Life expectancy for those with CF is rising, increasing the number at risk of developing CRC. When compared to other population studies the overall prevalence of CFTR mutations in the CRC population was significantly higher than expected. This study demonstrates an increased risk of CRC among individuals with CF and a higher than expected incidence of certain CFTR mutations among those with CRC. When age-standardized the incidence of CRC was five times higher in individuals with CF. This increase in

Continued on page 23

Kathy Russell's Obituary

April 17, 1944 - May 13, 2023

It is with great sadness that we share the passing of Kathy Russell at age 79. Kathy was so many things, aside from being a founder of USACFA and *CF Roundtable*. She was a loyal friend, a licensed practical nurse, exceptional cook, canner of her own grown tomatoes and vegetables. She had a clever sense of humor and could make a statue chuckle. She imbued her empathy and caring to share her stories of her own CF experience on the pages of *CF Roundtable*, but also edited other people's submissions so that their voices shone through. Kathy was also part of a strong bonded team with her husband Paul. Wherever Kathy was, Paul was not far away. They were married on Marsh 27, 1965 (58 years!), and had a very happy and loving life together throughout life's curves and bumps. They weathered it together.

Kathy Russell and a few other people with CF heard a call for assistance from Lisa McDonagh to help her publish what was then known simply as *The Roundtable*. She was publishing it all on her own and, after each issue, she would be hospitalized with pneumonia, so she knew she needed help. Along with Kathy, there were: Melinda Timmermans, from Florida; Joe Kowalski, from Connecticut; Connie Knoles, from Oregon; Ken O'Brien, from Illinois; and Larry Culp, from Pennsylvania. These five people became the founders of the United States Adult Cystic Fibrosis Association, Inc. (USACFA). They registered it to be a nonprofit in 1990. While Kathy was a founder, she was designated as the treasurer in 1990 and later elected to the board of directors in 1993. She held many positions at the organization by being elected to every officer position from treasurer, secretary, editor, and director. She was president from 1997 to 2005 and managing editor of this publication from autumn 2011 to autumn 2019. She started her column called *Speeding Past 50* in winter 2006 when she was 62. In this column, she shared her own life lessons living with CF: Topics ranged from senior aches and pains to to sinus disease and what she did to combat it to wearing comfortable shoes because they just felt better than the coveted polka-dotted high-heals she loved. She adapted to life as it came her way with a smile and a good sense of humor.

KATHY RUSSELL

She was much looked up to in the CF community as she was also very involved in planning conferences and almost always attending them all over the country (before stricter infection protocols did away with most of them). She was also one of the oldest living people with CF most people knew. Oddly, her parents were told when she was diagnosed in 1956 at age 12, she would only live another 6 months longer. That they should take her home and cherish the time they had left. She showed those doctors! Which is why she was a great problem solver. She didn't take no for an answer if she felt she could do it. If she wanted something, she worked hard to get it. Her favorite job was working as a nurse in the newborn unit, and actually, probably the safest for someone with CF because it was free from infectious disease. She was a smart cookie.

Her favorite accomplishment that made her proudest, according to Paul, was being part of *CF Roundtable* and the CF community itself. She loved nothing better than talking to people who reached out to her and who might have wanted guidance, support or just an available ear to discuss CF. She was a kind and interested listener. In later years as she needed more time for herself, she mainly emailed with people.

As the years advanced, Kathy loved sitting in her comfy recliner doing 1000-plus-piece puzzles, taking day trips with Paul, cooking extravagant dinners and deserts for just the two of them, visiting friends and family when she could. She will clearly be missed by all who knew her. The CF community has lost a loving and fierce advocate. She leaves behind a beautiful legacy that generations will benefit from even if they did not have any connection to Kathy. She paved the way in many ways to living a long and fruitful life that enriched so many. It was an honor to know her. ▲

It is Kathy's wish that if anyone is so moved to make donations to *CF Roundtable* (https://www.cfroundtable.com/donorbox), CFRI (https://41224.thankyou4caring.org/donate-now) and CFF (https://www.cff.org/donate).

PET'S PERSPECTIVE

A Tail As Old As Time

By* Axil *and* Orbit, *translated by Ashley Coleman

Once upon a time, my mom was very sad because her old dog crossed the rainbow bridge and she was super lonely. Her friend made her go to the shelter where they keep puppies and kitties.

Just over seven years ago, my mom picked me up from that shelter. She says it was dirty, but I liked all the mud and playing with my six other siblings.

We were found in the middle of the road, picked up by this lovely lady, and brought to a shelter. These people who fed me named me Tuesday and Mom says that's cute because she picked me up on a Tuesday. Then she started calling me Axil but rarely calls me that anymore. At first, it was Axil but her friend's son said I was like a jumping bean so he started calling me Bean. I still don't know what a jumping bean is, but it's probably something perfect and beautiful since that describes me. Also, I have been the best dog ever and never do bad things.

Mom might disagree. I don't mean to do bad things because I have the biggest heart and love my mom. She calls me her treatment buddy, except I haven't seen her do many treatments lately, and it's been a long time since I stayed in the hospital. Our hospital here in Austin let me stay with Mom while she was sick. It was great! All the nurses and doctors loved coming to our room and I got lots of extra treats and bones! Trikafta made Mom very healthy, which is nice. I still miss my friends at the hospital some days. I hear rumors from my mom that her friend's dog, Husker, was the reason the hospital changed the dog visitation policy. I don't think I like this Husker character. I digress.

We get to go on more adventures than ever before. She takes me running and swimming and hiking. We even go to Starbucks and get Pupcups! We have the best times together!

About two years ago, Mom decided to get me a brother. He's an orange cat and does a lot of bad things. It made me nervous when we first got him because he climbs on everything! He even jumps on the walls. I think he needs to see a doctor because something is not right in his little brain. He makes Mom very happy and I love having a little brother...sometimes. Mom says I have to let him write about his story too.

***

Hello, my name is Orbit and I am very brave and strong! When I was little I almost died from an unknown virus, but I sure as heck didn't come from the streets just to have a little virus take me down. I was also very sick until I had my eyes completely removed because of something called microphthalmia. It just means that my eyes never fully developed and they were useless. I didn't need those things, anyway. Mommy says I see with my heart, which is weird because

ASHLEY COLEMAN AND HER PETS, CLOCKWISE FROM TOP LEFT: WITH AXIL AT ANIMAL SHELTER; AXIL SLEEPS IN ASHLEY'S HOSPITAL BED; ORBIT GOES IN FOR A SMOOCH; AXIL AND ORBIT HAVE A TÊTE À TÊTE.

that's not how hearts work.

Things I like: beating up my brother, stealing his food, dressing up, getting treats for being cute, jumping on the walls, climbing on everything, going anywhere in the car with my mom and brother (like the brewery), going for walks, and so much more.

I love life! I sit outside on my patio and overlook my kingdom and remind any other animals or people they are being watched. Mommy calls us her "stinky boys" and that seems a bit unfair because Axil is much bigger so he obviously stinks more.

> *Mommy says I see with my heart, which is weird because that's not how hearts work.*

I am a really good helper to my mom. I help her stay happy and give her so many kisses and snuggles when she is sad. I am so glad I chose her to be my mom! ▲

Ashley Coleman is 36 years old and has CF. She is the Social Media Manager for USACFA. Ashley lives in Austin, TX, with her dog, Axil, and her cat, Orbit (@orbittheblindguy on IG). She enjoys going to concerts, running, and exploring new things thanks to Trikafta.

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predisposition is likely to be multifactorial due to a combination of factors such as CFTR dysfunction, increased local and systemic inflammation, altered gut microbiota and the use of high fat and low fibre diets. The study also suggests a higher predominance of right sided tumors and an increase incidence in females, compared to the population without CF. This is a significant finding as right sided tumors of the colon have been shown to have worse survival and are frequently diagnosed at a later stage due to their symptom profile.
https://tinyurl.com/bdf2ed5y

Human Papillomavirus Prevalence, Persistence And Cervical Dysplasia In Females With Cystic Fibrosis

A higher risk of human papillomavirus (HPV)-related cervical intra-epithelial neoplasia (CIN) is suspected among females with cystic fibrosis (CF). The researchers found that transplanted females had high-risk (HR-HPV), abnormal cervical cytology and CIN prevalence rates compared to the general non-CF population. Although HR-HPV prevalence and persistence were globally not significantly different in non-transplanted females compared to the general population, high frequencies of abnormal cytology and CIN were reported. Cervical cancer screening and prevention should be promoted among females with CF.
https://tinyurl.com/2hm2pvuu

Disease Severity Of People With Cystic Fibrosis Carrying Residual Function Mutations: Data From The ECFS Patient Registry

People with cystic fibrosis carrying residual function (RF) mutations are considered to have a mild disease course. This may influence caregivers and patients on how intensive the treatments should be. Demographic, clinical characteristics, lung function and death probability of patients carrying at least one RF mutation were analyzed and compared to patients homozygous to minimal function mutations (MF). Patients carrying RF mutations were older, diagnosed at a later age, had lower sweat chloride at diagnosis and better FEV1pp at each age group. However, their FEV1pp declined with age and rates of chronic *Pseudomonas aeruginosa* increased with age. 4.5% of RF patients were treated with oxygen and 2.61% had a lung transplant. With increasing age, 26.6% of RF patients were treated with pancreatic enzymes associated with a more severe lung disease. RF patients had shortened life spans, with mortality starting around the age of 20 years. Thus, patients carrying an RF mutation experience a decline of pulmonary function with age, leading to life-shortening. Standard of care therapies and augmenting CFTR function may improve their survival and quality of life.
https://tinyurl.com/46dzfrct

Lung Infection May Be Less Transmissible Than Thought

Recent research found that various strains of the bacterium *Mycobacterium abscessus* were genetically similar, stoking fears that it was spreading from person to person. But a new study by researchers calls those findings into question, offering an alternative explanation behind the genetic similarity of clinical clusters. This suggests that the pathogen may not be that prone to person-to-person transmission after all. The results argue against direct person-to-person transmission in clinical settings and instead point to *M. abscessus* infections being acquired from the home or other environmental exposures.
https://tinyurl.com/2hpdrf8s

Breakthrough Drug Combination Remains Safe And Effective Long-Term In Patients With Cystic Fibrosis

Patients 12 years and older who received the combination regimen of tezacaftor (TEZ) and ivacaftor (IVA) for

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CHAPTERED LIVES

Young Lungs, Old Soul

NEW COLUMN

By Andrew B. Corcoran

Most people in my life do not have Cystic Fibrosis. Inevitably, they are the ones who ask what it feels like. My reply is always the same…

Imagine breathing through a straw with your nostrils closed. Then, while breathing only through that straw, run up and down a flight of stairs—that feeling, that drowning, that breathlessness, that desperation for air, and that panic when it doesn't come—imagine that all the time.

I was 6'1" and 150 pounds at 17 in high school. I wandered through hallways and life with zero responsibilities and zero interference from cystic fibrosis. My senior yearbook quote read: "High School was a three-ring circus; I was the lion tamer." As far as I was concerned, CF was a disease in name only—it had no hold on my body or mind and, even though I was well aware of it, the effects of CF felt about as destructive or interfering as a landslide on a sand dune.

My freshman year of college was barely memorable for all the right reasons. It was 1998 and I was young and healthy. My lung function was near 100%. I was on the crew team at St. Joseph's University, doing two-a-day workouts and raced in the winter regatta on the Schuylkill River in November. There were few, if any, reasons to even think about CF. I had it, I knew it, and I ignored it.

By September of 1999, that changed. The summer between my freshman and sophomore years is when cystic fibrosis decided to demonstrate and illuminate precisely how different I was and precisely how formidable it was.

I was living in Galway, Ireland, for those few months and, when I returned to Philadelphia, my lung function had dropped by 50% capacity. Back-to-back-to-back cases of pneumonia took half of my lung function in less than four months.

Despite that, going back to college that second year seemed imperative. I was desperate to continue the life I had known. Still, every two weeks, I would drive to St. Christopher's Hospital in Philadelphia, where I would watch my FEV1 dip with every visit: three percent drop, five percent drop, eight percent drop. The PFT machine was my nightmare, slowly parading an objective truth that all CF patients know—as we lose our lungs, as we lose our breath, we lose ourselves, both body and mind.

There are different kinds of anxiety in life. There's the anxiety of running into traffic on your way to a meeting, the anxiety of misplacing your car keys or cell phone, or the anxiety of being late on a bill payment, for example. There's another altogether separate layer and level of anxiety—something otherworldly, supernatural, impossible to explain or understand unless you, yourself, have experienced it. That is the anxiety of watching your body being overtaken by cystic fibrosis, the anxiety of knowing your lungs and body are failing you, abandoning you, discarding your will and desire—your frenzied, frenetic, and frantic attempts to survive.

> *"My life had altogether taken on a new meaning and was now defined by terms that my peers knew nothing about. I couldn't relate to anyone."*

ANDREW CORCORAN

I finally left college my junior year. It was Thanksgiving and I drove myself to St. Christopher's Hospital. I remember crawling across the street from the parking lot to the automatic doors of the hospital. I remember dragging my feet over the asphalt and breathing so fiercely that I nearly lost consciousness. I remember feeling the cool air on my face when I stumbled through the hospital doors and collapsed into the arms of my pulmonologist.

As I lay in that hospital bed as the holiday came and went, a switch turned on in my mind. A simple switch with immediate effects and terrible consequences. I would no longer talk to people, I would no longer express love or joy, I would no longer smile or laugh or try and enjoy this life I was living. Instead, I would survive. I

would *not* die. And, if a miracle did happen, if a call came for a transplant, then, and only then, would I allow myself to feel again.

I was surrounded by mountains of love, but my mind told me, *express nothing, love no one, cut everyone and everything off–then, when you die, you won't be as missed, it won't be as tragic.*

Two and a half years. Thirty months. 913 days of waiting. Then, on July 21, 2002, the phone rang.

The gratitude for my transplant was all-consuming. It was immeasurable and still is. It is difficult to express this level of relief, difficult to find the words to speak or thoughts to think, though with this new life came new complications—survivor's guilt, PTSD, confusion, anger, and overwhelming joy.

Rather than taking the necessary time to heal emotionally, I tried returning to my old life. At 24, I returned to college. I was a "college kid" again, at least in name.

Sitting in those classrooms, I found myself wondering and whispering to no one and nothing in particular how meaningless it all seemed. I was no longer a kid, partying on the weekends, skipping class with a hangover from last night's bender. I was no longer living in some cramped dorm room with eight other boys pretending to be men. My life had altogether taken on a new meaning and was now defined by terms that my peers knew nothing about. I couldn't relate to anyone. On top of that, I felt like time had passed me by. Every person my age was already working and starting careers, getting engaged, moving onto the next phase of their lives. How could I catch up? What shortcuts could I take? Before my transplant, my life had become years of racing towards a red light and now, I was seeing green lights for miles, but with no direction in mind, so I just kept driving.

In 2005, I graduated from St. Joseph's University with a degree in literature. I worked for a few years in the television and film industry. The transplant allowed me to breathe. My physical health flourished, but the emotional distress that I had ignored had finally became a shadow that was impossible to escape. And so, one day, without much warning or consideration, I did the only rational thing I could imagine: I left.

I left my life behind. Everything. All of it. Family and friends, my possessions, my job and money, security and relationships, even my doctors. I packed a bag, bought a one-way ticket for Europe, boarded a flight, and began a new life. At 26, I was just beginning to understand what my life was going to be and who I would be in it. Cystic fibrosis was and will always be a part of my story, but only one chapter. The rest are for me. ▲

Andrew Corcoran is 43 years old, has CF and received a lung transplant in 2002. He now lives in South Jersey with his family and friends. He is a writer. He can be reached at AndrewBCorcoran@gmail.com.

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four years did not experience any serious safety concerns and tolerated it well, suggesting its appropriateness for long-term use. Not only did patients tolerate the combination regimen over the course of the 96-week study, but they also maintained the improvements they had achieved in an earlier trial. The trial assessed the long-term efficacy and safety of treating patients with a combination of two CF modulators, TEZ and IVA (TEZ/IVA). While this trial was still being conducted, a three-part regimen composed of TEZ/IVA plus elexacaftor (Trikafta) was approved. With TEZ/IVA, there is about a 2% to 3% increase in lung function, whereas with the triple, there is a 15% improvement in the lung function. Patients are also less likely to require hospitalization. Although clinicians recommend taking the three-drug combination regimen because of its increased effectiveness in improving lung function compared to TEZ/IVA, some patients cannot tolerate it and so must remain on the dual therapy. This study's findings that TEZ/IVA is safe and well-tolerated over the long term also builds confidence that two of the three drugs in the standard-of-care therapy can be used safely over time. Again, this is important reassurance for patients who will likely take the drugs for the rest of their lives. Should safety issues arise, these data will be useful in pinpointing the problematic agent in the combination regimen.

https://tinyurl.com/2lnsjd5l

AND

https://tinyurl.com/yrskmtyt

AND

https://tinyurl.com/3m6uaejd

AND

https://tinyurl.com/36bp9k85

Orkambi Eased Airway Inflammation, Boosted Bacterial Diversity

Treatment with Orkambi (lumacaftor/ivacaftor) for six months lessened airway inflammation and enhanced airway bacterial diversity in people with cystic fibrosis (CF), but only when administered before patients were chronically infected with the bacterium *Pseudomonas aeruginosa*. The findings suggest that CFTR modulators should be started as soon as possible, ideally before the patient is chronically colonized with *P. Aeruginosa*. Since bacterial infections

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Transplant: A Lesson In Me

By Matison Deaton

Six years ago, in March 2017, I packed all of my medications, medical supplies, and devices into three suitcases, put my mask on for a 12-hour day of flying (even pre-pandemic I always wore a mask when I flew), and headed off to the first stop on my journey to transplant: Duke University Hospital. I would spend six long days there, in North Carolina, going through evaluation for a double lung transplant. It was grueling and stressful and yet I had hope for the future. I was cautiously optimistic that this could be my time for a second chance at life. I would no longer need oxygen 24/7. I would soon not need the daily intravenous and oral antibiotics I had been taking continuously for eight years at that point. I would be able to start living life again instead of existing in a state of slowly dying. And then, after those six days of invasive testing and countless appointments with doctors, and just one day after returning home to Anchorage, Alaska, the phone call came that changed the course of my life.

I was not a candidate.

Now, let me take you back to 2009. I was 17 and in my junior year of high school. I should've been spending time with my friends before graduating, figuring out which colleges I was going to apply to, and prepping for my last year at Dimond High. Instead, I was on a mandated quarantine at my mom's house where I was living half the time, isolating from the people around me, and waiting to find out the results from a sputum sample I had given two weeks prior. I had recently been discharged back home after a week of debilitating fevers and body aches. My breathing had started to decline so badly that I had to quit my school's soccer team because I could no longer run. It was only the beginning of my decline with cystic fibrosis. In the hospital, after being stumped by the origin of my symptoms, my pediatrician brought in an infectious disease specialist—one who would later become like family to me. She, too, was completely stumped after running blood tests, collecting numerous sputum samples, and checking every organ in my body for the culprit.

MATISON DEATON WITH HER OLDER BROTHER, NICK, WHO WAS HER PRIMARY CAREGIVER AFTER TRANSPLANT. THREE WEEKS INTO HER RECOVERY, NICK BOUGHT THIS SET OF STUFFED LUNGS TO FOREVER REMEMBER THE JOURNEY THEY HAD SHARED TOGETHER.

After a slew of inconclusive results, her best guess was that I may have some kind of mycobacterial infection. At that time in Alaska, the most common of these was *Mycobacterium tuberculosis* (TB). After getting my fevers under control and starting me on a treatment regimen, I was discharged and sent home under a strict quarantine due to the extremely infectious nature of TB. That infectious disease doctor sent my sputum samples out to independent labs to further identify what bacteria was growing. She told me that there were many different kinds of mycobacteria and that her gut was telling her it wasn't TB. She predicted that it would come back as either *Mycobacterium avium* complex (MAC) or *Mycobacterium abscessus* (*M. abscessus*). For some reason, whether by intuition or just curiosity, I asked her, in her opinion, which outcome would be worse for me. Her reply came easily, "My hope is that it's not *M. abscessus*."

Flash forward two weeks, and the results were back. It was, to both her

dismay and mine, *Mycobacterium abscessus*. I was instantly anxious. What did this mean? I had never heard of this infection before. I had never even heard of non-tuberculous mycobacteria in general. I had no idea what would happen, what this would cause, and just how devastating this outcome would turn out to be.

This infection became the biggest hurdle in my life, especially when it came to transplant. In fact, years passed between my Duke rejection, the first attempt of many at a lung transplant evaluation, and the next attempt I made because I was so distraught and disappointed. For my next few referrals to transplant centers, my pulmonologist's office sent my records out only to receive rejections in the form of one-page letters, sometimes even just one paragraph in length. Each one was quick and to the point: "*Mycobacterium abscessus* infection is a contraindication for lung transplantation." There would be no further look into my history or who I was. I was, to put it simply, too high risk.

This infection in particular made surgery very dangerous for me. For one, *M. abscessus* can colonize not just your lungs, but also your trachea, sinuses, and mouth. This means there is potential for it to eventually infiltrate transplanted lungs. Secondly, there is obviously a lot of cutting that happens during a lung transplant. If they were to cut into a pocket of infection as they removed my original lungs, the bacteria could spill into my chest and infect the new lungs or even the incision site, as *M. abscessus* is also a commonly known skin infection. And the worst-case scenario: If it spilled into my blood stream, it would travel throughout my whole body, eventually infecting my brain. One doctor described this as a "slow, miserable death," your body basically wasting away as it tries to fight off the infection, to no avail.

So, for a while, after nine total rejections in one year, I put the thought of transplant away. It was too hard to hear that my life was not worth the risk. At least, that's how it felt. Trust me, I understood those valid reasons why I was an "undesirable candidate." I would also not want those things to happen to me. Additionally, I understand the concept behind "do no harm," and that they don't want to put a transplanted organ in someone who may not survive because organs are in high demand. But how could I not take this kind of rejection personally?

> "My life meant something to me. No matter how many times people told me no, I could not give up."

To me, it was my life. It was nothing but personal.

And then, two years after the last rejection letter I received, after enough time went by to heal my emotional wounds, something shifted in me. I was ready to try again. I had grieved those devastating blows, those losses, and regained clarity through them. My life meant something to me. No matter how many times people told me no, I could not give up. This is part of what I learned from this process: It's not over until it's over. I know it's a cliché and we hear it all the time from people trying to give us encouragement, but I came to see that it was true: If my heart is still beating, if I am still breathing, it's not over.

So I tried again.

I rallied my home team, my pulmonologist, his nurse, and got to work. This time, rather than just sending a referral to the biggest transplant centers, I started googling centers that actually had a history of transplanting patients with *M. abscessus*. I put together a list of the top 10 I felt most likely to accept me, and my records were sent to the first one. My pulmonologist had decided we would go down the list, waiting for one to say no before moving on to the next. Within a week, I heard back. First, I would travel to Baltimore, Maryland, to be evaluated at The Johns Hopkins Hospital. Unfortunately, but not surprisingly, it ended with another rejection. But I kept going. Next on my list was the University of California San Diego. Here I received a brief phone call letting me down, which was at least a step up from a generic letter in the mail. Third on the list was the University of California San Francisco. Here, I received a glimmer of hope.

I began my relationship with UCSF at the end of 2019. They were upfront with me that they did perform lung transplants on patients with *M. abscessus*; however, because I had never been able to reduce the burden of the infection in my decade-long battle against it, they weren't sure yet whether I was a potential candidate. But, they had hope and again I found myself cautiously optimistic. It was the first time I had heard a transplant doctor tell me in no uncertain terms that he thought they could do it. Yes, they needed to discuss as a team and review my history further and they would have to meet with me in person before making their final decision, but it was the foot in the door that I needed.

At first, the process with UCSF was slow. It started with virtual appointments and testing done locally, as I was

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still living in Anchorage. When the pandemic hit, my chance to fly to San Francisco to be evaluated in person came, but I was no longer comfortable getting on an airplane. My thought was it wouldn't be worth the journey if I caught COVID-19 on the way there and could no longer go through with the evaluation. At this point, the vaccines had just barely come out. I decided to wait, hoping my health would at least stay stable until the time felt right.

Finally, in 2022, that time came. UCSF came up with a plan to get me there, and it began to move quickly. In April, I was brought into my local hospital to be transferred via Medevac to UCSF Medical Center. My oxygen settings at the time were too high to take a commercial flight, so this was my only option. When I arrived, I was inpatient for five days, going through the formal evaluation and meeting their whole team. I was then discharged but had to stay in the area. If they were to accept me to their program, I would need to permanently remain in San Francisco, at least until two years post-transplant. On May 18, they called me with their decision.

This time, I was accepted. My heart leapt. I could not believe it.

I was placed on the waiting list for a double lung transplant and a kidney transplant as, by this time, due to my continuous antibiotic treatment for M. *abscessus*, I had injured my kidneys irreparably and developed renal failure. The goal was that I would receive both organs from one donor.

Because this opportunity had eluded my grip for so long, I naturally doubted it would come to fruition. Due to my previous experiences, it became instinctual to protect myself from further rejection this way. However, despite my disbelief, on August 20, 2022, after spending three months on the waitlist, I received my life-saving transplants form a deceased donor. It was the hardest, most emotional thing I have ever been through. Words cannot sufficiently describe the feeling of waking up that first day; of taking your first breaths after extubation; of being told you no longer need that oxygen cannula; of seeing your family's reactions as you begin to change before their eyes. It was so much to process; my brain could not keep up. Even to this day, I have a hard time wrapping my mind around the experience. Recovery was difficult, frustrating, and slow. And then, suddenly, it was fast, rewarding, and I began to see it for the blessing it was. I am forever grateful to UCSF, the incredible team there, and, most importantly, my donor and their family, for making the most courageous, selfless decision.

As I said before, I learned so much through my tumultuous journey to transplant. It was far from easy, and absolutely nothing happened the way I expected it to. But I learned so much about myself and gained so much knowledge about what's important to me. And that timing really is everything. In hindsight, I am thankful it happened when it did and not a moment sooner. I needed to go through, and grow through, those trials to become the person I was at the time of my transplants. I gained perspective in my life that I wouldn't have otherwise and became the person I am now.

My years of fighting and advocating for myself taught me that I am capable. I have power within me, even at my weakest. At one point during my recovery in the hospital, after I was brought to the step-down unit, I had a conversation with my lung transplant team. I felt the need to thank them for taking a risk on me, for giving me this second chance. As I finished my heartfelt statement of gratitude, the response from one of the three doctors sitting in front of me caught me off guard. His opinion was a little different than mine: I had myself to thank for this gift of life—"you got yourself here." He felt I had made them take that chance on me. He credited my grit, my perseverance, and my unwillingness to give up as the true reasons for my getting to that moment. I beat down their door and they let me stay.

I also learned that life is made of the small moments, the ones you might look over had you not been paying attention. It is not the big moments of excitement and adventure that I longed for all those years I spent too sick to fully live, thinking that my future was just my death. It was the tiny, miniscule ones. I wanted nothing more than to sit on the couch on a Friday night, watching a movie cuddled up with my cats, and have absolutely nothing else on my mind. I wished for the day I could play a board game with my family again, in the comfort of my own home, on my own time, instead of at my bedside in the hospital, when a nurse wasn't connecting me to an IV line. I always thought of the day I would be able to walk out the door again at a moment's notice, not having to think about how many oxygen batteries I needed to carry with me or when my next antibiotic was due, counting the minutes until I could get back home the whole time because just the action of getting dressed was so exhausting.

Before my transplants, every moment of my existence seemed to hang in the balance. To be able to exist quietly, softly, presently, within the simple, mundane moments of my day, is all I ever wanted. Transplant, my experience up to it, made me see what matters to me. It taught me to slow down and take in the view. And it showed me that even in my weakest moments, I am strong. ▲

Matison Deaton is 31 years old and originally from Anchorage, Alaska. She now lives in San Francisco. Matison has two black cat furbabies, loves puzzling, and walking in and connecting with nature. She also finds joy in being a part of the CF community. Matison is on the Rose Up Committee and has dedicated time to spreading awareness for CF and now organ transplantation.

What is the Boomer Esiason Foundation?

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

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

What does BEF do?

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

- **Educational Scholarships**
- **Lung Transplant Grant Program**
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Mailbox

I'm proudly enclosing a check in honor of my daughter, Debra Radler, who achieved the miracle of celebrating her 60th birthday. When she was born, the life expectancy was 18 years. None of this would have been possible without the dedication of USACFA. I am beyond thankful to everyone who has worked so hard to have made this miracle happen! God bless!

Loretta Mulatz

Kathy was like a sister to me and my sister, Paige. We have spent the last few days sharing stories and our sadness at her death. Paul called us and other cousins shortly after Kathy had died. I write as I receive your newsletter that Kathy and others started in 1990. What a journey it has been. I've learned so much over the years...I'm sure there will be an article about Kathy and though it will be sad to read about her passing/it's amazing the legacy she leaves behind.

Kathy was an amazing individual who lived and loved her life—especially with her best friend, Paul.

In tears,
Laurie Worth

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commonly occur in the lungs of those with CF, the benefits of Orkambi also may be linked with changes in the airways' microbiota, the community of microorganisms living in the lungs, and a reduction in inflammation. Importantly, a correlation between diversity of the microbiota and lung function was found, with patients who had worse lung function showing significant increases in the relative abundances of the bacteria *Pseudomonas* and *Lautropia*. Those with a ppFEV1 of 80% or higher, showed an abundance of the bacterium *Streptococcus*, *Porphyromonas*, *Actinomyces*, TM7x, and *Peptostreptococcus*. In terms of fungi (mycobiota) diversity, no differences were seen regarding age or lung function. Calprotectin — a measure of airway inflammation — levels significantly increased with age and were negatively correlated with lung function and bacterial diversity, meaning higher levels of calprotectin associated with lower lung function and less bacterial diversity. Calprotectin levels were significantly higher — indicative of enhanced inflammation — in patients with chronic *P. aeruginosa* infection when compared to those without it. After six months of Orkambi treatment, patients had a significant increase in weight when compared to baseline, as measured by the body mass index (BMI). In patients not chronically infected with *P. aeruginosa* at baseline, the levels of calprotectin were lower and the bacterial diversity significantly increased after Orkambi treatment. No differences were seen in fungi diversity.

https://tinyurl.com/munvhdn7

Orkambi's Anti-Inflammatory Benefits Seen In A Real-World Study

Beyond improving lung function, Orkambi (ivacaftor/lumacaftor) has potent anti-inflammatory benefits that may limit immune-related lung damage in people with cystic fibrosis (CF). Researchers found that one year of treatment significantly reduced the levels of pro-inflammatory signaling molecules in the bloodstream and airways of CF patients. Moreover, measuring these molecules in blood and sputum is a relatively non-invasive way to monitor disease-related inflammatory activity and the impact of therapeutics. In the lungs, mucus build-up can trigger an inflammatory response, resulting in lung damage. These responses are characterized by elevated levels of pro-inflammatory immune signaling proteins called cytokines in the lungs and bloodstream, likely contributing to tissue damage. After three and 12 months of therapy, there was an early and sustained improvement in mean lung function, and a significantly reduced sweat chloride concentration, an indicator of improved CFTR function. The need for intravenous antibiotics dropped by 68% in the first year of treatment compared with the year before, as did the mean number of pulmonary exacerbations, and hospitalizations for exacerbations, per patient. Blood tests showed Orkambi significantly reduced the levels of three pro-inflammatory cytokines — TNF-alpha, interleukin-1-beta (IL-1-beta), as well as interleukin-8 (IL-8), a cytokine well known to attract large numbers of inflammatory immune cells to the lungs in CF patients. No change in pro-inflammatory interleukin-6 (IL-6) or anti-inflammatory interleukin-10 (IL-10) was seen. Consistent with blood test results, Orkambi treatment significantly reduced sputum levels of TNF-alpha, IL-1-beta, and IL-8, but also of IL-6. No changes in IL-10 levels were detected. A comparison of bloodstream cytokines to clinical features showed patients with elevated IL-1-beta had worse lung function before and after one year of treatment. Likewise, those with high levels of IL-6 had a lower body mass index, a measure of body fat. Elevated TNF-

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Would you like to be featured in our publication?
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We would love to showcase you and your life in our publication. If you are interested in being interviewed for an upcoming issue of CF *Roundtable*, please email us at CFRoundtable@usacfa.org. We will pair you with an interviewer who will arrange a time to talk and then write questions based on your answers for you to fill out at your leisure. Each interview is crafted to bring out what each person wants featured about themselves. To go along with your interview, we ask for two photos that will go into the publication: one headshot and one photo of you with your peeps, family, pet, on vacation, graduation, etc. We want you to shine so that others can benefit from your experiences living with this shared disease.

TILLMAN continued from page 30

alpha, IL-1-beta, and IL-8 in sputum samples significantly correlated with more intravenous antibiotic courses, as did high IL-6 and IL-1-beta in blood samples, "suggesting subjects with high cytokine levels tend to exacerbate more compared with subjects with lower cytokine levels. The researchers concluded that in addition to ensuring significant improvements in parameters of lung function, Orkambi therapy produced sustained improvements in both circulatory and airway inflammation.
https://tinyurl.com/52hhaztt

Trikafta Rapidly Improves Lung Function In Real-World Study

Trikafta significantly improved the lung function of cystic fibrosis patients after only two weeks of treatment, with the benefits sustained after nearly four months. Early LCI [lung clearance index] measurements can help to assess the patient's response to this high-cost therapy. LCI is a test that involves measuring how long it takes for a tracer gas to be cleared from the lungs. No significant improvements were seen with Orkambi, however. Trikafta led to significant improvements in lung function within the first two weeks of treatment, as measured by both LCI and ppFEV1. These improvements persisted by weeks four and 16. Patients with more severe disease at baseline, as shown by a higher LCI, had the largest improvements after 16 weeks. LCI values after 16 weeks significantly correlated with baseline LCI, but no such relation was found for ppFEV1. Changes in LCI showed no association with baseline ppFEV1.
https://tinyurl.com/5n8vhr6y

Elexacaftor – Tezacaftor – Ivacaftor Treatment Improves Systemic Infection Parameters And Pseudomonas Aeruginosa Colonization Rate In Patients With Cystic Fibrosis A Monocentric Observational Study

The aim of this study was to methodically evaluate the effects of Elexaftor – Tezacaftor - Ivacaftor (ETI) treatment on clinical, biochemical data and *Pseudomonas* colonization in order to demonstrate its efficacy. Marked improvements were observed in biochemical markers of systemic inflammation as white blood cell count, levels of immunoglobulins A, G and M and albumin within 24 weeks of therapy. ETI treatment proved to be effective as seen by amelioration of lung function and sweat chloride concentration. Assessment of PsA colonization status revealed a conversion from a positive to negative detection in 36% of the cases after one year of therapy. Thus, ETI treatment effectively improves systemic inflammation parameters and shows promising results in PsA status conversion.
https://tinyurl.com/mrypjzr7
AND
https://tinyurl.com/kdc8v9ay

Eradication Of Mycobacterium Abscessus Infection In Cystic Fibrosis With Initiation Of Elexacaftor/ Tezacaftor/Ivacaftor

Mycobacterium abscessus is a nontuberculous mycobacterium that is often multi-drug resistant, difficult to eradicate and associated with a rapid decline in lung function in cystic fibrosis (CF). Elexacaftor/Tezacaftor/Ivacaftor (ETI) is a combination CFTR modulator that improves lung function and decreases exacerbations, but limited data exists

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about its impact on respiratory infections. A 23-year-old male with CF (F508del, unknown) was diagnosed with *Mycobacterium abscessus* subspecies abscessusinfection. He completed 12-weeks of intensive therapy, followed by oral continuation therapy. Antimicrobials were later discontinued for optic neuritis secondary to linezolid. He remained off antimicrobials with persistently positive sputum cultures. He then initiated ETI, and bronchoscopy eight months later suggested eradication *of M. abscessus*. By modulating CFTR protein function, ETI may improve innate airway defense mechanisms, facilitating the clearance of infections such as *M. abscessus*. This case highlights the potential positive implications of ETI on the challenging treatment of *M. abscessus* infections in CF.
https://tinyurl.com/zw44z834

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

Elexacaftor/tezacaftor/ivacaftor (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. The impact of ELX/TEZ/IVA was evaluated on the rate of lung function decline over time by comparing changes in ppFEV1 in participants from the Phase 3 trials with a matched group of people with CF 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 compared with a 1.92 percentage point annual decline in ppFEV1 in untreated controls. ELX/TEZ/IVA is the first CFTR modulator therapy shown to halt lung function decline over an extended time period.
https://tinyurl.com/2p8bp2j3

Real-World Safety And Effectiveness Of Elexacaftor/Tezacaftor/Ivacaftor In People With Cystic Fibrosis: Interim Results Of A Long-Term Registry-Based Study

To assess long-term effects of ELX/TEZ/IVA under real-world conditions of use, a 5-year observational registry-based study is being conducted. Interim results from the first 2 years of follow-up show that ELX/TEZ/IVA treatment was associated with sustained improvements in lung function, reduced frequency of pulmonary exacerbations and all-cause hospitalization, increased BMI, and lower prevalence of positive bacterial cultures. Additionally, there was a 72% lower rate of death and 85% lower rate of lung transplantation relative to the year before ELX/TEZ/IVA availability.
https://tinyurl.com/2x2hwsy3

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

Several people with CF (PwCF) cannot benefit from Elexacaftor-tezacaftor-ivacaftor (ETI) 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. Two main themes and six subthemes were identified. The first main theme (being deemed ineligible for ETI) had four subthemes (disappointment, information, happiness, and concerns). The second main theme (coping with a life without ETI) had two subthemes (lack of hope and continued hope). Thus, 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.

Continued on page 34

https://tinyurl.com/2un8cdwk

Sex Differences In Outcomes Of People With Cystic Fibrosis Treated With Elexacaftor/Tezacaftor/Ivacaftor

The effects of elexacaftor–tezacaftor–ivacaftor (ETI) use by sex prior to, versus after initiation, of ETI by pulmonary exacerbations (PEx), percent predicted forced expiratory volume in one second (ppFEV1), presence of *Pseudomonas aeruginosa* in sputum cultures, and body mass index (BMI) were evaluated. After treatment with ETI, there was a greater decline in PEx in males versus females. No statistical difference by sex for ppFEV1, presence of *Pseudomonas aeruginosa* or BMI pre- to post-ETI by sex was found.
https://tinyurl.com/yccd57tb

Personality Traits May Influence Quality Of Life With CF: Study

The personality traits of people with cystic fibrosis (CF) might influence their health-related quality of life (HRQoL). Scientists found that patients could be clustered into two distinct personality groups and that self-reported quality of life differed significantly between them. Personality — each person's unique pattern of characteristics and behaviors — may have a substantial impact on how they respond to stress and can influence life quality for people with chronic disease. But the potential role personality plays in patients' health and life quality hasn't been fully explored. Patients were split into two groups of five distinct traits: negativistic (pessimism/resistance to others' suggestions), schizoid (disinterest in social relationships), borderline (difficulty regulating emotion), depressed, and paranoid (being on guard). No specific personality trait was associated with clinical factors, including lung function, life quality, body mass index, or adherence to therapy. The patients in whom the traits were less accentuated were found to have higher quality of life than those in whom the traits were accentuated. Other health outcomes didn't differ between the groups. The researchers interpreted the findings as meaning that patients belonging to the cluster with lower scores for those five traits might be more "psychologically adjusted," whereas those in the other group are "vulnerable" to life quality declines. While the reasons underlying the relationship aren't clear, the scientists suggested it could be related to resilience, or the ability to "bounce back" from adversity and to cope well with difficult situations.
https://tinyurl.com/59x7ubct

Impact Of Chronic Medication De-Escalation In Patients With Cystic Fibrosis Taking Elexacaftor, Tezacaftor, Ivacaftor: A Retrospective Review

This study evaluated the effects of de-escalating cystic fibrosis (CF) supportive therapies in patients on elexacaftor/tezacaftor/ivacaftor (ETI). For many with CF, the clinical benefit of ETI exceeds that of supportive therapies. The primary objective was to assess non-inferiority of supportive therapies de-escalation by comparing the absolute change in percent predicted (ppFEV1) from baseline to month 1 versus the absolute change from baseline to month 12 after initiating ETI with patients. The investigators found that de-escalating supportive therapies for those on ETI was non-inferior to remaining on all supportive therapies. This suggests that medications may be able to be discontinued under the context of a de-escalation process, which may decrease medication burden and cost and increase quality of life.
https://tinyurl.com/mr2484d5
AND
https://tinyurl.com/4u9z88en
AND
https://tinyurl.com/bd3pzr6h

Medication Use In People With Cystic Fibrosis Before And After Modulator Therapy.

Changes in medication dispensings among people with Cystic Fibrosis (PwCF) following modulator initiation were investigated, using national prescription claims data in Australia. Immediately after modulator initiation, the mean number of dispensings was 0.9 higher in the modulator group, but then decreased to the level of controls after approximately 5 years. Modulator initiation in PwCF was associated with decreased dispensings of opioids and psychotropics, and increased dispensings of women's health medications, suggesting improved patient outcomes across multiple clinical domains.
https://tinyurl.com/yawf7bvu
AND
https://tinyurl.com/5brtt56t

Cystic Fibrosis: Management And Monitoring Of Respiratory Manifestations

This comprehensive article provides an overview of the pharmacological management of respiratory symptoms, highlighting the support that should be given to people with CF to enable them to obtain maximum benefit from their medication regime.
https://tinyurl.com/4r7zvsxy

Cystic Fibrosis: Management Of Non-Respiratory Manifestations

This article outlines the effects of CFTR dysfunction on the pancreas, liver and gastrointestinal tract. It also discusses the pharmacological management of these symptoms and describes the monitoring required for treatment optimization.
https://tinyurl.com/34akuuts

Acute Pancreatitis In Pancreatic-

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Insufficient Cystic Fibrosis Patients Treated With CFTR Modulators

The researchers present two cases of pancreatic-insufficient CF patients who presented with acute pancreatitis shortly after commencing elexacaftor/tezacaftor/ivacaftor modulator therapy. They propose that highly effective modulator combination therapy may restore additional pancreatic acinar activity, resulting in the development of acute pancreatitis in the interim until ductal flow is improved. This report adds to the growing evidence for possible restoration of pancreatic function in patients receiving modulator therapy, and highlights that treatment with elexacaftor/tezacaftor/ivacaftor may be associated with acute pancreatitis until ductal flow is restored, even in pancreatic-insufficient CF patients.
https://tinyurl.com/2zqc4jq9 ▲

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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United Network for Organ Sharing (UNOS): Phone: 1-888-894-6361 http://www.unos.org/
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