CF Roundtable®
A NEWSLETTER FOR ADULTS WHO HAVE CYSTIC FIBROSIS
WINTER 2023

How Writing A Novel About CF Taught Me It's Okay To Struggle

By Cindy Baldwin

My first book, *Where the Watermelons Grow,* was published in 2018. It was, in many ways, inspired by my life—about a disabled mom, a resentful daughter, and the ultimate recognition that families that look different from the mainstream can still be wonderful.

But it wasn't about cystic fibrosis. At that point, as a 30-year-old mother of a toddler, coming off a long period of frequent hospitalizations and health stress, I knew I wasn't ready to look deeply enough at my own baggage to write about a character with CF.

For years after writing *Where the Watermelons Grow,* I tossed around ideas for a book about a character with CF. I made notes, brainstormed, drafted a few chapters here or there. (In a painfully ironic twist, halfway through this process, a movie and tie-in book, *Five Feet Apart*, came out and both were eerily similar to one of my favorite ideas.) Nothing I started ever quite felt right. In the beginning of 2020, I decided it was time to write my CF novel, and I spent several days on a book tour trip studiously outlining the book I finally felt ready to write. And then, two weeks later, the world closed for the COVID-19 pandemic, and I wasn't sure I'd ever be able to write again.

When I finally did sit down to squeeze out a few words that spring, they weren't anything like the outline I'd carefully crafted. They were stream-of-consciousness poetry, and I followed where they took me. Eventually those words coalesced into a novel in verse that combined all of the things closest to my heart: poetry, dolphins, water, friendship, family... and cystic fibrosis.

CINDY BALDWIN AND HER NEW BOOK, "NO MATTER THE DISTANCE."

That book ended up being called *No Matter the Distance,* and it releases from HarperCollins Children's Books

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INSIDE THIS ISSUE

CF ROUNDTABLE
FOUNDED 1990
Vol. XXXIII, No. 1

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

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EDITOR'S NOTES

Happy New Year! 'Tis the season of new year's resolutions, yearly goals, intentions, and, for many, the start of another yearly health insurance deductible. This issue's focus topic beautifully dovetails that: accessibility with obtaining care.

For our focus topic, we're highlighting **Dr. Nowakowski**'s "Pearls of Wisdom" column. They discuss the importance of accessibility in virtual spaces for the CF community, especially in light of the ongoing pandemic, noting that accessibility is deeply *individual*, deeply *contextual*, and deeply *intersectional*.

Beth Sufian answers questions about COBRA extensions of coverage in her "Ask The Attorney" column. In this issue, **Isabel Stenzel Byrnes** writes about tools for managing and taming automatic negative thoughts, which she calls ANTs, as part of achieving peace and calm amidst our daily lives with CF. For our first installment of "Pet's Perspective," **Andrea Eisenman**, on behalf of Trixie, laments her forced hikes and walks in the big apple with her brothers Roscoe and Willie, and other adventures in her new home with "the Boss." **Colleen Adamson** writes about the various accommodations needed for both her pre-transplant and post-transplant life in the inaugural "Transplant Talk" column. **Maggie Williamson** shares her recipe for Mexican Potato Hash in her column this issue. You can also read about the latest CF research in **Laura Tillman**'s expertly collated "From the Internet" column. **Cindy Baldwin** notes that juggling CF care and parenting can be overwhelming. If you also feel pulled from both ends, be sure to read her four tips for making CF care accessible and manageable as a parent in the "Family Matters" column this issue.

Jerry Cahill, in the third installment of his series of interviews, talks about going back to work full-time for the first time in years and what that work/life/self-care balance will look like in the future. Additionally, you can read part one of a two-part story about **Rachel Johnston**'s climb to Everest Base Camp. Rachel writes about the challenges and wins of the first six days of her eight-day trek with 8,500 feet in altitude gain. Part two will be featured in our upcoming Spring issue.

Both the Arts Scholarship and our Higher Education scholarship are open and we're seeking applicants. For information on either of the two scholarships we offer, head to our website where you'll also find the application, requirements, and deadline for each.

USACFA is always looking for new directors and writers. If you're interested in joining our board or starting your own column in our publication, we'd love to connect with you. Send us an email at cfroundtable@usacfa.org so we can set up a time to chat about our mission.

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

Publication of *CF Roundtable* is made possible by donations from our readers and grants from Sustaining Partners - Fidelity Charitable Elliot Family Giving Fund (in memory of Shirley Althaus), Monaghan Medical, and Watson W. Wise Foundation; Pearl Sustaining Partners - Boomer Esiason Foundation, Cystic Fibrosis Foundation, and Scholarship for the Arts (in memory of Helen M. Eisenman); Diamond Sustaining Partner: Marina Day, Trustee of the McComb Foundation; and Endowment Partners - Nancy Wech (in memory of daughter, Lauren Melissa Kelly & in honor of son, Scott Kelly).

Information From The Internet...

Compiled by Laura Tillman

By 2040, Over 70% Of CF Patients In US Will Be Adults

According to a study, with continued advances in treatment, more people with cystic fibrosis (CF) are surviving well into adulthood and by 2040, nearly three-quarters of these patients in the U.S. will be adults and more than one-third of these adults will have minimal problems with lung function — specifically a score of 90% or higher on percent predicted forced expiratory volume in one second (FEV1pp). In the early 1990s, the expected lifespan for someone born with CF was around 30 years. Now, it's closer to 50. Today, more than half (57%) of people living with CF are 18 or older. It is likely that this growing, aging adult CF population will be more medically complex because of non-lung and age-related comorbidities, so coordination of care between CF care teams, subspecialists, and transplant centers will remain critical.
https://tinyurl.com/492tz8kd

LAURA TILLMAN

Contemporary Cystic Fibrosis Incidence Rates In Canada And The United States

The availability of new diagnostic algorithms for cystic fibrosis (CF), changing population demographics and programs that impact family planning decisions can influence incidence rates.

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LOOKING AHEAD

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

Winter (February) 2023: Accessibility and CF Care. (Current issue)

Spring (May) 2023: CF and the Pathway to Parenting. (Deadline March 15, 2023) What avenues did you explore in your pathway to expanding your family (becoming a parent, caring for children, playing a parental role in a young adult's life, etc.)? What were some of the biggest obstacles you faced? How have modulators impacted that process? What advice would you give to someone struggling to build their family? What does being a parent, guardian, or other adult role model look like for you? Did you look into multiple pathways to becoming a parental figure—for example, did you consider sexual reproduction, adoption, artificial insemination, surrogacy, in vitro fertilization, fostering, etc.? How did you decide which pathway you wanted to follow, and how do you feel about that choice now?

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

Autumn (November) 2023: The 10% Left Behind. (Deadline September 15, 2023)

ASK THE ATTORNEY

COBRA Extensions Of Coverage

By Beth Sufian, J.D.

Many people have submitted questions related to a COBRA extension of health insurance benefits. Nothing in this article is meant to be legal advice and is only information. Nothing in this article is meant to be a guarantee a person will receive an extension of health insurance benefits under COBRA.

If you have questions about laws related to health insurance, Social Security benefits, Medicaid, Medicare, employment rights, or education rights, you can contact the CF Legal Information Hotline (CFLIH) by emailing CFLegal@sufianpassamano.com or calling 1-800-622-0385 to schedule a time to speak with an attorney. Emailing is the quickest way to set up a time to speak to the CFLIH. All calls are confidential and there is no cost to the caller. The CFLIH can only provide information to callers. The CFLIH cannot provide legal advice and cannot provide legal representation to callers.

The CF Foundation provides funding for the CFLIH. CFLIH employees are not employed by the CF Foundation.

Question: If an employee and dependent child are enrolled in an employer-sponsored health benefit plan covered by COBRA and if both the employee and dependent child elect COBRA extension coverage upon a qualifying event, may the dependent child continue the COBRA extension coverage for the entire COBRA extension period even when the parent ends the COBRA extension coverage before the end of the entire COBRA extension period?

Answer: Independence of COBRA Election. The essence of this question is whether the covered child's COBRA rights are dependent on the parent's COBRA extension or is the covered child's COBRA election independent of the covered parent's COBRA election. The Code of Federal Regulation explains COBRA through a series of questions and answers. See 26 C.F.R. § 54.4980B-6. The regulation states that each qualified beneficiary (including a covered child) make an independent election to receive COBRA continuation coverage. Section 54.4980B-6 of the Regulation explains it in this manner: Can each qualified beneficiary make an independent election under COBRA? Yes. Each qualified beneficiary (including a child…)…must be offered the opportunity to make an independent election to receive COBRA continuation coverage.... An election on behalf of a minor child can be made by the child's parent or legal guardian. 26 C.F.R. § 54.4980B-6.

BETH SUFIAN

Question: If I am on COBRA, is there a way my health insurance coverage could end before the set amount of time that I am supposed to receive an extension of my health insurance benefits?

Answer: Specific Reasons COBRA Continuation of Coverage May End Before the Maximum Extension Period Expires.

Section 54.4980B-7 of the COBRA regulations specifies the events upon which a health benefit plan may terminate COBRA continuation of coverage before the end of the maximum extension period. A group health plan may terminate coverage earlier than the end of the maximum period for any of the following reasons:

1. Premiums are not paid in full on a timely basis;
2. The employer ceases to maintain any group health plan;
3. A qualified beneficiary begins coverage under another group health plan after electing continuation of coverage;
4. A qualified beneficiary becomes entitled to Medicare benefits after electing continuation coverage; or
5. A qualified beneficiary engages in conduct that would justify the plan in terminating coverage of a similarly situated participant or beneficiary not receiving continuation coverage, such as fraud.

See 26 C.F.R. § 54.4980B-7.

Significantly, a parent ending their COBRA extension period early is not one of the reasons that would permit a plan to end a dependent child's

COBRA extension period before the end of the maximum period.

A dependent child of a covered employee is a COBRA qualified beneficiary. The reasons specified in 26 C.F.R. § 54.4980B-7 apply to "qualified beneficiaries", which raises the question of whether a dependent child of a covered employee is a COBRA-qualified beneficiary. The COBRA regulations define "qualified beneficiary" as: any individual who, on the day before a qualifying event, is covered under a group health plan by virtue of being on that day either a covered employee, the spouse of a covered employee, or a dependent child of the covered employee. 26 C.F.R. § 54.4980B-3(i).

The COBRA discussion will be continued in a blog post on the USACFA website at the end of January 2023. Don't miss out on blog posts! You can sign up on our website to have blog posts delivered to your email address.

I'm wishing all of the *CF Roundtable* readers a healthy and happy 2023. The year 2022 was not one of my favorite years. I am looking forward to finding ways to make 2023 more peaceful, safer, and balanced. I am grateful to have so many friends in the CF community who provide me with love and support. I am so lucky to have so many friends around the country who brighten my days and provide joy and encouragement. As we enter our 25th year of the CF Legal Information Hotline, I look forward to continuing our work helping those with CF, their families, and CF care teams be informed of their legal rights. ▲

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.

SPIRIT MEDICINE

Accessing Our Inner World: Taming The ANTs

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

So much of our spiritual well-being is rooted in the energy in our heads and hearts. Our thoughts create emotions such as anger, sadness, resentment, or hatred, and those emotions, in turn, impact our actions. Our actions impact our relationships and our community.

Life with cystic fibrosis can be hard. We spend so much energy managing our bodies and keeping up with the demands of regular life. Since this is the one life we have, I believe we all want to live our best life possible. Cultivating inner peace and calm can parry the blow of life's challenges. And one way to find this peace and calm is to continually access and tame our inner worlds: our automatic thoughts.

We all have around 300 thoughts per minute, every day of our lives. Automatic thoughts are immediate, knee-jerk reactions to external stimuli. In relationships, when someone gives us a look, we interpret that person or their appraisal of us. If someone doesn't call us back, we create a story as to why. If we didn't get the job, it's because we are a failure. If we are hurt, we blame someone. Automatic thoughts are a rapid response to the world around us. They are usually learned as safe patterns early in life but are not always helpful in the present moment.

Usually, automatic thoughts are negative. They alert us to a problem. We catastrophize, judge, label, mind-read, blame, generalize, "should on ourselves," or personalize the issue at hand. We engage in unevolved defenses or revert to old unhelpful patterns about our worth and value. Automatic thoughts are also informed by learned biases and prejudices. Think of automatic negative thoughts as the acronym "ANTs." Your thoughts literally are like a row of ants, marching onward, chaotically, mechanically and mindlessly, sometimes multi-directionally toward something. They sniff out trouble and head for the garbage. Like ants coming indoors during the rain, ANTs become stronger during times of stress. And automatic thoughts *worsen* our stress by releasing more cortisol and adrenaline, hyping us up so we have the illusion of power and strength.

Often my automatic thoughts are about my body—anticipating a health crisis or doomsday. I recognize that many of my automatic thoughts are habits risen from my long history where these reactions were helpful to get treated when I was indeed in a crisis. When I was younger, many of my ANTs were social. Walking into a room of strangers, my ANTs told me people were thinking something negative about me based on what I looked like. Eventually, I learned I cannot mind-read and cannot be bothered by what others think and have since extinguished those ANTs.

> *"Many of my automatic thoughts are habits risen from my long history where these reactions were helpful to get treated."*

ISABEL STENZEL BYRNES

During my recent travels, I had to walk many miles to go to sites I wanted to see. My body was exhausted and weak, but I was traveling, and I wanted to push myself. My regular automatic thoughts included: "I'm diminishing," "This could be my last trip," "Something's wrong with my legs," "Maybe the cancer is spreading," or "I'm too slow for my family." The tangle of thoughts distracted me from what was in front of me. I noticed them, tried to relax around them, and told myself, "This is just a habit, and it's OK." Then I reminded myself compassionately that I am on chemotherapy, that I am out of shape, but that I am capable of walking, just at a slower speed with breaks. I felt gratitude for being able to be well enough to travel. I looked forward to my next nap, or my next chance to sit down. I thought of

the thousands of disabled and elderly who struggle to walk and yet keep going. I said a little prayer to God to help me develop the stamina and patience to be OK with what is. I communicated with my family about what their needs were, and whether there was another strategy for them to keep going at a faster pace. They told me they are fine with my pace, and I'm the one who is always in a hurry, and eager to do more and see more! And then I made up an excuse to stop for ice cream as a way to rest.

So what are some ways to cope with ANTs? True maturity is being able to pause, notice an automatic thought, assess whether it is helpful, and find ways to let it go or change it into more adaptive thinking. This pause turns down the dial of stress that ANTs turn on. The pause offers discernment; to wait for clarity on the thought at hand. This allows time and space before the next move to become clear. And for our minds to deliberately insert positive thoughts to refute the ANTs.

I have written in past articles that sometimes it can be helpful to find code words or phrases that we can use during an impulsive reaction like ANTs. These phrases serve as a yield sign to the brain and shift the focus to something more adaptive. A code word or image might be a stop sign or an untangling string or whatever helps hold the moment in space for a second. I like to imagine a magnifying glass to remind me to look more clearly at what's going on with my thoughts. If an ANT is about my body, I use the code word "*iiwii*" or "it is what it is." I try to remind myself what's working and going well. That doesn't mean dismissing the thought, and carrying it through to assessment and action, but rather to balance it with a reality-based thought about the situation at hand.

There are questions we can ask ourselves when an ANT pops into our minds. For example: *Is this thought true? What is this thought doing to my emotions and is that justified? Is this thought serving me or is this unhelpful? Is this a thought that is aligned with my values? What is my role or responsibility in having this thought? What am I learning about myself from this thought? Does this thought really matter? What does God want for me now?*

ANTs are often primitive animal instincts that serve to protect the ego from a perceived threat. They can be seen as survival instincts. Like any survival instinct there is a moment of decision making. Our conscious selves can decide whether to believe in the thought and then whether to act on it. Is there really a threat? What exactly is being threatened? (Many times, the threat is created in one's own mind.) If there really is no threat, ignoring the thought is good enough. If we act on the threat, the decision to take the high road or low road is in our hands.

Noticing automatic thoughts gives us access to our true, authentic, deeper Self. The deeper Self is the observer who is watching the theater of thoughts inside my head. The deeper Self is the healed Self who is in equilibrium, peace, and harmony. My true mind wants to be relaxed. If automatic thoughts are angry and resentful, then forgiveness is on the pedestal of the deeper Self. As people with CF, we ought to live in a way that supports the optimal functioning of our bodies, minds, and spirits. Through struggles, CF gives us an opportunity some healthy people don't get until much later in life: the potential to become more self-aware of our thoughts and control our well-being. To "know thyself" is life's greatest gift.

If we have cultivated a spiritual life, it can help to take automatic thoughts and pass them along to a higher being like Jesus, Muhammad, Buddha, God, Mother Earth, or our ancestors. We can accept our confused or broken ways and invite divine grace as a force in our lives. We can call on our values that guide us and help us to follow the loving path toward ourselves and others. There is so much more power and strength in love than in negativity! The spiritual paradox is both surrendering our control to something beyond ourselves while also owning our control in our lives. Taking responsibility for our own thoughts and feelings as well as for our actions and reactions is the key to spiritual freedom. Ultimately, controlling our inner world can help us be the person God wants us to be.

Afternote: If you have trouble accessing the parts of your mind that will allow you to notice and cope with automatic negative thoughts, I recommend seeking out a therapist to help you in this process. Therapy gives you undistracted space and time to focus on you and your betterment. Accessing our inner world first will help us become better advocates for ourselves in the larger world. And make us better people to others.

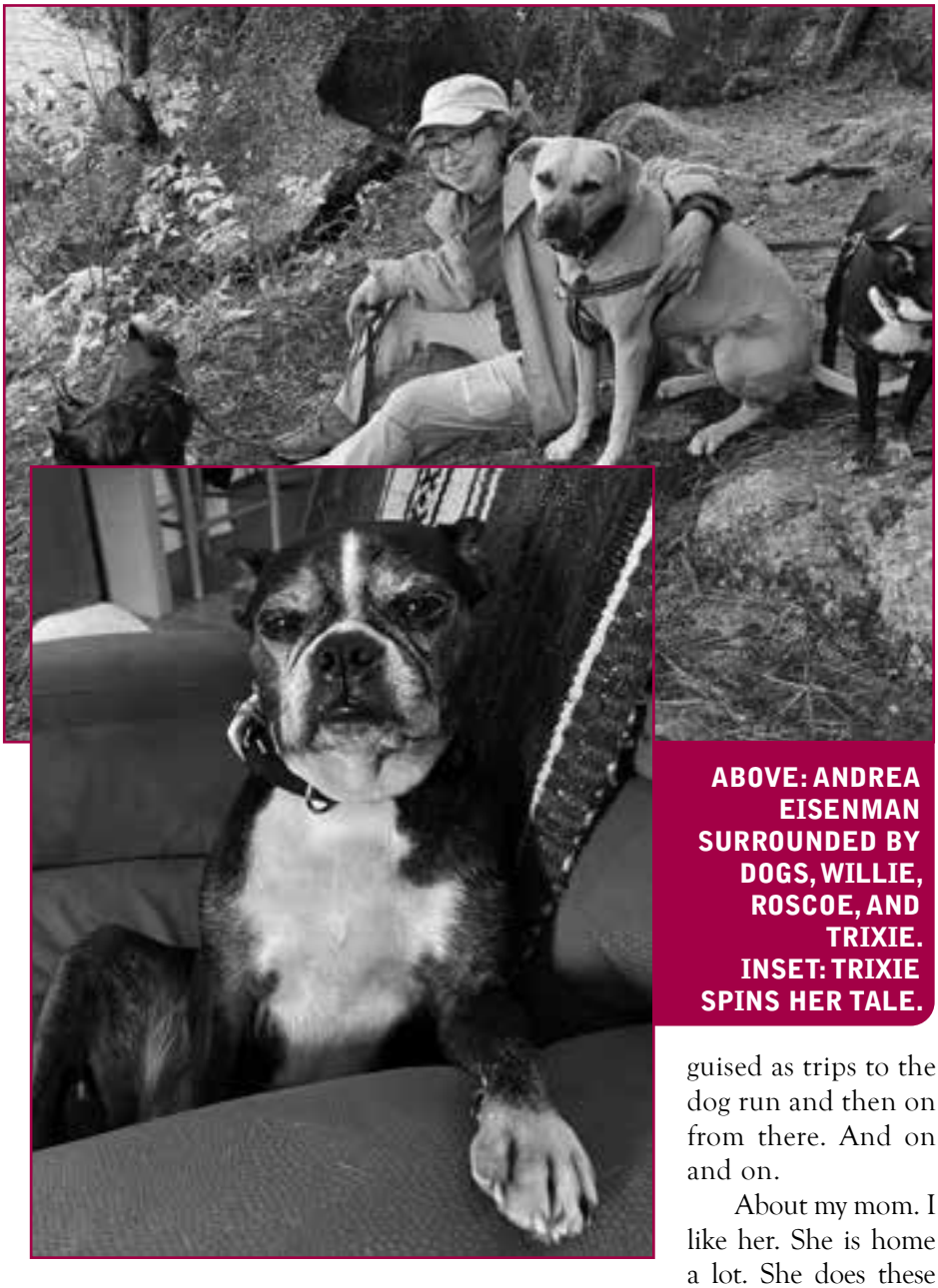
Accessibility to mental healthcare is a whole other challenge, that hopefully others have written about. Our best mental healthcare can be the safe and trusting people in our lives. Trained therapists are available on a sliding scale, remotely, and through organizations like CFRI. Spiritual counselors (chaplains, spiritual direction coaches, or Stephen Ministries) are also available. You can inquire at your local place of worship or check out psychologytoday.com. Always ask for a sliding scale if needed or call your insurance for a listing of covered therapists. I think I am preaching to the choir, but just wanted to offer a few tips on accessing mental and spiritual healthcare for this issue. ▲

Isa Stenzel Byrnes is 50 years old and has CF. She lives in Redwood City, California, with her husband, Andrew. She is 18 years post-lung transplant.

PET'S PERSPECTIVE

My Introduction To This Whacky Family

By Trixie, translated by *Andrea Eisenman*



ABOVE: ANDREA EISENMAN SURROUNDED BY DOGS, WILLIE, ROSCOE, AND TRIXIE. INSET: TRIXIE SPINS HER TALE.

It all seemed so innocent. I was living my life peacefully in Tennessee when I landed in the hoosgow somehow. I guess it was a kill-shelter because some nice people from NYC came from a rescue organization called Social-Tees and whisked me up north to the Big Apple. They believe they saved me from death row. Well, my tale (or tail), gets more involved from there.

A bit about me: I like a sedentary life—lying on the sofa, rolling on my back and waiting for belly rubs. Who doesn't?! And I like my food; just keep it coming!

This woman who adopted me—I will call her the mom of me—is pretty chill except when she decides we are going for walks. Not sure if they are for me or for her, or Willie, who I will get to shortly. I could easily spend my days lounging around. But no, we walk about one to two miles a day. It started slow, though, to trick me. I used to just refuse to go any farther than I absolutely needed to do my business—maybe two or three blocks each walk. But I now realize it was a ruse to get me used to walking farther. My mom's husband, who thinks he is the boss, and whom I will call the Boss, decided we should go hiking one day.

In this family, I am the last to arrive. First was Willie, the oldest at 13, who was owned by my mom's mom. He's pretty spry for an old guy. Then came Roscoe, the big lug, who is three. And I am around seven, but look a bit older. I will never tell my true age. A lady never tells. We make a Motley crew. I like them both, my brudders. Outside, Roscoe and I tear it up chasing after each other. Willie prefers a milder frolic so we leave him alone 'cos he old!

All of these walks that Willie, me, and my mom were taking got longer and longer so that we were out for almost an hour each time. I think it was also possibly to help me lose my "pandemic pounds" I put on (or the ones my mom did). It was disguised as trips to the dog run and then on from there. And on and on.

About my mom. I like her. She is home a lot. She does these "treatments" in the morning and at night. She cooks a lot and creates good smells in the apartment. She also has "health" appointments frequently and assures me and Willie she will be home soon to take us out again soon. Then we all nap together almost every day under the covers. Which I enjoy. Roscoe spends most of his day with the Boss.

I guess the hiking thing is what the Boss does. My mom takes me and Willie to Long Island and we go walking at the beach or in the park or in the yard—all level ground. So, I know nothing of hiking up a hill? No way. Why does anyone want to do this?

STEVE DOWNEY, "THE BOSS", MAKES LUNCH. EVERYONE PAYS ATTENTION.

One day we all load into the Boss's car and drive for five hours from New York City, probably due North. And we end up in the Adirondack mountains. We get there pretty late in the afternoon and, the next day, we hike up a one-mile trek to a nice little lake. It was not horrible. We ate some, walked around and then left to hike up an even bigger hill. Still about one mile up, but waaaayyyy steeper. I even helped pull my mom up the hilly parts. She was huffing and puffing and had to stop several times. I acted like I could keep going but was happy to take a breather with her, too.

Eventually we got to the top and I was not sure what the big deal was, but the two leaders—Boss and mom—were elated. The Boss was amazed I made it up there. I think my mom was even more amazed she did.

After this trek, I seriously considered packing up and heading back to Tennessee, from whence I came. But then I see how much I am loved and accepted and think I may stick around a bit more. No more pointless hikes, PLEASE!

All in all, this whacky brood is fun. I think I will stay, as long as we don't have to climb too many more peaks. I am happy just walking a few miles a day in Riverside Park or at the beach. I do see it is helpful for my mom to have a willing companion for walks, to clear her lungs, and someone to supervise her percussor after inhalations—also lung clearing. As well as someone to nap with. She needs me. And I think I am realizing, I need her, too, and even the Boss. ▲

Andrea Eisenman is 58 and has CF. She adopted Trixie in August 2022. She and Steve Downey share Roscoe, Willie and Trixie, their travel companions on the journey of life.

Please consider sharing your Pet's Perspective in upcoming issues of CF Roundtable.

TILLMAN continued from page 3

Thus, previously reported incidence rates in Canada and the United States (US) may be outdated. The objectives of this study were to estimate contemporary CF incidence rates in Canada and the US and to determine if the incidence rate has changed over time. Contemporary CF incidence rates suggest CF incidence is lower than previously reported and varies widely within North America. This information is important for resource planning and for tracking how programs (e.g., genetic counseling, modulator availability etc.) may impact the incidence of CF moving forward.

https://tinyurl.com/4c5n3swu

AND

https://tinyurl.com/mr4as9nx

Current Prices Versus Minimum Costs Of Production For CFTR Modulators

This study aims to estimate the minimum costs of production of CFTR modulators, assuming robust generic competition, and to compare them with current list prices to evaluate the feasibility of increased global access to treatment. Minimum costs of production for CFTR modulators were estimated via an algorithm validated in previous literature and identification of cost-limiting key starting materials from published routes of chemical synthesis. Costs of production for elexacaftor/tezacaftor/ivacaftor are estimated at $5,676 [$4,628-6,723] per year, over 90% lower than the US list price. Total cost of triple therapy for all eligible diagnosed CF patients worldwide would be $489 million per year. Comparatively, the annual cost at US list price would be $31.2 billion. Thus, elexacaftor/tezacaftor/ivacaftor could be produced via generic companies for a fraction of the list price. The current pricing model restricts access to the best available therapy, thereby exacerbating existing

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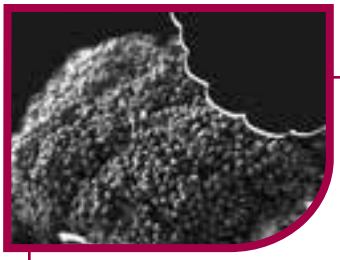

CULINARY CORNER

Mexican Potato Hash

By Maggie Williamson

Financially things are tough for a lot of people, including those in our CF community. Groceries have gotten more expensive and those receiving assistance from the Supplemental Nutrition Assistance Program (SNAP) are finding it harder and harder to make tasty meals on a budget.

The great thing about a dish like hash is that it is so versatile you can add almost anything in one pan. Hashes are, by nature, gluten free because potatoes are the base, and hashes can be dairy free. They can be vegetarian or vegan as well. I make a hash when I am running low on ingredients or am trying to finish up any items that may go bad soon.

My Mexican hash is great because it utilizes some canned items that are cheap and easy to come by. The notes at the end of the recipe offer suggestions of what to add or subtract, depending on what you have in your pantry/fridge and based on your budget. I have written this recipe for 4-6 servings, but if you are cooking for 1-2 people you can cut the recipe in half, freeze the other portions, or eat them as leftovers for the next couple of days.

Mexican Potato Hash
Yield: 4-6 servings
Prep time: 15 minutes
Total time: 45 minutes

Ingredients:
2 tbsp oil of your choosing
3-4 medium potatoes (any kind)
1 medium onion (white or red)
1 packet of taco spice mix or your own taco spices
1 15oz. can of black beans, drained
1 15 oz. can of corn, drained OR
1 cup of frozen corn kernels
2 cups shredded rotisserie chicken or canned chicken (optional)
Optional toppings:
Sour cream
Salsa
Green onions
Fresh cilantro
Preparation:

Step 1:

Chop potatoes into bite sized pieces leaving skins on for ease and more nutrition. Slice onion into ¼ inch pieces.

Step 2:

Add oil to sauté pan. Heat pan on medium to medium/low. Once oil is heated, add potatoes. Let them start to sizzle and then stir. If potatoes seem to be sticking, add a few tablespoons of water to help steam a bit. Once potatoes start softening (after 20 minutes) add your sliced onion. Let the onions cook for 2-3 minutes. When they start to become translucent add the spice mixture and stir.

MAGGIE WILLIAMSON

Step 3:

Add in the black beans and corn. If your corn is frozen you can also add it straight into the pan. Give everything a stir and test your potatoes with a fork to see if they are done. When the fork pierces through easily, they are cooked.

Step 4:

Finally, add your chicken and cook for another 1-2 minutes until chicken is heated through. Taste everything to make sure there is enough salt. (Store-bought spice mixes usually have plenty of sodium.)

Step 5:

Plate the hash and add any toppings of your choice. Salsa, sour cream, and cilantro are my favorite toppings to add.

Notes:

This dish is already a complete meal, but if you want to add some greens, add spinach or kale at the end until they are wilted down. If you need to get rid of any bell peppers, this is also a great addition. Just chop into small pieces and add with the onion. Adding a fried egg on the top gives it a more breakfast/brunch feel. You can also use any beans, including a can of chili beans if you prefer them over black. Also, omit the chicken if you want to keep it vegetarian or vegan. ▲

Maggie Williamson is 34 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.

BALDWIN continued from page 1

on February 21. It's the story of 11-year-old Penny Rooney, who has cystic fibrosis and bonds with a dolphin she discovers lost in her backyard creek.

Writing and editing this book was like no other writing experience in my career—I've joked that it was like 30+ years of therapy all rolled into one writing experience. It was difficult, but also deeply cathartic and, in many ways, joyful. Some of the things I learned through the writing of this story were ones I expected, but many were surprising.

I discovered, for instance, that marine mammals are prone to pneumonia caused by many of the same bacteria that colonize CF lungs, including *Pseudomonas aeruginosa* and *Mycobacterium abscessus.* I read articles in veterinary journals about researchers doing PFTs on captive dolphins and administering nebulizers to manatees. I learned that marine biologists do sputum cultures on sick whales and dolphins to better treat their infections.

I also discovered just how deep ran my own learned impulse to put a happy face on any situation, no matter how dire. One of my goals in writing *No Matter the Distance* was to deconstruct and push back on the way so many children with serious illness are raised to feel they must be upbeat, positive, and "inspiring," no matter how difficult their experiences may be. When I was a kid, I felt surrounded by adults—many of them medical professionals—who, during my most traumatic PICC placements or lonely hospital nights, tried to reassure me by telling me stories of CFers whose health was more critical than mine and pointing out that my situation *could* be a lot worse.

Instead of cheering me up, this often invalidated my own feelings of grief or pain and made me feel like I wasn't "strong enough" to deal with routine but traumatic CF experiences. In *No Matter the Distance,* I explored this idea, trying to show through Penny's story that hardship is hardship, even if it "could be worse." I let my main character be frustrated when she had to go to the hospital and be afraid when she had to get blood drawn or get a PICC placed, even though she and I both understood that these were routine difficulties for a CFer.

To my surprise, when I got notes back from my editor, her biggest critique was that I hadn't let Penny struggle *enough.* "These are hard things she's going through!" she wrote. "Penny seems unrealistically optimistic to me. I think you need to give her more room to be upset and angry about what she's experiencing."

It took me several days to digest and understand my editor's feedback. Hadn't I already done that?! Hadn't that, explicitly, been what I was trying to do with this book?

As I went back through and reread my draft, I not only saw all the places where Penny was brushing off her own struggle—I saw how much *I,* as the author, felt the need to brush off Penny's struggle as well. I found myself tempted to call my editor up and explain how really, a hospital stay and a PICC line weren't that big a deal, and Penny's situation could be so much worse.

But I remembered what it felt like when *I* was 11 and experiencing my first hospital stay since childhood and got my first PICC line. I remembered the anxiety, the anger, the feeling of unfairness at what I was experiencing compared to my friends without CF. I thought of all the kids with CF and other serious illnesses who might read my book one day, and, as I revised Penny's story, I hoped that reading it would tell them:

It's okay to be angry.

It's okay to be sad.

It's okay to struggle.

In the process of revising *No Matter the Distance,* all of this inner turmoil led to a light-bulb moment about something that has plagued me my whole adult life. Throughout my whole 15-year relationship with my husband, I've found myself irrationally angry when he gets sick. (And he's a very mild-mannered sick person—not a man-cold in sight!) As much as I try to be a generous and supportive spouse, I've always struggled with feeling impatient and unsympathetic whenever he or other "healthy" people were sick.

While working on my revision, I finally realized why this was. After a lifetime of being told that my own health challenges "could be so much worse," and rarely offered space to rage or grieve experiences that felt difficult to me, I instinctively passed the same attitude on to others. *Oh, you've got a cold? People are dying out there. Buck up and deal with it!*

I found myself needing to take a few quiet minutes to have a conversation with my child self: *I recognize that nobody honored your pain when you were experiencing it, but they should have. You do not have to be strong. It's okay to fall apart.*

And, surprisingly enough—the next time my husband had a health issue, I was able to remain neutral and loving and not be swallowed by my own resentment.

No Matter the Distance is not a universal CF story. There's no way to cover in one novel the myriad diverse experiences that different CF patients have. But I hope that reading the book will help give other chronically ill patients a little bit of the gift it gave me as I wrote it: the gift of validation and an acknowledgment that growing up with a disease like CF can be tough and traumatic, and that we all deserve space to struggle—no matter how much worse it *could* be. ▲

Cindy Baldwin is 34 years old and has CF. She is the author of several books with HarperCollins, including the upcoming No Matter the Distance *(releases February, 2023), which features a protagonist with cystic fibrosis. Cindy lives near Portland, OR, with her husband and daughter.*

FOCUS TOPIC

ASSESSIBILITY AND CF CARE

Accessibility Of Virtual Spaces: A Critical Pillar Of CF Community Justice

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

This issue we're taking 'Pearls of Wisdom' to the front page to shine a light on our Focus Topic of Accessibility! After three years of living with COVID-19, this seems like a great time to reflect on the many improvements the pandemic has fostered in virtual activity participation. We've come a long way in the CF community and have much to look forward to! Our future will only get better and brighter the more we prioritize accessibility in the virtual spaces where we spend so much of our time.

Let's zoom out a bit and think about what accessibility can mean—we've seen many good examples in the past few years of how this term can refer to many different experiences and needs. Many of us have seen things like reserved parking spaces that meet the minimum criteria under the Americans with Disabilities Act (ADA) for physical accessibility to people who use devices such as wheelchairs and crutches. These are certainly good examples of accessibility, but far from the full extent of what this concept includes! Broadly speaking, accessibility is what helps us live well with the functional limitations introduced by something like CF, while participating fully in activities that matter to us.

Accessibility is deeply *individual*; even within the CF community itself, people can have very different needs and preferences for how we approach tasks. Accessibility is also deeply *contextual*; what we need may vary quite a bit from one situation to another. And of course, accessibility is deeply intersectional; our needs are shaped by many different facets of ourselves and our position in society. These can include—race and ethnicity; culture and socialization; gender and sex; religion and spirituality; and much more.

Most importantly, accessibility is *realistic*. The standard for determining whether an activity is accessible is not participants being able to engage with considerable struggle and effort, but rather participants being able to engage with ease and comfort. This distinction has thankfully become clearer to many people during the COVID-19 pandemic as more neurodivergent people have "unmasked" ourselves in social spaces that better meet our needs. The past three years have been a time of tremendous learning about things like sensory accessibility, which makes a huge difference for many of us neurodivergent CF patients. It feels amazing not to spend the mental energy left over from the significant physical and cognitive labor of managing CF itself on trying to "act normal" when surrounded by sensory input that causes us terrible pain.

XAN NOWAKOWSKI

Closed captioning has also become much more commonplace during the pandemic. Videoconference software platforms like Zoom, Google Meet, and Microsoft Teams have greatly improved the functionality of their automatic captioning utilities. These platforms have also introduced additional ways to use third-party captioning tools and/or human live captioners during events. Captioning not only improves accessibility during synchronous activities for people with diverse hearing and auditory processing characteristics, but also helps people work together effectively in asynchronous contexts with less effort!

"Accessibility is for everyone" has become my mantra in encouraging new groups of people to embrace captioning as standard practice. Often, abled people become believers with astonishing speed when they realize how much easier captioning and auto-transcription makes the process of taking accurate meeting minutes. And of course, making space intentionally for Deaf and Hard of Hearing colleagues in our virtual spaces matters tremendously for the CF community, where many of our adult members have lost significant hearing due to sensitivity to harsh antibiotics like tobramycin.

Here's the thing about accessibility: By its nature, it makes space for tremendous diversity of both needs *and* preferences. Some folks find captions distracting. That's totally fine! In-meeting captions can always be toggled on and off by individual users to

optimize their own experience. Accessibility embraces the uniqueness of each person as an asset for justice in group settings. The standard of "reasonable accommodation" included in the ADA reflects this spirit of always making a good faith effort to fulfill as many different needs as possible, which requires flexibility and creativity.

Deliberately imagining the world from the perspectives of our peers with different lived experiences offers powerful support for accessibility in planning group activities. I always think back fondly on the earnest question I got from one of my students who had No Light Perception—a trait shared by about 10% of the Blind community overall. They asked if I was Blind myself because I always gave them directions that relied on non-visual types of input—like things they would feel, hear, or smell as they navigated a new physical space. This was just something I'd always done because I grew up with a parent who has Very Low Vision. However, I had never really thought about it until that moment.

What seemed so normal to me from growing up in my family and watching my mother use assistive technology to complete highly precise visual tasks in her neuroscience lab—and just being generally encouraged by my parents and teachers to think about how my fellow people might experience things differently than I did—often seems unusual to people who grew up in ableist environments. So I try to embrace this same energy in all of my advocacy for accessibility. Using our imagination to anticipate the kinds of questions we'll want to ask people about their specific needs and preferences not only saves a lot of time, but also communicates that we truly value the lived experiences of our peers.

On the visual accessibility front, many in the CF community deal with significant visual changes from secondary conditions like diabetes and vascular damage that our CF can cause. And of course, some folks with CF have been Blind their entire lives for reasons completely unrelated to this disease. A couple of basic strategies can make big changes for visual accessibility in CF community events.

Making consistent use of ALT text is an easy and remarkably effective way to support Blind community members as well as those with visual processing challenges. This simple utility helps screen readers describe images accurately for those who use them; people who don't use screen readers can also pull up the text directly and read it. When in doubt, right click on images you're adding to your presentations and communications. Generally you'll see an option to "edit ALT text" or provide an "image description." When you're writing descriptive text, keep it simple! You generally want a few words conveying the basics of the content in the image—for example, "great blue heron resting on a pier overlooking a lake at sunset."

Then there's the question of backgrounds and contrast. Here's where it gets tricky! What is optimally accessible for one person may be terrible for another when it comes to presentation and styling. If you participated in our Scholarships webinar on December 7, the slides we used for that session went through some revisions to improve functionality for as many folks as possible! Even with a Blind parent and some visual accessibility needs of my own related to extreme photosensitivity, I can't always anticipate what type of content may work best for a mixed group.

But getting things exactly right on the first try isn't the standard for justice. Rather, it's the commitment to continuous and enthusiastic improvement. I nailed the color contrast on my first version of those webinar slides and incorporated ALT text consistently for images, but the darker background on some of the more aesthetic slides might have made it challenging for some folks to distinguish different content elements. I restyled the slides to preserve the overall aesthetic on those portions while offering a lighter background for the text items. I had to put some thought into it, even though I've been thinking about this stuff my whole life. And that's okay—again, accessibility is about making our best faith effort to provide the broadest and most customizable functionality we can. It's not about being perfect or superhuman, just attentive and responsive.

> "Here's the thing about accessibility: By its nature, it makes space for tremendous diversity of both needs and preferences."

We've seen quite a few improvements these past three years on the *social* accessibility front that reflect this spirit of knowledge as power. It's now becoming commonplace to attend CF community events whose platforms intentionally make space for sharing things like our pronouns, prefixes, and name pronunciations. These types of information help folks know how to address us properly without having to ask. Feeling seen is absolutely an important part of being accommodated—and

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an excellent illustration of how accessibility really is for *everyone*.

Likewise, event platforms these days often give folks the opportunity to share about our sexualities, sex and gender identities, religious and spiritual traditions, racial and ethnic identities, cultural backgrounds, and much more. The COVID-19 pandemic and related global advocacy efforts in social justice have demonstrated the destructive power of exclusion. In the CF community, we have seen the devastating consequences of delayed diagnosis—or more often, preventable death from no diagnosis ever being made—in patients of color. And those of us from ethnic minority backgrounds still struggle for equitable access to highly effective drug therapies.

I was fortunate to get tested for CF as a preschooler and receive some appropriate treatment in childhood, even though my conclusive diagnosis came from genetic testing at age 32. As far as I know, I'm still the only diagnosed case of CF in the U.S. with my specific genetic profile—not an unusual situation for patients with substantial Indigenous lineage. Like others in "the final 10 percent," I can't take Trikafta or any other CFTR protein modulator. I feel lucky to thrive as much as I do on my existing care regimen. And, for the moment, I'm alive to say that; many of my peers no longer are.

I don't doubt for a moment that I would have gone untested and probably died if I hadn't been racially white. Our community is missing the voices and faces of many Black and Brown people with CF whose families were gaslit by medical professionals who believed the disease only impacted white people. And now many of our spaces remain socially inaccessible to racial and ethnic minority patients who rarely encounter meaningful representation of our own lived experiences. This has begun to change—and it will keep changing as we continue to advocate.

Importantly, many event platforms now ask registrants if we have specific accessibility needs for our participation! Even just asking this broad question and allowing people to respond freely in a comment box does wonders for inclusion and affirmation. Certainly it gives the event organizers vital information to use in crafting the virtual environment. Moreover though, it communicates to participants that our experiences, needs, and wishes matter. Starting from that baseline of intentionally valuing one another makes all the difference. It gives us room to work on the finer details of accommodating each other as best we can.

As you're thinking about how to improve accessibility in your own CF community activities, remember above all never to let the perfect become the enemy of the good. Make your best faith effort; embrace opportunities to do better; and share what you learn in the process. It really is that easy. ▲

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.*

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inequalities in CF care.
https://tinyurl.com/2fkwxzv2

The Impact Of Elexacaftor/Ivacaftor/Tezacaftor On Cystic Fibrosis Patients Who Acquire COVID-19 Infection

The combination of medication containing elexacaftor, ivacaftor, and tezacaftor (EIT) has dramatically impacted the treatment and prognosis for patients with cystic fibrosis (CF). Lung function, weight, and self-reported quality of life have improved for many of these patients, but little is known about whether this treatment will have a beneficial effect in preventing morbidity and/or mortality from respiratory infections such as COVID-19. This study shows that cystic fibrosis patients who were already receiving treatment with the combination of elexacaftor/ivacaftor/tezacaftor had a significantly decreased risk of developing acute respiratory failure after becoming infected with COVID-19. This study also showed a 50% increased likelihood of patients who were not on treatment with EIT developing ventilator dependence, but the low numbers of patients likely contributed to that difference not achieving statistical significance. By improving chloride ion flow through the CFTR channel, EIT substantially improves lung function and decreases respiratory morbidity in CF patients. This improved lung function results in improved tolerance to the pulmonary insult caused by COVID-19 infection. Overall, this data makes a compelling argument to consider this combination of medications for any CF patient who is a candidate.
https://tinyurl.com/yckj48pd

VX-522, For CF Patients Who Can't Use Modulators, To Enter Trial

Vertex Pharmaceuticals is launching a clinical trial to test VX-522, its inhaled messenger RNA (mRNA) therapy for lung disease in cystic fibrosis (CF) patients who are not eligible for treat-

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PHOTO BY STEPHEN BOYER

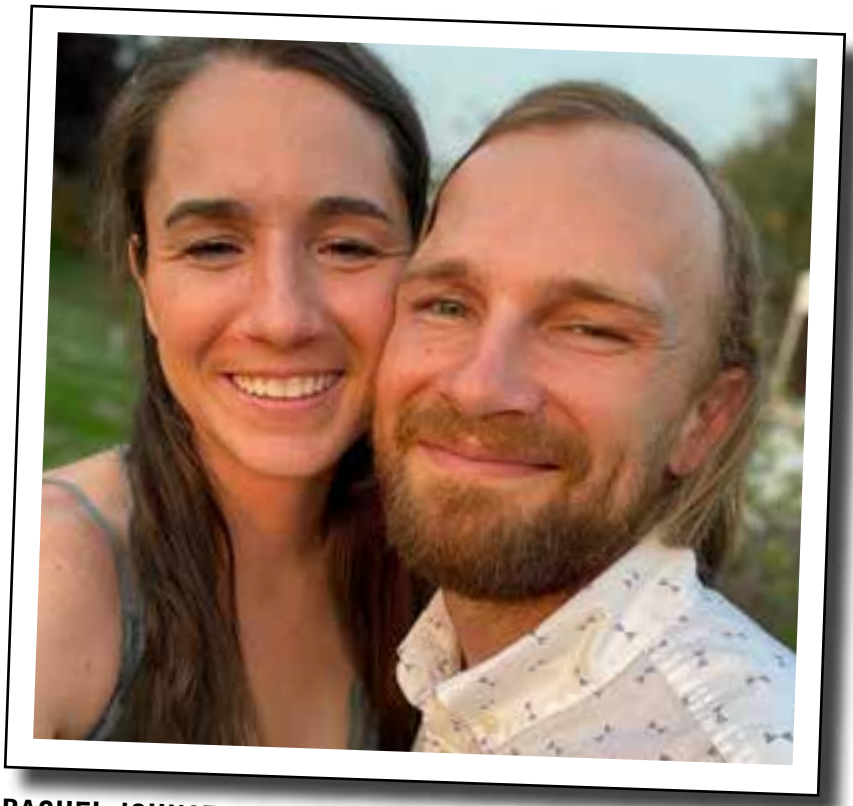
Terra Incognita

October 1980, (CF diagnosis)
December 1998, (Double-lung transplant)
November 2006, (Hemorrhagic stroke)

Pushing forward;
No guide to lead the way
Excited, yet Scary
The world is brand new
New sights, new smells, new everything
New medical terms, new medications, new physical limitations
Some former one or two, also
This is Terra Incognita for me, Undiscovered territory.

-M. Compton, 2008

FROM OUR FAMILY PHOTO ALBUM...



RACHEL JOHNSTON WITH HER HUSBAND, NATHAN JOHNSTON.

MAGGIE WILLIAMSON ENJOYS LAST SUMMER'S STRAWBERRIES.

SCOTT AND COLLEEN ADAMSON WITH THEIR DOG, PENNY.

ANDREA EISENMAN WITH HER DOGS, WILLIE AND TRIXIE.

SYDNA MARSHALL WITH HER COUSIN, MOLLY DUNCAN, AND NIECE, LUCY DUNCAN, AT MEANWHILE BREWING IN AUSTIN, TEXAS, BEFORE HER DAD'S WEDDING RECEPTION.

JERRY CAHILL WITH HIS FORDHAM PREP TRACK TEAM RUNNERS CHEERING ON THE TEAM BOOMER RUNNERS AT THE NYC MARATHON, NOVEMBER 2022.

CINDY BALDWIN WITH HER HUSBAND, MAHON, AND DAUGHTER, KATE, VISITING A PUMPKIN PATCH IN THE FALL OF 2022.

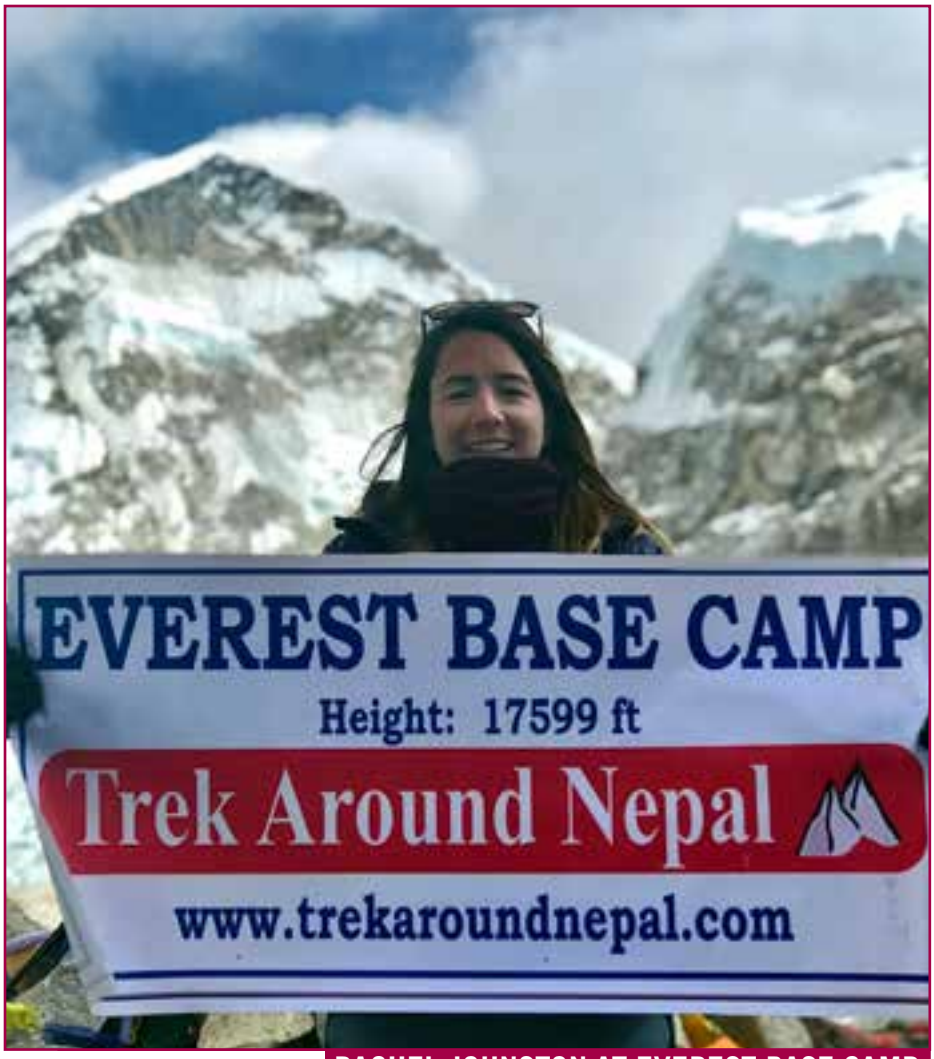
Part I: I Am A Miracle

By Rachel Johnston

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

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

On Sunday, February 11, 2018, after eight neverending days, 8,500 feet in altitude gain, frigid temperatures, and 40 miles trekking through the tallest mountain range in the world, I finally reached Everest Base Camp. Did I just write that? A small-town girl from Oregon hiked Everest Base Camp? A girl who was told at nine years of age that a disease I could not control would steal my breath from me and that I should make every effort to protect it. A girl who is the perfect poster child for how *not* to live with cystic fibrosis—pursuing a career as a nurse in the ICU who is continually surrounded by deadly, contagious bacteria every day, one who is always forgetting to take her medication, one who only does nebulizer treatments as a last resort, one who is always letting physical activity take a backseat to her very stressful job, and one who quit her job to travel to some of the world's poorest countries for months on end, filtering her drinking water, living in less than ideal conditions for even a healthy person—*this* girl? This girl just hiked to Everest Base Camp? Living a relatively normal life with CF without oxygen is rare, but hiking Everest Base Camp with cystic fibrosis and without oxygen? *That*. That is a miracle. *I* am a miracle.



RACHEL JOHNSTON AT EVEREST BASE CAMP.

Ever since the first week of 2018, when I began to ask the Lord for healing from cystic fibrosis, I have been waiting expectantly—I really felt like the Lord was going to do something miraculous in January. So, when January passed and we traded Africa for Nepal, I began to feel doubt creep in. The enemy attacked me with condemna-

tion. *You could have been praying today during the time you were playing cards with your team. You could have read your Bible more this morning. You could have worshipped with more intentionality tonight. You are asking the Lord for this big thing of healing, but you are so unworthy to receive it. You won't ever be able to walk that closely with God to receive a gift like that.* Wow. How quickly I believed and succumbed to the lies of the enemy without even realizing it. It wasn't until I received a message from my dad one morning that snapped me back into the reality of grace:

> *"I was praying this morning and you came to my mind. You talked about your reluctance to think you could ask God to heal you. We know that healing is something we can't earn. It's a part of God's grace that He dispenses according to His will. Keep asking, seeking, and knocking."*
>
> *"He rescued me because He delighted in me."*
> *Psalm 18:19*

Believe it or not, I did not put a lot of thought into what hiking to Everest Base Camp (EBC) would be like. While still in Africa, our squad received an email inviting us to hike EBC with our squad leader. We had two days to make a decision. Five of us said yes. And then we had 17 days to wrap our minds around what we had signed up to do *and* buy all our gear in Kathmandu. All of our traveling for the past seven months had been in countries with average temperatures of about 85 degrees, so we needed to buy a lot more clothes to stay warm.

Day 1:

Flying into Lukla alone is already one of the most dangerous places to fly into in the world, and that was just day one! The first day and a half of the hike was actually very pleasant. I talked with God and prayed and thanked Him for this incredible adventure. My mind went wild with thoughts of what crazy things He was going to do over the next 11 days. I was freshly showered, the sky was clear, the sun was warm, and the views were breathtaking. We traversed several beautiful suspension bridges over crystal blue water and the climb was initially a gradual downhill terrain until we came to this double suspension bridge. At this point, I now had to climb up to the very top bridge, cross it, and then proceed to climb 800 vertical meters (or about 2,500 feet) to get to Namche Bazaar. After eight hours of brutal switchbacks, with the sun sinking, we finally made it.

"Believe it or not, I did not put a lot of thought into what hiking to Everest Base Camp (EBC) would be like."

Day 2:

On only day two, I was forced to take a good, long hard look at what I had said yes to. How quickly my thoughts changed from what awesome things God might do on this adventure to survival mode.

Day 3:

The next day was a "rest" or acclimatation day. I was so relieved, but this feeling did not last long. Acclimatation day is not restful in the least. You climb up a few hundred meters and then come back down and stay the night at the same elevation, to make sure your body can adjust to the altitude and to prevent deadly altitude sickness or, as the Sherpas call it, "the mountain sickness." This was the hardest day of the whole trek for me. It was endless. It was cold, windy, and snowing. It was vertical. It was so vertical, they couldn't even make switchbacks; it was just straight up. The only thing that kept me from turning back was the promise of an incredible Everest view from the top—our first chance of the trek to see this legendary mountain. As I climbed, I prayed with every step. My prayer for the Lord to sustain me crept back in. It wasn't a prayer to thrive; it was a prayer just to survive. When we finally reached the top, my lungs were heavier than I had ever felt them and they felt like they were on fire. As we looked toward the north, our guide regretfully told us that we were unable to see Everest today because there were too many clouds. Out of desperation, I prayed, "*God can I at least just see it?*" At this point I couldn't imagine any possible way that I was going to make it another five days and thousands of vertical feet with thinner and thinner air. Tears came to my eyes as He quickly replied. *"No. Because I'm going to take you to it."*

Day 4:

I woke up the next morning with an unreal sense of renewed energy. The climb was a hard one, but, after the day I had before, this one seemed doable. It was a huge mental challenge, though, as we had to hike down 400 meters before lunch and then go right back up, almost 800 meters to get to Deboche, where we would spend the night. That night I struggled with sleep. My chest felt like it was a hundred pounds—it felt crushing. I assumed it was from working out my diaphragm

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FAMILY MATTERS

Making Parenting With CF Accessible

By Cindy Baldwin

By the time we reach adulthood, we CFers are well used to finding creative solutions to get things done while fitting in treatments, doctor's visits, exercise routines, and hospital stays. But even for the savviest of us, parenting can pose huge issues: Juggling parenthood *and* CF care can often feel overwhelming, and it can be next to impossible to get everything done. As with any other job, parenting with CF can require creative accommodations.

As a mom to an almost-ten-year-old girl, I've had a lot of time to try, fail, and try again to perfect the juggling act of caring for my daughter while keeping myself as healthy as possible. Although I don't always strike the perfect balance, the following tips have helped me get as close as I can.

1. Make it possible to do treatments on the go. While not everyone still does multiple breathing treatments a day on highly effective modulators, lots of us still do, and fitting those regular treatments in while raising a small kid can be grueling. When my daughter was four or five, battery-operated vests hit the market and helped revolutionize my ability to get therapy done in all kinds of situations. I also have two nebulizer compressors—one in my "normal therapy spot" in my family room, and one in my car—so that I can do treatments while driving if needed. These days, I often do 50% or more of my morning treatments in the car while driving my daughter to various activities. (An inexpensive compressor from Amazon can be a good car machine if your insurance won't cover one. I pair mine with a car inverter that plugs into my car's cigarette lighter and allows me to run my compressor while the car is on.)

2. Use the tools you need to keep yourself healthy. Parenting a newborn is overwhelming, especially when you tune into the many messages the world sends about the *right* ways to raise a little one. But parenting with CF sometimes requires compromises. When my daughter was little, I mostly breastfed her, but my husband gave her one bottle of formula every night so that I could get a few hours of uninterrupted sleep. Whether you need to use formula, baby swings, or other tools to help care for your child while you get what you need to stay healthy, almost nothing is more important for your baby than having a healthy parent.

"I have two nebulizer compressors—one in my "normal therapy spot" in my family room, and one in my car—so that I can do treatments while driving if needed."

CINDY BALDWIN

3. Let your kid join you in exercise! For several years when my daughter was little, the *only* way I could get a workout in was if she was heavily involved. We'd go for walks with her in the stroller, bike rides with her in a child seat, and do exercise videos on YouTube geared for kids. Cosmic Kids Yoga is a great YouTube channel with routines easy enough for kids to follow, but rigorous enough to provide a light yoga workout for busy parents. Did I get tired of Frozen-themed yoga? Definitely! But it was worth it to get my exercise in without it cutting into my rare moments of child-free time.

4. Minimize germ exposure where possible. Kids are serious germ factories, and the sad fact of the matter is that, as a CF parent, you won't be able to escape all of their plague-ridden coughs and sniffles. But for those of us who have immune system issues or for whom a cold is generally a ticket to the hospital, it's worth implementing germ-mitigation strategies during times of high viral transmission. When Vogmasks became popular while my daughter was a baby, I started making a practice of wearing one in crowded public spaces when there

were a lot of viruses going around, and it dramatically reduced the number of colds I got. These days, my husband, daughter, and I all mask up in high-risk settings, and we've all stayed much healthier as a result. We've also seen a reduction in the number of colds family members bring home when the person who's been in a high-risk setting, like school or work, showers and changes clothes immediately after coming home. Finally, communication is key: When planning play dates or get-togethers with other parents and their kids, I'm up-front about my viral susceptibility and honest in asking that they please let me know if their kids are sick before we see them. I've found that, in many cases, parents simply don't realize how serious toddler sniffles can be for someone with a condition like CF, and most people are happy to give a heads-up if a child is sick.

Parenting with CF requires a lot of creative accommodations—but being willing to be flexible and think outside the box can go a long way in making parenting accessible for us. ▲

Cindy Baldwin is 34 years old and has CF. She is the author of several books with HarperCollins, including the upcoming No Matter the Distance *(releases February, 2023), which features a protagonist with cystic fibrosis. Cindy lives near Portland, OR, with her husband and daughter.*

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ment with an existing CFTR modulator. The study will assess VX-522's safety and tolerability at single ascending doses in adults with CF, ages 18 and older. As an inhaled treatment, VX-522 is intended to deliver full-length CFTR mRNA directly to cells in the lungs. The mRNA is enclosed in lipid (fat) nanoparticles and, once inside the cells, is expected to give rise to the production of a working CFTR protein.
https://tinyurl.com/4aettex3

Dose Adjustments Of Elexacaftor/Tezacaftor/Ivacaftor In Response To Mental Health Side Effects In Adults With Cystic Fibrosis

In this case series, the authors report the use of sweat chloride as an indirect biomarker to guide dose adjustments of elexacafor/tezacaftor/ivacaftor (ELX/TEZ/IVA) in people experiencing significant mental health problems (depression, anxiety, brain fog and insomnia) after starting treatment. The researchers hypothesised that these adverse events (AEs) are a result of a combination of predisposition to anxiety and/or depression, the presence of a premorbid mental health condition, individual variation in elexacaftor metabolism and increased systemic CFTR expression. They found that dose adjustment of ELX/TEZ/IVA was associated with improvement in mental health adverse events without significant clinical deterioration.
https://tinyurl.com/4wdpmwv7

Elexacaftor-Tezacaftor-Ivacaftor Improve Gastro-Oesophageal Reflux And Sinonasal Symptoms In Advanced Cystic Fibrosis

Upper gastrointestinal and upper airway disease are common in cystic fibrosis (CF) and may contribute to lower airway infection and inflammation. In a longitudinal cohort study of 32 patients with advanced CF lung disease starting elexacaftor-tezacaftor-ivacaftor, the reflux symptom index score fell from a pre-treatment median (IQR) of 15 to 5 (2.8-7.3), the Hull airway reflux score fell from a median of 26.5 to 7.5, and the sinonasal outcome score from a median of 36.5 to 20 at 6 months on treatment. Mean FEV1% predicted rose by 9.2 points, the median respiratory domain score of the CF Questionnaire-Revised rose by 27.8 points and mean body mass index rose by 2.6 kg/m. In addition to improving lung function and weight, CFTR modulators improve upper airway and gastro-oesophageal reflux symptoms in advanced CF.
https://tinyurl.com/casmjape

Elexacaftor/tezacaftor/ivacaftor And Gastrointestinal Outcomes In Cystic Fibrosis: Report Of Promise-GI

Elexacaftor/tezacaftor/ivacaftor (ETI) improves pulmonary disease in people with cystic fibrosis (PwCF), but its effect on gastrointestinal symptoms is not clear. Gastrointestinal symptoms, evaluated by validated questionnaires: Patient Assessment of Upper Gastrointestinal Disorders-Symptom (PAGI-SYM), Patient Assessment of Constipation-Symptom (PAC-SYM), Patient Assessment of Constipation-Quality of Life (PAC-QOL)), fecal calprotectin, steatocrit and elastase-1 were measured before and 6 months after ETI initiation. After 6 months of ETI, fecal markers of inflammation decreased. Gastrointestinal symptoms improved, but the effect size was small. Pancreatic insufficiency did not improve.
https://tinyurl.com/ufdxxuar

Trikafta Helps To Resolve Inflammation In CF: Study

Treatment with the triple-combination therapy Trikafta (elexacaftor/tezacaftor/ivacaftor) reduces inflammation and promotes lung tissue repair in cystic fibrosis (CF). Results showed that levels of interleukin-6 (IL-6), a molecule

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TRANSPLANT TALK

Accommodations For Pre-Transplant And Post-Transplant Life

By Colleen Adamson

I had a lung transplant 24 years ago and a kidney transplant 16 years ago. Prior to my lung transplant, I experienced respiratory failure and was on a ventilator for five months. I was in a rehabilitation facility for three of those five months, where they were able to get me off the ventilator. Prior to going home, we (me, my family, respiratory therapists, and nurses) took a trip to my townhouse, which is three stories (lots of stairs!) to determine what accommodations I needed to live there in my weakened body. I almost cried when we got there; I hadn't been home in almost six months. One thing they determined I needed was a chair placed on the landing of the stairs going up to the bedroom. I couldn't make it the whole way up without getting completely exhausted and out of breath. I also needed a chair in the shower because I was so weak I couldn't stand very long. I had a wheelchair for the same reason. Not that I was going out much, but I needed it for doctor and lung transplant evaluation appointments. Since I was on oxygen, we decided to replace our gas stove with an electric one just to be on the safe side. My mother-in-law was the beneficiary of the gas stove; we were glad she was able to use it! I had a big tank of oxygen in our bedroom just in case the oxygen concentrator stopped working or we had a power outage. The team advised us to call our electric company to be put on a list of homes that need power in an emergency. We did lose power and had to switch to the oxygen tank. This was very stressful since we didn't know when power would be restored, but luckily the outage only lasted a few hours.

Since I was coughing a lot, we needed to prop up my side of the bed to a 45-degree angle. We used two big pillows and a bed rest pillow, which helped immensely. I was also dealing with a lot of anxiety. I had to take anti-anxiety medication every time I took a shower and used the Vest. The Vest especially made me very claustrophobic, as you can imagine. I could not bear to be alone, so my mom stayed in our bedroom if my husband was out of town. We also had a walkie-talkie next to my bed so I could communicate with my family downstairs. My family really went all-in on trying to make my horrible situation bearable, always thinking about ways to help me deal with my situation, both physically and mentally. I am very lucky to have such a great family who are always there when I need them.

COLLEEN ADAMSON GIVES A THUMBS UP.

A few years ago, I ended up with non-tuberculosis *Mycobacterium* (NTM) and *Aspergillus*. That was pretty rough, but I got through it with a lot of help from my CF, lung transplant, and infectious disease doctors. After that happened, my doctors suggested I start doing hypertonic saline nebulizers and using the Monarch Vest. I do these for maintenance purposes, to keep my lungs clear, and I have not had a lung infection since I started these therapies. I was not thrilled to be back on a nebulizer treatment and postural drainage, but I try to adapt to keep myself healthy. Additionally, I am also on Trikafta, which I also credit with me not having lung infections. I tried taking the full dose, but it caused my ankles to swell up. On a lower dose, I no longer have that issue and I still get the benefit of Trikafta clearing out my sinuses.

Skin cancer is also something that I've been dealing with for a long time, since people who are immunocompromised are more prone to getting skin cancer. This time I have squamous cell carcinoma on my right thumb. I started seeing a dermatology oncologist for this, since everything my regular dermatologist tried didn't work. She had me use several medicated creams, but the cancer kept coming back. My doctor also tried photodynamic therapy (PDT), which killed the cancer on my arms but not my thumb. PDT is very painful since you are basically frying your skin with a blue light for 15 minutes, but it was worth it since it got rid of at least some of the cancer.

After exhausting all treatment options for my thumb, my doctor sent me back to the radiation oncologist. He is the one who did my radiation on my head for another squamous cell carcinoma in 2011. I had seen him

about my thumb to talk about my options, and he is the one who sent me to the dermatology oncologist. He also (nicely) said he never wanted to see me again. Ah, famous last words! I actually apologized to him for coming back. He devised my plan of treatment and told me there was an 80% chance it would kill the cancer. Good odds, so I asked, when do we start? Within a couple of weeks, I started radiation therapy. I just completed six weeks of radiation on my thumb, going to the hospital every weekday. Not fun, but at least it was fairly close to my house, and the radiation treatment itself only took about five minutes. In the waiting room I saw people who had it much worse than me; I tried to keep that in mind as well.

My thumb became blistered and very sore as the treatment went on, and my thumbnail is half disintegrated and the rest is about to fall off. My doctor said it probably would not grow back. Ewww! Now we wait for a month for my thumb to heal and see if the cancer is still there. If the cancer is still there, they may have to amputate my thumb. The only good thing about this, besides getting rid of the cancer, is that I am left handed. I did, however, start paying attention to how much I use/need my right thumb. It does come in handy when opening jars or carrying things or driving or typing. I also took note of things like gloves and oven mitts which have a separate thumb section on them. Sigh. There will be plenty of other accommodation and accessibility issues that I will have to take into account and prepare for. I guess I will learn as I go, if it comes to that.

Fingers (and thumbs) crossed that I will be cancer free and not thumb free! ▲

Colleen Adamson is 53 and has CF. She is the Treasurer of USACFA, and lives in Alexandria, VA. Her contact info is on page 2.

JOHNSTON continued from page 19

the past four days, hiking for six to eight hours each day and struggling to breathe deeper as the air thinned.

Day 5:

This morning I woke up just not feeling well at all. I felt like I was sick. I didn't have an appetite, which, if you know me at all, is *unheard* of. I struggled hiking all day. It was miserable. I had to force my body to move. When we finally made it to Dingboche, doubt began to creep in again. Worry and anxiety gripped me as I sat in the tiny tea lodge with a few other trekkers and teammates that night. One of them had brought a pulse oximeter with them, which was such a brilliant idea and something I appreciated as a nurse! I felt a lump in my throat as I put it on my finger and watched the numbers pop up. Heart rate: 125. SpO_2 (blood oxygen level) 80%. Now, a normal heart rate is anywhere between 60-100 beats per minute. However, a resting heart rate should be closer to 60-80 beats per minute. These vitals were taken after I had been sitting and resting for several hours. Blood oxygen level should be higher than 94%. A Sherpa informed me that normal levels at 4,400 meters were 85%, which blew my mind as nurse. This slightly eased my anxiety, but not for long. The rest of my teammates' vitals were heart rates ranging between 70-100 and oxygen levels of 88-95%. That night, as I lay in my triple layers of clothing, hat, gloves, sleeping bag liner, -10 degree-thermal-rating down sleeping bag, and two giant comforters piled on top, I broke down in fear. I knew now why I had struggled to sleep the night before and why my chest was so heavy—it wasn't from my diaphragm: It was heart pain. My heart rate was way too high and I had no idea how to slow it down. I was lying in bed for crying out loud and my heart was ticking away like I had just gotten done running sprints.

The enemy shrouded me in fear. I began thinking about how many days I could stay in this cold tea hut alone and wait for my teammates to come back for me after getting to base camp. I knew if my heart rate was this high just lying here, there was no way I could possibly hike. The enemy sneered at me. *How could I have ever thought I could do something like this, with a lung disease? How foolish I was.* I peeled myself out of all my layers and desperately walked toward my other teammate's rooms, to ask for prayer. I lightly tapped on their doors. Nothing. *They went to bed over an hour ago and are warm in their beds. How could you possibly think or ask them to get out in the freezing cold just to pray for you? They need their rest. This is foolish. This is not their fight. This is yours. You are alone.* For as long as I could stand, I walked outside and stared up into the black starry sky through the frost of my own breath as I cried and pleaded for God to help me. *He doesn't hear you. What do you want? A miracle? He already gave you Everest Base Camp. You chose this, knowing your physical limitations. You want more now? You've made it this far, that is already far more than you deserve.* I went to bed defeated. Under a crushing darkness of the enemy's lies.

But nothing could have prepared me for what Day 6 would hold.

Rachel's story will be continued in the upcoming Spring 2023 issue. ▲

Rachel Johnston is 31 years old and has CF. She currently lives in Oregon with her husband. She has traveled all over the world as part of various mission trips. She came back to the United States in June 2019 and got married in November 2021.

TRANSPLANT TALKBACK

My Conversations With Jerry Cahill—Striking A Balance

By Andrea Eisenman

In our last interview, Jerry Cahill was losing his long-term disability at age 66 and a half. It ended on January 2, 2023. As Jerry now embarks on heading back to work full time, we sat down to go over what this might entail at this point in his life: after a long career in retail, volunteer work at Boomer Esiason Foundation (BEF), and coaching. Instead of retiring, he is reinventing himself yet again. He talks about his fears, his passions, and what propels him forward. He considers doing all this a juggling act worthy of the center ring at Ringling Brothers and Barnum & Bailey. Read on to learn more.

Explain what "reinventing yourself" means at this stage of your life at age 66.

Reinventing has been something I look at in a positive way. I worked, I played sports, I had to go off disability; I believe you are never done, that you have to keep pushing forward. I try to live in the realm of possibilities. Here I am again having to reinvent myself. I never thought, at 66, that I'd be in this position of having to go back to full-time work. I feel I am starting my career again, except at this late stage in my life. It's odd to me and I don't quite understand it. Even though I see people around me who are my age and I think they are old, I just don't think of myself as "old." It's an interesting time for me. I feel I am walking into a dark room and finding my way.

What do you see as pluses or minuses as far as reentering the job market at this stage of your life?

The minuses would be that growing up with CF, since I am ancient for someone with CF, I never thought there was a future and never saved my earnings to be able to fully retire. Even though I had great jobs with decent salaries, I didn't see the point of saving for retirement since I was not guaranteed a long future. The pluses of getting back into the job market include making money to support myself. Financially, it is not easy for anyone to live while working, especially someone who has CF and has lived through three transplants. Life is not easy. I feel as though I've run this marathon in life and I wanted to finish and enjoy, but I have to keep going [now back to work].

Due to my age, I cannot do what I did when I was 30. This concerns me as I have more fatigue and things take more of a toll on my body than they did 35 years ago. Whether it's exercise or daily living or coaching, which is my passion, it becomes harder to manage it all and stay on top of my health.

JERRY CAHILL GIVING JULIAN LYNCH A THUMBS UP FOR HIS WIN AND A PERSONAL BEST JUMP.

Will you retire?

Within a few years I will have to retire and live *very* frugally on a strict budget. But I think I will always do *some* work. I do not think I will do well being idle. I would definitely still coach, but that won't pay my bills. It is just something that is extremely gratifying to me.

What are your fears?

I may have to give up certain things in my life that I like so that I can go to work full time. I also have an A+ personality so I want to do the job at 200%. I do not want to give up things solely to survive financially; obviously, I want more life, more coaching, more of everything. Honestly, I am unsure of how much I can do, so this all brings anxiety.

My type-A personality helps propel me like a rocket flying through the air. I am always moving on to see what's next; what do I have to accomplish now. I keep pressing on to move forward. I don't give myself time to think of what I just went through—it's simply on to the next thing. It can also hurt me as I do not take time to enjoy my achievements. I like to keep busy and feel like I am

helping others. I try to lead by example so others with CF can see what is possible and possibly help them think they can do it, too. It's like we in the CF community are part of a sports team and if we see others doing well, it gives others hope that they can, too.

My fear is that my time and commitments may have to be balanced while I work and fit in the coaching, As it is now, my sleep suffers as does my personal exercise routine. I have to manage my time so I can fit everything in. Even now while I'm currently volunteering, I already feel as though I am walking on egg shells and I worry I will get sick from being too burnt out.

What do you do exactly when you coach?

When I coach, I do it at two different schools twice a week. One is Iona Prep and the other is Fordham Prep. I drive to each (I do not take public transportation for health reasons) and it entails paying for parking, tolls, and gas twice a week. I don't get home until 10:30 p.m. and this is all after a full day at work. It takes a lot of energy to rally these high schoolers, but I love it. On weekends there are track meets that I coach as well. Even though I am reimbursed for the travel expenses, I am basically volunteering my time. I find that volunteering this way is quite rewarding.

What I do is cheerleading and positive attitude while jumping around and encouraging them to be their best. It is exhausting, both mentally and physically. After my practices, the kids on the teams probably go home and sleep like babies. I get home at night and I am too amped up to sleep.

When I coach it is not just about teaching them a skill like pole vaulting; it is life lessons. It is about being responsible. If they cannot make a practice, they need to contact me. I also teach them respect for one another and that we have ground rules to adhere to. I educate them about being a better athlete and the importance of mutual respect and understanding others. However, because I am immune suppressed, there is no handshaking or group hugs. And if they have a cold or they are getting sick, they have to tell me. That was a big learning process for them and me. They are very good about it now. They are very aware of their health status and share that with me. I like passing on what I have learned in life.

> "My type-A personality helps propel me like a rocket flying through the air."

Share a teachable moment with us.

Educating them about having CF. I used to coach someone who is now a friend. I assumed he knew I had CF, but it turned out he didn't. He found out when I invited him to a BEF relay event. He told me, "You are unbelievable!" I was not sure what he meant. He told me he didn't know I had CF and yet I do all of this (volunteering at BEF plus running and biking). He also said, "I guarantee that no one you coach knows you have CF!" So now I tell my teams about CF. Once I had my lung transplant, I saw a teachable moment—I described the importance of organ donation. These kids had a lot of questions about issues we with CF take for granted—where do they get these lungs, how long does this surgery take, what is involved? Plus, I make sure they understand that they cannot attend a practice if they are unwell or sick.

Are you getting your strength back?

I've been building myself up post-liver and kidney transplants. I am seeing a difference in my strength level. I do feel stronger but it's a lot of work. I feel like I fit more into a day than most normal people so I can achieve all my goals. My friends are amazed at all that I do and I am slightly embarrassed when someone asks me what I did.

You have mentioned Endure More®, what is this?

It is similar to what I say about running a marathon and you've just got to keep running. A friend of mine with CF who is involved with sports said that we just have to endure more, like just keep going. And it is so true even though I had transplants. I just have to keep moving forward. You can give up or you keep on enduring and hopefully you get more life. I think it's a cool and true saying.

What keeps you going?

What keeps me going is my will and passion to make a difference in my own life and other people's lives. This applies mostly to the coaching I do. If I could afford to, I would coach all day long; it is that gratifying. But it is not realistic because I cannot survive on that salary. I am driven and, like an athlete who wants to break records, I am going for the gold metaphorically.

"Pass it on" is a line in the movie *History Boys*. A professor says to his students, "Pass it on. It's not for me, it's not for you, but it's for someone out there". And that is how I feel about coaching—I am passing on lessons of what I learned about training but also life lessons. And I continue to learn each day. I think it's important in life to be able to pass on what you have experienced. I have benefitted from others in this respect as well by learning from others about what they have been through. I continue to do this to support the communities I am part of. ▲

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

Andrea Eisenman
New York, New York
58 years old on November 28, 2022

Sydna Marshall
Austin, TX
42 years old on September 19, 2022

Dr. Xan Nowakowski
Lakeland, FL
39 years old on December 29, 2022

Wedding

Sydna Marshall and Adam Keys
Austin, TX
9 years on October 26, 2022

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implicated in inflammation, decreased significantly within three months of starting on Symdeko (tezacaftor/ivacaftor). Levels of IL-6 and C-reactive protein (CRP), an inflammation marker, were significantly reduced at one month with Trikafta, which was sustained through one year of treatment. Results at one year of Trikafta therapy showed significant reductions in the levels of IL-20 and MMP-10, both implicated in wound healing. Levels of calprotectin — a risk marker of pulmonary exacerbations decreased significantly within one year of starting treatment with Trikafta. A trend toward calprotectin decrease was also seen with Symdeko within one year. Radiological improvements were visible in all those who underwent CT scanning following a year of Trikafta.
https://tinyurl.com/3p7fz3uh

Magnetic Resonance Imaging Detects Improvements Of Pulmonary And Paranasal Sinus Abnormalities In Response To Elexacaftor/Tezacaftor/Ivacaftor Therapy In Adults With Cystic Fibrosis

Effects of Elexacaftor/Tezacaftor/Ivacaftor (ETI) on structural and functional lung abnormalities and chronic rhinosinusitis have not been studied by imaging. Researchers found that MRI detects improvements of chest MRI and chronic rhinosinusitis (CRS-MRI) scores in adult CF patients who first received ETI, demonstrating reversibility of structural lung and paranasal sinus abnormalities in patients with established disease.
https://tinyurl.com/bdffe9h3

Trikafta Aids Quality Of Life With CF In Multitude Of Ways: Study

Researchers analyzed the 11 non-respiratory domains (non-RDs) of the CF Questionnaire–Revised (CFQ-R), which assess general health-related quality of life (i.e., Physical Functioning, Role Functioning, Vitality, Health Perceptions, Emotional Functioning, and Social Functioning) and quality of life impacted by CF (i.e., Body Image, Eating Problems, Treatment Burden, Weight, and Digestive Symptoms), for participants in two Phase 3 trials. ELX/TEZ/IVA treatment led to higher scores in all CFQ-R non-RDs, with improvements in most domains compared with control treatments. These findings demonstrate that Trikafta improves a range of CF-specific symptoms and general functioning and well-being.

Orkambi's Benefits In The Real World Go Beyond Lung Function: Study

For a small group of people with cystic fibrosis (CF), one year of treatment with Orkambi (ivacaftor/lumacaftor) improved bone health and stabilized pancreatic function, nutritional status, reproductive hormone levels, and lung function.
https://tinyurl.com/26bfunuy

Orkambi Found To Improve Exercise Endurance In 3 CF Adults In Study

Treatment with Orkambi (lumacaftor/ivacaftor) for six months was found to improve exercise endurance in three adults with cystic fibrosis (CF). Moreover, by the end of the study, all three men experienced less leg discomfort and less dyspnea, or shortness of breath. After six months of treatment, exercise endurance time increased for all three patients — by 87%, 52%, and 23% among the men. Inspiratory capac-

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ity also improved for all three, and oxygen uptake increased in two patients. Additionally, all patients reported less breathing discomfort and leg fatigue.
https://tinyurl.com/4jxuz2y6

Discontinuation Versus Continuation Of Hypertonic Saline Or Dornase Alfa In Modulator Treated People With Cystic Fibrosis (SIMPLIFY): Results From Two Parallel, Multicentre, Open-Label, Randomised, Controlled, Non-Inferiority Trials

The SIMPLIFY study aimed to assess the effects of discontinuing nebulised hypertonic saline or dornase alfa in individuals using the CFTR modulator elexacaftor plus tezacaftor plus ivacaftor (ETI). Participants on both hypertonic saline and dornase alfa were randomly assigned to one of the two trials, and those on a single therapy were assigned to the applicable trial. All participants were then randomly assigned 1:1 to continue or discontinue therapy for 6 weeks. For participants randomly assigned to continue their therapy during a given trial, this therapy was instructed to be taken at least once daily according to each participant's pre-existing, clinically prescribed regimen. The primary objective for each trial was to determine whether discontinuing was non-inferior to continuing, measured by the 6-week change in ppFEV1. Participants across both trials had an average ppFEV1 of 96.9%. Discontinuing treatment was non-inferior to continuing treatment with respect to the absolute 6-week change in ppFEV1. The researchers established that in individuals with cystic fibrosis on ETI with relatively well preserved pulmonary function, discontinuing daily hypertonic saline or dornase alfa for 6 weeks did not result in clinically meaningful differences in pulmonary function when compared with continuing treatment.
https://tinyurl.com/2n63sl4y

Onset Of Systemic Arterial Hypertension After Initiation Of Elexacaftor/Tezacaftor/Ivacaftor In Adults With Cystic Fibrosis: A Case Series

Initiation with elexacaftor/tezacaftor/ivacaftor treatment might be associated with acute onset of systemic arterial hypertension. Careful monitoring of cardiovascular paramenters is recommended in patients starting elexacaftor/tezacaftor/ivacaftor treatment. Referrals should be made to cardiologists when patients present elevated blood pressure after the start with elexacaftor/tezacaftor/ivacaftor.
https://tinyurl.com/2fvm5xw5

Heart Failure Common In Cystic Fibrosis Patients

Roughly one in 10 adults with cystic fibrosis also had a diagnosis of heart failure. Among the roughly 15,000 cystic fibrosis patients, acute myocardial infarction (MI) and atrial fibrillation (Afib) were the other most commonly identified cardiac disorders, each present in about one in 20 patients. As patients have grown older, cardiac

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Be Creative: Apply Now For USACFA's Scholarship For The Arts

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

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

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

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

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

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

To apply for this scholarship, please specify what type of creative degree you are pursuing (e.g., fine arts, graphics, music, singing, photography, filmmaking, writing, poetry, dance, theatre, and other performance arts). Candidates should have a minimum GPA of 3.0. Please submit essay answers, electronic samples of your work for consideration, transcripts, and a letter from your physician confirming CF diagnosis. The application deadline is April 30, 2023. https://www.cfroundtable.com/arts-scholarship ▲

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.

Piper's Angels Foundation

Description:

Piper's Angels Foundation is a 501(c)3 non-profit organization dedicated to advocacy and innovation through supporting and improving the lives of families in the cystic fibrosis community by utilizing programs that offer life-expanding activities and provide urgent financial support.

Impact Statement:

Piper's Angels Foundation has recognized that as the life expectancy for people with cystic fibrosis increases, they have been given hope for tomorrow, but they still need hope for today and empowerment to not only live longer, but to live stronger. Through our innovative programs that offer mental, emotional, physical, and financial support, individuals fighting this life-limiting disease can utilize the various tools and resources that empower them to live their greatest life possible.

Programs:

- **Urgent Financial Assistance:** Cystic fibrosis can be incredibly difficult on many levels. Having financial stress can be overwhelming, especially while enduring the challenging treatments. We understand this need and are working to provide financial support to qualifying families to help carry them through these hard times.
- **Forever Stoke Scholarships:** The purpose of our "Forever Stoke Scholarships" is to provide positive experiences in a natural healing salt environment, while also empowering the person with cystic fibrosis to develop skill sets that can become a lifelong gift.
- **Unmasking Mindfulness:** Unmasking Mindfulness is an eight-week meditation program that utilizes live online webinars, educational video content, and in-person one-on-one instruction to provide a platform for the cultivation of mental health coping tools in the CF community. Unmasking Mindfulness educates the CF community on how to practice mindfulness and meditation with a proven scientific-based method.
- **Peer to Peer:** Piper's Angels Foundation's "Peer To Peer" (P2P) program was established to unite and empower the cystic fibrosis community through dedicated mentorship to inspire authentic living. This includes addressing the evolving physical, financial, and socio-emotional needs of individuals personally impacted by CF to achieve a sustainable desired quality of life.
- **Inspire Breathworks:** Our complimentary, sustainable, holistic, and educational health solution to balance the body, mind, and emotions while improving overall well-being of individuals with CF by increasing lung function.

Contact: pipersangels.org
info@pipersangels.org
1-883-PAF-X4CF (1-833-723-9423)

TILLMAN continued from page 28

sequelae, including pulmonary hypertension, right heart dysfunction, and cardiomyopathies have increasingly been linked to cystic fibrosis. Cystic fibrosis might affect the cardiovascular system directly, through CFTR gene dysfunction in the heart and coronary arteries, or indirectly through hypoxia, chronic inflammation, diabetes, and pulmonary hypertension.
https://tinyurl.com/mryev4ea

Acne May Be Side Effect Of Treating CF With Trikafta: Case Study

Researchers describe 19 patients with acne appearing or getting worse after starting Trikafta. All were eligible for Trikafta treatment. Nine had acne before starting treatment and 18 reported acne within eight months of initiating therapy, including nine who reported it within the first three months and four who reported it at six months or later. The face was the most commonly affected part of the body, with some patients referring also to involvement of the chest and back. No patient discontinued Trikafta due to acne as the therapy's benefits in lung function outweighed the risk. How Trikafta results in acne is unknown. The researchers hypothesized that it might be related to alterations in the salt content of sweat, due to changes in the skin microbiome. This would be similar to changes in the lung microbiome observed in patients treated with Trikafta. There is little research in CF patients on the skin microbiome, the collection of microbes, such as fungi, bacteria, and viruses, that naturally live in the body. An alternate hypothesis could be that the rapid decrease in sweat chloride itself precipitates an inflammatory response. The researchers emphasized that further research is needed to obtain a better understanding of various dermatologic changes that may occur with initiation of Trikafta.
https://tinyurl.com/yckmw7cx

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Oral Antibiotics Found To Be More Effective In Treating Cystic Fibrosis

A recent study analyzed the effectiveness of traditional antibiotic therapy as compared to intravenous antibiotic therapy in treating *Pseudomonas aeruginosa* infection among cystic fibrosis patients. This study was a 10-year trial. During the study, 137 patients were treated using intravenous antibiotics while 149 patients were given oral antibiotics. After medicine administration, 44% of patients given intravenous antibiotics recovered while a 52% recovery rate was reported in the case of traditional antibiotics. After the eradication treatment, fewer patients got hospitalized in case of intravenous treatment as compared to oral treatment. Not only were traditional antibiotics more effective than intravenous ones but they also cost less.
https://tinyurl.com/2p8hwkcx ▲

Laura Tillman is 74 years old and has CF. She is a former director and President of USACFA. She and her husband, Lew, live in Northville, MI.

REMINDERS

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

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

IMPORTANT RESOURCES

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

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

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

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

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