

When It Hurts—Deal With It!

By Kathy Russell

Pain is a part of life. Constant pain is a “pain!” It feels as if I have had pain all of my life. My joints, especially my hips, hands, feet, neck, and back give me trouble. When I was young, it usually wasn’t too bad, but, as I age, it can be quite unpleasant. Sometimes it is almost unbearable. Since I don’t use most pain medications, I have had to find other ways to live with it.

My favorite way to deal with pain is to find something that can take my mind off it. I need to be distracted by doing something rather than concentrating on the pain. Sometimes I read something that is very interesting. That’s a good way to be lost. Another distraction is to sew. I love sewing, so being at my sewing machine can help me pass several hours. If what I am making takes a degree of concentration, I am able to forget about my pain, at least for a while.

I used to do a lot of photography. I enjoyed doing portraits of people



KATHY RUSSELL

and getting good shots of scenery. I could lose hours working on my negatives and enlargements. If I was really into what I was working on, six or eight hours could just fly by without my being aware of any pain.

I mentioned that I don’t use pain medication. The reasons for that include having had bleeding ulcers, which make it problematic to take

meds such as aspirin. I can’t use ibuprofen because it causes me trouble with my platelets. Since I have a factor 11 deficiency, it doesn’t make sense to possibly compromise my platelet activity. I don’t use any opioids because I don’t want anything to compromise my breathing and most of them can do that. I avoid acetaminophen due to the potential liver damage from it.

I can use one topical pain reliever gel—diclofenac. The brand name is Voltaren. I use it sparingly, but it does give me some relief from my worst back pain, and it can offer some relief for my hands and feet.

I also use heat to ease the pain in my back. I find that if I have a heating pad on the low setting, I can sit with my back against it and get some relief, at least for a while. I usually fall asleep when I do this, so I believe that it must help the pain.

I recently had bone density scans, spine and neck X-rays, and foot X-rays done. I wasn’t surprised to learn that my bone density has decreased. My

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United States Adult Cystic Fibrosis Assn., Inc.
P.O. Box 1618
Gresham, OR 97030-0519
Email: cfroundtable@usacfa.org
www.cfroundtable.com

USACFA Board of Directors

Sydna Marshall, President Austin, TX smarshall@usacfa.org	Xan Nowakowski, Director Lakeland, FL xnowakowski@usacfa.org
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Beth Sufian, Vice President Houston, TX bsufian@usacfa.org	Beth Sufian, CF Legal Hotline Houston, TX 800-622-0385 cflegal@sufianpassamano.com
---	---

Sonya Ostensen, Secretary Melbourne Beach, FL sostensen@usacfa.org	Sydna Marshall, Managing Editor Austin, TX cfroundtable@usacfa.org
---	--

Colleen Adamson,
Treasurer
Alexandria, VA
cadamson@usacfa.org

Andrea Eisenman,
**Executive Editor/
Webmaster**
New York, NY
aeisenman@usacfa.org

Ela Castillo, Director
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ecastillo@usacfa.org

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EDITOR'S NOTES

Fall heralds change and with that a chance to let go of that which no longer serves us. It's apropos that our focus topic for autumn centers around the transition from pediatric to adult care.

Andrea Eisenman shares her experience of transitioning to adult care back when adult clinics were not yet part of the process—we simply had adult pulmonologists rather than a pediatric doctor. We also have an anonymous submission that details the many challenges of switching clinics along with the multitude of differences between the pediatric and adult clinics, most notably the hospital protocols during an admission. On the flip side, **Mariah Caise** recounts the joy in changing to the adult clinic as she was given the autonomy to manage her CF on her own. **Turner DeMott**, who is on the cusp of changing clinics, writes about his experience in pediatric clinics and his concerns about the change to adult care, especially now that he has undergone a liver transplant.

In this issue, you can also read about the latest CF research in **Laura Tillman's** expertly collated "From the Internet" column. **Dr. Nowakowski** focuses on the myriad ways the transition in care leaves the CF population in a lurch and where we have a great deal of room for improvement to provide the best care possible for our community. **Isabel Stenzel Byrnes** writes about both the seasons of life and the various seasons of health and the importance of finding the beauty in whichever season you find yourself. We're also introducing the return of two of our older columns starting with the winter issue: Transplant Talk and Pet's Perspective. This issue we've got a new column, Culinary Corner from **Maggie Williamson**. Her inaugural recipe, Cream of Broccoli Soup, looks divine and I for one can't wait to make it now that the weather is cooler. If you make it, send us your pictures and reviews.

Jerry Cahill, in the second of a series of interviews, talks about moving forward after his lung, kidney, and liver transplants and what life looks like now in the wake of those operations. Also in the autumn issue, **Rod Spadinger** writes about the mission and origins of CF Vests Worldwide and how the organization has helped CF patients around the world acquire access to much-needed therapy vests.

We're also pleased to announce the winners of the Higher Education Scholarship: Jacob Greene, Samantha Johnson, and Else Mayo. You can read about their accomplishments and accolades on page 6. The Arts Scholarship is 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 and deadline for each. Additionally, Speakers Bureau members, **Isabel Stenzel Byrnes** and **Dr. Xan Nowakowski** 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.

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

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Information From The Internet...

Compiled by Laura Tillman

CF Foundation Estimates Increase In CF Population

The CF Foundation announced that the population of people with cystic fibrosis has increased over the past decade, according to a new estimate. Close to 40,000 children and adults are living with cystic fibrosis in the United States and a total estimated 105,000 people have been diagnosed with CF across 94 countries. The CF population was last estimated in 2012 to be more than 30,000 people in the U.S. and



LAURA TILLMAN

70,000 globally. In the past few decades, evolutions in the CF care model have helped change the face of cystic fibrosis. Advancements in multidisciplinary care, treatments, nutrition, and universal newborn screening have extended lives. For a child born between 2017 and 2021, the median predicted age of survival is 53 years old — up from 38 years a decade prior 34, resulting in the expanded population. There are now more adults living with CF than children. People with CF are achieving milestones never before thought possible — attending college, building careers, getting married, and starting families of their own. As people with CF get older, they often experience more complications and a more complex course of this progressive disease.

<https://tinyurl.com/3frc822y>

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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.

Autumn (November) 2022: Transitioning from Pediatric to Adult Care. (Current Issue)

Winter (February) 2023: Accessibility and CF Care. What does accessibility look like for CF care in clinic? What steps should our healthcare providers take to make sure every patient can fully participate in their own care? How do we make community activities and resources for people with CF as accessible as possible? What are our biggest accessibility needs as a population and what can we do to meet them? Can you give an example of an event you attended that felt fully accessible for you? What made that experience work so well? What does accessibility mean for you at home? Have you and your family modified things in your physical space, sensory environment, social interactions, etc., to help you or others live better?

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



SPIRIT MEDICINE

The Seasons Of Life

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

I have been through major transition in the last months. I learned that my eye cancer has spread. I have ended my job of nearly nine years and applied for disability. I have gone from a high-paced, stressed-out, packed calendar to open and spacious days wondering what I was going to do. I have gone from months without going to clinic to now going every week for chemotherapy.

We all go through transitions. Many of them are positive ones: graduating from college or graduate school, marriage, becoming parents, getting a new job, or transplant. Transitions are part of life: They mark a new stage in our lives—a promotion to the next chapter. We have social rituals to mark certain transitions: the cap and gown, the bridal shower or wedding ceremony, the baby shower, or the retirement party. These are times to gather with friends and family to acknowledge the changes in our lives.

Reaching the end of life marks another transition to the unknown. We can believe in an afterlife, but, in reality, we'll only find out if one exists when we die. The most common social ritual when someone dies is the funeral. We gather to celebrate the life of the person who has "transitioned" or mourn together with their community the transition to their physical absence.

We have fewer rituals for undesirable non-death transitions. There are no rituals for transitions like divorce, getting diagnosed with a serious illness, or for going on disability. If we are lucky, a few friends may take us out to lunch or do something to acknowledge the challenge. Nowadays, it seems like

setting up a Gofundme or CaringBridge.org site is the newest ritual for a personal difficulty.

We all learn how to cope with life's transitions in our own way. What helps me spiritually and emotionally cope with transition is to see these transitions as seasons in life. Many people are familiar with Ecclesiastes 3:18 (or from The Byrds' "Turn, Turn, Turn" song), which says:

"There is a time for everything, and a season for every activity under the heavens: a time to be born and a time to die, a time to plant and a time to uproot, a time to kill and a time to heal, a time to tear down and a time to build, a time to

weep and a time to laugh, a time to mourn and a time to dance, a time to scatter stones and a time to gather them, a time to embrace and a time to refrain from embracing, a time to search and a time to give up, a time to keep and a time to throw away, a time to tear and a time to mend, a time to be silent and a time to speak, a time to love and a time to hate, a time for war and a time for peace."

Regardless of whether you follow the Bible, this age-old passage captures a universal truth for all of us. We all have seasons in our lives. After my transplant, I used to tell people, "I'm in the summertime of my life!" I have been blessed for 18 years to live in my favorite season of summertime. Though I had health issues arise here and there, for the most part, I was strong and healthy and capable of working full time. I was a bagpiper! I was a backpacker! I was a competitive swimmer! I enjoyed every bit of it. I reveled in the miracle of easy breathing. And then I remember the dark winter of my life with advanced CF, when the future was bleak and breathing—and everything, really—was an effort. Strangely, memories of hardship can fade and I hardly remember the struggles I had. I had my turns for winter and summer and now this season.

I spent most of my childhood dreading the fall. The autumn was my season of sickness. I spent almost every Halloween of my adolescence in the hospital. Most of my major health transitions happened in the fall. Halloween or pretty sunsets or cooler weather did little to change my opinion about the



ISABEL STENZEL BYRNES

fall. No matter how much I dreaded the autumn, it was guaranteed to return, year after year. I had no control over the seasons. They were nature's forward motion, the cycles of time as we travel around the sun.

And how I loved springtime. Every time I traveled somewhere beautiful, I'd tell my husband, "This place would be so amazing in the springtime!" I love the greenery, the wildflowers, the moisture, the promise of rebirth and the potential and possibility of life. Metaphorically, my life's springtime was between the age of six and ten, the longest stretch of my pre-transplant life where I did not go into the hospital. For four long years, my parents and I had hopes of a normal childhood, a healthy life despite cystic fibrosis. Perhaps we (my sister and I) had won the genetic lottery and our disease would be mild. We travelled to Japan and Germany during those years, went camping in the mountains, and traveled by boat with my uncle from Seattle to Canada. We soaked up the normalcy. Unfortunately, starting in the autumn of my tenth year, the chronic infections and hospitalizations started and would not stop.

Perhaps some of you are in your springtime of life. You may be starting college or graduating with the world full of opportunities ahead, coupled with your physical wellness. You are blooming and hope abounds! And then there is love... when we find the right person to share our life with, and our will to live gets rebooted and the future seems expansive. This springtime of life is truly beautiful.

There is temptation to worry about what the next season will bring. That anxiety is natural but can also diminish the present season's joys. It takes mental discipline to set aside worries of the future and be able to absorb what is happening now. The sun is shining, the grass is green, and

let's bask in the goodness of now.

Now I'm in a season of serious illness again. Just like nature's seasons, I have little power to prevent seasons from unfolding into the next. It just is, and that surrender and acceptance saves me so much energy. I could protest and wish it wasn't so, but I have to live in reality. Having had a little time to prepare for or expect this season has helped make the blow less intense. I do what I can to seek out treatments, stay compliant with my meds, do research for my situation, and collaborate with doctors. I eat/sleep/rest/exercise well and the rest is not in my control. I can appreciate the good that is in each season, even this one. I feel great relief that I don't have to push myself at work anymore; I don't have to juggle the demands of a job with the demands of healthcare. I was at my breaking point. Thank God that season is over.

I have witnessed so many of my post-transplant CF friends enter a difficult season of illness, including my twin's cancer 13 years after her transplants. They have shown me the way—to face the challenge ahead with hope, realism, grace, dignity, courage, determination, humor, and feist. Some of my friends have died and some are still living. I accept this season of cancer as inevitable after so many years of immunosuppression and my summertime of wellness. I will keep living as best as I can, for as long as I can, and I will appreciate each moment of health. Like chapters in my book of life, I don't know what the next chapter will be about. Maybe I will die, maybe I will live. The only thing that is guaranteed is that this season will change someday. The next chapter will be written.

I wonder what season of life you are in. Sometimes the older we get, the harder the season. But that may not be the case for us with CF. I believe Trikafta has brought summertime to

many friends with CF. But then the next blockage, lung bleed, or major infection can bring all the hope crashing down and we enter a colder season once again. We dance between hope and despair, between wanting the season to never end and hoping the season would change quickly. Such is the nature of these health transitions. And those who are unable to use modulators may be struggling with the wintertime of CF, desperate for new options. We all ride the waves of hope for something better around the corner.

Ralph Waldo Emerson said, "Each moment of the year has its own beauty." So I proclaim to myself that this is also the season of mental and spiritual wellness even if my physical body is not well. I wonder if you, too, can find beauty in each season that you are in. Do you have affirmations that help you get through the tough times?

In conclusion, here are some of my positive proclamations to boost my mental and spiritual health as I endure this season of my life:

I will not dread the future. I will stay in the present. I will surround myself with supportive and loving people. I will absorb positive and enriching books, podcasts, films, and lectures. I will engage in joy every day. I will spend time in nature to remind me of the greater life force and the seasons of all living things. I will believe in a greater Spirit watching over me and showing me the way. I will remind myself of how blessed I have been to have my turn to experience every season and to learn and grow from them all.

May we all practice gratitude each day for all the rich and delightful seasons of our lives. ▲

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.

Recipients Of The Higher Education (Formerly The Lauren Melissa Kelly) Scholarship Announced

The U.S. Adult CF Association (USACFA) is pleased to announce the recipients of the Higher Education (formerly the Lauren Melissa Kelly Scholarship).

In our evaluation, we look for students who demonstrate tremendous academic achievement, community involvement, and a powerful understanding of how their CF—matched with these achievements—places them in a unique situation to gain leadership roles within the community. Our scholarship is open to all pursuing any degree, from associates to Ph.Ds. We believe that any higher education is a strong foundation for advocacy and involvement in the CF community.

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

demonstrated outstanding potential, just like Lauren years ago. Lauren was an inspiration to all who knew her. An incredible leader and scholar, her drive and success are the foundation of her memory. She was transformative in

every aspect of her life. She had distinguished herself as a member of the Golden Key Honor Society, Mortar Board, Phi Upsilon Omicron, Gamma Beta Phi, Delta Gamma sorority, and was chosen as one of ten Senior Leads



JACOB GREENE



SAMANTHA JOHNSON



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DIAMOND ENDOWMENT PARTNER

In Memory of Lauren Melissa Kelly

at the University of Georgia. She acted as one of the re-founding members of the Phi Kappa Literary Society and was significant in the metamorphosis of the Z Club into the William Tate Society. Although Lauren lost her battle with cystic fibrosis late in her senior year, her hard work and memory continue to live on through her inspiring involvement.

We are pleased to announce **Jacob Greene**, **Samantha Johnson**, and **Else Mayo** as the recipients of the scholarships for this calendar year. They will each be awarded \$2,500.



ELSE MAYO

Congratulations to all three!

Jacob Greene is a determined and accomplished individual. He earned his Bachelor of Science with Honors from Stanford University in 2021. He has presented at multiple conferences in the last few years, in addition to his published works. He has been a research intern and assistant at various university labs and pharmaceutical companies. Jacob started his first semester at University of California, San Francisco School of Medicine this fall.

Samantha Johnson plans to graduate with a Bachelor of Arts in Global Studies and Women and Gender Studies with a concentration in Global Health from the University of South Carolina in May 2023. Samantha has authored multiple articles in various magazines in the last few years. She has also served as one of the South Carolina Maternal and Child Health (MCH) Leadership, Education, and Advancement in Undergraduate Pathways (LEAP) mentees at her university. The MCH LEAP program bridges trainees' transition to MCH-related graduate education or public healthcare jobs upon graduation.

Additionally, Samantha has served as both the regional director and writing intern of the Borgen Project in Seattle, Washington.

Else Mayo is earning her Bachelor of Science in Biology at University of Puget Sound in Alaska. Else is very active in the University of Puget Sound Symphony Orchestra as well as chamber music ensembles. She is an active volunteer and participant in various modulator research studies. Else is passionate about access to care and strives to bring both awareness and better access to care for everyone with cystic fibrosis.

All three scholarship winners demonstrated the leadership, intelligence, and drive of Lauren Melissa Kelly. All of us at USACFA look forward to seeing them further develop their leadership and advocacy in the cystic fibrosis community.

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

TILLMAN continued from page 3

Lupin Gets Tentative Approval For Cystic Fibrosis Drug

The drug manufacturer announced that it has received tentative approval from the US Food & Drug Administration (USFDA) for its Abbreviated New Drug Application (ANDA) for Ivacaftor tablets, 150 mg. Ivacaftor tablet is a generic equivalent of Kalydeco tablets manufactured by Vertex Pharmaceuticals. Ivacaftor tablets are used to treat certain types of cystic fibrosis in adults and children 4 months of age and older.

<https://tinyurl.com/556w2ysm>

New Research Could Prevent Hearing

CF Roundtable ■ Autumn 2022

Loss For 50% Of People With Cystic Fibrosis

People with cystic fibrosis are prone to recurring lung infections which need to be treated with aminoglycoside antibiotics. Aminoglycoside antibiotics are very effective against life threatening infections and are associated with low rates of antibiotic resistance, however they enter and kill the sensory hair cells in the inner ear that are vital for hearing. Researchers estimate it may be as high as 50% of adults with hearing loss. Researchers are attempting to develop new aminoglycosides that are less toxic to hearing. By the end of three years the researchers hope to have at least three

new aminoglycosides that can be moved towards clinical testing.

<https://tinyurl.com/2xvap35k>

Exposure To Air Pollution Particles May Add To CF Clinical Variability

Exposure to carbon nanoparticles – tiny air pollution particles that account for a substantial part of air pollution in urban areas – significantly reduces the levels of CFTR, the faulty protein in cystic fibrosis (CF). Being exposed to these air pollution particles also increased signs of cellular stress, DNA damage, and cell death. These findings suggest exposure to air pollution such as

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PEARLS OF WISDOM

Lost In Transition: The Long Shadow Of Missed Opportunities In Young Adult Care

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

It feels good to say that, as of late May, my face is no longer under construction. If you followed my journey through months of reconstructive work on my upper jaw, I appreciate it deeply. I also regret to note that this whole saga stands as but one example of the consequences of poor transitional care for adolescents with CF aging into adulthood. So for our current focus topic, I want to explore some of the other ways that inadequate transitional support for teenage and young adult patients continues to harm our community.

Getting accustomed to losing things is part and parcel of aging with CF. We hear early and often that, despite amazing advancements in treatment, CF remains a progressive disease. No matter how much we manage to slow down that progression in collaboration with our care teams, we still experience damage to many different structures in our bodies. Consequently, we often struggle to understand what is normal for our bodies versus what requires focused intervention. And even for people without CF, adolescence involves both volatility and change almost constantly. How do we achieve a meaningful frame of reference for normalcy in our journeys toward adulthood with CF?

Losing my teeth has been one of many things I have come to accept as usual despite it hardly being normal.

My adolescent and early adult years also found me losing what remained of my exocrine pancreatic function; sustaining permanent damage to my vaginal and cervical mucosa from repeated infections; developing severe scarring and interstitial inflammation in my bladder and kidneys from virulent bacteria; breathing almost entirely through my mouth due to my sinuses becoming completely obstructed with swelling

and hardened mucus; and struggling through over six years of antibiotic-resistant MAC pneumonia.

These changes in physical health predictably exerted mental health tolls as well. Medical trauma probably merits its own case file in this history. Years of being gaslit and ignored by different types of clinicians culminated in me having to dismiss a doctor from my room on an intensive care ward lest I

spent possibly the last day of my life fighting to be heard by someone who would not listen. In a distinctly cruel twist of irony, I later experienced suicidal ideation once I began making more progress with my physical health in collaboration with providers who did listen to and valued my input.

The “biographical disruption” in education and career that scholars in my field describe as one of the hardest things for chronically ill people to deal with hovered constantly in my mind. I learned the hard way that the consequences of trying to avoid such disruptions by powering through serious illness would prove equally devastating. Years later, I remain exhausted physically and mentally. Since the start of the COVID-19 pandemic these feelings have only increased, accompanied by a general sense of being left behind by the world. Constantly being shown how little our lives matter to others can make caring for ourselves feel insurmountably difficult.

Indeed, behavioral health also matters tremendously in transitional care as

“So, even as it poses substantial challenges, this time of increased longevity for many adults with CF also brings unprecedented opportunities in transitional care.”



XAN NOWAKOWSKI

we age into adulthood. Obvious areas like infection-control practices and nutritional support strategies matter immensely. So, too, do less obvious ones like sleep habits, substance use, intimate relationships, and boundary setting. I have shared openly in prior columns about my history of physical, emotional, and sexual abuse. I reflect now on the irony of easily saying “no” to drugs that could harm my lungs or liver while fearing the potentially deadly consequences of refusing sex or anything else to an abusive man. The red flags that my health-care providers during those critical transitional years missed were as numerous in the moment as they are devastating in retrospect.

It took a hospitalization and me being completely alone in my room with a social worker for someone finally to ask if I was being abused at home. Every time I see similar questions in the social history questionnaires for my CF clinic or other care providers, I feel my heart leap into my throat. I cannot help but think in those moments about how different my life might be now had someone asked those questions then. The social worker who rounded on me in the hospital needed only seconds to understand what questions they needed to ask. At the time, I could only avert my eyes and cry silently while they sat with me and held space for my pain. But that single supportive interaction changed the course of my life in ways that would take years to appreciate fully.

Barriers to effective transitional support abound. We should not need to land in intensive care to receive basic prevention and management services to help us transition healthfully and safely into adulthood. Yet in the fragmented patchwork that passes for a healthcare system in the U.S., many of us fall into the cracks between those disjointed pieces. And some of us

“Indeed, behavioral health also matters tremendously in transitional care as we age into adulthood.”

never come back out mentally even if we manage to survive physically.

Adult CF care also remains a rare specialty relative to population need. This disconnect between available providers and patients needing care will only grow larger as survival increases with CFTR protein modulators and other innovative therapies. The practice of separating adult and pediatric patients that some clinics use—including my own—also creates unfortunate consequences for transitional care. Having to start over with a new team further undermines the continuity of care we already struggle to achieve. It can also deepen our difficulty in building meaningful trust with our providers over time. Those longstanding trusting relationships prove critical in our adult years because nobody really knows what healthy aging with CF looks like.

Our providers learn that information primarily by dialoguing with us as we grow older. Although we often lack a defined sense of normalcy, we can often narrate changes in our health in great detail, including observing patterns that might easily escape notice by clinicians. So, even as it poses substantial challenges, this time of increased longevity for many adults with CF also brings unprecedented opportunities in transitional care.

As the primary stewards of our own daily care, we are educating our providers in uniquely valuable ways and actively charting what our care will look like in the future. And as more of us enter health-related professions ourselves, this process of collaborative education becomes easier and more dynamic.

Technology also provides a vital safety net as we continue to navigate transitions in our individual care amid uncertain global circumstances. Staying connected with our different

providers has never been easier or safer thanks to widespread innovation in HIPAA-compliant communication options. During the COVID-19 pandemic, many clinics have also switched to more person-centered medical records platforms that give patients greater control of both our own information and our correspondence with care team members.

My own clinic changing to the Epic EMR system has proven transformational for me and my providers. Having a medical record that highlights and affirms both the complexity of my medical history and the intricacy of my personal biography lets me spend more time on the content of my care and less time trying to establish an appropriate context for it. These changes have made my healthcare more accessible both physically and socially. They have also made balancing care and career substantially easier. I cherish any time I can spend sharing “pearls of wisdom” with my clinical professions students rather than repeating myself in educating my own providers. ▲

Dr. Alexandra “Xan” Nowakowski is 38 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.write-whereithurts.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.



CULINARY CORNER

Cream Of Broccoli Soup

By Maggie Williamson

Hi everyone, my name is Maggie. I'm 34 years old with cystic fibrosis and had a double lung transplant in November 2014 at Stanford in California. Not even a year later, my British husband and I found ourselves crossing oceans and moving to England to start the next chapter of our lives. I have always enjoyed cooking, but my passion skyrocketed post-transplant when everything started to taste better and I was voraciously hungry all of the time. I started cooking up a storm for anyone who would eat my food. I then went on to volunteer for a dementia day center cooking their lunches once a week. I enjoyed the fact I could make people happy with feeding them and began to realize this was what I wanted to do. After a few odd jobs in cafes and looking after people in their homes, I fell into being a private chef. I started cooking out of my own home and making meals for individuals and families based on their specific needs. I work very part time, but enjoy feeding people when I can. My motto for my style of cooking is "I want the food to feel like a hug." I hope you make and enjoy the recipes I will be writing about here in this new column.

Cream of broccoli soup and I go way back. I wasn't a big eater as a kid and my parents would do anything to get me to eat just a few mouthfuls of any type of food. My mom knew she could get me to eat one thing consistently, and that was Cream of Broccoli Soup at our local café, Town Kitchen. It always had a sense of warmth to it, not just because it was hot soup, but the cream and the hint of spiciness from the pepper and the chunks of broccoli made me feel full and satisfied. Of course, I make my own version now and every time I take that

first bite, it brings me right back to the plush, vinyl covered booths, with brown panelling on the walls and an unlimited supply of saltine crackers in a bowl on the table. It is that hug that I look for whenever I am eating.

Cream of Broccoli Soup

Yield: 4–6 servings

Prep time: 15 minutes

Total time: 1 hour

Ingredients:

- 2 pounds of broccoli; cut both florets and stems into small, bite-size pieces
- 1 medium-to-large onion, finely chopped
- 6 tablespoons unsalted butter
- ¼ cup all-purpose flour
- 3 cups of vegetable or chicken stock
- 2 cups of heavy whipping cream
- Salt and pepper to taste

Preparation

Step 1:

Melt 3 tablespoons of butter in a large, heavy-bottomed pot over medium heat. Add onion and a pinch of salt. Cook, stirring occasionally, until onion is softened and translucent, about 5–8 minutes.

Step 2:

Add broccoli stems and florets and season with salt and pepper. Cook, stirring occasionally, until broccoli is bright green and slightly tender, about 8–10 minutes. Using a large spoon, transfer broccoli mixture to a medium bowl and set aside.

Step 3:

Heat remaining 3 tablespoons of butter in the same pot over medium heat. Add flour and whisk consistently, cooking until mixture has turned a pale golden brown, about 2–3 minutes.

Step 4:

Gradually whisk in the stock until no lumps remain (it will thicken considerably at first), followed by heavy cream, and bring to a simmer. Add the broccoli mixture back to the pot.

Step 5:

Reduce heat to low and simmer until liquid has thickened and reduced by about a quarter and the broccoli is completely tender. Season soup with salt and pepper and divide among bowls. Top with some grated cheese or your favorite crackers. You can serve with a salad, sandwich, grilled cheese, or just a piece of bread for dipping. ▲



MAGGIE WILLIAMSON



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.

spine is more twisted and curved than before. The discs in my neck have degenerated even more than before and I have both osteoporosis and osteopenia at various spots in my back. My spine still resembles a pretzel. There is osteoporosis in the neck of the left femur, which helps to explain some of the trouble I have had with that hip.

I had the right foot X-rayed because there had been more pain in that foot recently and I noticed the big toe was harder to flex. I know there is arthritis in both feet, but the right one was giving me more trouble than the left. Both of them feel as though I am walking on marbles, which isn't pleasant. The film showed that there is a prominent bunion along with the arthritis. That explains the increased pain.

There is one thing that can give me some relief from that discomfort and it is a warm paraffin bath. My husband got me a special paraffin bath unit. I can warm it to the appropriate temperature and dip my hand or foot into it repeatedly, until I get a good coat of the warm paraffin. I then put the hand or

foot into a special plastic bag and wrap it with a towel to keep it warm. I am able to have the warmth on it for 15 or 20 minutes, which is quite soothing.

The pain in my hands keeps me from doing some of my favorite things, such as making bread. No longer am I able to mix the dough by hand and I can't knead the bread by hand, either. So, since I still want homemade bread, we got a bread machine. I can still use recipes that I like and let the machine do all of the hard work. We love our homemade bread.

I used to experience severe gut pain. It could wake me from a sound sleep. That pain and gastroesophageal reflux disease (GERD) caused me to lose a lot of sleep. I couldn't find any relief from it. That is, until I had half of my colon removed and *just like that* the pain was gone. I did have to have one more abdominal surgery after that, but it was unrelated to either the pain or the partial colectomy. After the surgeries, I started taking a generic form of Nexium and all of my GERD symptoms ceased. Wahoo!

I spent the first 50+ years of my life

suffering with severe sinus disease. That pain was horrendous and it really had a negative impact on my life. I had a succession of sinus surgeries, which made it better, but it wasn't until after endoscopic sinus surgery that the pain was gone. The relief from pain was so marked that I didn't even need any post-op pain relievers. That earns another Wahoo! It has been 27 years since that surgery, and I have had no recurrence of pain in my sinuses.

So, as I creak on into old age, I keep hoping that there will be some kind of relief for the aches and pains of arthritis and osteoporosis/osteopenia. I am not holding my breath waiting for relief. Instead, I am dealing with it and finding ways to live my life to the best of my ability.

Please stay healthy and happy.

Kathy ▲

Kathy Russell is 78 and has CF. She is a former director and officer of USACFA. She and her husband, Paul, have been living in the same house in Gresham, Oregon, for more than 45 years. In March 2023, they will celebrate 58 years of marriage.

carbon nanoparticles may explain the clinical variability among CF patients, including those carrying the same disease-causing mutation. Airway cells from CF patients have been shown to be more susceptible to nanoparticle uptake and accumulation than those from people without the disease. Previous research showed that the damaging effects of carbon nanoparticles are linked to inflammation and oxidative stress, a type of cellular damage implicated in CF. A team of researchers evaluated the effects of pure carbon nanoparticles on human bronchial epithelial cells grown at an air-liquid interface. Bronchial epithelial cells, those lining the main passageways into the lungs, form the interface between the external and internal airway environ-

ment. Results showed a four-hour exposure to aerosol-delivered carbon nanoparticles impaired the integrity of the cells' natural barrier when compared with before exposure. Carbon nanoparticle exposure also significantly reduced both the activity of the CFTR gene and CFTR protein levels by about 40% relative to exposure to clean air. A significant reduction in the activity levels of two CF genetic modifiers – TMEM16A by 60% and TNFAIP3 by 70% – also was observed. Genetic modifiers are genes or genetic variants that can increase or reduce the severity of a condition without necessarily causing the disease themselves. TMEM16A encodes a protein shown to be critical to CFTR's presence in the cell membrane. Meanwhile, lower TNFAIP3

activity has been associated with worse lung function in CF patients. These findings highlight that exposure to aerosol-delivered carbon nanoparticles reduces barrier integrity and CFTR protein levels, while promoting oxidative stress, DNA damage, and cell death in lab-grown human airway cells. Further studies are needed to confirm this association and understand the mechanisms behind carbon nanoparticle-induced reduced CFTR levels and increased cellular stress, damage, and death, the researchers noted.

<https://tinyurl.com/2y76vhnm>

AND

<https://tinyurl.com/mrcspmkz>

The Negative Impact Of Chronic

Continued on page 19



FOCUS TOPIC

TRANSITIONING FROM PEDIATRIC TO ADULT CARE

Time To Embark On A New Adventure: Adulting

By Mariah Caise

The time has come to transition to adult care—new doctor, new location, new CF team. Is this scary, exciting, overwhelming, nerve-racking, freeing, or all of the above? For me, it's all of the above.

You essentially have the same CF care team your entire pediatric life. These people have known you since you were a baby, and now they are sending you off to a whole new team with whom you will start from scratch. It's bittersweet but also very humbling because there was a point in my childhood where my CF doctor didn't even think I would live to see my teens, much less adulthood, and yet here I am transitioning to adult care. For that, I give myself a huge pat on the back. I would tell my younger self, the one who was sitting in that hospital room with IVs hooked up to her, oxygen in her nose, a feeding tube supplying her nutrients, and staring at the ceiling during that lonely night by herself: "Don't give up. You may not see the light now, but trust me it's there and you will see it very soon."

My transition to adult care started when I was still in pediatric care. My mother passed away when I was 17 years old, and she was my biggest voice when it came to advocating for my care. She was a pharmacist, so she knew the meds and what they could do to my body as well as the mental stress it could take on me. I didn't transition to adult care until six years after my mother's death, but I should have done so much sooner. I blamed my doctor for the lack of time I had with my mother in her last days on this earth. My mother had cancer—she was sick



MARIAH CAISE

and in the hospital. I had a doctor appointment that same week, and my lung function test didn't go very well, so my doctor decided to throw me in the hospital for a tune-up, even though I explained that my mother would die soon. He didn't seem to care. While I was in the hospital, my father showed up to tell me that my mother's oncologist said she had less than 24 hours to live. I did not see my mother that last week she was alive. Once she passed away, my father and I took on the mantle of advocacy. I began speaking up on my behalf and my father would always back me up. He looked at the facts and numbers, but he also knew how his daughter felt. Pediatric doctors want patients to have the highest lung function possible despite other factors or side effects. I fell into a state of depression and lost 75% of my hearing from medicine being pumped in me as

my pediatric doctors obsessed only about optimal lung function.

At some point, I got fed up and hated the fact that I was never listened to—I wanted to make my own choices regarding my CF care. So I disappeared. I stopped going to doctor appointments and stopped taking doctor calls or emails until I was in the hospital every other month and was forced to see them. They would only see me when I was sick and this was unhealthy. So, at the age of 23, I had a conversation to move to adult care. Pediatric care wasn't working for me. I had a toxic relationship with my doctor and I couldn't forgive him. I was ready for a fresh start and a healthier/more positive experience with my doctors.

During one hospital visit, I was able to meet the entire adult CF care team. I asked if they could stop by so I could formally meet them and ask them questions. I was here in the room, anyway, and couldn't go anywhere, so they might as well come by while making their rounds. The main physician came in, followed by the pulmonologist, and the nurse practitioner. I liked that they asked, "Who is Mariah?" and I answered: "Mariah is a sassy, God-loving girl who was diagnosed with cystic fibrosis at four months old. She graduated with her master's degree in accounting from University of Michigan and currently works as an accountant. She likes crocheting, kickboxing, writing poetry and traveling in her free time. She is also 75% deaf so, if she doesn't hear you the first time, she's not ignoring you—she just didn't hear you. She received a cochlear implant back in 2020, so she's taking her hearing back one decibel at a time. She has one motto she lives by and that is 'God saved my life, so I better live it.' She continues to strive

for the best and won't let cystic fibrosis get in her way."

The adult doctor also came to one of my clinic visits and introduced herself to me along with my current team. It was a casual visit, which I thought was cool because it wasn't an official "okay, bye, see you later" moment. I actually got to have an informal meeting with her and just talk about who I am. I shared my background, how sick I was, and she shared how adult life was different than pediatric life. Pediatric care is more of a forced process where you basically do what your doctor says. You go to the doctor appointments and they remind you all the time and are constantly on top of you as far as taking care of your health. Adult care, as I have learned, is the complete opposite. It's all on the patient now and there is more autonomy in managing your own health. There are pros and cons to both. I am stubborn and don't really like to accept help even if it's necessary. I like to be in control and be my own boss. But the pediatric nature of doctoring made me angry because I would be admitted to the hospital when I wouldn't want to go and forced to go to doctor appointments. Adult care is weird—if you don't want to go, they won't force you, they won't check up on you, and they leave it to you. During one visit I straight up said I didn't want to do PFTs and, to my utter shock, they said, "Okay we won't do them. You're an adult, and you don't want to know your lung functions, so we won't force you." And that humbled me a lot. This whole time I was complaining about my pediatric doctor basically forcing me to do things but then, when the adult doctor listened to me, I was like, "Wait, no fight back? No hard time? No explaining to me why it's important and why I should do it?" Just an "Okay, we won't do it." I ended up doing PFTs because I was just so taken aback. Call it reverse psychology or call it the reality of adulting.

“At the age of 23, I had a conversation to move to adult care.”

Transitioning to adult care came with hurdles to overcome as well as a new sense of freedom. I had a full-time job as an accountant and, while transitioning to adult care, I was also transitioning to adult life in general. I had recently graduated with my master's degree in accounting, so I was transitioning from being in school to having a professional job, and having my first car (which I bought for myself), all while managing doctor appointments, visits, and my own health. My father is still a huge help and supporter while I'm negotiating adulthood, but my friends, who would always come to visit, are also all adults and have moved to different states. When I'm sick or hospitalized, they can't come to visit like they used to when we were adolescents in high school. However, going from pediatric to adult care, I will say that the focus is not just solely on my lungs but on other health goals as well. They explain what the numbers on my chart and lab results mean, and I found out that I had asthma. I didn't know I had asthma! Certain things that were ignored in pediatrics weren't ignored in adult care; in adult care they want to be as transparent as possible, whereas in pediatric care, I realized they will hold back certain information so that the family/parents will agree with certain treatment or go with the treatment that the doctor wants. Adult doctors want to have a relationship with you and your input matters. They care about my mental health, my life goals, my aspirations, and all sorts of concerns unrelated to CF. They are not just focused on a number on a paper; rather how the person is feeling is just as important. I'm not saying my pediatric experience was terrible, but I'm also not saying it was great. In the end,

however, I made it to adulthood because of my care team, for which I'm forever grateful.

If there was one thing, I would want to change about my transition it would be the scare tactic. I was fully transitioned to the adult care team by age 23, but that was later than I intended because doctors and nurses would scare me into not going into adult care. They would say they are not as patient as us, you can't pull any crap with them, they don't care whether you show up or not, and they won't baby you. I wish I didn't have that ingrained in my head before going in because it made me scared my care would be worse and that these people had cold hearts, which wasn't the case at all. Don't fill the patient's head with scary stories or scare tactics just so you can manipulate them.

New doctors equate to new perspectives. Change is good; change is scary; change is inevitable; but it's how you adapt to change that makes the whole experience worthwhile. Transitioning can be smooth or bumpy, but so can cystic fibrosis. It's truly not about the destination, but about the journey. Each journey has different paths, detours, hiccups, and end results, but that's what makes it special along the way. So here I am—fully into adult CF care—still learning as I go, but I feel prepared and ready for the long haul. ▲

Mariah Caise is 26 years old and has CF. She was born and raised in East Brunswick, NJ, where she currently resides. In her free time she likes to sleep! On the rare occasion when she's not sleeping, working, or doing treatments, she likes to crochet, write poetry, go bowling, hang out with her friends, and watch her reality tv shows about housewives.



Pediatric To Adult Care: Change Is Hard

By Anonymous

As a kid born in the 80s, and one who is part of the late-diagnosis cohort of people with CF, the transition from pediatric to adult care was less than ideal. Back then we didn't have organized pediatric clinics, organized adult clinics, a national registry of clinics, nor any real method to the madness when it came to coordinated care.

I started out seeing a pediatrician when I was diagnosed in 1991. This pediatrician specialized in CF but wasn't a certified pulmonologist, to my knowledge. He was also the only option in my town. He was demanding of his patients and his bedside demeanor wasn't warm and fuzzy, to say the least. However, he had a long-standing policy that all CF patients were seen immediately, regardless of his schedule. I was always able to get in to see him within the hour. It wasn't until the late 90s or early 00s that they divided the waiting room lobby into "well patient" and "sick patient" sides, with CF patients always sitting on the well-patient side to avoid further exposure from other sick kids. Beyond that, we didn't really have infection protocols in place. We also had maybe two inhaled drugs for CF back when I was diagnosed—the proliferation of nebulized medication didn't come until I was graduating high school and starting college. Nebulizers could be rented from local pharmacies for use at camp, medications were pulled from the big amber bottles with syringes and put in the nebulizer cup, and we rinsed cups with water. Long before Xopenex (and other medications) came in plastic vials, it came in glass vials that had demarcations on the end to easily snap off the ends to pour into the nebulizer cups. However, sharps containers weren't part

of that process. Things were so different in the early days of CF!

Shortly after I graduated college in 2001, I was admitted to the hospital for IVs and a pulmonary tune-up for the first time. Because my hospital didn't have a children's wing and the existing children's hospital wasn't even in the cards back then, I was admitted to the main hospital on the respiratory floor. At the time, and still to this day, that hospital and floor are the only floors people with CF can be admitted to as they have the resources and protocols

to break—they live with me, they share the same germs, why did they need gowns?! After a few years, the four-hour passes were revoked. Insurance, and likely doctors, decided those of us admitted for 14 days who felt well enough to leave weren't that sick and didn't *need* to be admitted and therefore insurance didn't want to pay for the tune-ups in the hospital.

Over time, I developed drug resistance and drug allergies so we had to try newer, stronger drugs that had to infuse over longer periods of time

“That first hospitalization was scary! I sobbed. Now, I'd give anything for the easy, breezy admits back then.”

to take care of us better. That first hospitalization was scary! I sobbed. Now, I'd give anything for the easy, breezy admits back then. I was given a four-hour pass to leave the hospital every day! My mom would come visit and we'd sit downstairs in the gift shop and have coffee; she'd take me to lunch; we would go get pedicures, etc. She brought me snacks and stuffed animals and called me every day, multiple times a day. There were no contact precautions back then. The PICC lines at that time were stitched into the arm—the stat lock to hold the catheter in place wasn't a thing in the beginning.

Over time, these things slowly changed. More awareness around infection control meant keeping the door to your room closed. Masks were worn outside the room. Eventually, I cultured *Pseudomonas* and I was put on contact precautions so everyone had to gown up to enter my room. This was a hard habit for family members

more frequently so hospital stays weren't "spa" stays anymore. They were full days of rigorous nebulizer treatments four times a day, IV antibiotics every eight hours, plus labs and PFTs and little to no sleep. The respiratory floor can be loud at night.

In my late 20s, we started to get more coordinated care and specialized clinics. The pediatric clinic was the first to officially exist—after all, CF was still a pediatric disease then. At clinic we were checked in to the hospital, essentially, but we saw the pulmonologist, dietician, respiratory therapist, and eventually a social worker. When the pediatric clinic was organized, talks of an adult clinic began circulating. And, with that, I would eventually have to switch clinics and pulmonologists. I had been with my pediatric doctor this entire time. He was the one-man-show! He alone visited every single patient he had who was admitted to the hospital every single day.

Continued on page 24



PHOTO BY ALEJANDRA CHAVERRI

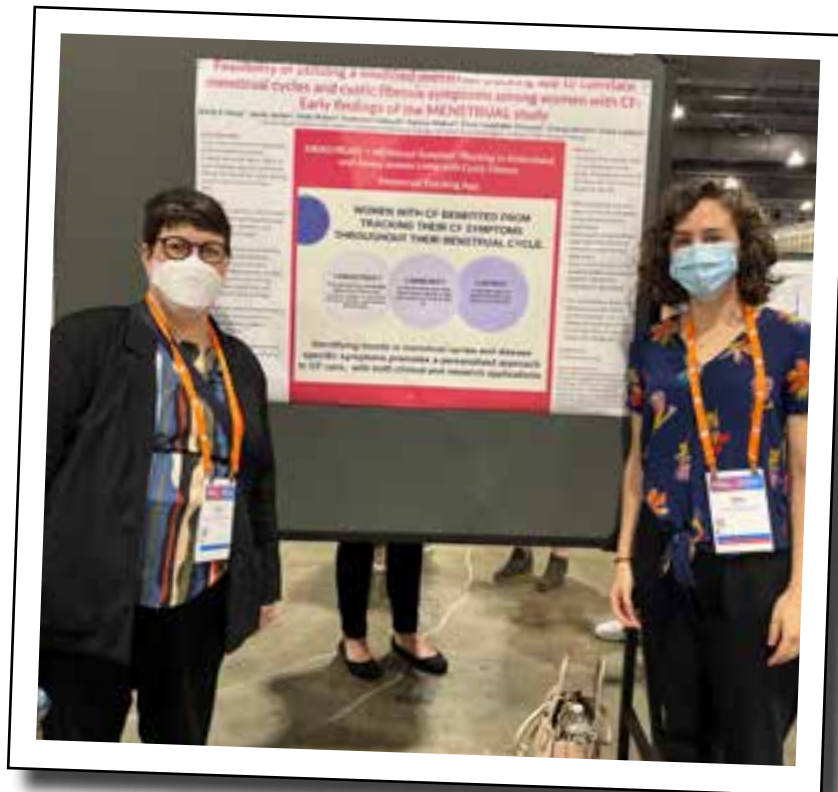
Through My Eyes

Touched by my hands
Nurtured by my words
Comforted by my smile
Encouraged by the sparkle in my eye
My heart has embraced many young lives
The memories are vivid, as is the pain
The paths taken: unique; the journeys: inspiring
Research rewards us
Hope sustains us

But, in the end, the final destination is always the same.

-K. Shelton, 2002

FROM OUR FAMILY PHOTO ALBUM...



SANDY SUFIAN, PH.D., M.P.H., AND MOLLY RUBEN, M.P.H., AT NACFC WITH THEIR POSTER, "FEASIBILITY OF UTILIZING A MODIFIED MENSTRUAL TRACKING APP TO CORRELATE MENSTRUAL CYCLES AND CYSTIC FIBROSIS SYMPTOMS AMONG WOMEN WITH CF—EARLY FINDINGS OF THE MENSTRUAL STUDY."



ANDREA EISENMAN WITH HER DOGS. LEFT TO RIGHT: TRIXIE AND WILLIE.



THE DEMOTT'S FAMILY PHOTO AT JOE T. GARCIA'S IN FORT WORTH: WILLING, MARIAN, TURNER, JASON, BRIGGS, AND ELLA.



MAGGIE WILLIAMSON IN CHARGE OF THE BBQ AT A FRIEND'S BIRTHDAY CELEBRATION.



JERRY CAHILL AT A LUNG TRANSPLANT VISIT WITH HIS NURSE COORDINATOR, MAGGIE CARROLL, AND HIS DOCTOR, DR. SELIM ARCASOY AT NEW YORK-PRESBYTERIAN.



MARIAH CAISE



On The Cusp Of Transitioning To Adult CF Care

By Turner DeMott

When I was an infant, my parents visited many different doctors in an effort to figure out what was going on and why I wasn't healthy. It wasn't until I was 16 months old that my gastroenterologist diagnosed me with CF. For as long as I can remember, I have regularly seen various specialists and doctors as a routine part of my CF care.

At least one of my parents has always accompanied me to my doctor appointments and, whenever possible, both will come. They are always very involved and have lots of good questions for the doctors. I think they will continue to be involved as I move into adult care.

In the summer of 2021, at 16 years old, I was scheduled to have a feeding tube (a G-tube) placed. However, the surgeon was unable to proceed, because a scope revealed varices (swollen blood vessels) around the stomach and esophagus. I would have been at risk of internal bleeding if they had placed the feeding tube. I was then referred for a liver transplant.

Later that summer, I went to Texas Children's Hospital for a transplant evaluation. Over the course of several days, I met with a team of doctors and had various tests. They recommended that we proceed with the transplant. However, I was starting my junior year of high school, with multiple advanced classes. I asked if we could postpone until the end of the school year. They agreed. I went on the organ donor list on June 3. Just 12 days later, we got the call that there was a match. I was in the hospital for two weeks.

After my discharge from the hospital, I had to stay in Houston for

another month for close monitoring, while I recovered from the surgery. My parents and I stayed in an apartment near the hospital district. It was nice and I had my own room, but it wasn't what I am used to. There was less space for medicine. I couldn't do my breathing treatments in my room. And I didn't have a very good bed. However, I was close to my transplant



TURNER DEMOTT

team, and I think that helped me recover faster than if I had gone straight back home to Fort Worth. Being back home now is nice because I have my own room and plenty of space for all my medical equipment. I can do my treatments in my own room and I get better sleep without all the interruptions. I used to share a room with my little brother, which made everything a bit more difficult, but he never got in my way too much. I defi-

nately think it is better to have my own room now, though.

The liver transplant has given me a lot of new medicines to take for about one year. One medicine has to be taken at both 9:00 a.m. and 9:00 p.m. every day for the rest of my life. Transplant has also impacted my diabetes—I now have to take fast-acting insulin before I eat, and I closely watch my blood sugar with a continuous glucose monitor. This has been very complicated due to one of the medicines I have been taking. I wasn't able to get back into physical activity straight away.

The first three months after transplant are probably the most challenging. I just hit that milestone. Now, I am going to start going back to school on campus and I can start playing tennis again. This is my senior year. Fortunately, I have completed most of my high school credits already and will be taking a light course load and won't have to be at school until about 10:00 a.m. This will give me more time for treatments and to ensure my health continues to improve before starting college.

With my liver disease, I had to stop playing contact sports. Both my liver and spleen were enlarged and the doctors were concerned about the risk of impact. This was very disappointing. I had played soccer since first grade, until switching to tennis last year. Recently, three different doctors have noticed that my spleen has significantly reduced in size as a benefit of the transplant. I don't think anyone expected this to happen so quickly or as much. With a new liver and a smaller spleen, I might get to play soccer again one day.

I have been seen by multiple doctors for many years. Throughout all of

my childhood, I have had five ENTs, four gastroenterologists, three endocrinologists, two pediatricians, and only one pulmonologist. One of the biggest changes was when we switched hospitals for some new providers at Children's Health in Dallas when I was 14 years old. When my liver disease began to worsen, my pulmonologist (the lead doctor on my CF clinic team) referred us to a new gastroenterologist in Dallas, who had more expertise in CF and liver disease. Over time, we changed to an endocrinologist and an ENT in Dallas as well. All of these doctors work closely with the CF clinic there. The Dallas doctors helped me by monitoring my liver disease more closely and getting my diabetes under control with less medication. We are able to see the gastroenterologist and endocrinologist on the same day in the same clinic. My gastroenterologist in Dallas referred me to the transplant team at Texas Children's in Houston because of their experience with liver transplants for CF patients. My pediatric doctors have taken a genuine interest in my health and we have developed a team that works well together. This has been reassuring through all

of these challenges.

As I mentioned, I am currently a senior in high school and will be going to college in a year, so I will be at school while I transition to adult care. I am considering some colleges that are out of town; however, my current top choice is a local university, which would allow me to live at home. I think being at home would be good because I could still have parents to help me with my medical needs. For a while longer, I will be able to work with my current team of doctors.

At 17 years old, I do not have to transition to adult care just yet. But, before long, I will. So far, my medical needs have always been met and I anticipate that this will continue to be the case. Because I already have experienced going to three different hospitals to find the care that I needed, I understand that a beneficial transition is possible, even if I have to make changes or go some extra distance.

I hope that my future doctors will be able to make my transition into adult care as smooth as possible, but that will not be easy. I have two rare mutations (so I'm not eligible for any of the modu-

lators available right now) and I have complications that come with these mutations. I regularly see specialists at three different hospitals. My pulmonologist is the only one who has been part of my CF care team from the beginning. I will continue to see a lot of doctors frequently into the future. It is going to be a lot to change all of these doctors. I just hope it goes smoothly and all the new doctors collaborate, communicate, and really listen to my concerns and understand my situation.

When I transition to adult care, I hope my new doctors will see things similarly to my current doctors. I hope I don't have to change doctors as much or as often as I have at times. I hope they understand everything that is going on and can give the same help and care I have received in the past. ▲

Turner DeMott is 17 years old and has CF. He was born and raised in Fort Worth, Texas. He is a senior in high school. As a former soccer player and current tennis player, his primary hobby is playing and watching sports. His favorite team is Manchester United and he once saw them win at Wembley Stadium.

TILLMAN continued from page 11

Rhinosinusitis On The Health-Related Quality Of Life Among Adult Patients With Cystic Fibrosis

With improved survival in cystic fibrosis (CF) patients, it is crucial to evaluate the impact of chronic co-morbidities such as chronic rhinosinusitis (CRS). The objectives were 1) To determine the prevalence of CRS with a large series of CF patients 2) To evaluate the impact of CRS on the Health-Related Quality of Life (HRQoL) of CF patients and 3) To compare CRS-specific, CF-specific and general HRQoL instruments. CF patients were recruited and completed the 22-Item

Nasal Outcome Test (SNOT-22), Cystic Fibrosis Questionnaire-Revised for adolescents and adults over 14 years of age (CFQ-R), Cystic Fibrosis Quality of Life Evaluative Self-administered Test (CF-QUEST) and the 36-Item Short Form Survey (SF-36). Out of 195 patients eligible for analysis, the prevalence of CRS with positive endoscopic findings was 42.6%. CRS patients reported significantly lower HRQoL with higher SNOT-22 scores and lower scores in the respiratory domain of CFQ-R and physical health domains of CF-QUEST and SF-36. The physical and mental domains of SF-36 and

CF-QUEST had a strong correlation with SNOT-22. Higher scores of SNOT-22 nasal subdomains correlated with lower scores of SF-36, CFQ-R and CF-QUEST. Thus, CRS is a prevalent co-morbidity of CF patients, which significantly reduces HRQoL. SNOT-22, CFQ-R, CF-QUEST and SF-36 were strongly correlated. Severity of sinonasal symptoms have a strong correlation with HRQoL in CF patients.
<https://tinyurl.com/2ctbputf>

Impact Of Antibiotic Eradication Therapy Of *Pseudomonas Aeruginosa*

Continued on page 24



My Transition: The Not Good, The Bad, And The Ugly

By *Andrea Eisenman*

Waking up in a hospital bed to sounds of gurgling from a tracheotomy being suctioned in the next bed was a bit shocking. But now, as I look back in disgust at that memory, I am even more horrified that there were no infection protocols in place during my old hospital stays. I was placed in rooms that had three other adult patients in them, on an adult ward. This was in the late 1980s, and it boggles my mind as to what I was unknowingly exposed to back then.

This was all striking because, whenever I was admitted to the hospital in my late teens to mid-twenties, I was in what was termed the “juvenile wing” of Mount Sinai. I had my own room and the nurses were well versed in treating CF. I had a chest physiotherapist come to my room twice a day after my inhalations. We had art therapy sessions, group therapy chats, or cooking classes a few times a week. My doctor and CF care team were pediatric pulmonary folks. I felt they knew all about me and CF and felt confident in their care from the dietician to the PFT lab tech to my favorite nurse who wore many hats.

My first hospitalization was when I was about 17, before I went to college. My doctor felt I could benefit from a tune-up. I went in and it was my first bout of getting IV antibiotics for two weeks. This was when a person with CF had to remain in the hospital for a full 14 days to do IVs—home IVs weren’t offered or part of the protocol back then. Of course, while there, I suffered through boredom, but many people visited me and I was very close to the age of the nurses so we had a lot of fun together, too. We played Pictionary with other patients when they had down

time. There were sad times, too, because some patients would die and all of the nurses were down. They got to know their patients, especially those with CF—we were the “repeat offenders.”

I was admitted yearly after that until my senior year and then it began



ANDREA EISENMAN

to be every six months. I was always admitted to the same floor. At some point, my pediatric pulmonology team told me it was mandated that adults with CF be seen by an adult pulmonologist. I then met with a very nice one who was aligned with my CF doctor. She was more of a consulting doctor—my primary care was handled by my original CF doctor, where I continued to go to a CF clinic for PFTs, to meet with the dietician, to see the social worker, etc.

I didn’t mind seeing the adult pulmonologist outside of the hospital setting, but I didn’t know that I was no longer allowed to go to the juvenile ward when admitted—I had to “graduate” to the adult hospital floor. This

was a big shock. Not only did I have three other women in a room with me, I had to share a bathroom with them, those that were ambulatory at any rate. Also upsetting, every time the nurse came into the room to administer my IVs or inhaled aerosols, the other three patients wanted this or that so the nurse hated coming in the room. The adult floor had a shortage of nurses working. Nurses were charged with taking care of triple the number of patients compared to those on the pediatric floor. They were overworked, and I almost got the wrong IV medication a few times, which is terrifying. I was lucky to get my medications on time, but forget about getting my enzymes before meals, so I brought my own stash. Also, there was no chest physiotherapy, the food was barely edible, and there was no art therapy or cooking classes. But I would say the worst thing was the wild rantings and screaming coming from the other three beds after 10 p.m. I barely slept.

So why was I still seeing an adult pulmonologist if I continued at my pediatric pulmonary CF center? Good question. The adult pulmonologist had the adult hospital rights for when I was admitted. My pediatric team, thankfully, still came to see me but could not write any medical orders on my chart when I was hospitalized—the adult pulmonologist was the only one who could. This was so badly managed that I am sometimes surprised I am still alive. I was exposed to people cleaning the radiators by expressing their exhaust, which were filthy, next to my bed. Right after that, I blew my nose and black gunk came out. I don’t even want to think about what went into my lungs! With those traches being suctioned, other patients coughing, and,

at that time, no thoughts of cross-infection precautions, I shudder to think of what could have been with infectious effluvia flying about the room. This nightmare scenario must have helped keep me out of the hospital and staying well. I somehow managed to keep hospitalizations to a bare minimum. And then, suddenly, I heard about people getting home IV antibiotics. I was elated, although my doctor was less so. I had to sell him on it. He was used to dealing with pediatric patients, not a 20-something woman who would no longer do in-hospital IVs if her life depended on it, which soon it would. I told him I would be open to doing IVs more often without the hassle of being admitted. This was in the nascent period of the health industry allowing patients to care for themselves at home. Eventually, I was persuasive and he agreed to have me start IVs in the hospital for five days and then finish the rest of the 14-day treatment at home. I then was allowed to do the whole treatment at home.

While I still saw the adult pulmonologist, my care eventually went back fully to my pediatric CF clinic. I was no longer hospitalized until I became deathly ill.

Then I was back in the adult unit. But because I was so sick, I was not that upset because I was barely aware.

After a major lung bleed that entailed an ER visit, I was told to go back to see the adult pulmonologist because it was time to contemplate a lung transplant. She was in charge of Mount Sinai's lung transplant center. Through her, I did all of the pretesting that was needed at that time to get into their transplant program. But because they had performed very few lung transplants, my family and I decided to look elsewhere that had more experience and for a center than had a proven success rate for those with CF. With my testing done at Mount Sinai, my family and I went to Columbia Presbyterian where they had done many transplants and I liked the pulmonologist who was to be my doctor there. So, in a sense, I still saw an adult pulmonary doctor at Columbia and remained under the care of my pediatric pulmonary team until I received my transplant at age 35.

I am fully aware that adult CF centers are now the norm and I can see the great benefit that they offer. But when I was transitioning, it was a haphazard piecemeal approach. There was no such

thing as an adult CF center, only adult pulmonologists. Maybe that is why things run so smoothly now and kids are transitioned from a pediatric care team to an adult care team in their early to late teens. It takes a village because CF is not just a lung disease, but intestinal and endocrine as well. A team approach is required.

While my doctors were all great, no one knew better back then because adult care was still a new concept and going from pediatric to adult care was all uncharted territory. So much has improved since my early 20s with hospital care and obviously new treatments for those with CF. Most likely many with CF are not hospitalized so much. I guess I was part of the learning curve. I look back in amazement that I am still alive and, frankly, I think my CF pediatric pulmonologist does, too. ▲

Andrea Eisenman is 57 and has CF. She lives in New York, NY, with her husband Steve and dogs, Willie, Roscoe and new girl, Trixie. Andrea is the Executive Editor for USACFA. She enjoys cooking new recipes, playing pickle ball, biking, tennis when possible, and staying active as her health allows. Her contact information is on page 2.



Announcing The Return Of Our Transplant Talk Column

We at CF Roundtable are also bringing back one of our previous columns, "Transplant Talk!" Transplant Talk is a column that provides personal experiences, insights, and knowledge related to having CF and a transplant or multiple transplants. Members of our board will be writing about their transplant

experiences and we invite our CF readers to share their transplant stories as well! From pre-transplant to post-transplant, what have you experienced? What made you decide or not decide to have a transplant? What have you learned about getting a transplant? What have you learned about yourself? What side effects have you had?

Inquiring minds want to know!

We want to hear all about your experience with transplant, and, best of all, you will be helping people with CF who are pre-transplant understand what all is involved in having a transplant, hopefully alleviating some of their anxiety along the way.

Send articles to: articles@usacfa.org



CF Vests Worldwide: Reaching Out To People In Need

Arshaad: a four-year-old boy in Bangladesh whose parents were broken when they found out their son carried a fatal disease they had never heard of before. Yasmin: a seven-year-old girl in the Netherlands whose uncle died at the age of thirteen from the same lung disease she has. Natalia: a sixteen-year-old girl in the Ukrainian city of Mariupol, which was bombed and occupied by Russian troops, who fled the blockade of her city after three weeks of Russian occupation, hungry and cold. All three of these children have cystic fibrosis and each one lives in a medical environment where access to a therapy vest, a basic health essential in the United States, is painfully difficult. These children are trapped in the geography where the possibility of obtaining a \$20,000 piece of equipment without the assistance of robust health insurance coverage is grossly unattainable. Rod Spadinger, President and Founder of CF Vests Worldwide (CFVWW), established this charity in June 2020 with a mission of providing vests to CF patients around the world. As of October 2022, CFVWW has donated 155 vests to CF patients in 40 countries, ranging from Ecuador to Thailand, with dozens of countries in between.

Rod, born and raised in Honolulu, Hawaii, has navigated his way through the many bumps and hurdles CF brings since his diagnosis at three months of age. At four years old, Rod became very sick—it was 1976 and both CF knowledge and care in that period were limited. Desperate for an answer, Rod's parents contacted the CF Foundation seeking the best cystic fibrosis doctor in the United States. The Foundation recommended seeing Dr. Robert

“Upon Mohamad's vest request, Rod reached out to the online CF Community asking if anyone could donate a vest to Mohamad, expecting that no one would ever respond.”



ROD SPADINGER

Kramer in Texas. Dr. Kramer's sage advice for Rod to run five miles a day, each day, is the reason Rod remained alive and relatively healthy—relatively healthy in the world of cystic fibrosis, that is.

In August 2017, a week before his 45th birthday, with 17% lung function, Rod was graced with a double lung and liver transplant from his angel donor, Joni Marie. As a way to express his heartfelt appreciation to her, Rod published his memoir titled *A Collection of 50 Stories Inspired By My Angel Donor, Joni*

Marie exactly two years after his transplant. This was a means of giving back to his donor's family by honoring their angel daughter. A year later, Rod discovered another method of giving back.

In January 2020, Rod received a message on social media from Vale, a young lady from Chile whom he did not know. Vale asked Rod if he could donate a vest to her. He thought at first this person was likely a fraud. “Who is this person, how did she get my contact information, and why in the world would anyone ever give someone they don't know a vest worth tens of thousands of dollars?” were his immediate thoughts.

When Rod realized he no longer needed the Hill-Rom enclosed in the dark blue duffel bag that had been sitting for two and a half years unused under a table, six feet away from his laptop, on which he received Vale's message, he decided to FedEx his vest to her. That turned out to be the very first vest the soon-to-be-created CFVWW would donate. The second vest request came two months later from Mohamad, a CF patient and a Palestinian refugee living in Lebanon. This was another message to Rod by someone he did not know. Mohamad, like Vale, was a stranger. Upon Mohamad's vest request, Rod reached out to the online CF Community asking if anyone could donate a vest to

Mohamad, expecting that no one would ever respond. "Who just gives vests away to strangers? No one," Rod mused. However, within a week, Danie from Ohio heard his call. In a matter of days, Rod was able to facilitate Danie's vest to Mohammad in Lebanon. Danie became the first CFVWW vest donor, with Mohamad being the second CFVWW vest recipient.

Two strangers living 8,500 miles apart contacted, within months of each other, a CF patient in Texas whom neither knew. These two strangers unwittingly spurred the creation of CFVWW. Thirty-four months after the message from Vale to Rod, CFVWW is gaining momentum as it drives to the next milestone of 200 vests to CF patients around the world. CFVWW will not stop, as the need is too great.

CFVWW's mission to continue donating life-saving vests to those living in medical deserts accelerates forward as there are thousands of CF patients across the globe who are without reliable care and no means for proper airway clearance. Want to become a member of CFVWW's worldwide mission? If you have a vest machine and/or vest garment sitting unused in a closet or resting under a table, please contact Rod at rod@cfvww.org or visit www.cfvww.org. Your donation of a vest will make an unimaginable difference in extending the life of a CF patient unknown to you in a far-off country. What's more, your donation will elevate you to hero-status for that CF patient and their family, as your vest will truly be a gift to them worth more than gold. ▲

Rod Spadinger is 50 years old and has CF. He was born and raised in Honolulu, HI, but now lives in Dallas, TX. He enjoys following the financial markets, catching football games on the weekends during the season, and reading when he can find the time.

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Scholarship for the Arts
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IN MEMORY OF
LAUREN
MELISSA KELLY

He missed one visit one time in all those years. He referred me to some of my absolute favorite specialists in that time. He saw me in his office every single day for months before my first admit because I was that sick and he was that persistent in trying to get me healthy. He was always available to see me, even last minute, and he took the time to call me back personally with any results from cultures. It took us a long time to reach an understanding, but I respected his no-nonsense attitude and he heard my concerns. He also delivered quite a few devastating one-liner comments in those years, but, ultimately, I needed the harsh truths. Mostly, he kept me alive.

The adult clinic was also structured, even more than the pediatric clinic—more specialists at each appointment, more forms to fill out before appointments, and more processes and protocols to follow, etc. Some of these changes could have been general changes across all clinics, not just the adult. Regardless, switching to the adult pulmonologist also meant I would have a team of doctors who have an on-call rotation for visiting patients admitted to the hospital so it wasn't guaranteed that I'd see my CF doctor during any admits. I was assured that records were shared and this would be an easy and seamless

transition. I would also lose my pediatric nurse coordinator, who was an absolute joy and made the frustrations of CF and clinic easier. I waffled on what was the best approach—wait out the change until my doctor retired or jump ship and learn to swim.

On my 30th birthday I switched to the adult clinic. It's still one of the most poignant moments of my entire life of CF care. I wrote my doctor a long note thanking him for helping me make it to 30! At that time, I had outlived my life expectancy twice.

The switch to the adult clinic was a huge adjustment for me. The day-to-day clinic experience was essentially the same. The hospital admits, however, were another story. Contrary to what I was assured, even though records were supposedly shared, the on-call doctor rarely wanted to make "the call" for any change in treatment or protocols or plans as I wasn't their patient when outside of the hospital setting. This was infuriating, especially when you have a complex medical history and putting all the pieces together like a jigsaw puzzle is a challenge. During one of my admits, I had an on-call doctor threaten to pull and discontinue every single medication on my chart if I wouldn't produce a urine sample for a pregnancy test, despite

my continued arguments that I'd had an Essure and an ablation and pregnancy was impossible. I'm still irate over that!

Now that we're a decade plus into this transition, I'm happy with the change. As the CF population changes and ages with the advent of these new therapies, new modulators, and new medications, so does the care. The infection control, the communication between specialists, the online patient portals, and the push for a primary care physician to handle regular aging has all been a tremendous step forward. Do I miss seeing the same doctor each day in the hospital? Yes. Does he visit me just to say hi when he's in the hospital for another patient? YES! In the end, I benefit from the specialty care that comes with a pulmonologist and a clinic that stays up on all the latest research and trends in the community.

We're on the precipice of a huge learning curve across the community. CF is no longer just a pediatric disease. People with CF are living longer, fuller lives and the adult care will be critical in this next chapter. And the transition from pediatric to adult, in theory, should be much smoother for the younger patients as we, the doctors, specialists, and patients, know more now than we did when I was first diagnosed about managing CF. ▲

On Long Term Lung Function In Cystic Fibrosis

While antibiotic eradication therapy (AET) of early *Pseudomonas aeruginosa* infection is considered standard of care, its long-term effect on the subsequent course of cystic fibrosis lung disease remains unclear. CF patients who were *P. aeruginosa*-free for at least a year and had a minimum of 10 years of pulmonary function measurements were included. Subjects were categorized as Never if they never had *P. aeruginosa* isolated from a respiratory tract sample.

Subjects changed to the Eradicated group if they had a *P. aeruginosa* infection, were treated with AET, and subsequently cleared their infection. Subjects changed to the Chronic group if AET did not clear their *P. aeruginosa* infection. The primary outcome was absolute FEV1 decline over time, with age as the time variable. The researchers concluded that AET against *P. aeruginosa* improves lung function trajectory in CF patients.

<https://tinyurl.com/3urwrda>

Regular Antibiotic Use In CF Linked To Hypersensitivity Reactions

Regular use of antibiotics to treat persistent infections is linked with hypersensitivity reactions in individuals with cystic fibrosis. Specifically, the risk is highly associated with the number of cumulative exposures to antibiotics and increasing age. And intravenous administration of antibiotic treatments was tied to a much higher percentage of hypersensitivity cases. However, most cases involved mild to moderate skin

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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 other 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 build-

ing 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 silk-screen 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., PM.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 US CF community.

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



TRANSPLANT TALKBACK

My Conversations With Jerry Cahill—Moving Forward

By Andrea Eisenman

What are your thoughts post-transplants (liver and kidney)?

Getting two organs at once is pretty rare and doing so brings with it many complications, like blood loss, rejection, and possibly death. I am amazed and humbled at what it takes to go through this experience.

I just celebrated my one-year anniversary of receiving the two organ transplants (July 22, 2021). It has been a long, slow process to heal and recover. But I am thankful and grateful for more life. There are a lot of realizations after receiving two more transplants. I thought after getting a bilateral lung transplant, I am done and I can get on with my life. So, it was jarring to have to be listed as needing two more organs. I can't even imagine what it must be like to receive a second lung transplant. This experience has made me realize that life is short and precious.

What is exercise like for you now?

Getting stronger is a lot more difficult than with my lung transplant. I am older, and, at 66, I can no longer do things I did post-lung transplant, like running a marathon or biking centuries. Life is still good, though. I still volunteer some at Boomer Esiason Foundation and coach track and pole vaulting at Iona Prep. To remain strong and fit I now walk and hike. I don't jog or run any longer, but I decided to start back to biking outside. Instead of road bikes and 100-mile bike rides, I own a bike that is more upright and I don't go so far. I went from biking on high-end road bikes to a lower-end hybrid bike. Luckily, the bike store guy sold my road bikes for me and suggested I buy a more comfortable hybrid. I keep having to reinvent myself to keep mov-



JERRY CAHILL ON HIS NEW UPRIGHT HYBRID BIKE.

ing forward.

My transplant pulmonologist, Dr. Arcasoy, and I have different ways to measure my success with the new transplants. We have this back and forth on most of my visits because I am frustrated with my level of fitness and he is elated I am still alive. He laughs when I lament my strength level. In his opinion, I am doing better than 99% of all transplant patients he knows, and he feels I am fortunate I got the transplants when I did. That sets me off and I say, "Well if they weren't going to do it here (at Columbia), I would have gone elsewhere!" He smiles and says, "Nobody would have taken you. I had to argue with liver and kidney teams for you to get it." He says he doesn't want to hear how strong or weak I was. In the end, though, he was my fiercest ally in pushing for me to get these two transplants. Speaking honestly with me he said, "No one expected you to get through this surgery." Obviously I was sicker than I realized.

What are you up to aside from exercise?

With three transplanted organs, my biggest job is staying on top of my health: being aware of rejection if it starts; constantly going to doctor appointments; and getting lab work weekly to make sure all is right. Initially, right after the liver and kidney transplants, I had to be seen once a week with each of those two transplant teams on different days. Now I see each of them once a month and, as I am farther out from the surgeries, it will probably become every three months or so. I continue to see the lung transplant folks every two months, but, if there are issues, I have to see doctors more frequently.

It is a balancing act, as you well know. For anyone with CF or specifically transplant, a small infection can bloom into a major problem quickly due to being immunosuppressed and not having the ability to fight things off on my own.

Do you get biopsies to check for rejection for these two new organs like you did for your lung transplant?

They have not done biopsies yet. Blood work apparently is the be-all and end-all. So they can see a lot of information about adjusting my immunosuppressants through blood work. I do deal with high potassium levels in my blood, which can be dangerous for my new kidney. So, to keep it in check, I am prescribed a powder that I ingest to reduce potassium in my system. I am no longer allowed to eat bananas and tomatoes, which are high in potassium.

Maintaining my health also means seeing my skin doctor every few months (to remove skin cancers, which have become prevalent post-transplant); checking in with my sinus doctor every few months; and seeing the GI doctor for an endoscopy and colonoscopy. It's a juggling act to fit it all in. The medical stuff is not my life but becomes a major part of it. I enjoy my volunteer work, but, at times, it is hard to do it all. I have started napping in the afternoons after the liver/kidney transplants because I am more fatigued. Sometimes it is overwhelming, but I do try to do what I can and remain positive.

How do you view transitioning from pediatric to adult care and then to transplant?

In regards to transitioning, I look at it as a promotion because most people with CF do go on to adult care. I compare it to sports. If you are a freshman or sophomore playing a sport, your goal is to be in varsity or the Olympic team. Similar to when you are in pediatric care, going to adult care is the ideal goal. If you do well, you move onward. It's part of maturing. In high school I was going to the CF doctor on my own, but, if there were issues with my lungs, they contacted my parents. I feel like most people with CF grow up sooner than their healthy peers because we are forced

to deal with life-and-death issues that must be dealt with—it is unavoidable. It forced me to grow up faster and be more responsible for my own care.

My experience with transitioning was difficult. I was in the dark ages of CF treatment, and I saw only a pediatrician into my 30s. It seemed like all of a sudden it became mandatory to see an adult CF pulmonologist. But there really were no adult pulmonologists who specialized in CF care like I was used to at my clinic. Once my pediatric pulmonary physician, Dr. Gorvoy, retired, I was forced to see a few other adult pulmonologists. At that time I was 30-something. That didn't work out well. One was mostly asking me about CF and for me to explain to him what this or that meant as if he didn't know anything about CF or how to treat it. I then saw a few others and none worked out, so I continued seeing Dr. Gorvoy until he finally retired. He insisted I see an adult pulmonologist and finally sent me to another one at Columbia. He was also a transplant pulmonologist as well as the director of the transplant program at that time. Because I was told I had to be seen at a CF center, I started seeing an adult CF pulmonologist at Columbia named Dr. DiMango, whom I liked a lot. She knew CF well and I felt I was getting good care and even taking part in clinical trials for drug studies. I finally ended up with proper care by an adult pulmonologist, but it was a lot of stops and starts to get there.

What were your thoughts about lung transplant?

Eventually, with my declining PFT scores, it was recommended to me by the CF center that I get listed for transplant. That, too, took a circuitous route, but I ended up back at Columbia after being seen at UNC Chapel Hill.

In the end, I didn't list with UNC because I would have had to move there with a family member to wait for a trans-

plant. I am very independent and I said I would not be comfortable doing that. I don't like to have to rely on others, even if it is my own family. Thankfully, around the same time, one of the nurses at the transplant center at Columbia called me and said they have a transplant team with a new surgeon. She told me about Dr. Sonett and the transplant pulmonologist, Dr. Arcasoy. I met them both and liked their approach. I then decided to list solely at Columbia Presbyterian in New York City.

Dr. Sonett is still the surgeon I see if I need any surgical corrections, but, thankfully, I have not. I am happy I was able to remain close to home in New York and not have to move to get my lung transplant.

Are you taking Trikafta again?

Prior to receiving my new liver and kidney transplants, I could not be on the prescribed dosing because it was damaging my liver. I was only on it twice a week so it was not ideal, I was only getting partial results. I am now on a full dose because my liver is fully functioning. Now that I am on a full dose of Trikafta, I see a huge difference in my sinuses. The incidence of infection of my lungs due to my sinuses so far has reduced to zero. Even my ENT is amazed at how great my sinuses look.

What side effects, if any, do you have from Trikafta?

So far, I have had no side effects from Trikafta. I don't know that I am really human or alive! Sometimes I think I am a robot. I have had so many COVID-19 vaccine injections and Evusheld shots and have had zero reactions. There are a lot of medications and vaccinations coursing through me and yet I don't have any side effects. But I actually did get COVID-19 in May.

What was your experience having COVID-19 as a transplant patient?

One Saturday in May I started feeling run-down and thought I was over-

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doing it with volunteering. I felt achy and had a sore throat. By the next day I felt slightly worse and someone suggested I test for COVID-19. I tested positive on two home tests. Because I have a great relationship with my transplant pulmonologist, Dr. Arcasoy, I texted him immediately. Within a few hours, I had arranged for monoclonal antibodies through Maggie Carroll, the transplant coordinator. After filling out the prerequisite paperwork online, a nurse administered the antibodies that evening via IV push over a five-minute period. The nurse said I would be fine the next day. I was dubious because I woke up in the early morning with a

fever of 101 degrees. But by late morning the next day I felt completely fine. I did quarantine at home for ten days and was told I might remain positive for COVID-19 for up to two months.

What is your life view?

Living with transplants is different. Living on disability is challenging—nothing is easy. And transitioning to retirement is not going to be possible for me now.

Currently I still have physical struggles as I don't feel as strong or as in shape as I would like. There are struggles financially because the Long Term Disability I was blessed with will be ending December 31, 2022. I will be start-

ing on Social Security and I can't live solely on that. Since I grew up so long ago when CF was only a childhood disease, I was never taught to save for a "rainy day." Most people with CF that I grew up with are now no longer alive. I was never taught to save for my future as I was never assured I would have one. It is scary to think I might have to get a job at this age. But I am ready for whatever I have to do and to keep moving my life forward. ▲

Andrea and Jerry will continue their conversations about life after transplant. If you have questions for Jerry Cahill, email us at: cfroundtable@usacfa.org.



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

Blake Caven

Kansas City, MO

31 years old on June 21, 2022

Transplant

Andrea Eisenman, 57

New York, NY

22 years on April 25, 2022

Wedding

Colleen and Scott Adamson

Alexandria, VA

25 years on June 28, 2022

Andrea Eisenman and Steve Downey

New York, NY

14 years on September 13, 2022

Dr. Xan Nowakowski and Dr. J Sumerau

Lakeland, FL

6 years on June 25, 2022

reactions. Severe reactions, such as anaphylaxis, were rare. People with CF are prone to infections with *Pseudomonas aeruginosa* and often need 10- to 14-day intravenous treatment courses with antibiotics. Among antibiotics of choice are beta-lactams such as ceftazidime and piperacillin, aminoglycoside antibiotics

including tobramycin, or polymyxin antibiotics such as colistimethate sodium. However, CF patients are known to be susceptible to antibiotic hypersensitivity — though it is unclear how frequently such reactions occur and what are the factors increasing the risk. A team of scientists led a retrospective study that

addressed this knowledge gap. The rate of antibiotic hypersensitivity reactions was 31%. A vast majority of cases (98%) were skin reactions — 30% itching, 23% skin eruptions or rashes, and 18% hives. Some patients experienced tingling sensations, and rare cases of nausea, vomiting, diarrhea, shortness of breath, cough-



NEW PROGRAMS COMING THIS FALL AND WINTER FOR THE CF COMMUNITY
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ing, dizziness, and low blood pressure also occurred. Three reactions were potentially life-threatening and classified as anaphylaxis. In 48% of cases, hypersensitivity reactions occurred between two and 10 days after the antibiotics were given, but in 28% of cases, the reactions happened occurred within the first hour of antibiotic injection. Other reactions occurred after one hour but before 24 hours of the first antibiotic dose. Beta-lactam antibiotics were responsible for 71% of all reactions and 81% of non-oral

courses. Piperacillin was the most common cause of intravenous reactions, followed by ceftazidime, colistimethate, and then meropenem. Regarding oral antibiotic treatments, cotrimoxazole was the most common cause of hypersensitivity reactions, followed by amoxicillin and ciprofloxacin. Skin eruptions or rashes, angioedema (swelling under the skin), hives, itching, and flushing were the most common reactions to oral antibiotics. The researchers noted that no patients developed anaphylaxis after oral

treatment. Intravenous antibiotics were almost six times more likely to cause hypersensitivity reactions than oral antibiotics. The number of antibiotic courses significantly predicted having these reactions, with patients taking the most courses displaying the greatest risk.
<https://tinyurl.com/f36kjt5u>

Antibiotic Switch With CF Pulmonary Exacerbation Not Likely To Help

Pulmonary exacerbations in CF

Continued on page 30

patients are often caused by bacterial infections. Usually, after a test of which microbes are responsible, patients are treated with antibiotics to kill off the pathogen. In individuals who don't respond to a first antibiotic, healthcare providers may switch treatments. Switches included antibiotic additions, subtractions, substitutions, or regimen changes. Common reasons for such switches included a change in antibiotic administration for discharge patients, drug reactions, and targeting additional microbes. In other cases, a switch was made because the exacerbation failed to respond to initial treatment, as evidenced by a lack of symptom improvement or by FEV1 values not returning to at least 90% of what they were prior to the flare. Researchers then assessed whether the switch benefitted lung function in these non-responders by comparing them with non-responders who did not change their antibiotic treatment. Most clinical features did not differ between the switch and no-switch groups. Baseline lung function, or that prior to a pulmonary exacerbation, was poorer among those given an antibiotic switch than those who were not. But lung function was similar among patients in both

groups at the time a first treatment was initiated. Overall, those in the switch group saw significantly smaller absolute improvements in FEV1 by the end of treatment, even after switching antibiotics, than those in the no-switch group. In a final analysis, the researchers determined that an antibiotic switch did not have a significant positive or negative influence on lung function at the end of treatment or at a follow-up about three months later. The proportion of patients whose FEV1 values returned to at least 90% of baseline values also was not significantly different between those who switched antibiotics and those who did not. However, given the lower baseline FEV1 values among patients who switched antibiotics, findings imply that these individuals may have more substantial lung damage which may affect their treatment response. Gains in lung health noted with an initial treatment in some patients may be slower to arrive for others, which in turn influences the response of healthcare providers. It's also possible that in some cases, the wrong bacteria is being targeted for treatment, or that the cause of the pulmonary exacerbation was not infectious in origin.

<https://tinyurl.com/bd36bpa6>

Duration Of Intravenous Antibiotic Treatment For Acute Exacerbations Of Cystic Fibrosis: A Systematic Review

Acute exacerbations of Cystic Fibrosis (AECF) are associated with significant morbidity. Recommendations are to treat for 2-3 weeks despite limited data. Spirometry is a measure of clinical response yet appears to plateau at 7-10 days. While durations <9 days have been associated with poorer outcomes, a duration of 10 days may be as effective as 14 days, potentially conferring advantages in terms of cost and adverse events. A 2019 Cochrane review did not identify any randomised controlled trials (RCT) comparing durations of treatment. Utilising data from non-randomised studies (NRS), researchers reported a systematic review of intravenous antibiotic treatment, exploring changes in FEV1 (Forced Expiratory Volume in 1 second), CRP (C-reactive protein) and peripheral WBC (white blood cell) count in studies with different treatment durations. Studies were categorized according to their duration of treatment, of 10-12 days and 13-15



Announcing The Return Of Our Pet's Perspective Column

We at *CF Roundtable* are bringing back an older column about people with CF and the pets that support them through their journeys—Pet's Perspective. Please consider sharing your stories about your side kicks. It can be from the pet's perspective of our daily treatment schedules: how it affects their napping or play times; how they attend to our needs; get us motivated to exercise; their snarky

side eye/stink eye; their concerns when we are in the hospital or doing IV treatments, etc. For example, "why is my human up at all hours and too tired the rest of the day to play with me? Why am I not getting my afternoon walkies?" You can write about their vacations with us and our families, their daily observations of our coughing, or the lack of coughing after Trikafta or a lung transplant. Or you can share how your pet

helps you cope and keeps you feeling well in whatever way that they do.

This can be any pet, from four-legged to the feathered to the scaled (no pet rocks!). All contributions must be accompanied by a photo of said pet with you. Or, to illustrate their attitude, one photo of just them.

Let's share our joyous bestie stories!

Send articles to: articles@usacfa.org

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days. No significant differences in change in FEV1 were observed based on duration of treatment. They also determined that treatment setting may affect outcome. When the entire treatment course was as an inpatient, no significant difference in change in FEV1 was observed. However, where some of

the total treatment duration was administered as an inpatient, studies with longer treatment durations had a significantly greater change in FEV1 compared with shorter durations. This systematic review provides evidence that shorter durations of treatment may be associated with similar changes in

FEV1, CRP and WBC compared with longer durations.

<https://tinyurl.com/t2d3xe7w> ▲

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



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IMPORTANT RESOURCES

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

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

Transplant Recipients International Organization, Inc. (TRIO): Phone: 1-800-TRIO-386 <http://www.trioweb.org/index.shtml>

An independent, nonprofit, international organization committed to improving the quality of life of transplant recipients and their families and the families of organ and tissue donors. For information, write to: TRIO, 7055 Heritage Hunt Dr, #307, Gainesville, VA 20155 or email them at: info@trioweb.org

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

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