CF Roundtable®
A PUBLICATION FOR ADULTS WHO HAVE CYSTIC FIBROSIS
WINTER 2024

Riding The Rollercoaster Of Life Post-Transplant

By Jenna Strickland

On the outside I look like the typical 30-something-year-old woman, but my insides tell a very different story. I was diagnosed with cystic fibrosis at birth after being born with meconium ileus, which is a bowel blockage in a newborn's first stool. I continued to have stomach issues and blockages throughout my childhood and started suffering from frequent lung infections in high school. After a long battle with chronic lung infections, which got a lot worse in college, my lungs were greatly scarred and, by the time I was 25, I was dealing with a rare bacteria, *Pandoraea*, in my lungs. This bacteria made me extremely sick and, unfortunately, there wasn't much research on it or how to treat it. My CF doctors contacted other CF centers to try to get any information on this bug. I blamed myself for contracting *Pandoraea*, but we weren't even sure where I had come in contact with it.

JENNA STRICKLAND

I started to need IV antibiotics monthly in addition to oxygen at night and during exercise. Eventually I needed supplemental oxygen 24/7. I even had to wear oxygen in the shower and remember feeling so short of breath walking just a few feet. I had a feeding tube because maintaining weight was impossible with all the coughing and lung infections. I was underweight my whole life and people were always telling me to eat more, assuming I was anorexic. Little did they know I was coming home from school and eating fast food daily and choking down high-calorie Boost nutrition drinks.

Around 2014, my CF doctor told me I would need a double lung transplant to survive and he referred me to Columbia Presbyterian in New York for lung transplant evaluation. After what seemed like a never-ending list of tests, both physical and mental, I was finally deemed a candidate for a double lung transplant. I was listed and it felt like a big relief even thinking about transplant being a possibility, since there was no guarantee I would get a call for lungs. After one month

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INSIDE THIS ISSUE

CF ROUNDTABLE
FOUNDED 1990
Vol. XXXIV, No. 1

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EDITOR'S NOTES

This issue we're focusing on CF and transplant. For the focus topic, **Paul Albert** details not only the myriad complications he endured after his double-lung transplant in 1992 but also how the transplant journey, despite the challenges along the way, was worth it. He has definitely lived a full life after his first transplant and is continuing to do so after his kidney transplant in 2022! **Piper Beatty Welsh** writes about all the ups and downs she's encountered after each of her double-lung transplants and the lessons learned along the way. We're also featuring the art of **Dominic Quagliozzi** who showcases both his transplant process and life post-transplant in Blue Morpho and Monarch.

We're excited to introduce two new columns this issue! **Marcus Miller** pens his reflections on running a 50-mile run in Moab, Utah, for the first "Adventures Abound" column. **Matison Deaton** writes about the many logistical roadblocks on the journey to her double-lung and kidney transplants in her first "Piecing Life Together" column. In their "Pearls of Wisdom" column, **Dr. Xan Nowakowski** shares their perspective in watching their dad undergo a liver transplant and how they were able to take part in his transplant journey. **Beth Sufian**, in her "Ask The Attorney" column, answers more questions about social security benefit overpayments in Part II of the series. For our "Pet's Perspective" column this issue I, on behalf of Husker, rebut Axil's assertions that the hospital dog policies were revoked due to Husker's behavior. **Maggie Williamson** shares her recipe for butternut squash, carrot and ginger soup in her "Culinary Corner" column this issue. **Aimee Lecointre**, our research guru, recaps exciting new CF research in her "Research Roundup" column. In the third installment of "Chaptered Lives," **Andrew Corcoran** writes about his sister's lung transplant nearly a decade after his own. **Katie Lockwood** talks about the challenges of being part of the sandwich generation in her "Salty Parenting" column. For our "Transplant Talk" column, **Colleen Adamson** revisits her squamous cell skin cancer in her thumb and the subsequent Mohs surgery to address the recurrence.

We're seeking nominations for both the Jacoby Angels Award and the Founders Award. You can learn more about prior winners, how to nominate, and the deadline for nominations on p. 40 of this issue. We'd like to say congratulations to the first recipients of the newly established Coon scholarship! Additionally, USACFA now offers four scholarships! You can read about our newest Stenzel scholarship on p.41. You can head to our website where you'll also find the application, requirements, and deadline for all four. As always, we're looking for new writers, new columnists, and people to interview for our "In The Spotlight" column. We love hearing from our readers! Reach out to us if you're interested in sharing your story in an upcoming issue. You can also find a list of focus topics both on our website and on p.3 of this issue.

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

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

Research Roundup

Compiled by Aimee Lecointre

A Systematic Review Of The Clinical Impact Of Small Colony Variants In Patients With Cystic Fibrosis.

A literature search was conducted in April 2020 to identify articles of interest. Data pertaining to demographic characteristics of participants, diagnostic criteria of SCVs, SCV prevalence and impact on lung function were extracted from included studies for analysis. Twenty-five of 673 studies were included in the systematic review. Individuals infected with SCVs of *Staphylococcus aureus (S. aureus)* were more likely to have had prior use of the broad-spectrum antibiotic trimethoprim sulfamethoxazole ($p < 0.001$), and the prevalence of SCVs in patients infected with *S. aureus* was estimated to be 19.3%. Additionally, patients infected with SCVs of Gram-negative and Gram-positive pathogens were identified to have a lower forced expiratory volume in one second percentage predicted than those infected by NCVs. The findings of this systematic review demonstrate that SCVs of *S. aureus* have a high prevalence in the CF community, and that the occurrence of SCVs in Gram-positive and Gram-negative pathogens is linked to poorer respiratory function. Further investigation is necessary to determine the effect of infection by SCVs on the CF population.
http://tinyurl.com/f3bmhw82

AIMEE LECOINTRE

Food Insecurity Screening And Local Food Access: Contributions To Nutritional Outcomes Among

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LOOKING AHEAD

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

Winter (February) 2024: Organ Transplants (Current issue)

Spring (May) 2024: CF and Cancer. Have you been diagnosed with cancer, either pre-transplant or after? If so, what kind? Has CF made it more complicated or challenging to treat? Were there chemotherapy or radiation treatments? Have you had recurrent skin cancers either pre- or post-transplant? What coping strategies have you used to process being diagnosed with cancer in addition to having CF? Were you angry, resigned, or sad and how do you deal with those emotions? What advice would you give others who are fighting cancer? **(Deadline March 15, 2024)**

Summer (August) 2024: CF and Travel. What tips can you share about traveling with CF? How do you manage all the medications and machines while traveling? What packing tricks do you have to make sure you remember everything and that your trip goes as smoothly as possible? What do you do differently for international travel? How do you prepare for a potential CF emergency while traveling? What lessons have you learned the hard way as far as travel? Are there certain places you cannot go or don't feel comfortable visiting because of your CF? **(Deadline June 15, 2024)**

Autumn (November) 2024: CF and Dating. (Deadline September 15, 2024)

ASK THE ATTORNEY

Social Security Benefit Overpayments: Part II

By Beth Sufian, J.D.

In the last issue of *CF Roundtable*, this column discussed Social Security Administration (SSA) overpayments and some common causes of overpayments. In this issue, the column will provide information about what happens after an overpayment occurs. Nothing in this column is meant to be legal advice and is only meant to be legal information. SSA rules and regulations can change, and the following column provides information related to SSA rules in effect as of December 2023.

If you have questions, you can contact the CF Legal Information Hotline at CFLegal@sufianpassamano.com. The CFLIH is sponsored by the CF Foundation. Employees of the CFLIH are not employees of the CF Foundation. All contacts to the CFLIH are confidential and there is no cost to the caller. The CFLIH can also provide information related to Social Security benefit eligibility, applications, and Continuing Disability Reviews, as well as education and employment rights and access to health insurance issues.

I. Notice of Overpayment.

An overpayment is simply a payment from Social Security to a beneficiary over the amount the beneficiary is due to receive. When an overpayment occurs, the Social Security Administration (SSA) will calculate the amount overpaid and will send a notice of overpayment to the beneficiary or to the beneficiary's representative payee. The notice of overpayment ordinarily includes:

1. The amount of the overpayment;

2. The reason for the overpayment; and

3. Repayment options.

Usually, the notice of overpayment will include information on how to appeal an overpayment or request a waiver of overpayment. However, some notices of overpayment make a demand for repayment without instructions on how to appeal or request a waiver. The description of how the overpayment was calculated is important to understanding whether the amount of an overpayment claimed can be successfully challenged by an appeal or a waiver.

II. Dealing with an Overpayment.

Generally, there are three ways to deal with a notice of overpayment: (1) an appeal; (2) a request for waiver; or (3) repayment.

BETH SUFIAN

A. Appealing an Overpayment.

Appeals must be filed timely within the time limit set out in the SSA Notice of Overpayment. The time limit on filing an appeal is one of the most important aspects of any appeal. Generally, a beneficiary (or their representative payee) has only 60 days from the date the notice of overpayment is received to file an appeal. The SSA assumes the addressee receives a notice within five days after the date stated on the notice. If the last day to appeal is a Saturday, Sunday, or national holiday, Social Security usually accepts an appeal on the next regular business day. If the beneficiary does not file their appeal within the 60-daytime limit, the right to appeal may be lost and the decision made by Social Security becomes final.

The consequence of a final decision is that the beneficiary: (1) can no longer challenge the reasons for the overpayment; (2) can no longer challenge the amount of the overpayment; and (3) may lose the opportunity to have a hearing by the administrative law judge and the SSA appeals council. Occasionally, Social Security will grant additional time to file an appeal if the beneficiary can show good cause for not filing an appeal within the 60-daytime limit. A request for more time must be made to Social Security in writing, stating the reason an appeal was not timely filed; however, a request for additional time must show a good cause. Social Security may deny a request for additional time if SSA determines the request fails to state a good reason for the delay in filing the appeal.

A beneficiary who disagrees with the reasons or the amount stated in the notice of overpayment may appeal the

determination of overpayment. Social Security also refers to an appeal as a request for reconsideration. Appeals must be filed within the required time limits and must be in writing. There are several ways to file an appeal of an overpayment. An appeal can be filed in person at a local SSA office provided it is in writing and timely filed. The SSA has a form to make a written appeal. The SSA may say the form is not required but it is best to use the form. The forms are available online. An appeal may also be requested online at www.ssa.gov/benefits/disability/appeal.html. Generally, in an appeal, Social Security needs information that shows that the reason stated in its notice of overpayment is incorrect or that the calculation made to determine the amount of overpayment is incorrect.

"A beneficiary who disagrees with the reasons or the amount stated in the notice of overpayment may appeal the determination of overpayment."

In a recent case, some unknown person improperly reported wages using a person's Social Security number. Looking at the reported income, Social Security assumed the beneficiary had returned to work and issued a notice of overpayment based on the improperly reported wages. In the appeal, the beneficiary showed that he had not actually returned to work and that the reported wages were not actually paid to him. This is an example of an appeal that successfully showed that the reasons SSA stated for the overpayment were incorrect.

In another case, SSA claimed that a beneficiary's earnings were just over the allowable amount of earnings. On appeal, the beneficiary showed that he had work-related expenses that Social Security failed to deduct from its earnings calculation. When SSA considered the work-related expenses, its recalculation showed that the beneficiary had not exceeded the earning limits and no overpayment was owed. SSA will stop recovering the overpayment from the beneficiaries SSI or SSDI payment while the appeal decision is pending. However, sometimes Social Security does try to have the SSA recipient make a payment plan to pay small amounts each month toward reducing the overpayment.

B. Requesting a Waiver of Overpayment.

Requesting a waiver of the overpayment is another way to avoid paying the amount of overpayment claimed by Social Security, which is separate from an appeal. A waiver is not a direct challenge to whether the overpayment is correct. Rather, a waiver is a request that Social Security not collect an overpayment that is determined to be due.

In a request for a waiver of an overpayment, the beneficiary asserts that (even though the amount of overpayment is correct), he/she was not at fault for causing the overpayment and the collection of the overpayment would cause financial hardship on him/her. Generally, there is no time limit for filing a request for a waiver of an overpayment.

A beneficiary may submit a waiver form (SSA-632-BK) to the SSA. If the amount of the overpayment is $1000 or less, SSA may grant the waiver request after an in-person visit to the local SSA office or a telephone interview with the beneficiary, without requiring the beneficiary to file a completed request for a waiver form. However, it is best to submit a written request for a waiver so there is proof a person has requested a waiver. A waiver is usually only granted upon showing evidence of financial hardship and that the overpayment was not due to any fault of the beneficiary.

Hardship usually requires evidence of genuine hardship rather than an inconvenience. For example, evidence of the absence of financial resources or even the risk of homelessness if the overpayment were collected may be good evidence of financial hardship.

C. Repayment.

If an appeal or request for waiver of an overpayment is unsuccessful, the beneficiary will have to repay the overpayment. A person can request a payment plan and Social Security will determine how much a person can pay each month toward paying back the overpayment. In most cases, Social Security will come to an agreement of a reasonable amount for the person to pay back each month, based on the person's monthly income form benefits or work earnings, even if it will take a long time to pay back the overpayment.

1. Repayment from Current Benefit Payments.

When a beneficiary is continuing to receive benefit payments, Social Security will typically collect an overpayment by withholding all or part of a current benefit payment due until the whole overpayment is collected. Withholding all of a current monthly benefit payment to recover an overpayment can cause a financial hardship for many people, especially if the overpayment is large and would take several years to repay. However, Social Security is usually willing to arrange a payment plan to recover an overpayment. The actual amount of withholding under a repayment plan depends on the individual case.

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SALTY PARENTING

The Sandwich Generation —A Salty Club Sandwich

By Katherine Lockwood

Many people with cystic fibrosis are living longer and having children. We're entering a new phase of life with challenges arising that we just didn't expect. In case you are unfamiliar with the term sandwich generation, it represents those who are caregivers for both aging relatives and children at the same time. For those of us lucky enough to have had loving and supportive parents who are still with us, we may need to consider that our aging relatives may need our help in the future.

Statistically, when a child is born with a disability, the parents' likelihood of divorce is dramatically increased and, 40 percent of the time, one parent either quits their job or reduces their hours to become the full-time caregiver, which is usually the mama[1]. These two factors can have devastating effects financially and, while it is common in many households for one parent's career to take a backseat while raising young children, that is generally only temporary. When a child is disabled, it is more likely to be a permanent shift.

When you have children there are many costs, and raising children with disabilities is especially expensive and time consuming. Our community faces unique characteristics that may exacerbate this. As both a person with CF and a mother to children with their own medical needs and disabilities, I see firsthand the five to ten plus hours per week that I spend scheduling and attending appointments for my wonderful babies on top of my own care, and the five plus hours a week I spend arguing on the phone with insurance/pharmacy/doctors on repeat. Adding in my needs as an adult who has CF turns a regular sandwich of care into a club with three pieces of bread.

As someone who assumed I would not outlive my loved ones, this shift feels a bit unsettling. Planning for long-term care, accompanying my parents to their doctor appointments, advocating for their needs, assisting with errands, and supporting them through their aging process, while also raising my children and caring for myself, is an adventure that I am both nervous and so honored to be a part of. I have had the privilege of learning how to do so from the very best—the parents of a CF patient. ▲

KATHERINE LOCKWOOD

Katherine Lockwood is 35 years old and has CF. She lives on Cape Cod with her husband Arden and her girls, Rose and Magnolia. She is a therapist for Verge Therapy and focuses on supporting individuals and couples experiencing disability in the family. She is the author of Why Me, Mama?, *an award-winning children's book about the disability experience. She is currently working on two picture book projects: Salt & Roses–supporting families when a parent has cystic fibrosis–and* OUCH! OOPS! & OH NO!, *a set of three books to support pre-k to third graders in reducing bullying. You can follow Katie's picture book projects at acorncottagepress.com and on Instagram @acorn_cottage_press.*

[1]Uribe-Morales BM, Cantero-Garlito PA, Cipriano-Crespo C. Fathers in the Care of Children with Disabilities: An Exploratory Qualitative Study. Healthcare (Basel). 2021 Dec 22;10(1):14. doi: 10.3390/healthcare10010014. PMID: 35052178; PMCID: PMC8775232.

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Children And Adults With Cystic Fibrosis In The United States

As the nutritional status of people with CF (PwCF) is associated with their socioeconomic status, it is important to understand factors related to food security and food access that play a role in the nutritional outcomes of this population. The contributions of CF program-level food insecurity screening practices and area-level food access for nutritional outcomes among PwCF were assessed. A cross-sectional analysis of 2019 data from the U.S. CF Patient Registry (CFFPR), linked to survey data on CF program-level food insecurity screening and 2019 patient zip code-level food access were conducted. Pediatric and

SUFIAN continued from page 5

Withholding 10% of the monthly benefit is usually agreeable to Social Security. However, even 10% of the monthly benefit can cause a financial hardship—especially for SSI beneficiaries. If a withholding of 10% of the monthly benefit causes a financial hardship, the beneficiary can request a repayment plan that is reasonable under the circumstances, but the monthly repayment plan withholding cannot be less than $10 per month.

2. Repayment When No Longer Receiving Current Benefit Payments.

The SSA will attempt to recover an overpayment even if the beneficiary is no longer receiving a monthly benefit payment. In the absence of a repayment plan, Social Security can seek to recover the whole amount of the overpayment by other mechanisms, including attaching IRS tax refunds and garnishing wages.

D. Ways to Avoid Overpayments.

1. Reporting Changes in Income.

One way to avoid overpayments is promptly reporting changes in income to the SSA. A beneficiary—either an SSI and SSDI beneficiary—should inform SSA when they begin work or return to work.

SSDI beneficiaries can report their work earnings to their local SSA field office by phone, or by presenting copies of their pay stubs to their local SSA office. SSA also has an online system for SSDI beneficiaries reporting monthly earnings called MyWageReport. SSI beneficiaries can also mail or bring copies of their pay stubs to their local SSA field office during the first six days of the month. The Social Security Administration has separate automated wage reporting tools for SSI beneficiaries, such as:

a. SSI Telephone Wage Reporting (SSITWR) System;

b. SSI Mobile Wage Reporting (SSIMWR) application, which allows a beneficiary to report the total gross monthly wages for the prior month using an Apple or Android mobile device; and

c. MyWageReport, which allows beneficiaries to report their monthly earnings online with their online Social Security account. Regardless of which method an SSI or SSDI beneficiary chooses to report wages, the beneficiary can sign up online to receive monthly e-mail or text message wage-reporting reminders.

2. Reporting Changes in Living Situation or Marital Status.

SSI beneficiaries can also avoid overpayments by reporting changes in living situation or marital status as soon as they occur. Changes to living situation or marital status can affect the SSI payment amount due to a beneficiary, but do not affect SSDI payments. ▲

Beth Sufian is 58 years old and has CF. She is an attorney who focuses her law practice on disability law and is the Vice President of USACFA. Her contact information is on page 2. You may contact her with your legal questions about CF-related issues at CFLegal@sufianpassamano.com.

adult populations were analyzed separately. Analyses were adjusted for patient sociodemographic and clinical characteristics. The study population included 11,971 pediatric and 14,817 adult PwCF. It was found that food insecurity screening and local food access are independent predictors of nutritional status among PwCF. More frequent screening is associated with less underweight among children with CF, whereas screening in writing (vs verbally) is associated with higher BMI among adults. Limited food access is associated with higher odds of being underweight in both children and adults with CF, and additionally with lower BMI among children with CF. Study results highlight the need for standardized, evidence-based food insecurity screening across CF care programs and for equitable food access to optimize the nutritional outcomes of PwCF.
http://tinyurl.com/37k6efun

Cystic Fibrosis-Related Mortality In The United States From 1999 To 2020: An Observational Analysis Of Time Trends And Disparities.

The national CF-related mortality rates have declined, and the median age of death among CF decedents has significantly increased, indicating improved survival. These changes were relatively slow during an earlier period of the study, followed by a rapid decline more recently. Patterns of disparities based on sex, ethnicity, race, and geographical factors were observed. These disparities were associated with a widening gap between ethnicities, a narrowing gap between races and rural vs. urban counties, and a complete closer of the gap between sexes over the study period.
http://tinyurl.com/6djuey48

Inflammatory Activity of Epithelial Stem Cell Variants From Cystic Fibrosis Lungs Is Not Resolved By CFTR Modulators.

CFTR (cystic fibrosis transmembrane conductance regulator) modulator drugs restore function to mutant channels in patients with cystic fibrosis (CF) and lead to improvements in body mass index and lung function. Although it is anticipated that early childhood treatment with CFTR modulators will significantly delay or even prevent the onset of advanced lung disease, lung neutrophils and inflammatory cytokines remain high in patients with CF with established lung disease despite modulator therapy, underscoring the need to identify and ultimately target

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PEARLS OF WISDOM

Three Months Apart: Reflections On My Father's Liver Transplant

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

My dad has much cooler surgical scars than I do. He's very proud of this! One of the first photos I saw of him after his liver transplant in March 2019 was him grinning and brandishing an enormous stapled incision on his abdomen that looked roughly like an inverted letter Y. His transplant surgeon at the Mayo Clinic in Jacksonville called it a "chevron" shape. My mom and I disagreed on the semantics—but it definitely looks impressive, regardless of the name.

Nobody had it on their 2019 bingo card that my dad would be getting a major organ transplant. At least not before 2018, when we learned completely out of the blue that he had end-stage cirrhosis of the liver. Not the kind of thing one expects just to sneak up. Especially not for a family well accustomed to progressive disease and the catastrophic outcomes of things that show warning signs for years. My parents paid thousands of dollars out of pocket to have my gums reconstructed so that I wouldn't lose my lower jawbone to infection with the same bacteria that destroyed all the tissue around my teeth. Or what's left of them, anyway. It's been thousands of dollars out of pocket for those too, between my parents and me and whatever dental insurance deigns to cover.

You might say being rebuilt because the technology exists is a way of life in my family.

The business about my dad's liver came as a surprise, though. I distinctly remember talking in mid-September with my mom, who'd just been liberated from a sling on her arm after extensive repair of a snapped bone just beneath the shoulder, about something not seeming quite right with my dad's behavior. He just seemed like he wasn't himself. He'd been helping my mom with basic tasks over the summer after her injury; even after she got the bone repaired with a huge metal plate, it took time and physical therapy to regain the range of motion in the rotator cuff that operates the shoulder. But we didn't think that would make my dad seem depressed and withdrawn, or confused about what was happening around him.

About a week later, my mom sent a message telling me she was at the emergency room with my dad. He'd suddenly started slurring his words and swerving on the road while driving them to campus from their house. With every bit of regained motion in her right rotator cuff, my mom leaned over and grabbed the wheel, pulling the car off the road and yanking the emergency brake with her free hand. She managed to get them to the hospital and call a neurologist colleague of ours for a consult right away.

> *"What I was prepared for was the opportunity to help my father open his mind about transplant."*

XAN NOWAKOWSKI

When you've been raised around hammers, things predictably look like nails on first sight. She and I were sure this was some kind of neurological issue, likely Parkinson's disease or something similar that would explain everything we'd observed previously, combined with the sudden decline in speech and motor function. So when a diagnosis of hepatic encephalopathy came back, we were stunned—and terrified.

The liver that came out of my father's body a few months later didn't look much better than a lot of those photos we see in our community of people's original lungs after transplant. He managed to get a perfect match liver just in time to save his life.

I wasn't prepared for almost losing my father. I'm still not prepared for it, frankly. It's been over five years since all this started and I still feel shattered by what happened to my family that year. I wound up so severely anemic from all the stress and its impact on my GI functioning that I turned blue and

experienced warping and infections in the nail beds on both hands. I've got some new scars, too. But they're not as cool as my dad's.

What I *was* prepared for was the opportunity to help my father open his mind about transplant. Initially, he wouldn't consider it. Which I get, completely. I'm not keen on the idea, either. I wasn't keen on it when my lungs were going badly downhill in my 20s. I haven't been keen on it since being diagnosed with chronic kidney disease in my 30s. But if things come to that, I'll consider it along with whatever other options are available. There's no honor in dying prematurely. And there's no grace in saving an organ for "someone who deserves it" or "someone who wouldn't be a waste," either.

I know how it feels to think that way. I've been there. We tell a lot of jokes in the CF community, to ourselves and to others, about how hostile our bodies are to any sort of life. It becomes almost a point of pride. Certainly we've earned that. It also masks tremendous pain beneath.

It's hard, even now, not to think of myself as useless and a waste of resources. I've gotten that message beaten into me all the more these past few years with COVID-19, a pandemic many people think is over and now irrelevant. In some ways life has become easier for me. At least I can wear a KN95 in the grocery store without people treating me like I shouldn't be at the store in the first place. But I've also watched the world leave me and many others behind, and had to remind people constantly that CF doesn't ever allow respite from constant vigilance against infection. Not if we want to live. Not if we want to thrive.

Many years ago, when I was 23 and he still had a functioning liver, my dad saved my life. I was in the cardiac intensive care unit at a local hospital in New Jersey, hooked up to several machines and entering the beginning stages of multisystem organ failure. My potassium levels had crashed so low that from a medical standpoint I shouldn't have been alive at all, let alone conscious or lucid. But there I was, stubbornly persisting in life. My parents were halfway around the world when I was admitted to the hospital, at one neuroscience conference in Germany and about to head to Hungary for another. They dropped their poster on the floor of the exhibit hall and raced to the airport instead. My mom was so badly in shock that she had trouble speaking. So my dad handled most of the communication—with me, and with the ticket agents at the airline counter to get himself and my mom back into the country.

My dad told me to hang on, that they were coming. I held on with everything I had. I told myself just to focus on getting through one day and the next until I could see them again. My potassium levels wouldn't budge until I heard his and my mom's voices on the phone. And my parents didn't rest until they were at my side. I got out of the hospital and went to public health school so that I could make a difference for other people who've been in this kind of situation, dying from a poorly managed progressive disease they've had since birth despite what passes for good access to care in the US.

I never expected to find my dad in a similar position. But we already knew some kind of mysterious vascular and metabolic condition ran in his family. Most of his male relatives died young. He'd already had one close call with stroke risk in his early 50s, but was able to get treatment not available to his ancestors. None of us knew something more sinister was brewing. The official diagnosis from Mayo Clinic after his transplant—and examination of his original liver by 16 different pathologists—didn't help much. *Idiopathic non-alcoholic cirrhosis*. That's what they tell people who don't have alcoholism, HIV, or cancer when their liver kills itself.

If my dad had a sense of fatalism, he certainly came by it honestly. I understood, on both an intellectual level and an emotional one. I also couldn't stand by and listen to him say his life wasn't worth saving. I'd been down that road before when, after my 2007 intensive care stay and my 2017 diagnosis of CF-related chronic kidney disease, it looked like I might need a kidney transplant sooner rather than later. So far I've still got those original parts—and the lessons that period in my life taught me about my value in the world as a chronically ill person; lessons I was able to discuss meaningfully with my father toward the end of 2018 as he cycled in and out of emergency care before beginning treatment at Mayo.

Loving parents will do almost anything for their children. I played that card without a trace of shame. I implored my dad to at least allow himself to *consider* a transplant—to get information about the process, to talk to people with donated livers, and to make a genuine decision based on all those factors and his own preferences. If he'd chosen not to pursue transplant, I would have supported him completely. What I couldn't abide was him dismissing the idea that he could *deserve* a transplant.

Life isn't about what we deserve, and neither is organ donation. My father, a difficult match for liver transplant due to his not having the CMV pathogen that lives in most people's bodies, managed to get an organ perfectly suited for him after only about six weeks on the transplant list. He'd gotten his consult with Mayo that January, with the first words out of his mouth to the care team being, "My

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child has CF and virulent bacteria in their respiratory system. If getting a transplant means I'll never be able to see them again, I'm not doing this."

The doctors and nurses at Mayo Clinic found this hilarious. They told my dad to sit down and that they'd make a targeted plan for protective antibiotic therapy during the time that he would still be heavily immunosuppressed after an initial period of no direct contact. So he got listed and then massively surprised when a call came through a few weeks later telling him that Mayo had a liver for him. His odds of getting a CMV-negative liver were infinitesimally low. That opportunity was only going to come around once—and that liver would not have been an ideal match for anyone who wasn't CMV-negative themselves.

At 1:43 a.m., the new liver was fully connected and immediately began making bile. And when he woke up from sedation, my dad was his old self again. Just like that. Foods tasted as they used to; his cheerful disposition and robust sense of humor returned. He'd been very well managed by Mayo prior to transplant with a special diet and medications that helped him excrete the excess ammonia that would otherwise have built up in his brain. He was even well enough to swim regularly again—his favorite hobby and something that had been out of reach before beginning care with his transplant team. He was actually in the pool when he got the call about Mayo having a liver for him! They had to call twice because he was underwater the first time and couldn't hear the ring.

But things were still rough. And it was very disorienting, especially for my mom, to have no idea what kinds of foods would even appeal to him. My dad has had very consistent taste preferences his entire life, all of which shifted dramatically when he developed end-stage liver disease. This would be like me not liking hot sauce anymore. I am a notorious consumer of hot sauce who is not above eating it directly from the bottle at times. I have an entire shelf full of the stuff in my refrigerator—nothing better for clearing the sinuses!

My dad got that new liver and became fully himself again. It was quite literally like plugging someone back into a wall socket after their battery got depleted. Neither my mom nor I had ever seen anything like it in several combined decades of experience teaching medical school. Me having CF and chronic bacterial colonization in my lungs and face meant I couldn't see my dad in person for a while. It took about three months for his doctors at Mayo to clear him to interact with me directly again. Time well spent in the utmost. When I was finally able to hug my dad, his incision had long since healed and he was back at work full-time.

> *"And there's no grace in saving an organ for 'someone who deserves it' or 'someone who wouldn't be a waste,' either."*

This marked the end of a long and horribly ironic period of never being able to see him—first because emergency room settings are horribly unsafe for people with CF, then because he was so immunocompromised from infected bile backing up into his abdominal cavity that I posed a danger to him at home, and finally because he was recovering from transplant and my respiratory bacteria could harm his own lungs and possibly contribute to organ rejection.

All of this made me reflect on what it really means to care for another person.

I couldn't be there physically at all; it was too dangerous for both of us to attempt that. My spouse, who's absolutely the teammate I would want in any catastrophic scenario, volunteered to help my mom with whatever she needed. This made me realize I didn't need to be present physically to make an impact in that sphere. J could be my hands while I took more of a leading role in social and emotional caregiving—for my dad, and also for my mom as his primary caregiver.

This wound up being a really powerful way to participate in his transplant journey. About three weeks before getting his new liver, my dad said he felt he could talk to me about his illness and the prospect of facing death in ways nobody else really understood. We were able to connect over that shared experience of reckoning with our own mortality. He said that helped him process the dramatic changes from the rapid destruction of his liver tissue and accept whatever outcome he would wind up getting.

Happily my dad wound up having a very easy recovery in their hotel room onsite. Mostly he was just bored, which he coped with by sending large quantities of unnecessary emails until my mom begged him to stop. He went home after the last drain came out and worked part-time for a couple of weeks before going back to full-time. Over four and a half years later, he remains one of Mayo's most successful liver transplant stories with no evidence of rejection whatsoever.

I consider myself a success story, too, for still being alive and distinctly kicking despite a lot of missteps and challenges in my care over the years. CF isn't an easy disease to live with. I'm lucky that with proper care my lungs function pretty well. My GI functioning is fairly bad at the best of times—but I still have all the original equipment!

And at least so far, my kidneys are still hanging in there.

It's all much easier to manage because I have such great support from my family. My parents and my spouse, as well as others close to us, have come together for me consistently over the years. Regrettably, I'm not a great surgical patient. I'm more like a tiny terrorist when heavily sedated. Controlling me physically is usually the bigger priority than supporting me emotionally when the going gets tough. I'm well accustomed to dealing with gnarly health challenges and have a very high pain tolerance. My spouse has perfected the strategy of rolling me up in blankets like a burrito to stop me doing all manner of unhinged things whenever I've needed additional surgeries. It's a bit like giving pills to cats—angry, gangly, sinister cats that know a lot of four-letter curse words.

My mom still remembers the time I alternated between cursing her out and floundering like a disoriented fish after some colon biopsies. This is one of the few times in my life where I've been absolved by amnesia—sorry, Mom. Thankfully my dad didn't attempt to destroy the world while still under anesthesia. But recovery from organ transplant is still serious business, one that requires support on many fronts. Being there for my dad throughout his journey has shown me how profoundly and impactfully people who are constantly sick ourselves can care for others.

Besides, who else is going to laugh at my father's dad jokes? As he likes to point out, nobody else ever seems to get them quite like I do. So I'm very glad that I'm still here. The care a parent receives from a child is precious and unique. And no matter my own body's foibles, or how much I might struggle with the insidious voice in the back of my head saying my parents got a raw deal, I remain the only one qualified for the job of giving that specific kind of support to my dad.

Doing that job has reframed my own thinking about my CF being a grave misfortune for my parents. I've sometimes been inclined to see myself as an expensive mistake and a fundamental source of suffering for how they've had to watch me struggle and cope with the very real possibility of me predeceasing them. But when death came calling at their own door, my disease began to feel like a superpower of sorts. Although it has almost taken my own life several times, CF let me make a difference in my dad's when he needed it most.

I'm okay with the rest if it means I still get to laugh at his jokes. ▲

Dr. Alexandra "Xan" Nowakowski is 40 years old and has CF. Xan is a director of CF Roundtable, *in addition to being a medical sociologist and public health program evaluator. They currently serve as an Associate Professor in the Geriatrics and Behavioral Sciences and Social Medicine departments at Florida State University College of Medicine. They also founded the Write Where It Hurts project (www.writewhereithurts.net) on scholarship engaging lessons from lived experience of illness and trauma with their spouse, Dr. J Sumerau. You can find their contact information on page 2.*

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the sources of this inflammation in CF lungs. Objectives of this study were determine whether CF lungs, like chronic obstructive pulmonary disease (COPD) lungs, harbor potentially pathogenic stem cell "variants" distinct from the normal p63/Krt5 lung stem cells devoted to alveolar fates, to identify specific variants that might contribute to the inflammatory state of CF lungs, and to assess the impact of CFTR genetic complementation or CFTR modulators on the inflammatory variants identified herein. The emergence of three proinflammatory stem cell variants in CF lungs may contribute to the persistence of lung inflammation in patients with CF with advanced disease undergoing CFTR modulator therapy.

http://tinyurl.com/y5zyazma

Changes In Fecal Elastase-1 Following Initiation Of CFTR Modulator Therapy In Pediatric Patients With Cystic Fibrosis.

Improvement in exocrine pancreatic function in persons with CF (pwCF) on cystic fibrosis transmembrane conductance regulator (CFTR) modulators has been documented in clinical trials using fecal pancreatic elastase-1 (FE-1). This group endeavored to evaluate real-world data on FE-1 in children on CFTR modulator therapy at three pediatric cystic fibrosis (CF) centers. 70 pwCF were included for analysis. 53 had baseline and post-modulator FE-1 values. Age was negatively correlated with change in FE-1. 15 pwCF had post-modulator FE-1 values ≥200 mcg/g, consistent with pancreatic sufficiency (PS). The PS group was significant for younger age at initiation of first CFTR modulator and a higher baseline FE-1. Most pwCF experienced an increase in FE-1 while receiving CFTR modulator treatment and a small percentage demonstrated values reflective of PS. These data suggest that PS may be attained in those that initiated modulator therapy at a younger age or had a higher baseline FE-1. FE-1 testing is suggested for children on any CFTR modulator therapy.

http://tinyurl.com/2mut72sd

Triple Therapy for Cystic Fibrosis

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ADVENTURES ABOUND

The Romance Of Running

NEW COLUMN

Well, I believe beauty is the physical reality that functions as the sparkplug to set fire to our emotional and spiritual being.

By Marcus Miller

In the town of Dover, Ohio, on the second day of July 1992, my twin sister Maria and I were welcomed into this world by our caring parents and one brother, Matthew. I was diagnosed with cystic fibrosis when I was two years old. Those first couple of years my parents noticed that something wasn't quite right as I wasn't gaining weight like my sister and always seemed to have stomach and bowel issues. They tried feeding me goat's milk and making other dietary changes; however, nothing seemed to make a significant difference. Finally, heeding the advice of an aunt who also has a son with CF, my parents scheduled an appointment for me at the CF center in Akron, Ohio, with Dr. Robert Stone. My diagnosis was confirmed with a positive sweat test, while a negative test came back for both of my siblings. After a short stay in Akron, I was sent home with the proper medications and care instructions to embrace this newfound reality. Of course, I didn't know anything had changed but undoubtedly there was much uncertainty about the future flooding my parents' minds.

While I've had my share of ups and downs with my health, I've been fortunate enough to have a semi-slow decline in lung function. I was also able to start on Trikafta soon after its release. While the "miracle drug" has brought about its own host of problems, from mental health challenges to insurance nightmares, I'm still grateful every day for my physical health and overall quality of life.

As an adult I've become very passionate about health and fitness. I also love exploring, hiking in the mountains and spending time in the wilderness or desert far away from civilization. Much of my writings here will continue to focus on stories and anecdotes from those adventures.

> *"I learned none of it gets easier; your goals just become bigger and what was difficult yesterday becomes the standard today."*

MARCUS MILLER

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I'm writing this story from 34,000 feet on a flight from Wilmington, North Carolina, to Sarasota, Florida, to celebrate Thanksgiving with my family. I'm excited for several reasons. Some much-needed time off work, lots of delicious food and, most of all, precious time with family members. As I grow older, I appreciate this day more and more. Sometimes I find myself being discontented in life, but this time of year helps me ground myself and really appreciate my life and all the good things in it. As I reflect on this year, I realize it's truly been a year full of adventure and wonderment for me, and I'm so extremely grateful. There are so many stories to tell, but, in this moment, what stands out in my mind has been my continued running journey and the myriad lessons I've learned along the way.

The journey really started in the latter part of 2022 after running my first half marathon. I sure wasn't breaking any land speed records that day; however, I was successful in completing the race and running the entire distance. I wondered where was my limit? How far could I actually push myself? I thought, "Maybe if I just kept training and got lucky, I could get to the point where I could run a marathon." Almost immediately, though, negative thoughts would always fill my head and remind me how painful it was to run 13.1 miles and that I'd be stupid to think I could run twice that distance. So I would slowly talk myself out of it and kick the can down the road with a simple, "Well maybe sometime later." Fast forward a couple months to January 2023, and I

found myself in south Texas traveling for work. My mind wandered as I drove along the desolate highways and my ambitions of running kept nagging at my brain. I still don't know if that lonely Texas desert was getting to me or if my stubbornness finally won out the battle that raged in my head, but I made a snap decision. "Forget doing a marathon," I exclaimed audibly, "I'm going to run an ultramarathon." I immediately called up both of my running partners and explained to them how this was a terrific plan, and they should join me as well. At first, they both questioned my sanity and well-being, but, after some coaxing and convincing on my part, both accepted the challenge. The kicker was the race I picked out was only a short eight weeks away. Little did I know that single decision would set in motion a trajectory of growth and discovery that would enrich my life beyond my wildest dreams.

Now, of course, running ultramarathons is no cake walk for anyone, but throw a chronic lung disease like cystic fibrosis on top of that, and it is sure to add to the complication and technicality of it all. Diminished lung capacity means greater preparation for the race is needed. So, I trained and trained, spending countless hours running and lifting and stretching my tired muscles and joints. I dialed in my diet to ensure my body was receiving all the proper nutrition to function as effectively as possible. I used hypertonic saline in my nebulizer several times a day to keep my lungs as clear of mucus as possible. Those eight weeks flew by, and the end of March was upon me quickly. Race day came and went and although there were some challenges along the way, I was quite grateful to have successfully completed my first ultra. You can read the full story of that first race in the Summer 2023 publication of the *CF Roundtable*.

After that first race and a taste of success, I developed some faith in my ability and wanted more: a bigger challenge, another goal to crush, a new seemingly impossible obstacle to overcome. So, I chose a 50-mile run in beautiful Moab, Utah, in late October. I signed up, paid the entrance fee, and started making plans. Again, I trained and trained. Fortunately, I was doing a good bit of travel over the summer, so I had the opportunity to run some really amazing trails, including hiking and running the trails on Grays Peak in Colorado. To say it was mesmerizing is an understatement. The views from atop a 14,278-foot mountain are otherworldly and words fail to describe the euphoric feeling from being atop a peak like that. I got to run some desert trails in the middle of nowhere Wyoming, where the fear of rattlesnakes made for heightened senses and an extra spring in my gait. I ran on days with the most perfect of weather, days that were laden with sweltering heat and days in the torrential rain and darkness of night.

Race weekend in Utah arrived and, this time around, I was lucky enough to have my brother with me as my crew, chauffeur, cheerleader, and motivator. We spent several days in Colorado and Utah exploring and just taking in all the glory of the mountains. On the morning of the race, I woke up around 3:00 a.m. to fuel and get prepared for the day. I was excited but fidgety and anxious, not knowing for sure what lay ahead of me.

That anxiety quickly dissipated as I left the starting line in a sea of people. I focused solely on managing my pace and maneuvering the rocky terrain beneath my feet. I felt so utterly alive as the sun came up over the rim of the canyon and lit up the valley below. My imagination went wild thinking about how this landscape must have looked in the Jurassic period when dinosaurs and other now-extinct creatures roamed this land. Along the way, I had some wonderful conversations with other amazing athletes. The encouragement and support from others in the running community is truly genuine and such a rare thing to find.

Step by step, mile by mile, the day passed by so quickly and my body grew more and more fatigued the closer I got to that finish line. I crossed over the line with a small crowd cheering me on and my brother there to greet me with a hug. We both fought back tears as we began to process that I had just run 50 miles. Even as I write this mid-flight, I'm fighting back tears again. Thank you, Matt, for being there by my side that day. That memory will stay with me forever. I crossed the finish line with a total time of 12:07. Overall, I felt good—my body of course fatigued and hurting but all things considered, not too terribly. My lungs were my biggest concern. They had performed great all day but after the race it took about an hour to get my heavy breathing back down to a normal cadence. As we drove back to the hotel, my mind filled with celebration and anticipation of what the next goal will be. One hundred miles, perhaps?

While much of this journey and the milestones achieved can be romanticized, there were also many difficult things I've had to contend with along the way. The physical demands of course are monstrous; however, the emotional and mental battles were much more difficult to overcome. There are times of great loneliness in such a journey: the multitude of hours spent training alone coupled with battling with one's body and mind to continue on when every fiber in you wants to quit. I learned none of it gets easier; your goals just become bigger and what was difficult yesterday becomes the standard today. The way I figure, the personal growth and discovery far outweighed all the sacrifices made along the way.

One discovery that left the greatest impression on me was all the beauty I found along the way. There is

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PET'S PERSPECTIVE

Fiercely Protecting My CFer

By Husker (mostly) and Cutty (but not really), translated by Sydna Marshall

Frenz! I think I'm pretty famous already but, in case you haven't seen my handsome face on the cover of *Pibble Weekly* or in any of momstaff's socials, I'm Husker! Staff did one of those genetic tests and it turns out I'm part pitbull, part rottweiler, and part chow-chow. Dadstaff, aka beardstaff, likes to joke that I'm three parts a**hole but I'm really not. Ok, chow-chow gives that name a run for the money. I digress. Pitbull-loving owners have adopted the term pibble as a term of endearment to combat the negative stereotype that pitbulls are mean, aggressive dogs who are usually bred and raised for dog fighting rings. Pibbles love their people and, back in the day, they were often nicknamed "nanny dog" or "babysitter dog" because of their sweet temperament around children (when properly trained and socialized). Anywho, I'm the source of most of the shenanigans in the Marshall-Keys household and, honestly, I'm a big deal. I've been waiting eleventy-billion dog years to rebut Axil's assertions in a prior issue of *CF Roundtable*. My time has finally arrived!

I'll give you the skinny on how I came to be the *most* important part of this household. Staff rescued me from the animal shelter on the Ides of March, 2015. I secretly love that my gotcha day is the anniversary of the death of Julius Caesar. *chuckles* Well, momstaff says she picked me out online back in December 2014 and then fell in love with me at first sight. Beardstaff wanted to shop around for other pups but momstaff and I had already made the connection and she was not to be swayed (thankfully)! I had spent a year and a half at the shelter, nearly half my life. I came with an 18-page rap sheet according to staff but, honestly, if no one saw it did it even happen? Skip forward a zillion dog years and staff came home with Cutty, aka Cuts. Cuts was grandma's dog and we got to keep her when grandma passed away. Staff forgot to run this by me for approval but alas, Cuts is the best little sister in the world, mostly because she lets me boss her around and I've only growled at her once or twice, which she deserved. Staff says that's a stretch.

CLOCKWISE: MOMSTAFF (SYDNA MARSHALL) AND HUSKER SNUGGLE IN BED; SANTA POSES WITH BEARDSTAFF (ADAM KEYS), HUSKER, SYDNA, WITH CUTTY CLOWNING ABOUT; HUSKER AND CUTTY SIT AND CATCH THE RAYS.

We all know momstaff has CF. We've had a lot of adventures together. She used to walk me on the nature trails, but I'm so protective of her that it became cumbersome for her to walk me around other dogs. Did I mention that I don't like other dogs? I don't. Staff is mine and I don't share. Now we just walk in the neighborhood, uphill both ways, barefoot, in scorching heat and snowy mountains for millions of miles (ok, ok, just four miles). I had an uncanny knack for racing down the stairs during zoomies and leaping over

the end table directly onto her port while she lay on the couch. That takes serious skill! I still don't think I'm properly rewarded for nailing that maneuver countless times. I love laying toes up with belly rubs in the bed so that staff can tell me the story about my gotcha day, the day I was adopted! I also really love doing yoga on the mat with momstaff. You've probably all seen the videos on the book of faces (FB, you know) where the Australian Shepherd mirrors her mom and does acrobat yoga with her. I'm not that adept but I'm excellent at the Sphinx pose. Gotta start somewhere, amiright?

Remember how I said I'm fiercely protective of my people, namely momstaff? Many, many moons ago she was hospitalized for two weeks. They were the *longest*, *worst*, weeks of my life. It was sooooooooo boring but I did enjoy snoring on her pillows in the interim. During that time, I was allowed to go visit momstaff at Spa Seton (aka the hospital, which is decidedly *not* a spa). I got to snuggle with her on the tiny hospital bed with all the blankies and pillows. She had this weird tubing coming out of her arm (the aforementioned port-landing incidents hadn't transpired yet) and, after eavesdropping in the hallway, I learned it's a PICC line. So we were all comfy and cozy until Nurse Ratched came in abruptly and started messing with her tubing and the beepy machine. The nerve! All reasonable dog-loving people know that you have to get permission from staff and *moi* first. So, what does a pibble do? I barked, let out a low growl, and snapped to let Nurse Ratched know that she needs to back away from my person! This seems like Psychology 101 behavior here—not sure why I have to reiterate this all the time. Needless to say, she ran out of the room and returned with the charge nurse who permanently banned me, and all dogs, from Spa Seton. Le sigh. Thankfully, momstaff started Trikafta so the trips to the hospital have been much less frequent (dare I say, rare) and this is much less stressful for me. It's hard to protect her when I can't get to her!

I have read so many articles in this publication about all the trials and tribulations that CFers endure and it seems pretty obvious to me that the support from loved ones is crucial for their health and wellbeing. I take my job very seriously! I patrol my fence and secure the perimeter of the house multiple times a day. No one gets past me! If you want to get on my good side, bring all the treatz, especially the cheese and chimcken variety. No salad ingredients. Yuck. If you'd like to get routine updates of my shenanigans and photo essays for *Pibble Weekly*, you can find me on momstaff's IG: @sixty5roses. ▲

Husker is almost 13 years old and loves walkies, treatz, and naps. Cutty is almost 12 years old and has less refined taste, except when it comes to her pillows.

Sydna Marshall is 43 years old and has CF. She is the President and Managing Editor of USACFA. She lives in Austin, TX, with her husband Adam and their two furbabies, Husker and Cutty. She loves yoga, reading, cooking, and working on puzzles. Her contact information is on page 2.

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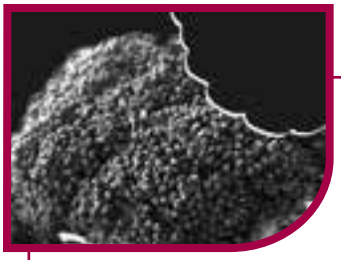
tremendous beauty in the world and we never fail to marvel at the ocean or a magnificent mountain range, but it seems in our daily busyness and increasingly stunted attention spans, we often miss the tiny details of life: The morning song of a Carolina wren, or the tiny yellow wildflowers growing on the side of the highway often go completely unnoticed. But in all my hours of running, I learned that beauty is a seashell with a living creature inside. It's the smell of the forest after a summer rain. It's a kind word from a stranger. It's the warmth we feel when the sun's rays touch our face on a cold winter day. Beauty is truly present all around us, but oftentimes, in order to see the beauty, we must stop, adjust our lens to a far less frequented perspective, and take it all in from a new angle.

So what is it about beauty exactly that makes such a lasting impression? I believe beauty is the physical reality that functions as the sparkplug to set fire to our emotional and spiritual being. From a psychological perspective, I find that perfect beauty is found in the balance between chaos and order. Excess chaos, of course, is rocky—it's uncertain and induces anxiety and vast amounts of stress. Excess order, on the other hand, is that place where we stop growing and tend to fall into complacency and resentment. But in that place where storm and calm meet, that's where we find our purpose. It's where we find fulfillment and true joy in life. As John Keats, the English poet said, "A thing of beauty is a joy forever...." ▲

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



CULINARY CORNER

Butternut Squash, Carrot And Ginger Soup

By Maggie Williamson

I'm in soup mode right now and this is one of my favorite soups to make in a pinch. Everything gets roasted and very minimal chopping is required. I even refuse to de-seed the squash before I roast because it is so much easier to just scoop out the seeds when the squash is fully roasted. However, you will need an immersion blender or blender for this one, but otherwise no fancy equipment needed. The ginger in this soup gives it a little heat and warmth to complement the sweet root vegetables. This is great for any meal and also a great soup for a dinner party starter. Grab some crusty bread or make a simple green salad and you have yourself a great meal! Enjoy!

MAGGIE WILLIAMSON

Butternut Squash, Carrot and Ginger Soup
Yield: 4 servings

Ingredients:
2 tbsp olive oil
1 medium-sized butternut squash
3-4 medium-sized carrots
1 onion
2 large garlic cloves
1-inch fresh ginger, minced
1 tsp cinnamon
½ tsp nutmeg
6 cups vegetable stock
Salt and pepper to taste

Optional Garnishes:
Cream
Olive oil
Crème fraîche
Bacon
Pepitas (pumpkin seeds)

Preparation:

Step 1:
Preheat the oven to 400 degrees. Cut the squash in half and drizzle with olive oil, salt, and pepper. Lay squash flesh side down onto foil-lined baking sheet.

Step 2:
Peel the carrots and cut into large chunks. Add them to the same baking sheet and drizzle with olive oil and a little salt and pepper.

Step 3:
Cut the onion into quarters; set aside with garlic cloves (keep skins on garlic cloves).

Step 4:
Roast the squash and carrots in the oven for 30 minutes. Add the onion and garlic to the baking sheet and continue baking for another 30 minutes or until the squash is fork tender.

Step 5:
Scoop the seeds out of butternut squash. Scoop the flesh of the butternut squash into a big pot with carrots, onion, and ginger. Squeeze the roasted garlic out of the skins and add to the pot. Add cinnamon and nutmeg and cook on medium for a minute or two. Add the vegetable stock and let it all come to a boil. Cook for 5 minutes. Take off the heat and blend with an immersion blender or stand-up blender until smooth. Check seasoning and add more salt and pepper if needed.

Step 6:
Serve immediately or let cool down and store in fridge up to 3 days or in freezer for up to 3 months. ▲

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

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LUNG TRANSPLANT GRANT PROGRAM

The BEF Lung Transplant Grant Program offers financial assistance to individuals with CF who are in need of an organ transplant.

The BEF IVF Grant Program is dedicated to ensuring that adults with CF, in their family building journey, have the financial resources to utilize assisted reproductive technology.

BEF
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The BEF scholarship program provides financial assistance to students in the CF community who are pursing higher education opportunities.

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The financial burden on families affected by CF can be significant. The BEF is committed to providing individuals and families with the assistance needed to achieve stability in times of need.

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FOCUS TOPIC

ORGAN TRANSPLANTS

Journey To Tomorrow

By Paul Albert

Living with CF and undergoing a lung transplant are journeys that are not for the faint of heart. If not for my loving family, friends, and incredible medical professionals, I would not be writing this article. In 1962, when I was diagnosed with CF, my life expectancy was nine years. I do not know what the life expectancy was after my lung transplant, but I learned those are just numbers and averages, not what will happen to me. I have a strong faith in God and a lot of people praying for me. I am very thankful for the way my life turned out.

Although the idea of a transplant was first mentioned to me in 1987, it was not until 1992 that I seriously considered it. That May, I experienced a serious lung infection and was faced with the prospect of being intubated to give my lungs a chance to heal. Fortunately, that was not necessary, but it was the start of continuous oxygen and using a BiPAP machine during sleep to improve my blood gasses. Dr. Stanley Fiel, my CF doctor, encouraged me to consider a transplant. At my request, I came home from the hospital with a list of nearby transplant centers. I made up a list of 20 questions and spoke to coordinators at the three closest transplant centers and decided on the University of Pittsburgh Medical Center because they were performing over 60 lung transplants yearly and were experimenting with a new anti-rejection drug, FK506. That summer I was evaluated at UPMC Presbyterian and accepted for their transplant list. What a relief that was! For me, deciding to pursue a transplant was more difficult than the wait. There were 111 people on UPMC's waiting list when I was accepted. At that time someone's place on the waiting list was determined by blood type, body size, and time on the waiting list. I was the only one with B-positive blood who could accept a donor between 100 and 200 pounds—I was at the top of my own little list. However, because B-positive blood is less common, they said it could be either a short wait or long one, depending on the possible donors. Five months of waiting included at least a dozen wrong-number calls to my pager. They always occurred around the lunch hour. My coordinator concluded that the pager number must have been similar to that of a pizza place in Pittsburgh. Eventually, I received a new pager.

PAUL ALBERT

Waiting for a transplant in 1992 was much different than it is today. I had the opportunity to speak with other lung recipients, but it was before the internet and there was little written information about lung transplants. I did speak to my cousin who was a kidney recipient and he said that taking the prescribed medication on time and keeping good notes of my transplant medical history were important. I took his advice on both of his recommendations and I would highly recommend other recipients do the same. By January 1993, my lung function was down to 16% as I continued to work full time as an internal auditor in county government. I needed the health benefits that the job afforded me and, just as important, I had a handful of people at work who were part of my support team. I think I would have gone nuts if I was at home thinking about a transplant all day.

By February, I was fairly comfortable with the idea of a transplant. I knew that there were no guarantees with a transplant, but I also knew what would happen if I did not receive one. On February 9, around 11:00 p.m., the phone rang during my evening chest percussion. It was my coordinator Dan. He asked me how I was feeling and

then said that they thought they found a pair of lungs for me. He asked how I felt about that and I said, "That's great; that's what I've been waiting for." At the same time, I was shaking—knowing that if it happened, my life would never be the same. The transplant could be successful, I could have a lot of health problems and poor quality of life, or I could die before coming home from the hospital. I knew I had to put my life in God's hands because it was out of my control. Dan called back at 11:30 p.m. to say that it was a go, that he had arranged for MedEscort, an air ambulance service, to fly my parents and me from Allentown to Pittsburgh on a small turbo prop plane. It was a clear, cold night, and it was also my first time on an airplane. No shortage of drama!

My transplant took about 10 hours and I was doing well post-op. By then my brother and sister had arrived. I was fortunate to be put on the anti-rejection drug FK506, now commonly known as Tacrolimus. On the third day I was off the vent and eating and the CTICU doctor told me that I received a perfect pair of lungs. By the fifth day I was able to walk to the family waiting room. My family and I were both really encouraged with my progress. I thought I should be out of the hospital in a few weeks. Well, that didn't go as planned. I then hoped for release by St. Patrick's Day, then Easter, and then Memorial Day, but those didn't happen, either.

I had many bouts of infection and rejection, including a sinus surgery to try and alleviate the main source of the infections. Toward the end of February my left lung collapsed and I went into cardiac arrest. After seven minutes of CPR and meds to restart my heart, my heart was beating on its own again. However, from that day on, it seemed like I received blood transfusions quite frequently. The doctors could not figure out why this was happening. I was intubated on and off for about six weeks. At one time I was intubated for 21 straight days and fought them when they wanted to trach me. Eventually I was able to be weaned off the ventilator. April came and the doctors were convinced that I had a bowel obstruction because my abdomen was hard and distended. Attempts to correct that did not work so I had emergency exploratory surgery on my abdomen on Easter. The surgeon found that it was not a bowel obstruction: my abdomen was filled with fresh and old blood. Apparently, my lower rib punctured my liver when they gave me CPR back in February and, as a result, I had a slow bleed for about seven weeks. After that was repaired, I felt better for about a week until I became septic. At that time, my parents were told to expect the worst. A couple weeks later I continued having trouble with rejection and was given a series of horse serum shots, which caused extreme pain and something called serum sickness. That only lasted a few days but come May, my kidneys were not doing well and I started dialysis. Finally, on June 1st, after 111 days, I was discharged from the hospital. I was still very weak, but I made it! It was such a strange feeling to be out of the hospital. I stayed on dialysis until mid-August. By then the doctors said that my kidneys had repaired themselves and I could discontinue dialysis.

> "I thought I should be out of the hospital in a few weeks. Well, that didn't go as planned."

I share all these details about my hospitalization to point out that some people have rough starts to their transplant journey but that does not mean that things will not improve or that it will be a short journey. Everyone's journey is different—just keep fighting.

In September, I went back to work part time and, by October, I was back to full-time work. I retired in May 2023 after 41 years as a lead auditor with a short stint as audit manager. The work was very rewarding, but it was all the people whom I worked with over the years that made work enjoyable. Not just people in my office but the many people whom I met doing audits in different offices in the county. I made a lot of friends and have a lot of good memories.

The quality of my life was completely different after the transplant. I started golfing again and that got me ready for my first US Transplant Games as a member of Team Philadelphia in Atlanta in 1994. It was just incredible to meet so many recipients and donor family members. On the golf course I met someone who visited me after my transplant and gave me a good pep talk. Mickey Hart from Team Buffalo, who also had CF, was transplanted two years before me at UPMC. We hit it off and became very close friends, along with his wife Sara. Mickey was a mentor to me and many other people. Mick passed away in 2016, a few months after the transplant games in Cleveland. It was a big loss for me. My journey would have been much more difficult and less enjoyable without Mick. I continue to attend as many games as I am able, missing only two since my first Games. Everyone should try it at least once, even if you are not athletic.

Through Team Philadelphia I became a volunteer and later a board member with the Gift of Life Donor

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Program in Philadelphia. It brought things full circle for me to be able to educate the public about the importance of organ and tissue donation in addition to helping people on the waiting list. It also provided a lot of interaction with other recipients and donor family members, something that meant a lot to me since I never heard back from my donor's family. I also volunteered with the CF Foundation and currently volunteer with a non-profit organization that supports our local National Guard and reserve units.

When I was inquiring about a transplant, I was told that I would be exchanging one set of health problems for another. And while that is true, it was a deal worth taking and one that I won. After release from the hospital, I had a couple bouts of acute rejection that were easily treated. I still have sinus issues and trouble with digestion, two common CF symptoms. Starting about 15 years post-transplant, I began having occurrences of skin cancer and that continues today. Stay on top of this! Protect your skin and see a good dermatologist regularly. Traces of blood in my urine in 2016 led doctors to find that I had cancer in the lining of my bladder. Treatments with a chemo drug directly into the bladder eliminated that. In 2017, when I was being evaluated for a kidney transplant, I had a reoccurrence. Use of a different chemo drug took care of it that time and it has not returned. However, my need for a kidney transplant remained and I received a deceased donor kidney on July 9, 2022. I was in the hospital for a week and although it was not easy, it was nowhere near as challenging as the lung transplant. Thank you to my donor's family!

I went into the lung transplant process not knowing what to expect. Thanks to many people, especially my organ donors and family, I still have a very good quality of life, but one that is not without its obstacles. I just take things day by day and enjoy the little things in life that much more. ▲

Paul Albert is 64 years old and has CF. He lives in N. Catasauqua, PA. He received his lung transplant 31 years ago on February 10, 1993, and his kidney transplant on July 9, 2022. His hobbies include golfing, walking, gardening, volunteering, and spending time with family and friends. You can email him at palbert5@ptd.net.

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(Elexacaftor, Tezacaftor, and Ivacaftor): Desensitization After Skin Rash.

ELX/TEZ/IVA (Trikafta) is the current gold standard of treatment for the majority of patients with CF, and health professionals must be aware of common side effects, and it is important to establish and make available desensitization protocols to manage these side effects. Several desensitization protocols can be used, and presented here is the one that worked successfully with this patient. As patients strongly benefit from treatment with ELX/TEZ/IVA and in the absence of effective alternatives, the reintroduction of the same drugs with the risk of rash recurrence is acceptable, given the benefits in the quality of life of patients with CF.
http://tinyurl.com/ycxmp38x

Identification Of Cystic Fibrosis Transmembrane Conductance Regulator Gene (CFTR) Variants: A Retrospective Study On The Western And Southern Regions Of Saudi Arabia.

To investigate the geographic distribution of common cystic fibrosis (CF) variants in the western and southern regions of Saudi Arabia a retrospective study was conducted on 69 patients diagnosed with CF at King Faisal Specialist Hospital & Research Center, Jeddah. Patient data were collected retrospectively between June 2000 and November 2021. Various parameters were considered, including patient demographic information, CFTR variants, and respiratory cultures. 26 CFTR variants in 69 patients with CF, including one novel variant that had not been reported or published before (1549del G) in 2 patients with CF were identified. This study highlighted features of patients with CF residing in the Western and Southern regions of Saudi Arabia. Six of the 26 CFTR variants were common in these patients. It was also reported, for the first time, a novel variant and other CFTR variants that are yet to be reported in Saudi Arabia. These findings could help establish a foundation for cystic fibrosis screening in Saudi Arabia and may assist in clinical diagnosis and prognosis.
http://tinyurl.com/yk9f2vdx

Effects Of Elexacaftor/Tezacaftor/Ivacaftor On Liver Fibrosis Markers In Adults With Cystic Fibrosis.

There are limited studies to date on the effects of elexacaftor/tezacaftor/ivacaftor (E/T/I) on markers of liver fibrosis in adults with cystic fibrosis (CF). This study aims to analyse changes in makers of liver fibrosis before and after initiation of E/T/I in CF adults. Outcome measures of liver fibrosis, including liver stiffness measurement (LSM) using FibroScan, AST-to-platelet-ratio index (APRI) and gamma-GT-to-platelet-ratio (GPR) were available in 74 CF adults following initiation of E/T/I. This was compared to historical data collected in 2018 prior to UK availability of E/T/I. Apart from APRI, no changes were found in liver fibrosis outcomes after initiation of E/T/I in adults with CF. Those with a historical

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Gratitude

I had to do it or I would die.
I might die anyway,
but I had to take the chance.

They said "Yes" without even being asked.

On May 24, 2000, my uncle Rickey
and my friend Jerry went into surgery,
and had part of their lung removed.

They were put into me.
I lived.
They are fine.

There are no words to describe...
feelings too deep to express.
All I can do is say "Thank You."

-M. Thompson Reta, 2002

PHOTO BY STEPHEN BOYER

FROM OUR FAMILY PHOTO ALBUM...

MARCUS MILLER AND HIS BROTHER, MATT, IN FRONT OF MAROON BELLS IN COLORADO, IN OCTOBER, 2023.

MATISON DEATON AND HER BROTHER NICK, WHO WAS HER CAREGIVER AFTER HER LUNG AND KIDNEY TRANSPLANTS. THEY WALKED ACROSS THE GOLDEN GATE BRIDGE AND BACK TWO MONTHS INTO HER RECOVERY.

SYDNA MARSHALL WITH HUSKER (LEFT) AND CUTTY (RIGHT).

PATRICK WELSH, HOLDING WINSTON, AND PIPER BEATTY WELSH, HOLDING NEW PUPPY "SNOOPY." SNOOPY GOT HIS NAME BECAUSE HE BEARS A CLOSE RESEMBLANCE TO HIS CARTOON COUNTERPART!

SCOTT AND COLLEEN ADAMSON, CHRISTMAS 2023.

PAUL ALBERT (RIGHT) WITH HIS SISTER, JANE AND BROTHER, CHUCK.

ARTWORK BY DOMINIC QUAGLIOZZI

DOMINIC QUAGLIOZZI

The butterflies series is an exploration into how my life is transformed by chronic illness, debility and rejuvenated by life-saving organ transplantation. I received a double lung transplant in 2015 and have been examining and rebuilding my identity ever since. These butterflies are made out of deconstructed hospital gowns, scrubs, moving blankets, and the scraps of failed paintings. I make gestured marks with colored pencils on the hospital gowns and sew together the materials—out of that grow the butterflies.

As a reflection of my transplant process and now life post-transplant, I am using hospital gowns and moving blankets because these two materials have so much emotion driven into them through our human experience, individually and collectively. Each stands as embodied past, present, and future—representations of our physical history, a moment of transition in collective physical and emotional labor, and the hope and mystery of a future unknown. ▲

Dominic Quagliozzi is a 41-year-old visual artist living in Massachusetts with his wife and four-year-old son. He received a double lung transplant in 2015 from Stanford Hospital.

BLUE MORPHO, 2023
COLORED PENCILS ON HOSPITAL GOWNS, PARTS OF PAINTINGS, MOVING BLANKETS, NITRILE GLOVES
38" X 26"

MONARCH, 2023
COLORED PENCILS ON HOSPITAL GOWNS, PARTS OF PAINTINGS, MOVING BLANKETS, HOSPITAL GRIPPY SOCKS, MYLAR BALLOONS
30" X 58"

Benefactors

BRONZE BENEFACTORS

(UNDER $250)

Paul Albert
Anonymous
Bridget Barnes (in honor of Lucy Larkin Barnes)
Robert & Robin Barrett
Benevity Fund
Douglas Booten
Nancy & Robert Coleman (in honor of their daughter, Claire Herriot)
Julie Desch
Tina Dimick
Michelle Duran
Nina Ferrell
Linda Fried
Doreen Gagnon (in memory of Sally Drury)
Kenneth & Judy Greenberg
Paula Grey
Barbara Harison
Philip & Cynthia Hays (in honor of Zach Hays)
Douglas Holsclaw, M.D.
Gertrude Huntington
Douglas Johnson
Lawrence Lafary
Philip & Johanna Libbert
Cathleen Marceau
Peggy Martinet (in memory of Kathy Russell and Cathy Martinet)
Robert & Donna McLaughlin
Laura Mentch (in memory of Isa Stenzel Byrnes)
Loretta Mulatz (in honor of her daughter, Debra Radler)
Paypal Giving Fund
Maureen Perlette
Tim Peters
Richard and Karen Quinn
Judith Riley
Reid Seilheimer Family Trust
Connie Smith
Michael & Dianna Smith (in memory of Michael Edward Darrar)
Laura Tillman
UK Online Giving Foundation
Robert Young

SILVER BENEFACTORS

($251-$499)

Scott & Colleen Adamson
John & Jeanie Hanley (in honor of Mary Ellen Sampson)
Karen Scott (in honor of Nicholas Scott)

GOLD BENEFACTORS

($500 -$999)

Susie Baldwin (in honor of Kathy Russell)
Terry Craft
Dick Marshall (in honor of his daughter, Sydna Marshall)

PLATINUM BENEFACTORS

($1,000 -$4,999)

James Yankaskas, M.D.
Barbara Nechis (in honor of her granddaughter, Ela Castillo)

SUSTAINING PARTNERS

($5,000-$9,999)

Elliot Family Giving Fund (in memory of Shirley Althaus)
Monaghan Medical Corporation
Watson W. Wise Foundation (in memory of Watson W. Wise)

PEARL SUSTAINING PARTNERS

($10,000-$19,999)

Boomer Esiason Foundation
Cystic Fibrosis Foundation
Helen M. Eisenman Fund Scholarship for the Arts

DIAMOND SUSTAINING PARTNERS

($20,000 AND UP)

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

DIAMOND ENDOWMENT PARTNER

($20,000 AND UP)

In Memory of Lauren Melissa Kelly

The Good, The Bad, And The Unexpectedly Beautiful: The Highs And Lows Of 13 Years With Transplanted Lungs

By Piper Beatty Welsh

When I was first listed for transplant in the fall of 2009, I remember my doctor talking me through a seemingly endless list of possible side effects and complications. My mother, who was also acting as my primary support person through the transplant process, sat next to me, a look of growing discomfort on her face as we progressed from "little" issues like the need for a lifetime of immunosuppression to bigger concerns like kidney failure, chronic rejection, and cancer. The rhythmic hum of my portable oxygen concentrator provided fitting background music as I jumped in to reassure both my doctor and my mother (and, quite honestly, myself) that everything would be okay. "I know the risks," I declared, with as much strength in my voice as my battered lungs would allow. "I know the side effects. But I also know that none of that is going to happen to me. My life with new lungs is going to be beautiful—just you watch!"

I didn't know at the time just how wrong I was. Or how right.

My first transplant was rocky from the start, with near-constant infections that kept me in and out of the hospital and often on home IV antibiotics. After living 28 years with CF lungs, none of this felt particularly new or scary to me, so even though other people I knew with transplants kept talking about how great it was to live a hospital-free life and not spend so much time at clinic, I wasn't really worried. My doctors and surgeons emphasized that every recovery looks different, and most of my issues were treatable. It wasn't until the end of

PIPER BEATTY WELSH

2012, just over two years out of my surgery, that things seemed to get more complicated. A routine surveillance bronchoscopy found acute rejection and, after the usual round of intense IV and oral steroids, it was still there, which was confirmed in a second procedure. My doctors admitted me to the hospital to receive rATG (an IV infusion specifically formulated to combat the immune cells that can cause transplant rejection). When that still didn't solve the problem, I started photopheresis, which is kind of like dialysis for the immune system. I would spend several hours attached to a machine designed to pull out my white blood cells, render them unable to attack my new lungs, and then place the treated blood back into my body. I would stay on this treatment for several months until continued decline in my lung function finally led us to try a one-time chemotherapy infusion that my doctor described as a "nuclear bomb" for my immune system. When the treatment was over, I was left extremely immunocompromised, but unfortunately my lung function still didn't stabilize. The only option left was a second double-lung transplant. I couldn't help feeling like I had failed my body, my family, and, most importantly, my donor. How could my own system destroy this beautiful gift? How could I be "one of those people" who needed another transplant so soon (just three years!) after my first?

As it turns out, I may never know the real why behind my fast and unexplained rejection. My doctors at a top transplant center certainly did all they could to save my lungs, and I never missed a dose of medication or a single doctor's appointment throughout the process. Sometimes, it seems, life just isn't fair or easily explainable in transplant, just like in cystic fibrosis. And so, despite my hearty assertions to the contrary before I received my lungs, I found myself learning to accept that some of those scary complications were, in fact, my reality. Throughout the three months that I spent preparing my body and waiting for my second double-lung transplant, I slowly came around to the understanding that just because things don't turn out the way you hoped or expected, that doesn't mean your life can't still be full of beauty and light. I made wonderful CF community friends going through my second transplant. When I got my second new set of lungs, I jokingly said to my mom that at least now I could begin my "complication-free" post-transplant life.

Wrong again, Piper.

It would be nearly six years out of

that first major complication that I would get my second reminder that life post-transplant is full of surprises. After an aggressive battle with skin cancer, a common post-transplant complication thanks to the intense immunosuppression drugs, I developed what we thought was a swollen lymph node on my neck. A few tests later, we learned that this growing bulge was metastatic squamous cell carcinoma, and that it had spread into both sides of my beautiful, transplanted lungs. My doctor was honest with me that survival rates for these types of cancer post-transplant were often low, but with the support of my family, friends, and work colleagues at the Cystic Fibrosis Foundation, I decided to begin intense chemotherapy. Much like my life before transplants, I once again found myself losing weight, struggling with daily tasks, and spending most of my time in the hospital. Emotionally, it was the lowest point of my life, but my support system buttressed me up until I finally found my footing on this new ground of being a CF, transplant, and cancer patient. A few months later my scans revealed no evidence of disease. Four years later, I'm grateful to say that is still the case. I had once again faced one of the scariest complications on that list from so long ago and come out standing.

So what can I say about my time post-transplant so far? Well, for one thing, I've definitely learned not to be so cocky about my ability to avoid side effects! More importantly, though, I have come to see that a beautiful life doesn't mean avoiding all the hard or scary stuff. Through my highs and lows with new lungs, I have (re)learned the value of slowing down and focusing on my health. I have taken stock of the people and things most important to me. I have learned not to view my complicated body as a failure, and instead to celebrate the fight and strength that has allowed me to get this far despite the obstacles. Best of all, I met my husband during this period of my life, the man who chose to stay and step in as my partner, despite the litany of health challenges and complications, and the guy who still stands by my side ten years later. To paraphrase Charles Dickens, my time post-transplant has truly been "the best of times and the worst of times."

And it has been beautiful every step of the way. ▲

Piper Beatty Welsh is grateful to be living and thriving with cystic fibrosis, double-lung transplant (two times over!), and chronic cancer at age 42. A former attorney, she now works full time as part of the Community Partnerships department at the Cystic Fibrosis Foundation, where she has the privilege of serving the CF community daily. When not in front of her computer, Piper spends most of her time reading, playing outside with family and friends, and planning where to travel next. She lives in Redding, CT, with her husband, Patrick, and their two silly dogs, Winston and Snoopy.

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diagnosis of CFLD had no significant worsening or improvement of liver fibrosis markers. A reduction in LSM in those with liver nodularity, with an initial highest result suggesting a potential positive treatment effect of E/T/I in this category of those with severe CFLD was observed.
http://tinyurl.com/bdf598m5

Electronic Home Monitoring Of Children With Cystic Fibrosis To Detect And Treat Acute Pulmonary Exacerbations And Its Effect On 1-Year FEV1.

This study aimed to investigate the effect of the use of electronic home spirometry in children with cystic fibrosis on 1-year FEV1 change. This is a randomized, one-year prospective study including children with CF between 6 and 18 years of age. Subjects were randomized into home spirometry group (HSG) and usual care group (UCG). Children in HSG performed two pulmonary function tests (PFT) per week. Data regarding acute pulmonary exacerbations (PEx) was obtained from patients' records. At baseline and 12th month, health related quality of life questionnaires for CF patients (CFQ-R) and lung clearance index (LCI) were performed. Sixty children were recruited with a median age of 13.3 years. Absolute change in FEV1pp from baseline to 12th month as median was +1% in HSG and -2.50% in UCG. In conclusion the study found electronic home monitoring of children with CF by spirometry may result in improvement in lung function.
http://tinyurl.com/2sjare3y

Telerehabilitation And Telemonitoring Interventions Programs Used To Improving Quality Of Life In People With Cystic Fibrosis: A Systematic Review.

The objective is to evaluate the effect of telemedicine programs in people with cystic fibrosis. Before the COVID-19 pandemic, telerehabilitation and telehealth were used, but it was after this that there was a boom in these types of assistance in order to continue caring for cystic fibrosis patients. For the search, the PubMed, Scopus, Web of Science, PEDro, Cochrane, and CINAHL databases were used.

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of waiting, I got the call that there was a match. My parents drove me to the hospital and we waited for hours until finally a doctor told us that the surgery was a go. For some reason I wasn't scared, I knew I was in good hands. I trusted the surgeon and doctors around me and my faith assured me that I would be okay regardless of what happened.

I remember waking up in the ICU extremely uncomfortable on the ventilator with my parents by my side. I had so many machines around me, and my dad said that they were testing my lungs and they were getting perfect results. I remember feeling anxious, uncomfortable, and thirsty. I couldn't talk so my parents gave me a pen and paper to write. I kept trying to ask for water and the nurse put little wet sponges on sticks in my mouth. I was very agitated and something felt wrong; eventually my nurse realized my sedation had been lowered so they adjusted the dosing to put me back to sleep. The second time I woke up I was much calmer—I was finally extubated and the long tracheal tube was removed. My throat hurt so much and my mouth was unbelievably dry. I felt a big sense of relief knowing that the intubation was over and I had survived the surgery. I expected to be able to take big deep breaths but that wasn't possible because the staples were so tight that my chest felt restricted—I was a little disappointed.

During my three-week hospital stay, I got physical therapy, pain management, education concerning all my new post-transplant medications, and the time to adjust to the feeling of my new lungs. I had some complications at first—I had atrial fibrillation (Afib) a couple nights after being on the transplant floor and that was a really scary experience; my heart was racing the fastest I've ever felt it while I was just lying there doing nothing at all. I also started seeing and hearing things because of the pain medicine and they had to switch it to something else. They said what I was experiencing were hallucinations that often appear on high doses of IV pain medicine, which I had never had before. I had to pass a swallow test to be able to eat and drink and soon I was able to eat my first meal, which was a cheeseburger and french fries from a nearby restaurant. The hospital food looked disgusting and one thing about

JENNA STRICKLAND, MARCH 12, 2016, AT THE BONE MARROW TRANSPLANT UNIT AT COLUMBIA PRESBYTERIAN.

me is I'm always going to get good food from somewhere, no matter what! There are a lot of food restrictions, especially the first year after transplant. The most important restriction was that everything had to be cooked well done. My mom made sure the restaurant cooked my burger well done and that there were no raw vegetables on or near it. It was hard to keep remembering what I could and could not eat since I was so used to eating whatever I wanted in hopes of gaining weight.

The rest of the time in the hospital was focused on physical therapy and walking without a walker. I had six chest tubes, but they removed four by the first or second week and the last two I had pretty much until the day I was discharged. The doctor said it was because my lungs were very large so that's why they continued to drain. Someone from thoracic came every day at 6:00 a.m. to check the drainage; if the volume of drainage was under a certain amount ,they could pull the tubes. The chest tubes were the most uncomfortable part of transplant recovery for me. They weighed me down and it looked like a tiny string was holding them in and that they could rip out with any sudden movement. I couldn't get out of bed on my own, which was very frustrating especially since PT encourages you to walk multiple times a day. Another hard part of recovery was that I had to sleep on my back. I'm a side sleeper and I wanted so badly to roll on my side but I couldn't because of the chest tubes.

I felt like I was in a dream when I finally got to go home—all my CF equipment that I had used to try to keep myself healthy was put away. My large oxygen concentrator and portable oxygen concentrator were given back to the rental company, and my house felt so much more like a home and not a makeshift hospital. I felt like my body had superhero strength—I could walk without getting short of breath and go upstairs without feeling light-headed and my heart pumping out of my chest. I never realized how much effort it took for my body to go up a flight of stairs or walk a couple miles. I was obsessed with walking and working out—it felt so easy and I was so happy to have healthy, clear lungs. My main goal now was to get back into the gym; I wanted to see how far I could push myself now that I could breathe. I had follow-up transplant appointments early in the morning and they involved x-rays, pulmonary function tests (PFTs), blood work, and a visit with both my transplant nurse and doctor. Over the next few months, I got my staples out, my feeding tube pulled,

sinus surgery that involved removing bone from my cheeks to open everything up for optimal drainage, and a bronchoscopy to check on my lungs and take samples to see if anything was growing in my sputum.

My first bronchoscopy was an absolute nightmare—I ended up having a pneumothorax (collapsed lung); I had to get an emergency chest tube; and I was admitted until my lung reflated. When we went over the potential risk factors before the bronchoscopy, there was a 1% chance of pneumothorax and I was unlucky enough to have it happen to me. My sputum also grew *Pseudomonas* but it is extremely common post-transplant because it can be colonized from the sinuses. I still had a PICC line in my arm from getting IV antibiotics in the hospital after transplant so I was able to do them at home. After I recovered from the pneumothorax, I was feeling really great and enjoying waking up in the morning and not having to sit there with a vibrating vest coughing up mucus for an hour. I also had to do a nebulizer treatment for a couple weeks but, compared to doing three nebulizers twice a day, this was really easy. My transplant clinic made me keep track of my oxygen saturation levels, my blood pressure, my weight, and my peak flow numbers in a big binder I brought to my transplant check-ups. Everything was looking good and I felt like my body was healed. I knew I still had CF but it didn't feel like I did anymore.

Around four months post-transplant I began having a terrible sore throat. I assumed I had strep throat so I asked transplant if I could go to my primary doctor in my town and get a strep test done. It came back negative; they thought I had thrush or tonsillitis. My transplant team decided that I should see my ENT so he could examine my throat. I remember him saying he didn't think it was cancer because it was such a short time post-transplant and that if someone is diagnosed with cancer post-transplant, it would usually be many years out. I was still really worried that it was cancer—I couldn't even swallow my saliva and the pain was so intense. It felt like sharp needles were stabbing the back of my throat.

One night it got so bad that my mom drove me to the ER at Columbia where I was admitted and had a biopsy done. A few days later after I was discharged, my transplant doctor called and said that I had PTLD (post-transplant lymphoproliferative disorder). I had cancer. I couldn't believe it. It was non-Hodgkin's lymphoma. After being sick my whole life, I finally felt what it feels like to breathe like a normal person and now this. I kept thinking "why me?" You always hear that life is unfair but I felt like this was beyond that; it was almost so surreal that my brain couldn't even process the thought of it. My transplant team referred me to an oncologist. My parents and I decided I would continue with my cancer care and treatment at Columbia because then they were able to work directly with my transplant team if I needed them or if I had any symptoms that affected my new lungs.

I started treatment right away and it consisted of six rounds of 96-hour chemo, meaning I had to be admitted into the bone marrow transplant unit of the hospital and I had 96 hours of straight chemo going into my PICC line. The first round went okay, but after that my blood counts began to drop and I had to get multiple blood transfusions while I was admitted. I also received a shot to stop my menstrual cycle and put me into menopause because I was bleeding so much. Within a very short time on chemo I became neutropenic, meaning my white blood count was dangerously low and my body wouldn't be able to fight infections of any kind. I could get an infection just from flossing my teeth too aggressively or getting a paper cut. I had to get daily injections to increase my white blood cell count, which caused severe bone pain. I can remember being in so much pain in between chemo and being at home that I had to go to the ER. I was in agony—even lying down and doing nothing hurt.

It felt like my back was being electrocuted and the pain shot down my hips and legs. I had to have a bone marrow biopsy so my oncologist could make sure that there were no cancer cells in my bone marrow. While they collected a sample of my bone marrow, they also injected chemo right into my spine in case there was cancer there. I felt overwhelmed and exhausted. The good news was the spot in my throat looked like it was melting away so it responded quickly to the chemo. I lost my hair, my eyebrows, and my eyelashes in addition to gaining about 30 pounds from being on very high doses of steroids. I responded so well to the chemo that my doctor let me do four rounds of 96-hour chemo and then two rounds of IV methotrexate, which was still done inpatient. I ended up being admitted for about a week because they had to wait until the medicine was completely out of my bloodstream to discharge me. My PET scan showed no cancer and I was so ready to heal my body again and get back in the gym.

I felt like I was my own nurse and physical therapist at this point. I joined a gym that wasn't crowded so I was able to clean the machines before and after I used them to prevent any germs. I also started working per diem at a company in the accounting office. It felt so good to be working and be a productive member of society. Unfortunately, I had that feeling again that something was wrong. I kept getting stomach pain at work and I couldn't figure out why. Eventually my gynecologist found a cyst on my uterus and we scheduled surgery to remove it. It was an easy surgery and I was relieved that it was such an easy fix; however, I still had the stomach pain, which progressively got

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worse. I went to the ER near my house and they diagnosed me with colitis.

I then had both a scan and a colonoscopy done at my transplant hospital because the pain kept worsening. Finally, they saw something on my sigmoid colon and again it turned out to be PTLD—the same type, which was Non-Hodgkin's Lymphoma. It had returned. I met with my oncologist and we came up with a treatment plan—this time it would be outpatient and consist of two medicines that aren't considered chemotherapy but are used to treat certain types of cancers.

Everything was going fine until I started getting migraines, my left eye started swelling, and I was super sensitive to light. I had to wear sunglasses everywhere and I couldn't open my right eye either because then my left eye would try to open as well. It was so painful. I found myself back in the ER again and my doctors all agreed it was sinus related and that I needed emergency sinus surgery.

My ENT cleaned out my sinuses and eye socket and also sent samples to pathology. He came in my hospital room the next day and I knew by the look on his face that it was cancer. I couldn't believe this was happening—PTLD, the same non-Hodgkin's lymphoma in the third place! I really didn't have time to dwell on it for long before my oncologist came in my room and said we were starting a new chemo regimen right away. He said I could stay on the lung transplant unit and a nurse from the oncology floor would come start it here for me.

A nurse came and started the drug and by morning someone found me unconscious—I was immediately rushed to the ICU. When I woke up I saw doctors standing around me, who told me I had a toxic reaction to the chemo drug and I would have to wait for it to get out of my bloodstream and hopefully mentally and physically go back to normal. I couldn't talk; I couldn't even spell my own name. It was one of the scariest things I've ever been through. I was shaking all over—especially my hands—and I had electrodes all over my head. I had an MRI because the doctors were worried that I had had a seizure but thankfully it was clear.

JENNA STRICKLAND, JULY 19, 2016, AT COLUMBIA PRESBYTERIAN, RECEIVING HER FINAL TREATMENT.

Eventually they released me from the ICU. I went back to the transplant floor, and my speech went back to normal. The scary part was I could no longer receive chemo. So how was I going to survive this cancer? I remember sitting in my room praying to God that they would come up with some kind of treatment. Shortly after that a research doctor visited my room and told my mom and me about a research study using donated cells that are brought to a lab and basically turned into "soldiers that attack the cancer cells" as he explained it to me.

There was a huge packet of information that I had to go over and sign to enroll in this study. I wasn't sure if this was the right thing to do because one of the side effects could be rejection of a transplanted organ. My heart sank when I read that thinking about my donor's beautifully working lungs. If I didn't enroll in the research study, I was going to die from cancer. I didn't have the choice to opt out of the study. This was my last chance at life. I began going to outpatient research study appointments and getting donated cells injected into my PICC line. I wasn't sure if they were working but the day after an injection I always felt incredibly tired and had more stomach pain than usual so that was my clue that they were doing something.

During one cell injection the pain got so bad they gave me IV pain medicine and decided I should be scanned. The scan was shown to a colorectal surgeon who said my colon had been perforated and my body tried to wall it off. This was so strange to everyone since I'm on prednisone, which can affect how the body heals. I needed surgery to fix it, but that meant I would also need a colostomy bag for at least six months so my colon could heal properly. During the surgery a portion of my intestine was removed and a colostomy bag was put in. My surgeon said it looked much worse than he thought; my insides were stuck together and my intestines had to be pulled apart. I don't remember much because I woke up in agony. I got IV pain medicine around the clock and I told my mom the pain was so bad that I didn't want to be alive anymore. I was brought to the bone marrow transplant unit, which was basically my second home now.

The nurses taught me how to change my colostomy bag and there was a whole process to putting it together that resembled an arts and crafts project. I was in disbelief that this was my life. Things were happening way too fast and I was just in awe that a few weeks ago I was diagnosed with colitis when it

was actually cancer. I was trying to see the good in the situation—any glimmer of hope that would help me get through these next few months with this colostomy bag. The best thing about being chronically ill is that you see firsthand how resilient you are; you see pictures of yourself from when you're in the thick of an illness and then you see yourself years later and realize how much your body is able to heal. I got my colostomy bag reversed six months later; the cell therapy worked and put me in remission. I've been in remission since 2018 and I'm so thankful for my family, friends, and all the healthcare professionals who who helped me cope with the many treatments and surgeries to get me where I am today

If there's one thing I have learned from having cancer it is to always be your own advocate. If you think your doctor is missing something, get a second, third, or even fourth opinion. Also, just because something is statistically unlikely doesn't mean that it shouldn't be looked into as a possibility. You know your body and you know when something feels off.

For the next year I was neutropenic and had to give myself injections every day to raise my white blood cell count. I lived with chronic bone pain and was getting frequent sinus and throat infections. My sinuses were completely clogged with thick mucus and my throat was so red and inflamed that I had to spit out my saliva rather than trying to swallow because it was so painful. I had fevers often and I couldn't sleep because of the pain. I went to the ER near my town and they admitted me for IV antibiotics, fluids, and pain meds. About a year later, my white blood cell count had recovered and I was able to slowly stop my injections. I haven't had a throat infection since so they must have been linked to being neutropenic.

My eighth lung transplant anniversary was on July 17, 2023. I feel beyond grateful to still be alive and have my donor's lungs but, at the same time, I feel a deep sense of guilt. I feel guilty because many of my CF peers have passed away in these eight years, many of whom I became close with and who were younger than me and weren't able to experience as much in life as I have. I feel blessed that I was able to have a somewhat normal life while my CF was manageable. My own lungs lasted me 25 years; I was able to graduate with a college degree and get into the graduate program of my choice. I have a lot of limitations now and I often feel overwhelmed by my health issues but I'm so lucky to have amazing family, friends, and the best parents I could ever ask for who make me feel so seen, loved, and supported.

Ever since my colostomy bag reversal it's been hard for me to eat and go to the bathroom without pain. I used to love trying new restaurants and new food and it was something I really enjoyed but now I have to be strategic when I eat, and I try to eat things that I know won't upset my stomach as much as others. It's been a long journey and I've felt a lot of judgment from certain doctors and nurses because of the stigma of pain medication. I've been talked down to and denied pain meds multiple times, but I have the best transplant and palliative care doctor who understands my stomach pain and how I want to have the best quality of life that I can. I've been on palliative care since around 2019 and I have a set pain medicine regimen for when I'm home and when I'm in the hospital. I think, because of the way I look, those who don't know my story may not understand why I need pain meds or they might think I'm not in pain and just drug seeking.

I've suffered from blockages since I was little and needed surgery for a twisted bowel when I was six. I was diagnosed with distal intestinal obstructive syndrome (DIOS) in 2012, which is when thickened stool blocks the small intestine. I've been admitted many times for blockages. During my treatment for lymphoma in my colon, I suffered from severe pain, especially after the reversal of my colostomy bag. My palliative care doctor explained that this is the result of scar tissue. When I'm on antibiotics the pain gets a lot worse, especially after eating. Some of my nurses don't understand my pain and they wonder why I don't eat much while I'm admitted. I try to eat bland food so I stay away from the hospital food and usually eat cereal, bagels, yogurt, and peanut butter and jelly sandwiches. I have constant diarrhea when I'm on antibiotics so I'm often in the bathroom for long periods of time and going to the bathroom can be very painful. I'm on high doses of pain medicine because I've been on them for so long that my body has become tolerant and pain is harder to treat and prevent. I've had to leave social events early, including the wedding of one of my best friends, because of my stomach pain. My family and friends know when I'm starting to have pain by just looking at my face. I will bring medicine with me for breakthrough pain if I know I'll be eating or if I'm on antibiotics. There is definitely a stigma associated with pain medicine; it's unfortunate that, because of those who abuse it, the ones who actually need it get judged or looked down upon.

I still struggle with sinus symptoms and my scans show that my sinuses are always inflamed which my doctors say is from PTLD being in that area. I started taking the CF modulator called Trikafta in the summer of 2020 it has definitely helped some of my CF symptoms. Even though I don't have CF lungs anymore, my other organs are still affected. Since I began Trikafta I've gained weight, which means I'm absorbing my food better; my stools are less greasy; I don't sweat salt; I have way more energy; my ENT says my sinuses look better; and I haven't had polyps in my sinuses since I started taking Trikafta.

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Unfortunately, the negatives of this drug are all mental for me. Everything I've been through has made me a very anxious person but, since starting Trikafta, my anxiety is much worse. It's hard for me to concentrate on anything and, instead of feeling sad sometimes, I feel a lonely and empty feeling deep inside at all times that I can only describe as depression. I've talked to many of my CF peers who also feel this way on the drug. Like any drug there are always pros and cons, but it's very hard to suffer from depression while constantly having to take care of yourself and stay on top of medicines, doctors' appointments, exercising, and eating, as well as frequent hospitalizations, procedures, and surgeries, when you don't feel good.

> "The scary part was I could no longer receive chemo. So how was I going to survive this cancer?"

My CF clinic has hired a psychiatrist and has weekly therapy for patients, which has really helped me develop coping skills for my anxiety. I'm still processing a lot of trauma from being in the hospital so much, especially in the ICU and on a ventilator. I had sepsis in 2020 and almost died—I was rushed to the hospital in an ambulance. My blood pressure was extremely low. I had pneumonia in my lungs, and the only way for me to survive was to be intubated. The doctor told my mom that they might have to put me in a medically induced coma but thankfully that didn't happen; I was intubated and put on IV antibiotics. The hardest thing about what happened was hearing everything and everyone around me but I was unable to move or open my eyes. I remember everything that was said and how I felt and I was scared to sleep for a long time after.

I was diagnosed with diabetes after my colostomy bag reversal. I had to learn how to check my blood sugar and give myself insulin injections. Because CF affects the pancreas, many of my CF peers have had diabetes their whole lives, except for those who usually get diagnosed after transplant from being on steroids. This was a huge change for me because I always ate whatever I wanted without even thinking about it. My diet has changed from eating as many carbohydrates and calories as possible to learning how to count carbs in the hospital and limit things like soda and dessert.

Now, to manage the diabetes, I use a sliding scale for my Humalog, which is short-acting insulin that I take with meals and I have a standing dose of 12 units of Lantus, which is long-acting insulin at bedtime. Having diabetes was one of the hardest things for me to get used to, and I still struggle to remember to check my sugars before I eat and when I'm out with friends as sometimes it's hard to want to give myself insulin in a public place. This is when I rely on CF friends to help me navigate my diabetes in the best way. The CF community is so supportive and offers such a wealth of information. I've met so many of my CF peers on Facebook and in person at transplant clinic and it's these friends whose advice I seek often and with whom I can vent my frustrations. It's crucial for me to have people in my life who understand firsthand what I'm feeling. It helps ease my anxiety when I can talk to someone who has already been through it about a new medicine or procedure. Since I was diagnosed while inpatient, I never really had an endocrinologist, but I'm now looking into one to discuss the best way to monitor my blood sugar. The new diabetic technology is truly amazing! Things like the Omnipod, which is a wireless insulin pump that you can even swim with, makes managing diabetes so much easier. This is something I want to discuss with my endocrinologist to try to make my life easier and make sure my blood sugar is being managed at all times.

On my birthday this year I felt like I was numb the entire day. How am I alive? I tend to joke about that a lot because I've realized if I'm not laughing about it then I'm crying my eyes out. The truth is dying scares me a lot—I love my life, despite how hard and painful it can be. I've had to give up a lot and that often makes me bitter and extremely jealous of my peers. I had to give up earning my master's degree, a career, having children, and a lot of my independence since I can't drive on my pain medicine and I live with my parents since I'm on disability. The one thing that having a chronic illness has taught me is to cherish every little thing and that the little things really are the big things. It also taught me to appreciate every single day that I'm able to wake up in the morning. My therapist taught me that I could be both angry at my illness but also feel extremely blessed and appreciative of my new lungs and my life in general.

I'm just trying to take it day by day and not worry about what the future holds. Regardless of what happens I'm going to keep fighting for my health so I can keep making memories with the people I love. ▲

Jenna Strickland has CF and is 34 years old. She lives in Stratford, Connecticut. She had a double lung transplant on July 17, 2015. You can contact her at Stricklandjr2@gmail.com. Some of her favorite things are cooking, working out, barre and Pilates classes, her dog Lilly, spending time with friends and family, watching HGTV, and anything involving fashion.

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Randomized controlled trials, pilot studies, and clinical trials have been included. A total of 11 articles have been included in the systematic review. No improvements have been found in quality of life, forced expiratory volume, and forced vital capacity. Good results have been found in increasing physical activity and early detection of exacerbations. Adherence and satisfaction are very positive and promising. Despite not obtaining significant improvements in some of the variables, it should be noted that the adherence and satisfaction of both patients and workers reinforce the use of this type of care. Future studies are recommended in which to continue investigating this topic.
http://tinyurl.com/mr3f2h2w

Quality Of Dietary Macronutrients Is Associated With Glycemic Outcomes In Adults With Cystic Fibrosis.

Poor diet quality contributes to metabolic dysfunction. This study aimed to gain a greater understanding of the relationship between dietary macronutrient quality and glucose homeostasis in adults with cystic fibrosis. Individuals with CFRD consumed less total fat and monounsaturated fatty acids (MUFA) compared to those with normal glucose tolerance. In Spearman correlation analyses, dietary glycemic load was inversely associated with C-peptide. Total dietary fat, MUFA, and polyunsaturated fatty acids (PUFA) were positively associated with C-peptide. Plant protein intake was inversely related to HOMA2-IR. Associations remained significant after adjustment for age and sex, Improvements in diet quality are needed in people with CF. This study suggests that higher unsaturated dietary fat, higher plant protein, and higher carbohydrate quality were associated with better glucose tolerance indicators in adults with CF. Larger, prospective studies in individuals with CF are needed to determine the impact of diet quality on the development of CFRD.
http://tinyurl.com/yh6pu7nd

Prevalence of Low Testosterone in Men With Cystic Fibrosis and Congenital Bilateral Absence of the Vas Deferens: A Cross-sectional Study Using a Large, Multi-institutional Database.

The objective of this study was to investigate the prevalence and treatment rates of low testosterone (T) in men with cystic fibrosis (CF). Despite the well-described association between CF and infertility secondary to congenital bilateral absence of the vas deferens (CBAVD), men with CF report further sexual and reproductive health concerns, many of which are often associated with low testosterone. Serum T levels were measured in 10.1% of men

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TRANSPLANT TALK

To Keep Or Not To Keep The Thumb

By Colleen Adamson

The last time I wrote for "Transplant Talk" I mentioned that I had squamous cell skin cancer in my right thumb. I had just completed six weeks of radiation, and the cancer had not come back.

Ah, those were the days: I was cancer and pain free for about five months after my radiation treatment ended. I was back to playing golf without pain. It was very nice...until the pain started to come back and got worse over time. The cancer had returned! This cancer was as stubborn as a person with CF, believe me. Nothing had worked to eradicate it. Topical chemotherapy creams and ointments, blue light therapy, more creams and ointments for longer periods of time, and now radiation had all failed to kill this cancer on my thumb. Did I mention it is uncommon to have skin cancer on a thumb? I get all the rare/weird stuff and it gets old being an outlier all the time.

In November 2023, I had Mohs micrographic surgery, which is a specialized surgery for skin cancer. If you have had a transplant, you might be familiar with this surgery as skin cancer is common in immunocompromised people. Mohs surgery, developed in 1938 by general surgeon Frederic E. Mohs, is a microscopic surgery used to treat skin cancer. Here is a description of Mohs surgery from my Mohs surgeon's office website (https://aadermatology.com/services/mohs/): "The visible portion of the tumor is removed, then working outward from the affected area, a thin layer of skin is removed at a time. The tissue is immediately evaluated under a microscope to determine the extent of the cancer. Successive layers are removed only where necessary, until no tumor is present. This complete, systematic microscopic search for the "roots" of the skin cancer offers a 97%–99% chance for complete removal of cancer without an excessive loss of normal tissue. As a result, Mohs micrographic surgery is very useful for large tumors, tumors with indistinct borders, tumors near vital functional or cosmetically important areas, and tumors for which other forms of therapy have failed." During the initial evaluation of the tissue, the patient waits in the waiting room for about one hour. If there are still signs of cancer, the patient goes back to have that surgically removed. The process keeps going until there are no signs of cancer, which means the patient could be there for several hours, going back and forth between the procedure room and the waiting room. It is done in the office, not at a hospital, and patients just go home when it's done. I've had more of these surgeries than I care to count, but luckily I have only had to go back to the procedure room once or twice during each surgery. For my thumb surgery, the surgeon had to go down to the bone to get clear margins, so he ended up taking out the entire top left side of my thumb! Since the surgical area was too big to close up, a few days later a hand surgeon placed a (fake) skin graft over the surgical area. These two surgeries were all planned out ahead of time because it was obvious to my two surgeons that this was going to be a big surgical area. I was very lucky to have two wonderfully accomplished and accommodating surgeons!

> *"You absolutely need to nip things in the bud before they get out of control and are harder or impossible to treat."*

COLLEEN ADAMSON GIVES THE THUMBS UP.

So far, the healing is going well. I see a wound care management team once a week, and I am learning so much from these wonderful people. They are, of course, very knowledgeable about what I need to be doing,

eating, drinking, etc., to help with the healing process (eating a lot of protein, which is very hard to do actually, and drinking water mixed with therapeutic nutrition powder), and what works best when wrapping my thumb (collagen dressing with oxidized regenerated cellulose (ORC) and silver, and polyester mesh dressing with hydrocolloid and petroleum jelly particles). Since my thumb is wrapped all the time, that means no washing dishes for Colleen! There—I found a silver lining in all this. Plus, I managed to keep most of my thumb.

There is a bottom line to all of this. As you may know from some of my previous *CF Roundtable* articles, I had a bilateral lung transplant on 07/04/1998 and a kidney transplant on 03/07/2006 (the kidney was donated by my best friend Kelly). I have had a lot of medical issues since my transplant, and I will have more in the future. Skin cancer is here to stay. However, I have found that people with transplants, like people with CF, need to be keenly aware of their bodies. If you think something is "off," please tell your doctor. You absolutely need to nip things in the bud before they get out of control and are harder or impossible to treat. I tell people that having CF trained me for this life post-transplant—all of the medications, doctor appointments, etc. It's a lot. If you are thinking about having a transplant, or are post-transplant, know that our disease has prepared you for this. It doesn't prepare you for everything, but you will have a great foundation to build upon as you undergo your own journey. And it is worth it. ▲

Colleen Adamson is 54 years old and has CF. She is 25 years post lung transplant and almost 18 years post kidney transplant. She is the Treasurer of USACFA, and lives in Alexandria, VA. Her contact info is on page 2.

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with CF and 8.9% of men with CBAVD. Within each group, 464 men with CF (32.7%) and 132 with CBAVD (43.0%) demonstrated low T. The majority of men with T < 300 ng/dL went on to appropriately receive TT: 59.3% of men with CF and 78% with CBAVD. The data suggests that hypogonadism is highly prevalent in men with CF and CBAVD. Investigation and appropriate treatment of testosterone deficiency may significantly improve quality of life.
http://tinyurl.com/yckrjse4

Chronic Rhinosinusitis In The Era Of CFTR Modulator Therapy.

Chronic rhinosinusitis is a common manifestation of CF that is associated with impaired quality of life and can be difficult to treat. CFTR modulator therapy has resulted in significant improvements in lower respiratory and nutritional outcomes for people with CF however their impact on chronic rhinosinusitis has received less attention. While an overall improvement in symptoms, imaging and endoscopic appearances is seen in response to treatment, limited impact is documented on olfaction. Outcome measures employed were heterogeneous, limiting comparison of findings. There is a need for well powered prospective real-world studies with standardized outcome measures.
http://tinyurl.com/cs9e5uvy

Characteristics Associated With Cystic Fibrosis-Related Pulmonary Exacerbation Treatment Location.

Previous studies indicate that hospital rather than home treatment of pulmonary exacerbations in people with cystic fibrosis can improve outcomes. Characteristics of adult participants from the Standardized Treatment of Pulmonary Exacerbations (STOP2) trial were evaluated with two separate comparisons: (1) those who were treated initially in hospital to those treated initially at home and (2) those treated only in hospital to those who were treated only at home or both at home and in hospital. Participants who had Medicaid insurance, were treated for shorter duration, and traveled longer to reach treatment centers were more likely to have been treated initially in the hospital. Having Medicaid insurance, being treated for a shorter duration, and being male were associated with being treated only in the hospital. This analysis suggests decisions about the location of treatment are based on pragmatic factors rather than on clinical characteristics.
http://tinyurl.com/2ju2sy8t

Red Ginseng Aqueous Extract Improves Mucociliary Transport Dysfunction And Histopathology In CF Rat airways.

It was previously discovered by these researchers that Korean red ginseng aqueous extract (RGAE) potentiates the TMEM16A channel, improved mucociliary transport (MCT) parameters in CF nasal epithelia in vitro, and thus could serve as a therapeutic strategy to rescue the MCT defect in cystic fibrosis (CF) airways. The hypothesis of this study is that RGAE can improve epithelial Cl– secretion, MCT, and histopathology in an in-vivo CF rat model. Seventeen 4-month old CFTR–/– rats were randomly assigned to receive daily oral control or RGAE for 4 weeks. Outcomes included nasal Cl– secretion measured with the nasal potential difference (NPD), functional microanatomy of the trachea using micro-optical coherence tomography, histopathology, and immunohistochemical staining for TMEM16a. In conclusion RGAE improves TMEM16A-mediated transepithelial Cl– secretion, functional microanatomy, and histopathology in CF rats. Therapeutic strategies utilizing TMEM16A potentiators to treat CF

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CHAPTERED LIVES

Port In A Storm

By Andrew B. Corcoran

In 2011, I was living in a tiny, one-horse town in the middle of Brazil. My apartment was on a dirt road and, to the left of me, was the beginning of the Brazilian jungle. My girlfriend, Veronica, and I were teaching English at local ESL schools. Life was about as uncomplicated as it could get. We would work, eat, drink, blow all our money on day trips to a remote beach, head back to our tiny village in the middle of nowhere, and on and on it went. It was simple. It was boring. It was exactly what I wanted. The closest doctor worked out of a dirt-floor clinic in the center of town. There were no hospitals for miles. I had no appointments to attend, no pulmonary function tests to do, no checkups, no thoughts of transplant or trauma, no reminders of CF or clinic, and no coordinators or nurses. I just had my life. My normal, everyday, easy-going existence.

I had finally gotten to a point where I no longer woke up and went to bed with stirring thoughts of dread. I was grateful for my gift of life, but I wasn't beholden to the guilt I previously experienced. I substituted that sorrow and penitence with love and gratitude.

For Christmas 2011, Veronica and I went south in Brazil to an island called Florianopolis. On Christmas night the two of us sat on the beach, staring out into the south Atlantic Ocean, listening to the soft waves crash, and looking at stars so bright and magnificent, so enchanting and wondrous, that I remember feeling *this is why I'm alive; this is why I got my transplant.*

I had fallen in love with living. For so long, surviving had been my only strategy, my only approach to life. Sometime, somewhere, somehow, over many years, living slowly and quietly replaced simply surviving. Once again, I was proud of myself—proud of the fact that I was healthy but, more so, proud of myself for enjoying all the little things that make life worthwhile.

I got my transplant on July 21, 2002. It wasn't until a decade later that my sister Maura, who also has CF, needed a transplant of her own. Even though Maura and I were both born with CF, her needing a lung transplant was somehow more difficult to process than my own. She was always healthier than me and took much better care of herself. My own transplant was complicated enough, not for the physical nature of it, but the aftermath. The emotional scars and trauma of knowing that a young man had died and that I had his lungs as a result was nearly impossible to process, especially those first few years. Even now, this many years later, I get overwhelmed when I think about the notion that I'm breathing through someone else's lungs.

> *"Sometime, somewhere, somehow, over many years, living slowly and quietly replaced simply surviving."*

ANDREW CORCORAN

And so, in the summer of 2012, after having traveled from one country to the next for six years, it was finally time to come home. In September 2012, my parents held their annual CF fundraiser. That year, my whole family attended and my sister gave a speech. At the time, despite the chaos that comes with having twin toddlers, she had CF exacerbations infrequently and, more often than not, her CF was largely controlled.

Within a month of her speech, Maura was on life support at Columbia-Presbyterian Hospital in NYC. Veronica and I were then living in Queens and, each day, we would take the train up to Columbia, where Maura's health continued to decline rapidly. In a very short amount of time, it was necessary for Maura to have a transplant of her own. It all happened extremely fast. I was on the transplant list for two and half years before my call finally came, whereas Maura's life expectancy was only a matter of days or weeks. She was deathly ill. My entire family, both immediate and extended, was at the hospital daily.

Recently, an uncle of mine told me a story about those days. My uncle is a surgeon and was speaking with another uncle, who is a pulmonologist. On a drive home, after seeing Maura in the hospital, they had a conversation:

"Have you ever seen anyone that sick before in your life?"

"No."

"Have you ever heard of someone that sick surviving?"

"...No."

In late October of 2012, I received a call from my father. He was a wreck. He told me to get to the hospital immediately because that day might be her last. I was in a daze, looking at the world through a thick mist of ethereal illusion during that train ride to the hospital. When I walked into Maura's ICU room and saw her, I collapsed. We all knew how sick she was but seeing her in that state was shocking—just the day before she was talking and communicating and now she was just *there*. Alive, but unconscious; warm and cold at the same time. She was occupying two separate planes of existence. Her body was in front of us, but she was slipping away so fast. I remember taking her hand and daring her to wake up—begging, crying, shaking. She was going someplace none of us could follow.

Maura needed a transplant *that* night. Her chances of surviving another day were so slim and so fragile that to even whisper it out loud would melt it away.

As my family gathered, we got word that a set of lungs were available. A collective sigh of cautious relief swept over us. For all of us who have received an organ transplant, we are acutely aware of the number of stars that must align in order for that miracle to happen.

However, my sister had another obstacle to overcome. One that no one could have seen coming. This was late October 2012 in NYC and the obstacle's name was Hurricane Sandy. Sandy had just hit lower Manhattan with winds over 80 mph. I remember walking outside, into the rain, as the winds were howling down 168th Street. Trash cans were being lifted off the ground; lower Manhattan was under water.

I stood outside alone and watched as the surgical recovery team was flying out into the hurricane to procure the organs. Hours passed. Stillness hovered like a cold cloud around and over the room. Individually, we all wandered the hallways. I watched my brother-in-law shake, pray, and silently utter for God to just "give her a chance."

"Maura needed a transplant that night. Her chances of surviving another day were so slim and so fragile that to even whisper it out loud would melt it away."

Then, a nurse entered the room. "The lungs are not good." *Confusion. Anger. Hopelessness. Despair. Devastation. Complete and total devastation.*

To this day, I cannot say why I did this, but my eyes and ears never left the doctor's mouth. A feeling came over me. A calmness. Perhaps it was mere hope. I like to think it was more than that, but to say for sure would be a lie. Either way, I simply continued to stare at Maura's doctor. He was on the phone; hurried yet collected. He turned to Maura's nurse as they tucked themselves into a corner and muttered into her ear. In that moment, she looked over at me. She was stoic, but a gleam in her eye made me realize something. Something was *happening*.

I turned to my dad as tears poured down his face, "Just wait...it's not over." *I always imagined that one day, God would make his way into my life. He would reach down from the heavens and let me know he's been watching and things would be ok. This was the feeling that crept across my body from that nurse's smile. Relief swept over me with a force higher than I remember, before or since.*

Maura's doctor entered the room, "there's another set of lungs...the team is on the way now...we're doing this tonight." Two different sets of lungs. One hurricane. A family's love. And my sister Maura. A mother, a daughter, a wife, a sister, a friend, a patient, and a CFer. *We're doing this tonight.*

For many people with a serious disease, there usually comes a time when a question is asked: what would our lives look like without the disease? For me, it was in the quiet, still moments of starry-skied, early-morning, insomnia-ridden, sleepless contemplation—those 3 a.m. moments where sleep seems impossibly distant and insomnia seems unforgivingly present. It's those nights when your mind is racing so fast and hard that the anxiety of not sleeping *tonight* turns into the anxiety of not sleeping *tomorrow* night as well. The one or two times I have allowed myself to entertain the idea of life without CF were always and quickly met with feelings of shame and guilt.

I have never met anyone with cystic fibrosis who would trade their life for another's. In essential ways, it defines a significant part of me. A resilience and survival that allows me to continue, knowing that whatever may come, my family and I will accept that challenge and persist with hope, with resolution, and, ultimately, with unconditional love. ▲

Andrew Corcoran is 43 years old and has CF. He received a lung transplant in 2002. He now lives in south Jersey with his family and friends. He is a writer. Andrew's email is acorcoran@usacfa.org.

PIECING LIFE TOGETHER

The Journey To My Transplants

NEW COLUMN

By Matison Deaton

I had never been to San Francisco before moving there, over 2,000 miles away from the home and life I knew, to receive combined double-lung and kidney transplants in 2022. At the time of pursuing transplantation, I was living in my hometown of Anchorage, Alaska. I had been searching for a center to accept me as a transplant candidate for years and with there not being one in the entire state, I knew from the start I wouldn't be able to stay there if it were to happen. The closest transplant center was in Seattle, Washington, a three-to-four-hour plane ride away. But until I heard the news that the University of California, San Francisco wanted to bring me down for an in-person evaluation in April of 2022, it hadn't hit me just what being accepted at an out-of-state center would mean.

I first met with a lung transplant doctor on the UCSF team over Zoom in August of 2020, during the height of the COVID-19 pandemic and almost two years exactly before I would receive my transplants. I had been through so many rejections from transplant centers due to the active infection in my native lungs, *Mycobacterium abscessus*, that I had essentially no hope that meeting with UCSF would bring any other outcome. So I was shocked, delighted, and truthfully a bit afraid when I heard the words, "I think we can help you" come out of the doctor's mouth; delighted because this was a sentence I truly believed I would never hear and horrified because now the anxiety I had been feeling up to this moment suddenly multiplied.

For me, the planning and anticipatory phase of transplant was the worst. I've always struggled with anxiety when it comes to not knowing the outcome of a situation and all the things that must be done to get to said outcome. And in the scenario in which I suddenly found myself, there was so much to figure out and line up for a move to another city. Before I could even be evaluated, potentially accepted, and then listed, I had to relocate fully to San Francisco. For the transplant team's confidence in the success of my transplants, I couldn't be in Alaska when listed, nor for at least two years post-transplant because of the risk of not getting transplant-specific care fast enough if a complication were to arise. While I understood, and even appreciated, the seriousness with which they held my situation, it was going to be a lot to plan.

> *"How was I to give up total control over my body, my organs, my life, to a whole team of people I barely knew and some whom I'd never met?"*

MATISON DEATON

First, there was the financial side: figuring out insurance coverage, copay expectations, hospital stay coverage, and my insurance company's approval of a Medevac flight, given that my liter flow rate of oxygen was too high for commercial flights. At that time, despite being 30 years old, I was still under my parents' medical insurance through their employers as a disabled dependent. My primary insurance company, through my mom, did not consider UCSF as a covered transplant center. Moreover, there was no "out-of-network" coverage for transplant care. I would've had to pay for it entirely myself, which was not even imaginable. In researching a solution, I found that I had the option of asking my mother's employer directly to make an exception in their offered policy and adding UCSF as a covered transplant care center. If they were to agree and make this change on my behalf, my pre- and post-transplant care at UCSF would be covered. It felt like a big and improbable ask, so I was pleasantly surprised when we received that phone call that my request was approved. While great news, my stress was only temporarily relieved—its source now shifting to the process and impact of leaving behind the only home and support system I knew.

While I'd been away from family when I went to college in Arizona for two years, I was relatively healthy then and not facing a challenge like organ transplants. Now, even though I main-

tained as much of a sense of independence as I could, I was reliant on physical help from those around me. Thinking of being in a city I'd never been to, alone, while in end-stage respiratory disease and preparing for a major life event and medical surgery, it was overwhelming to say the least. I was mentally prepared to do whatever I had to for this opportunity at life-saving care, but I didn't know how I was physically going to get through any of it. I was stepping the furthest away from my comfort zone I ever had in my life.

On a somewhat positive note, traveling for medical care wasn't a new concept to me. There's no official CF center in the state of Alaska. At the time of my living there, we had one CF specialist who worked out of a local pediatric pulmonology clinic in Anchorage, overseeing almost all CF patients in the state. For some years after my diagnosis at 18 months old, that's where I was followed, but, at one point, my parents made a change. We started traveling by plane every three to six months to Seattle, where I was then followed at the closest CF Foundation-affiliated center at Seattle Children's Hospital. I was even hospitalized there a number of times, one of my parents remaining with me until my discharge. So, while still not my idea of stress-free, traveling out of state for my health was at least a familiar hassle. The difference while anticipating it in 2020, though, was the enormity of the reason: a multi-organ transplant and the commitment that would follow for the rest of my life.

From 2020 to 2022, I constantly thought of what moving would be like. I played out every possible failure, loss, and disappointment that could happen. I had no clue where I would live, no way to know how I would get help if I needed it quickly, and, most importantly in my mind, no relationship yet with the team to whom I was handing my life. Anyone with experience navigating the healthcare world knows how difficult it can be to find providers in whom you have full trust and complete confidence. How was I to give up total control over my body, my organs, my life, to a whole team of people I barely knew and some whom I'd never met? The thought of dropping my present life to pursue a future was one thing but giving up all control, even to a team of experts, something I'd never been comfortable with, was entirely nerve-racking.

What got me through that stressful time, all my over-thinking and catastrophizing (other than having no choice in life but to move forward and through what comes) was this thought I'd carried with me, that one day I'd be looking back at all that had to happen, thinking, "Look, all of that is over. I'm here now." I'd be alive in a new chapter of life, facing the new challenge of shifting from surviving from moment to moment, to being present in each one. That gave me a goal to reach. My fears, instead of barriers to my steps forward, propelled me forward to a new destination, one I was determined to reach at all costs. While the path was difficult and at times less than enjoyable, I can now, three, almost four years after that initial Zoom meeting with UCSF, see that I made it, and more importantly, in one piece. While I may now be piecing life together in my post-transplant world, I'd choose every part of that journey again. Transplant, with its terrifying and beautiful unknown possibilities, was worth it. It brought me to the person I am today. ▲

Matison is 31 years old and has CF. She was born and raised in Alaska, and currently lives in San Francisco, where she received combined lung and kidney transplants in 2022. She's on the CF Foundation's Rose Up Committee; dedicates her time to advocating and spreading awareness for CF, organ donation, and kidney disease on her social media pages; and, in her spare time, she enjoys jigsaw puzzles. She can be contacted at mdeaton@usacfa.org, *and found on TikTok @onebreathatatime_ and Instagram @matisondeaton.*

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airway disease are appropriate and provide a new avenue for mutation-independent therapies.
http://tinyurl.com/3cnbzs2f

Impact Of Refrigeration And Freezing-Thawing Of Breast Milk On In Vitro Digestibility And Liposoluble Vitamin Bioaccessibility In Breast-Fed Infants.

There is little information about the impact of refrigeration and freezing-thawing on breast milk digestibility under gastrointestinal conditions of healthy infants or those requiring pancreatic enzyme replacement therapy such as infants with cystic fibrosis. This study assessed the impact of refrigeration and freezing-thawing on fat and protein digestibility and liposoluble vitamin bioaccessibility of breast milk. In vitro digestion models mimicking both healthy infant and CF infant conditions were applied. Freezing-thawing significantly increased the fat globule particle size. For CF digestion, this change had a more negative impact when using a freezing-thawing process than when using refrigeration of breast milk, reducing lipolysis (up to 18%), proteolysis (up to 28%), and vitamin A and E bioaccessibility. Under healthy conditions, no significant effects were detected. An adequate pancreatic enzyme replacement therapy dose would enable the same level of lipolysis (55%) as in the healthy scenario. In conclusion, breast milk is the only source of energy and nutrients for breast-fed

Continued on page 41

Call For Award Nominations

USACFA has presented two separate awards to members of the CF community for over 20 years. *CF Roundtable* readers submit nominations, and the USACFA board members vote on who will receive each award for that award cycle.

Please send your nomination, including a description of not more than one page, listing what that person has done for the CF community to awards@usacfa.org by March 1, 2024.

The Jacoby Angel Award recognizes **an adult with CF** who has followed in the footsteps of Dr. Jack Jacoby by dedicating themselves to helping others. The person could have helped one or more people who either had CF or did not. Any nominee must be living and cannot currently be serving on the USACFA board.

The Jacoby Angel Award is named in memory of Dr. Jacoby, who served as a medical advisor to *CF Roundtable* for 15 years and was a physician at the St. Vincent's CF Center in New York City. He worked to find innovative ways to treat CF at a time when treatment options were very limited. He had a heart of gold and was a hero to his colleagues, patients, and their families. Some previous recipients of the Jacoby Angel Award include Robyn Petras, Susan Burroughs, Michele Compton, Jerry Cahill, Pammie Post, Isabel Stenzel Byrnes, Dr. Paul Quinton, and Terry Wright.

The Founders Award recognizes a person, **with or without CF**, who has made an outstanding contribution to the adult CF community. Any nominee must be living and cannot currently be serving on the USACFA board. The award was named in honor of the group of adults with CF who founded USACFA and created *CF Roundtable*. They worked tirelessly to bring information to the adult CF community at a time when there was no internet and no efforts to connect adults with CF in order to provide information and support for the adult CF community.

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

An example of a prior winner is Pammie Post. Pammie was the recipient of the 2007 Jacoby Angel Award. She gave so much of her time and energy to a variety of CF-related causes. She served for nearly ten years on the USACFA Board of Directors. Her constant positive outlook has inspired many others with CF to look beyond the trouble and hardship that CF may cause them and to focus on the good things in their lives. She has given guidance and counsel to hundreds of families with children who have CF, and to adults with CF who are struggling with the disease. For her extraordinary efforts on behalf of people that have CF, we presented the 2007 Jacoby Angel Award to Pammie Post, of New Canaan, CT. ▲

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

Alexandra "Xan" Nowakowski
Lakeland, FL
40 years old on December 29, 2023

Andrea Eisenman
New York, NY
59 years on November 28, 2023

Sydna Marshall
Austin, TX
43 years on September 19, 2023

Wedding

Sydna Marshall and Adam Keys
Austin, TX
10 years on October 26, 2023

Announcing Our Newest Scholarship: The Stenzel Scholarship

The Stenzel Scholarship was established in 2023 in memory of two amazing women with CF, Isa Stenzel Byrnes and Ana Stenzel.

Isa was a remarkable woman of grace, wisdom, strength, and compassion. Isa was a licensed social worker and had a Master's of Public Health. She imparted her wisdom to *CF Roundtable* readers for 17 years in her *CF Roundtable* column, "Spirit Medicine." Isa worked for the past eight years as a bereavement social worker for Mission Hospice in the Bay area. She was 51 years old and 19 years post-lung transplant when she passed away on July 12, 2023.

Ana Stenzel was Isa's twin sister. Ana was a genetic counselor at Stanford hospital for 16 years. She was a strong advocate for people with CF and educated people on the importance of organ donation. She had her first lung transplant in 2000 and then a second lung transplant in 2007, at a time when second lung transplants were extremely rare. She was 41 years old when she passed away in 2013.

Isa and Ana dedicated their lives to helping others. They provided education, hope and comfort to thousands of people throughout their lives. They showed the world that people with CF could find meaning in their lives by making a difference in the lives of others.

Isa and Ana, wrote a best-selling book, *The Power of Two–A Twin Triumph*, in 2007. *The Power of Two* was made into a documentary about their lives and won numerous film festival awards in 2011. It has been seen and appreciated by thousands of people over the years. Isa and Ana were very active on Team NORCAL at the Transplant Games.

The Stenzel Scholarship aims to provide a scholarship once a year to a person who embodies both Isa and Ana's commitment to social justice and a commitment to helping others.

Description of the Scholarship:

The Ana and Isa Stenzel Scholarship may be awarded once annually to a person with cystic fibrosis who is, during the period for which the scholarship award is paid, enrolled in a course of higher education leading to a degree granted by an institution in the United States in either health science, social work, mental health science, genetic counseling, or environmental science.

Application Requirements:

An applicant must submit all of the following items to be considered:

- One essay, not more than 1000 words, separately addressing: (a) the applicant's motivations or inspirations to pursue their course of study; (b) the applicant's goals or plans after completing their course of study, especially those related to the CF community; (c) the applicant's activities and achievements, especially within the CF community; and (d) how this scholarship award, if received, would affect the applicant's ability to pursue the course of study and the effect of not receiving the scholarship award on their ability to pursue the course of study.
- Photograph of the applicant (head and shoulders).
- Resume or curriculum vitae.
- Evidence of enrollment in a degree course (either an associate's degree, a bachelor's degree, a master's degree or a doctorate) in a U.S. degree-granting institution of higher learning in either: health science; social work; mental health science, genetic counseling, or environmental science.
- Evidence of a diagnosis of cystic fibrosis (such as a statement from a physician confirming the diagnosis).

The application and further details, including the deadline, can be found on our website. ▲

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infants, so to prevent the loss of nutrient absorption, those with CF should not be fed with frozen-thawed breast milk.

http://tinyurl.com/yc59kw6p

When Is *Burkholderia Cepacia Complex* Truly Eradicated In Adults With Cystic Fibrosis? A 20-Year Follow Up Study.

Burkholderia cepacia complex (BCC) infection in cystic fibrosis is associated with increased morbidity and mortality. Current UK guidance recommends segregation of people with CF according to infection status. To date there is no universally agreed consensus on the number of negative samples or time interval since last isolation of *BCC* for eradication to be deemed successful. All

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Scholarships Offered By USACFA

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

The Stenzel Scholarship (03/30/24):

The Ana and Isa Stenzel Scholarship may be awarded once annually to a person with cystic fibrosis who is, during the period for which the scholarship award is paid, enrolled in a course of higher education leading to a degree granted by an institution in the United States in either health science, social work, mental health science, genetic counseling, or environmental science.

The Stenzel Scholarship was established in 2023 in memory of two amazing women with CF, Isa Stenzel Byrnes and Ana Stenzel. Isa was a licensed social worker and had a Master's of Public Health. She imparted her wisdom to *CF Roundtable* readers for 17 years in her *CF Roundtable* column, "Spirit Medicine." Ana Stenzel was Isa's twin sister. Ana was a genetic counselor at Stanford hospital for 16 years. Isa and Ana dedicated their lives to helping others. They provided education, hope and comfort to thousands of people throughout their lives. They showed the world that people with CF could find meaning in their lives by making a difference in the lives of others.

William Coon Jr. Scholarship (04/30/24):

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

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

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

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

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

Higher Education Scholarship (06/30/24):

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

Any student seeking a degree in higher education, from associate's to doctorate, is welcome to apply. We look for students who demonstrate tremendous academic achievement, community involvement, and a powerful understanding of how their CF—matched with these achievements—places them in a unique situation to gain leadership roles within the community.

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

WE HAVE A NEW ADDRESS! PLEASE TAKE NOTE...

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Please email us with your subscription changes: cfroundtable@usacfa.org

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WI 24

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To keep our records up to date, please be sure to complete and return a subscription form, on this page, to us or register online with any changes to your information, www.cfroundtable.com. (Any issue of the newsletter that is returned to us costs us about two-and-a-half times the first-class postage rate for that piece. Currently that runs about $3.73 per returned copy.) **Thank you for helping us with this.**

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cases of new *BCC* isolation at Manchester Adult Cystic Fibrosis Centre were followed-up between May 2002–May 2022. The number of subsequent positive and negative sputum samples for *BCC* were recorded, as well as eradication treatment received. Eradication was deemed successful if there were ≥3 negative sputum samples and no further positive sputum samples for the same species and strain ≥12 months until the end of follow-up. Of 46 new *BCC* isolation, 25 were successfully eradicated and 21 resulted in chronic infection. A cautious approach to segregation should be maintained after new isolation of *BCC* in CF, as some individuals with ≥3 negative samples 12–24 months after initial isolation had subsequent sputum samples culture-positive for *BCC*.
http://tinyurl.com/mvwy4fe6 ▲

Aimee Lecointre is 38 and has CF. She lives in Salt Lake City, UT. She loves reading, cooking, writing, and spending time with her husband.

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