CF Roundtable®
A NEWSLETTER FOR ADULTS WHO HAVE CYSTIC FIBROSIS
WINTER 2022

Disabled In The Time of COVID-19

By Katherine Lockwood, M.Ed.

The COVID-19 pandemic has changed everything—the price of used cars, shortages of both food and household supplies, time spent at home, and how and where we work.

I have cystic fibrosis and while I'm relatively healthy—thanks to my excellent medical care, health insurance, access to medications, and adherence to my health routine—the pandemic poses a real risk to my life. In the beginning of the pandemic, the school districts in Massachusetts allowed everyone to work from home. I worked safely from home. In year two of schooling during the pandemic, my employer, a public school district, allowed me and other employees with health concerns, or those who were expecting additions to their families, to continue working remotely. It was a very productive year and I supported students who were in the building and at home. I was grateful to be able to prioritize my health while working as a school adjustment counselor with students and staff in likely the greatest global mental health challenge of our lifetime.

At the end of the last school year, the pandemic appeared to be waning, vaccines were available, and I looked forward to returning to normal life and to the physical workplace in the fall of 2021. Unfortunately, I was being too optimistic—my doctor advised me two weeks before the start of the school year that he didn't recommend me returning in person as the Delta variant was raging and it wasn't safe for me or, as I found out a couple weeks later, for the little one starting to grow inside me. I put in a request to continue working remotely.


KATHERINE LOCKWOOD

I received a letter back immediately saying that students were attending school in person and they therefore needed me in person as well. My accommodation request was denied. I met with my Superintendent and tried to explain my situation and how I could effectively serve my role remotely. Nothing I said mattered—the district had decided that no one could work remotely and that was that. She told me that their lawyer said they didn't have to allow me to work remotely so it was a no. Oddly, she didn't disagree that it was unsafe for me, nor did she recommend any additional or alternative accommodations.

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CF ROUNDTABLE
FOUNDED 1990
Vol. XXXII, No. 1

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

EDITOR'S NOTES

Here in Texas, while I'm writing this issue's notes, today's cold winter weather I once used to love now brings back memories of last year's "Snowvid-21" and with that an accompanying dread. At the same time, I'm also happy that my mom passed away before that—she would have been miserable, to say the least, and getting her to a hospital for the oxygen she needed would have been impossible. I also feel guilty even having those thoughts because, naturally, I wish she were still alive. Over the course of the last 18 months since she passed away, I've often wondered if that cold I gave her at Thanksgiving, the one she never recovered from, was the straw that broke the camel's back. Would she have lived longer? I'll never know but somewhere I do logically recognize that, at the end of the day, she had COPD so her days were numbered, regardless of that cold I gave her. In that vein, the focus topic this issue—survivor's guilt—is not only timely, but especially relevant as we watch the median age of survival inch higher with the advent of newer and better genetic modulators. **Tré LaRosa**, writes about the grief and suffering surrounding the death of his sister, who also had CF, and how he has finally stopped blaming himself for surviving. **Andrea Eisenman** shares the conflicting emotions of gratitude for the gift of life and the ensuing sadness at the life lost in exchange for that gift, both of which she experienced after her double-lung transplant. **Julie Desch** weighs in on the basis and meaning of the word guilt and how the notion of survivor's guilt centers around the need to assign blame.

In this issue, you can read about the latest CF research in **Laura Tillman**'s expertly collated "From the Internet" column. **Xan** interviews Raven Aragon about her experiences as a transsex person with CF in our "In The Spotlight" column. In their own column, Dr. Nowakowski also addresses the use of survivor's guilt as a reason to take care of others with chronic illnesses in lieu of self-care and, even more importantly, what has worked for them in breaking that pattern. I know I'm guilty of those patterns in my own life and working through them, for me at least, is an ongoing process. In our "Ask the Attorney" column, **Beth Sufian** explains the ins and outs of the Social Security work incentive program called §1619(b). **Isabel Stenzel Byrnes** talks about her experiences as a grief counselor and how grief and guilt go hand in hand. **Cindy Baldwin** writes about survivor's guilt in the context of parenting, while **Mark Tremblay** shares what has worked for him as he has worked through survivor's guilt over the years. We also bid adieu to **Lara Govendo**, who advises us to do more of what brings us joy in her final column.

We're ramping back up with our Speakers Bureau in 2022 and don't forget to apply for our Arts Scholarship. Details are on pages **12** and **23**, respectively. 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 - AbbVie, Arrowhead Pharmaceuticals, Maxor Specialty Pharmacy, Monaghan Medical, Nestlé Health Science, and Viatris; Pearl Sustaining Partners - Boomer Esiason Foundation, Cystic Fibrosis Foundation, Gretchen Van Bloom Budig in memory of Sydna Marshall, and Scholarship for the Arts in memory of Helen M. Eisenman; Diamond Sustaining Partner: Marina Day, Trustee of the McComb Foundation; Endowment Partners - Carroll Groeger, Trustee for Paul Schnackenberg Trust, Nancy Wech (in memory of daughter, Lauren Melissa Kelly & in honor of son, Scott Kelly).

Information From The Internet...

Compiled by Laura Tillman

Cystic Fibrosis(CF): Life Expectancy

In the 1940s, most babies born with CF died in infancy. Today, people living with CF can expect to live into their 40s. Of those already over 30, half are expected to live into their early 50s. lifespan. The above statistics are the expected lifespan for a population, not for an individual. They are also based on people who were born decades ago, when many therapies currently in use were unavailable. A given person's life expectancy may be higher or lower than these numbers, depending on their overall health, specific CF mutations, and responses to treatment, as well as other factors. Babies born with CF in the last five years have a median survival age of 48—meaning half are expected to live to age 48 years or older. If treatments continue to improve at their current pace, at least half the babies born with CF today will live into their 50s and 60s. Men with CF have a median survival age that is two to six years greater than women with CF. Why men with CF tend to survive longer than women is not fully understood. One theory is that estrogen, a hormone women have in higher amounts starting in puberty, plays a role. Estrogen worsens mucus dehydration in the lungs and impairs the immune system's ability to clear common lung infections like *Pseudomonas aeruginosa*. Women with CF may also experience higher levels of lung inflammation. People with a single copy of the F508del mutation have a higher median survival age than those who carry two copies. Cystic fibrosis is most common in people of European ancestry, and less common in those of Asian, African, or Hispanic ancestry. However, Hispanics living in the United States have a lower median survival age than do people of European ancestry. An individual's lung function, as assessed by their mean baseline forced

LAURA TILLMAN

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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/newsletter.

Winter (February) 2022: CF and Survivor's Guilt. (Current issue)
Spring (May) 2022: Aging and CF. How has aging changed for you with the advances in CF drugs? Where and how are you getting care for issues arising out of aging rather than CF? As people with CF are living longer, what other issues besides pulmonary, GI, and sinusitis are you facing? Whom do you see for these issues and how is your care coordinated between doctors? Do you feel heard at your clinic? How proactive is your clinic in treating and early prevention of common issues with aging?
Summer (August) 2022: Dealing With Pain. What types of pain do you deal with regularly? What has helped you with pain management? How does pain affect your quality of life? What alternative therapies have helped manage your pain? What, if any, problems do you have with tolerance to medications or addiction and how are you dealing with it? What advice do you give to someone dealing with addiction to pain medications?
Autumn (November) 2022: Transitioning from Pediatric to Adult Care.

ASK THE ATTORNEY

Working While Receiving Social Security SSI Benefits

By Beth Sufian, J.D.

Some people with cystic fibrosis who are currently not working but receiving disability benefits want to work more than the allowable amount of less than 20 hours a week and make more work earnings than the Social Security rules allow. Yet they do not want to lose the Medicaid coverage they receive because they are eligible for Supplemental Security Income (SSI) benefits. This issue's column will provide information on a Social Security work incentive program called Section 1619(b), which is a program that encourages beneficiaries who are able to work more than 20 hours a week to undertake a work attempt while retaining their Medicaid benefit.

Section 1619(b) of the Social Security Act applies only to individuals receiving SSI benefits. The 1619(b) rules DO NOT apply to recipients of SSDI or Medicare. There is a separate set of rules that applies to SSDI recipients who have Medicare and this will be discussed in the second part of this series that will appear in the next edition of *CF Roundtable*.

Nothing in this column is meant to be legal advice and is only meant to be legal information. If a reader has a question about the information provided in this column, the reader can contact the CF Legal Information Hotline at CFLegal@sufianpassamano.com or call 1-800-622-0385. All contacts are confidential and there is no cost to call. The CF Legal Information Hotline is funded by the CF Foundation, but staff are not employees of the CF Foundation.

Continued Medicaid Eligibility Under §1619(b).

Some Supplemental Security Income (SSI) beneficiaries want to return to work after they are determined to be disabled by the Social Security Administration and after they begin receiving SSI benefits. Section 1619(b) of the Social Security Act (42 U.S.C. 1382h(b)) provides a way for individuals to remain eligible for Medicaid even when they engage in substantial gainful activity despite having a severe medical impairment. Section 1619(b) is intended to encourage SSI beneficiaries to return to work and allow them to retain Medicaid benefits, although the monthly cash SSI benefit will stop.

> *"Section 1619(b) is intended to encourage SSI beneficiaries to return to work and allow them to retain Medicaid benefits, although the monthly cash SSI benefit will stop."*

BETH SUFIAN

A. How Income Affects SSI Benefits.

Before looking at the details of §1619(b), it is helpful to see how income affects SSI eligibility to understand the benefits of the §1619(b) program.

A typical SSI beneficiary receives a monthly cash benefit and Medicaid. The maximum federal SSI monthly amount in 2022 is $841 per month. Some states add a state supplement to the federal amount. However, the SSA may reduce the monthly cash benefit based on the beneficiary's living arrangement. Also, the SSA will reduce the monthly cash benefit for each dollar of other income the beneficiary receives.

Unearned income reduces the SSI benefit by a 1:1 ratio—each dollar of unearned income offsets the benefit by one dollar. Earned income reduces the benefit by a 2:1 ratio—every two dollars of earned income offsets the benefit by one dollar. To maintain the Medicaid benefit, the beneficiary must have at least one dollar of SSI monthly cash benefit. If the monthly cash benefit is offset and reduced to zero, eligibility for SSI ends.

Many people who look at the Social Security website and publica-

tions will see a reference by Social Security to an amount of $1,310, which is considered substantial gainful activity and the maximum allowable monthly earnings from work activity. Many people incorrectly conclude that a beneficiary can retain their SSI benefits as long as their earned income is less than $1,310 per month.

What matters is the individual's monthly benefit amount and whether it is offset by income. For example, if a beneficiary received a monthly SSI cash benefit of $524 and had earned income of $1,050 per month, then the individual's monthly benefit would be reduced to zero by a $525 offset, since every two dollars of earned income offsets the benefit by one dollar. The result is the individual would become ineligible for SSI and lose both the monthly cash payment and Medicaid, even though the monthly earnings were less than $1,310.

However, an SSI beneficiary who is working may avoid the loss of SSI benefits due to earnings if the beneficiary enrolls in the Section 1619(b) program. By enrolling in §1619(b), the beneficiary's Medicaid eligibility will continue, and the monthly SSI cash benefit will stop. There are limits to the amount of income a Section 1619(b) beneficiary can earn and still retain Medicaid eligibility. However, the earning limit for an SSI beneficiary enrolled in Section 1619(b) is higher than the allowable earnings for an SSI beneficiary not enrolled in Section 1619(b).

B. Section 1619(b) Is NOT Automatic.

An SSI beneficiary must request enrollment in Section 1619(b) when returning to work and must make this request before the Social Security Administration suspends or terminates SSI eligibility for exceeding the monthly income limits.

If SSA suspends or terminates benefits because income or assets exceed the limits, then the individual loses SSI eligibility—both the cash benefit and the Medicaid eligibility. However, if SSA stops monthly payments because of Section 1619(b) enrollment, then the cash benefit stops but the Medicaid may continue if the individual otherwise qualifies.

C. Section 1619(b) Eligibility.

1. The beneficiary must be receiving SSI benefits.

To enroll in continued Medicaid eligibility under Section 1619(b), the individual must, of course, be eligible for SSI. That means the beneficiary must have received at least one month of an SSI monthly cash benefit, and the beneficiary must have been eligible for Medicaid coverage in the month before the beneficiary became eligible under Section 1619(b).

2. The beneficiary must continue to be disabled under Social Security rules.

The individual must also continue to have the disabling impairment on which the individual was found to be disabled by Social Security under its rules. Being disabled by cystic fibrosis under Social Security rules is not the same as having a diagnosis of cystic fibrosis. Social Security rules require specific symptoms or combinations of symptoms that are clinically documented for the person to be considered disabled by Social Security.

This can sometimes be a narrow path because often the degree of improved health that enables the beneficiary to work may also form the basis for Social Security to find the individual is no longer disabled. Social Security generally is permitted to reassess a beneficiary's health status to determine if the beneficiary continues to be disabled. In fact, the Social Security Administration directs its field office to reassess the disability status of any SSI beneficiary who applies for Section 1619(b) within 12 months after the disability onset date determined by Social Security. See POMS SI-02302.010; DI 13010.110

3. The beneficiary must need Medicaid benefits to maintain employment.

Also, the continuation of Medicaid benefits must be necessary for the individual to maintain employment. Generally, this means that the individual's earnings from work are not sufficient to replace SSI, Medicaid, and any publicly funded attendant care services. If the individual is earning less than the maximum allowed under Section 1619(b), this requirement is typically met.

4. Earnings from employment must be less than the maximum allowable annual amount.

Once an individual becomes eligible to participate in §1619(b), they may retain Medicaid eligibility even if they earn an amount greater than their monthly SSI cash benefit and more than $1,310/month, which is usually considered substantial gainful activity. Although SSI beneficiaries may earn more than these amounts while enrolled in §1619(b), their earnings must still be below the §1619(b) earnings "threshold." The earnings threshold is the amount of earned income that is sufficient to replace SSI and Medicaid (including publicly funded attendant care services, if any) in the state where the beneficiary resides. The threshold amount varies by state and the threshold amount in each state may be found on the Social Security Administration website at: www.ssa-gov/disabilityresearch/wi/1619b.htm

5. The beneficiary must remain within the SSI resource limit.

To be eligible for 1619(b), the beneficiary must also meet the SSI financial resource requirements. The financial eligibility requirements count both current income and other financial resources. If a beneficiary exceeds the

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limits, the person will no longer be eligible for SSI.

Although §1619(b) income limits are higher than ordinary SSI, the resource limit is not higher. SSI is a needs-based program for individuals who have countable resources worth not more than $2,000 for an individual or $3,000 for an individual and spouse. The countable resource limit does not increase if a beneficiary enrolls in Section 1619(b).

Countable resources are the things the individual owns that count toward the resource limit, such as cash, bank deposits, stocks, savings bonds, land, insurance with a cash value, personal property, vehicles, and anything else that could be exchanged for cash. Some resources owned by others can be deemed by Social Security as a resource available to the individual.

Some assets are not counted as a resource by Social Security. For example, the value of a house if used as the beneficiary's primary residence is not counted as a resource. Also, the value of one vehicle is not counted as a resource, but the value of any vehicles other than the first one are counted as a resource.

D. Meet All Other SSI Eligibility Rules.

There are other eligibility rules for Section 1619(b) and for continued SSI eligibility that are not mentioned here, and a beneficiary must continue to be eligible for SSI to participate in 1619(b). ▲

Beth Sufian is 55 years old and has CF. She is an attorney who focuses her law practice on disability law and is the Treasurer of USACFA. Her contact information is on page 2. You may contact her with your legal questions about CF-related issues at CFLegal@sufianpassamano.com.

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expiratory volume in one second (FEV%), is an important predictor of survival and their need for a lung transplant. Health problems arising from cystic fibrosis, such as poor nutritional status, liver failure, and diabetes, can shorten a person's lifespan. Lung infections worsen lung function in CF and contribute to early mortality. Common respiratory infections—such as those caused by *Pseudomonas aeruginosa* and *Burkholderia cepacia*—can cause severe lung damage in people with CF.
https://tinyurl.com/mw7ha3he

Promising Drugs In Clinical Development For Cystic Fibrosis

GlobalData focused its cystic fibrosis (CF) market forecast on five late-stage Phase II products expected to be licensed for the disease between last year and 2030. These include two transmembrane conductance regulator (CFTR) modulator regimens from Vertex Pharmaceuticals, VX-561 (deutivacaftor) alone and in a triple combination with VX-121 and tezacaftor; two CFTR modulator regimens from AbbVie, ABBV-3067 alone and in combination with galicaftor (ABBV-2222); and an anti-inflammatory agent in development by Laurent Pharmaceuticals, LAU-7b (fenretinide). Vertex has completed one Phase II study (NCT03912233) of its next-generation triple combination therapy, VX-121 + tezacaftor + VX-561 (deutivacaftor), in CF patients aged 18 years and older, with either one F508del mutation and one minimal function CFTR mutation or two F508del mutations. The regimen was reported to be well-tolerated and achieved the primary goal of improved lung function and the secondary goal of reducing sweat chloride. Data from the Phase II dose-ranging study of VX-561 (deutivacaftor) alone are due to be reported in the near future. Two Phase III trials are now being planned to evaluate VX-121 + tezacaftor + VX-561 in CF patients aged 12 years and older. The main goal of both Phase III studies is to achieve lung function improvements similar to or superior to Trikafta/Kaftrio. AbbVie's ABBV-3067, a CFTR potentiator, alone and in combination with ABBV-2222, a CFTR corrector, is being studied to target CF patients who are homozygous for the F508del mutation. Since these pipeline agents are not expected to treat new mutations or greatly improve efficacy or safety, it is likely that these drugs will mainly serve as alternatives to currently marketed CFTR modulator drugs. However, they may be useful options for patients who develop intolerances or allergies or experience adverse events in response to established products. In addition, further competition could help to drive down drug prices to the benefit of consumers. Laurent Pharmaceuticals' LAU-7b (fenretinide) is expected to be the first approved anti-inflammatory agent for CF. The ongoing late-stage Phase II APPLAUD trial (NCT03265288) is evaluating the safety and efficacy of LAU-7b for the treatment of CF patients aged 18 years and older. Prospects for this drug, however, remain uncertain. The drug would need to compete with other anti-inflammatory drugs recommended by guidelines, including azithromycin, ibuprofen and prednisolone, all of which have a well-established reputation, commercially and clinically, and lower cost.
https://tinyurl.com/yzn6n9fc

Better Understanding Of Cystic Fibrosis

A research team found that sodium transport is abnormal in lungs with CF. The researchers studied the swine model of CF and used a specialized microelectrode technique that allowed them to perform experiments with very high resolution. They discovered there is excessive sodium absorption in the small airways, a previously unstudied site in the body.
https://tinyurl.com/5n7fubyp

Women With Cystic Fibrosis Have

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To add insult to injury, the district also neglected to give me my sick leave days at the beginning of the year and, only at the prompting by the union, did they find the error. They then denied my request for time from the sick bank. Unfortunately, they have decided to discard me because I have an illness and have deemed me, at least for the time being, to be professionally worthless.

There is a Federal Law, the Americans with Disabilities Act, which is supposed to protect people like me. People who have a disability, who can work, but also need an accommodation to do so. The key pieces of the law relative to my situation are:

1. The ADA requires that the employer has at least 15 employees. YES! My employer meets that requirement.

2. The disability is a physical or mental impairment that substantially limits one or more major life activities. YES! Breathing qualifies.

3. The employee can perform the essential functions of the job with an accommodation. YES! I can perform all parts of my job via Zoom or Google Meet and I did so for a year and a half.

While this is financially devastating to my family, we have no choice but for me to stay safe. My school district initially offered me a year of unpaid leave with the guarantee that I could remain on the district's insurance. However, I was expected to pay the full cost of the premiums, including the portion the school normally covered for their employees. That privilege costs about $2,200 monthly. How can anyone afford to both lose their income and pay that much money to keep their insurance? We cannot. When I respectfully let them know that they were in violation of my rights under the ADA, I received multiple notices pressuring me to resign.

The pandemic has caused "The Great Resignation" and the ensuing disproportionate effects on moms. Over 3.5 million American mothers have left the workforce. Some good will come of this—people are leaving unsafe work environments where they are poorly paid for better work opportunities. People are reevaluating their priorities and shifting some of the focus from career to home and family. However, many (disproportionately women) will not be able to return to the workplace for many years, and their opportunities for career growth and compensation will be negatively impacted when they do return.

People with disabilities are more likely to have become unemployed due to the pandemic. People with disabilities who lose their jobs may be less likely to return to work. More may become newly eligible for or choose to file disability benefits and seek support from social services. People with disabilities face barriers to remaining at work. People like me with disabilities are more likely to have an underlying medical condition that puts them at increased risk of severe illness from COVID-19. I wonder how many others are in a similar situation.

I work in education as an adjustment counselor; 17.1% of students in 2020 in my school district have a disability and we are charged with preparing them for life after K-12. We create 504 plans and IEPs and tell their families (and ourselves) that we are preparing them for a bright future full of opportunities. We attempt to give them the skills and tools necessary to be productive members of our society. I find it hypocritical that this same educational system thinks that staff with disabilities are professionally worthless and easily disposable. We pay lip service to the concepts of equity and the importance of diversity. I am a young, white, middle-class (usually) woman living in Massachusetts. I am highly educated with two masters degrees and have spent years working at the Cape Organization for Rights of the Disabled, advocating for individuals and students with disabilities in the areas of housing, access, equity, employment, education, and social services. These experiences made me fully aware of my employment rights when this situation occurred. Personal discrimination is a new experience for me and being discriminated against because I have a disability feels terrible, is demeaning, and is illegal.

Most have entered a new phase of this pandemic where they are returning to a new normal. Some people, like me, are not able to do that yet. Our family has done everything possible to both stay safe and to also protect others. While never testing positive for COVID-19, we have been disproportionately negatively affected during the pandemic and we will continue to be so affected. We are trying to survive, as I am sure many others are.

I spent my career thus far envisioning a future where we create employment opportunities with inclusion and equity, where a diverse workforce with true representation is an actuality, and the ability to stay working is valued equally to obtaining employment. For that to happen, employers need to look critically at their practices and values and operate equitably and ethically. I have recently been denied extended unpaid medical leave and received an intent-to-terminate letter from the school district due to a claim of incapacity. My disability has not incapacitated me; my employer has. In the meantime, if you are in a similar situation or know of an awesome place to work remotely, please let me know. I'd love to connect with others going through these same battles. ▲

Read the Spring 2022 issue of CF Roundtable *for updates to Katie's story.*

Katherine Lockwood is 33 years old and has CF. She lives in Massachusetts with her husband Arden and their daughter Rose, with another daughter, Magnolia, on the way. When not surviving a pandemic, you can find them swimming, boating, renovating, drinking champagne, and eating potluck dinners with friends. Katherine has spent this time out of work crafting a children's book about differences. To read more about her project visit https://www.acorncottagepress.com/

SPIRIT MEDICINE

The Spirituality Of Guilt

By Isa Stenzel Byrnes

In this article, I'd like to ponder all kinds of guilt, not just survivor's guilt. Guilt is a prosocial emotion and burden for every conscious human being. How we carry guilt and what we do with it has a profound impact on our psychological and spiritual lives. I have a very close bond with guilt. It is drilled into me by my Japanese culture. I feel bad for my privilege, for burdening others, for needing attention, for hurting the environment with all my medical waste. You name it, I've got it. Most of the time, I look the other way, or I try to let it go.

As I have started losing friends to CF, I have often struggled with guilt about whether I was a good enough friend. I think to myself: *I should've called them more; I should've sent a gift or card; or I should've visited them and brought a casserole*. I feel guilty that I was too busy dealing with my own issues to pay attention to someone else's struggles.

The timing of the topic of survivor's guilt is especially pertinent to me. By the time you are reading this, I've celebrated my entry into my sixth decade of life. This is a milestone I never imagined that I could reach. While I celebrate this miracle, I leave behind my twin sister who barely experienced her forties. My birthday will always come with a sting of survivor's guilt. Hundreds of my CF peers never came close to this milestone. Why am I still here?

On an intellectual level, I've denied that I've had survivor's guilt for years. It's not my fault my friends died earlier. I couldn't control Ana's cancer. Everyone has their story. And I have a mantra: "guilt implies intent." This was never meant to be a competition for the survival of the fittest. If I didn't intend to outlive so many, then I don't need to feel guilty.

But the intellect and the heart and spirit are not always congruent. This is the hallmark of internal conflict. If I pause and be still, and allow the emotions to arise, I feel it. I carry survivor's guilt with such a profound weight. It seems so unfair that I've been given more time than so many of my friends with CF and my organ donor. It makes me sob.

> *"Atonement is a release of our guilt to a higher power or to a greater good. A spirit of love trumps all things."*

ISABEL STENZEL BYRNES

In my work as a grief counselor, guilt and grief go hand in hand. There is the guilt that one is not grieving hard enough. There is the guilt that one is not coping well enough. Guilt can be a form of self-inflicted suffering by the survivor so they can suffer as much as the person who died. The could've/should've/would've thoughts are incredibly natural in grief. This counterfactual thinking occurs when survivors feel they have responsibility for the death of a loved one. When my sister was dying of cancer, I gave her a very high dose of opiates and she died. Initially I felt guilty that my hands ended her life. But then I returned to my intentions. The dose was to relieve her of suffering; cancer killed her.

I see guilt as an attempt to control. Without control, the universe is truly unpredictable and chaotic. So grieving people often wish they could rewind the tape and "do more" to change the outcome. This tendency is a way to process the death and accept what did happen.

Another way I see guilt is as a form of caregiving. Parents and spouses are especially vulnerable to guilt, because of the intensity of their bond and their roles as protectors. Surviving family members are often the primary caregivers of the dying person. They care because they love the person, and we all have an innate instinct to care for people we love. After the death of a

loved one, we want to keep taking care of the deceased, and our minds trick us into thinking we can still do that—if we had only taken her to the doctor earlier; if I had forced him to do treatments; if we had fed them a healthier diet, etc. Eventually, we learn that this kind of caregiving is futile. We can redirect this caregiving into honoring and remembering the person instead.

Yet, truthfully, sometimes guilt is justified. Sometimes our actions do deliberately hurt someone. Yelling at the insurance guy because my insurance denied another medication makes me guilty for blaming him. Feeling guilty after driving drunk and killing someone is justified. Guilt after suicide is intensely real when you could've helped a friend who expressed thoughts of self-harm.

There are many ways to cope with grief-related guilt. We can write letters or talk to an empty chair or photo of the person who died. We can sit at the cemetery and express our apologies. This act of conscience is a gift to ourselves to relieve this burden. We can also share our feelings with others who understand us and ask them to be a mirror to us. Other people can help us see if the perceived wrongdoing is, in fact, real.

Guilt can be spiritually toxic. In fact, Buddhists regard guilt as a negative emotion that hinders spiritual growth. Guilt can lead to dangerous beliefs as a person such as "I'm a horrible person," or "I am unforgivable. It's my fault. I'm inadequate/weak." In the short term, some of this can be humbling or awakening, but in the long term, these beliefs will damage our souls. A spiritual practice can help us cope with guilt. Over time, it's important to convert dangerous beliefs to spiritually healthier approaches such as "I did the best I could. I can forgive myself and move on. I am okay despite my mistakes."

Many religions use guilt to control our human nature, often with threat and fear. Guilt gives us moral culpability and controls. As a Japanese person, I think my cultural guilt is only topped by Jewish and Catholic guilt! But seriously, religious practices can absolve us of guilt. They ask us to stand before God and repent. Our spirituality can highlight our intentions and give us perspective. It can help us see that our acts or lack of acts are so small in the grand scheme of things. Atonement is a release of our guilt to a higher power or to a greater good. A spirit of love trumps all things.

A spiritual life invites us to act on our guilt. We can do something with it. We can do good deeds to redeem a mistake; we can pay it forward and help others; we can turn to God. And we can learn to forgive ourselves. That doesn't mean condoning a behavior that gave us guilt and forget what happened, but, rather, accept our human actions with regret and make a commitment to live well going forward. In other words, guilt can play a powerful, spiritual role in our lives: to motivate us to follow a moral compass and to live the best life we can do. This facilitation of integrity is perhaps why a low dose of guilt can serve us well.

Living with CF is hard enough. I do not want to inflict any more negative energy into my life with guilt. I look for the teachings of guilt. It keeps me in check and keeps me from boasting. I express gratitude and glow in the awe of all I've received in this one precious life. I know how fortunate I am compared to so many others with CF. I don't need to punish myself for my blessings. ▲

Isa Stenzel Byrnes is 50 years old and has CF. She lives in Redwood City, California. She is 17 years post-lung transplant.

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Higher Rates Of Pulmonary Exacerbations

Women with cystic fibrosis have higher rates of pulmonary exacerbations compared with men, with differences appearing early in life. Researchers conducted a retrospective cohort study utilizing data from the Cystic Fibrosis Foundation Patient Registry from 2006 to 2019. Among 40,782 individuals with cystic fibrosis, the mean age was 14.6 years and 48% were women. Researchers evaluated the association of sex with the number of annual pulmonary exacerbations requiring IV antibiotics, both overall and among subgroups defined by nonmodifiable characteristics. Women with cystic fibrosis experienced 0.18 more pulmonary exacerbations per year compared with men, for a 29% higher relative rate of pulmonary exacerbations. In addition, pulmonary exacerbation rates were higher within subgroups stratified by age, race, ethnicity, delF508 mutation and CFTR functional class. Identifying subgroups at highest risk for pulmonary exacerbations is a high priority among the cystic fibrosis community.
https://tinyurl.com/y69reo37

Urinary Incontinence Can Rise As Women Age, Study Finds

Urinary incontinence, known to affect both girls and women with cystic fibrosis, is increasingly prevalent as patients age. Estimates of incontinence among women with CF range from 30% to 76%, but its exact causes are not clear. CF patients who reported urinary incontinence in this study were older on average and had many of the main characteristics of CF, such as pancreatic insufficiency, chronic colonization by *Pseudomonas aeruginosa*, and the presence

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FAMILY MATTERS

Carrying Their Legacy

By Cindy Baldwin

The topic of survivor's guilt is a weighty one when you grow up with a disease like cystic fibrosis. Nearly all of us have experienced death more frequently and closely than an average person as we watched CF friends succumb to disease progression, organ transplant rejection, and sometimes even out-of-the-blue complications like a pulmonary embolism or pneumothorax. We know the complex blend of emotions that come with these deaths—grief laced through with guilt that the often-random hand of fate came for our loved one and not for us; anxiety about our own condition; and gratitude for our relative health.

When it comes to parenting with CF, there are two types of survivor's guilt I struggle with: the deep stab of pain that comes when a fellow CF parent loses their life and leaves their children without them, and the dull ache when beloved CF friends who wish for parenthood are unable to achieve it—whether due to health status, infertility, or financial constraints. Some days, it's hard to look at my own beautiful eight-year-old daughter without being overwhelmed by this guilt.

Several years ago, I learned, to my complete shock, that Kristi, one of my closest friends with CF, had died of an unidentified pulmonary embolism. One of the things that had connected us was the fact that our daughters were almost exactly the same age, and that both of us had been in the first wave of women to conceive after starting Kalydeco in 2012. We loved comparing notes on our "Kalydeco twins" as they grew up, sharing photos and stories and sometimes even dressing our babies in matching outfits. For days, every time I looked at my daughter (then three years old), I thought about Kristi's own three-year-old, now growing up without her mother. I wondered over and over again what it would be like for *my* daughter if she were to lose me in such an unexpected way. I felt swallowed by my own guilt about being able to share small moments of beauty—snuggling together to read books, taking walks in the spring sunshine—with my toddler, when Kristi was no longer with hers.

As time passed, my grief became less acute. But even five years later, I think often of Kristi and her daughter. These days, when the two of them come to mind, I try to send a wish into the universe for her little girl—that, even though her mother is not with her, she might be surrounded by people who love and cherish her just like her mama did. And on those days, I hug my own daughter extra tight, determined to help her grow up understanding what a precious gift life is. In some small way, teaching my daughter to dwell in gratitude for the beautiful world and its blessings feels like a way to honor the legacy of my CF friends who do not get to raise their own children. When we do not take our lives for granted, but rather make time to recognize and give thanks for the many gifts life brings us, we are affirming the value of every life—whether it be long or short.

> *"When I know a friend is struggling with childlessness, it's easy to feel guilty that I* **was** *able to achieve my dream of parenthood."*

CINDY BALDWIN

There is also survivor's guilt that comes along with having been able to successfully bring a child into the world when so many would-be parents with cystic fibrosis aren't able to, whether because their health status prevents it or because of insurmountable issues with fertility or finances. Because it took me several years to get pregnant, I'm acutely familiar with the intense sorrow that comes from longing for a child without being able to have one. Even almost a decade after my own pregnancy, I remember the anguish of my infertile years with vivid clarity. When I know a friend is struggling with childlessness, it's easy to feel guilty that I *was* able to achieve my dream of parenthood.

Regardless of the reason for my survivor's guilt, I try to give myself time to work through the complicated feelings it stirs up. First, I allow myself to recognize that usually, this guilt is a manifestation of sorrow or grief—

whether because of the death of a beloved friend, or because of the understanding that they are in pain. For me, at least, I need to let myself feel and acknowledge that sorrow before I can move forward.

Next, I focus on gratitude and appreciation for the blessings that I have—recognizing that the happy moments in my life do not take away from or diminish anyone else's, and that no friends who love me would wish for me to deny the good things in my life in a misplaced attempt to make them feel better. I do, however, work hard to learn and respect any boundaries that a loved one might create—for instance, if I know that a friend who's struggling with infertility is triggered by talking about kids, I'll refrain from conversation focused on my daughter.

And finally, just as I've done over the past five years since my friend Kristi died, I try to help my daughter learn to feel and express gratitude for the blessings in her life. In addition, I work hard to make sure she knows that the world is filled with many kinds of people and many kinds of families—and that there are many ways to live a "good" life. In teaching her to respect others' differences, I hope that she may grow up to be a person who will create safe spaces in *her* life for the motherless children, childless adults, and unconventional families *she* comes to know. When I emphasize to her the importance of respecting and loving others, it feels as though I am carrying the legacy of the friends I love who have lost their lives or lost their dreams—working to make the world my daughter will inherit a kinder, gentler place for those who have experienced great sorrow. ▲

Cindy Baldwin is 32 years old and has CF. She lives near Portland, Oregon, with her husband and daughter. Cindy is the author of several middle grade books published by HarperCollins, including The Stars of Whistling Ridge, *which released in June 2021.*

FOCUS TOPIC

CF AND SURVIVOR'S GUILT

Is Survivor Guilt Sane?

By Julie Desch, M.D.

My body has long surpassed its expiration date, so when I see that a young kid (or anyone, frankly) has died from cystic fibrosis as I continue to enjoy my family, friends, and health, I cringe. It sets off a cascade of emotions that has been formed over the years of living with the fact that two of my siblings died decades before the idea of a "CFTR modulator" existed.

It sucks to outlive a sibling. It super-sucks to outlive a sibling who died of CF. And it super-sucks squared if this happens twice. But at the same time, as my father used to say, it beats the alternative.

It seems like the emotion of guilt is an inevitable consequence when we lose our friends or family members who have CF. Of course, we "should" feel guilty, right? It really never dawned on me to question the validity of this guilt. It seemed like a no-brainer type of reflexive feeling, over which I had no control.

JULIE DESCH

Until fairly recently, it seemed like something with which I just had to live.

But the emotion of guilt is not pleasant. If we had the ability to choose an emotion to feel, guilt would never be at the top of the list. So, in a sense, I have allowed myself to kick my own ass for decades without actually examining the basis for the emotion itself.

The basis for guilt is the thought, "I did something wrong" or, even worse, "I am intrinsically wrong." Clearly, being born into a family with other children with CF was not something I actually "did." Outliving another human is not something I "do." It just happens over and over until it stops happening. That two of those humans I have outlived happened to be my siblings is not something over which I had control. Logically, all of this makes sense.

This is what therapists have told me, friends told me, my parents told me, even my deceased siblings themselves told me. At some point, I finally began telling myself these things. It makes absolute sense and I cannot find a single objection to the obvious rationale.

CF Roundtable Speakers Bureau

As 2022 has begun, we are looking to ramp up our event schedule for future talks. The Speakers Bureau was developed to provide a free service to the CF Community by sharing the voices and personal perspectives of adults living with cystic fibrosis through in-person or virtual speaking events. We have a wonderful group of speakers who talk on a large range of topics, such as daily life with CF, finding a balance of work/college/family, mental health, clinical trials, and how CF affects sexuality. We are proud to sponsor our speakers for your group, fundraiser, community education, conference, and other events for those who are looking to understand the patient experience better. There are no costs for the hosting organization. It is our hope to educate audiences by providing seasoned adult speakers who have CF and will share their stories in order to improve the world for the entire CF community.

Please contact us at cfroundtable@usacfa.org if you or your organization would like to request a speaker. You can find more information on our wonderful speakers and topics on our website: https://www.cfroundtable.com/speakers-bureau.

If you have CF and are interested in joining our Bureau, contact us at cfroundtable@usacfa.org!

So why did I still feel so bad for so many years after the logic of this had finally clicked in my head? I felt bad because it felt better to feel bad (guilty) than to admit to the fact that I have no control over life now, that I never controlled life in the past, and that I have no control over life in the future. This initially felt terrifying. The idea of control has provided a sense of security over the years, and to see it as illusory is more than a little unsettling. Accordingly, it feels better to believe in a false sense of control of life and the idea that life "should" be a certain way.

The sense of control over life is part of necessary psychological development. For me, it was the coping mechanism that worked best while growing up in my family. I'm fairly certain that it is a coping mechanism that most people with chronic illness keenly develop. If we didn't feel somewhat in control as young kids who had CF, the stress would have been the end of us. Of course, this doesn't manifest as control over health. We are clearly screwed in that domain. Instead, it sneaks in as control over grades, or work, or fitness, or relationships, or you-name-it. In any arena, it feels good to believe we are at least somewhat in control.

> "Survivor's guilt is what happens when we outlive a loved one and look for someone to blame."

With that in mind, imagine what happens when life throws a really lousy curve ball at you. The first instinct is to find a person, place, or thing to blame. Ascribing blame provides an explanation for what happened and why. We tell ourselves the story of where things went wrong so that we believe that the next time around we can intervene and make life work the way we desire. This provides the sense of control that feels essential.

Survivor's guilt is what happens when we outlive a loved one and look for someone to blame. We look and look, but when there is nobody who did anything wrong, we blame ourselves. The alternative is unacceptable. But can it even make sense that we are in control of a life, either our own or that of another person? What if there is nobody to fault when a death occurs? What if there is nobody to blame because blame makes no sense? What if the fallacy is in believing that we "have" a life, as do others? The way I see it, we don't own a life and we don't lose a life. We *are* life...all of us. Death is not the opposite of life. Death is the opposite of birth. Life has no opposite, and nobody owns it. When we see this clearly, survivor's guilt ends. Grief remains, of course. Grief has to make its way through you, but guilt doesn't need to be part of this. ▲

Julie Desch is 59 years old and lives in San Rafael, CA. She enjoys meditation, reading, writing, exercise of every variety, and hanging out with her partner, two boys, and three dogs. She can be reached at Juliedesch@gmail.com.

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of CF-related diabetes. Compared with previous reports, females with urinary incontinence in this study also had more severe respiratory issues and longer CF-related hospitalizations, suggesting greater disease severity. Neither constipation nor exercise, both of which have previously been linked to urinary incontinence in the general population, were more common in CF patients reporting urinary incontinence. Higher body mass index was associated with a greater risk of incontinence, in line with a reported link between obesity and urinary incontinence in healthy people. The findings also indicated that undernutrition could also be associated with more severe disease and, by consequence, to urinary incontinence. A prompt referral for assessment and treatment with a specialist urologist should be available at each CF clinic, in order to investigate the onset and type of urinary incontinence early, even in younger patients.

https://tinyurl.com/2p8eumrs

Lung And Nutritional Health, Like CF Mutation, Can Affect Lifespan

Inadequate nutrition, poorly working or chronically infected lungs, and mutations leading to more severe disease can shorten the lifespan of people with cystic fibrosis (CF). All these factors interact with each other. Therefore, it is important to take into account all the analyzed factors when monitoring the course of the disease. In this study, the researchers looked at how a handful of factors may determine survival in CF, focusing on adults. The answer to the question of why some CF patients live longer than others is still unclear, which, together with the increasing number of adult patients, generates the need for research on the factors affecting the survival rate in this group. The period between 30 and 40 years of age is the

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I've Finally Stopped Blaming Myself For Not Being Able To Save My Sister's Life

By Tré LaRosa

It was a relief when I watched my sister finally take her final breaths. It's a relief to even finally write that sentence. And yet, for four years, every time that thought has entered my mind, shame immediately follows. I hated myself for being relieved, despite it also meaning she was no longer suffering.

I stopped believing in miracles a long time ago once I realized that miracles meant somebody was choosing to save some people but not others; it was easier for me to believe that life and nature are unfair, unthinking, and random. Cystic fibrosis didn't "happen" to me, my sister, my cousin who died in 1966, my aunt who died in 1978, or anybody who has the misfortune of a genetic quirk. Cystic fibrosis just is; it is a disease that encompasses countless experiences with thousands of different causes, and all of this together results in the suffering of people with CF and those who love us. CF touches everybody in its vicinity because, while the people who have it are the protagonists of the story, watching somebody whom we care about experience anguish creates additional anguish. Humans care about one another and we do not enjoy watching others suffer. But this isn't just about the CF community. This is also about anybody who has ever had a chronic disease or anybody who has ever suffered under the hand of other people or the randomness of the universe. Suffering, then, is universal and cannot be quantified. No methodology can cover the full scope of what it means to suffer, but everybody has suffered in one way or another.

Suffering comprises the things we experience and the things we don't. We are allowed to grieve the lives we didn't have, especially when those lives ended prematurely because of something as random as two deletions of a microscopic phenylalanine. Suffering is also not one dimensional. Suffering can result in progress.

I feel both gratitude and despair that I watched my sister die. In the moment, I felt horror as I realized I might be staring into my own future. I felt deep anger that while other 29-year-olds were buying houses and getting married, my sister lay in a hospital bed aware that her life was ending while being watched by her brother with the same disease and by her parents, one of whom had already lost a sister and cousin to CF many decades earlier. I felt relief that her suffering was coming to an end. I felt like it was my fate to watch my sister die since her suffering was so much worse than mine.

TRE AND ALYSSA LAROSA IN 2018.

I first thought about the possibility of my sister's death when I was a young child. I now realize how observant I was as a child as I reflect back on my younger days where I first confronted the realization that everybody we'll ever know and love, including ourselves, would one day die. I consider myself fortunate to have been there for my sister's final moments. Some people don't get that. Some people don't get to tell somebody they love them before they lose them.

Grief and suffering force us into precarious states of mind. My sister died from chronic rejection of her second double lung transplant. Hope, always my family's northern star, should remain flexible. We must carefully reflect on what optimism means in times of difficulty. Optimism is not usually wishing for things to be perfect. Mostly, optimism means recognizing what we are confronted with and determining which possible outcome will result in the least amount of suffering.

About six weeks before my sister died, Alyssa was declining rapidly. Neither of her bilateral lung transplants had gone as smoothly as we had hoped and we found ourselves in the same position as we had been in less than two years before: chronic rejection. It was clear Alyssa was not an ideal candidate for these procedures so a third transplant would not be an option. In some ways, Alyssa had suffered more after her transplants than before; the specter of hope of a healthy life with new lungs was never fully realized for her. We did not speak about it openly, but we knew her time was coming to an end.

We decided to take a final family vacation to LA. This trip was painful. We couldn't pretend that things were normal. We all knew these memories would forever be tainted by what they were sure to be followed by—her death.

During this trip, one day my mom and I drove out to get coffee. By this time—I was 23 and was working in a CF lab—I had assumed the role of the one who understood the medical and scientific complexity of our lives. I knew the prognosis for people with chronic rejection. The only hope is that you can prolong their life by maybe months to years, but with how rapidly my sister's health was declining and with no treatments working, I knew we were in the months range. But we also knew, since she had dealt with chronic rejection before, that her suffering would only worsen and result in more palliative care.

I explained this to my mom. I think my mom knew all of this, but my mom clung closely to hope. How could she not? My mom could not accept that Alyssa's life, after being on the precipice of death several times before, was truly nearing its end. We avoid death so much; we push it to the side, acknowledging that it's a part of life, but we never have the honest, difficult conversations with one another that we should have. I look back at this young version of myself and find myself, now at 27, shocked by just how young I was to be having these conversations. I laid out what I viewed to be the only possible futures for my sister: continued suffering for months on end followed by her death, or a shorter timeline of suffering followed by her death. Since a transplant wasn't an option and it's not possible to reverse the condition she was in, suffering for years in that state would've been worse than death.

Six weeks after that conversation, we would embrace as a family as we watched my sister take her final breaths. Something that you don't know until you experience somebody's death is how inconsequential the surrounding circumstances can feel. When people die at the hospital, you leave the room with your and their belongings. You walk out to the parking garage, just as you would if you were walking to go pick up lunch for a family member who just had a routine surgery, except, in this circumstance, you are preparing for life without them. The next phase begins with informing others and planning the funeral. Within two days of her death, I was writing my sister's obituary and her eulogy. These formalities were greatly therapeutic for me. At the time, I was still dreaming of becoming a writer, but my sister was one of my biggest supporters and never doubted writing was in my future. To be able to use this part of me to encapsulate my sister's life was more meaningful than I could've ever realized, partly because of the already devastating survivor's guilt I felt.

An unexpectedly difficult part of grieving my sister has been the dulling of memories. I have replayed the majority of our memories so many times since her death that I no longer really remember what it was like when she was with us. Her life—and mine while she was alive—feels like it was in the distant past. Sometimes it doesn't even feel like those memories are real; they feel like half-remembered dreams. My memories of her are fixed: I can't seem to remember anything new. What I do recall most frequently is how much I wish I could've done things differently when she was alive. I think about how much more I wish I had told her I loved and admired her; I wish I had told her I was proud of her more often; I wish I had spent more time with her. I wish I could've loosened up; I took my role as protective little brother so seriously that I would pressure her with tough love, often challenging her to take better care of herself. I know that I did every bit of that out of unconditional love, but there were many moments when she didn't need tough love; she needed gentle love and support. I frequently wish it would have been me instead of her.

Survivor's guilt is complex, strange, and frequently unreasonable. I feel guilty that I was born six years after my sister or that I never struggled with weight like she did. These feelings, unreasonable as they might be, are valid. When we are forced to grieve and reconcile something as inexplicable and painful as the death of somebody we love, our minds operate in ways that might not make a ton of sense to us. That's an element of survivor's guilt that I never expected; my mind, already at times feeling foreign to me, felt even more divorced from my being. I didn't realize how much my worldview would come to change as I recounted the time of my life when my sister was around. I didn't realize it would drive me to love people more fiercely; I feared that it would do the opposite. I did not anticipate that the rollercoaster of guilt, shame, and self-loathing would strengthen my resolve to love myself. I did not realize just how transformative survivor's guilt could be, if I just took a step back and stopped resisting the lessons these reflections were teaching me.

> "I did not realize just how transformative survivor's guilt could be, if I just took a step back and stopped resisting the lessons these reflections were teaching me."

I don't wish survivor's guilt on anybody, and I also don't believe people

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need suffering and trauma to be empathetic toward others, or that everybody has to find some grand romantic lesson in every bit of trauma they experience. I do believe that my survivor's guilt was rooted in something different than the death of my sister; for me, it was ingrained in my own struggles and insecurities. It was through the many tears I shed that I realized my survivor's guilt was rooted in my inability to save my sister's life. When she was alive, I believed death was something that we could turn away when it was on our doorstep. We could refuse death if we just tried hard enough. We could even help others turn death away.

I could not save my sister's life. For a long time, I blamed and hated myself for not being able to save her life. This feeling, while valid, doesn't make a ton of sense. Grief and suffering force our minds into precarious positions. I would've rather offered myself to death than not be able to save her life. I felt that, if I could've chosen to die myself, that would be a valiant sacrifice and a cause worth dying for, and it would've relieved me of the 23 years of guilt I felt when I was healthy while my sister was not. We had the same mutations and environments, so why did she spend 99.9% more time in the hospital than I?

I don't hesitate to express my love for others anymore. Life can change in the blink of an eye and I never want to regret not telling people I care about them. I don't blame myself anymore because I know I was trying my best out of love for my sister. I just wish I could tell my younger self that. ▲

Tré LaRosa is a writer and scientist currently residing in the Washington, DC, area. He spent four years in a CF lab before moving to another role as Research Lead where he focused on patient-reported outcomes. Currently, he is a consultant. He writes on his personal blog at trelarosa.com and also has a newsletter at trelarosa.substack.com where he writes about subjects including science, social commentary, politics, life with cystic fibrosis, grief, and anything else that comes to mind. His hobbies include spending time with friends and family, playing basketball, reading, skiing, conversing with people, reading Wikipedia pages, and also conversing with people about how much he loves Wikipedia. He also has a mini golden retriever who loves wearing bandanas (or so Tré says) whom he loves very much.

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most critical and most exacerbations occurred over the period between 20 and 35 years of age. The type of CF-causing mutation, FEV1%, BMI, and the presence of *P. aeruginosa* all determined the probability of living for a lesser or longer time. Patients who died at younger ages had two copies of a mutation leading to more severe disease (Class 1–3 mutations in both gene copies), an FEV1% lower than 40, a BMI lower than 18.5 kg/m2, and the presence of *P. aeruginosa* resistant to antibiotics. Next, the researchers used a method of analysis that allowed them to predict the likelihood of survival based on observations of this patient group. They found that the risk of death increased by two times for patients who had a mutation leading to severe disease, almost six times when FEV1% fell below the normal range, and more than four times when BMI fell below the normal range of 18.5 to 24.9. It also rose by two times in the presence of *P. aeruginosa* and by eight times if the bacteria were resistant to antibiotics. The researchers concluded that the probability of survival among adults with CF is a result of both genetic and environmental factors contributing to the course of the disease. In this study, patients living beyond the age of 40 had an unknown mutation or a mutation leading to mild disease, normal or moderate lung function, a healthy weight, and were free of infection by *P. aeruginosa* or infected with strains responding to antibiotic treatment.
https://tinyurl.com/y3r4lskj

Results Of A Self-Management Intervention For Cystic Fibrosis

In patients with cystic fibrosis (CF), the use of a self-management intervention involving data-logging nebulizers, a digital platform, and behavioral change sessions attained higher adherence rates than usual care. Investigators conducted a 2-arm, parallel-group, open-label, randomized controlled trial at 19 CF centers to assess whether a self-management intervention designed to support treatment adherence would decrease exacerbation rates among patients with CF. The primary study outcome was pulmonary exacerbations. Secondary endpoints included percent predicted forced expiratory volume in 1 second (FEV1); BMI; and Cystic Fibrosis Questionnaire-Revised (CFQ-R) score, which measures quality of life and includes a perceived CF treatment burden subscale. Results of the study showed that the mean exacerbation rate was not statistically significant. Likewise, the percent predicted FEV1 was not statistically significant. With respect to objectively measured adherence, adjusted mean differences favored the intervention group compared with the usual-care group. Adjusted mean differences in BMI also favored the intervention group. With respect to CFQ-R scores, although 7 of 8 different CFQ-R subscales revealed no between-group differences, the CF treatment burden subscale scores were reduced in the intervention group. Among the other 11-patient-reported outcomes at 12 months, which

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PHOTO BY MICHELLE COMPTON

Rooftop

I find myself alone . . . again on the roof.
Missing yet another friend, who didn't make it through.
We'd been here together, in better times for fun.
But here I sit, alone again . . . missing another one.
Their lives were inspirations, of which I'm honored to have shared a part.
Now their legacy is what comforts me . . . for of it, I am still a part.
I wake each day and thank the Lord, for sharing them with me.
And, before I lay, to him I pray, use me for others to see. . .
Each day is a gift we share,
With those who really care,
For tomorrow has no guarantee.

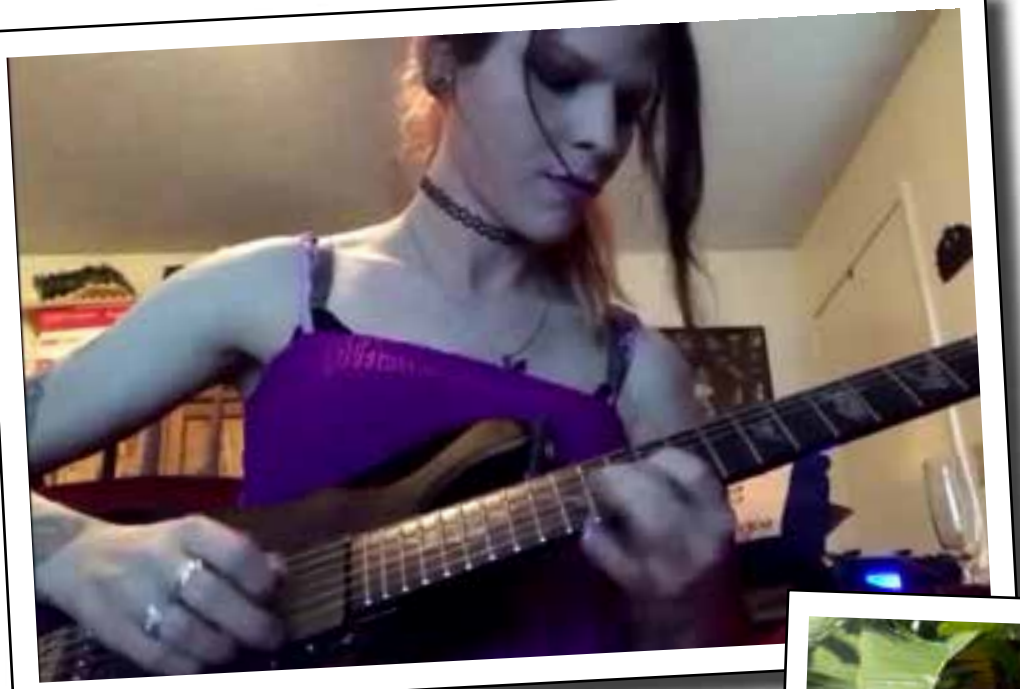
-J. Lindic, 2000

FROM OUR FAMILY PHOTO ALBUM...

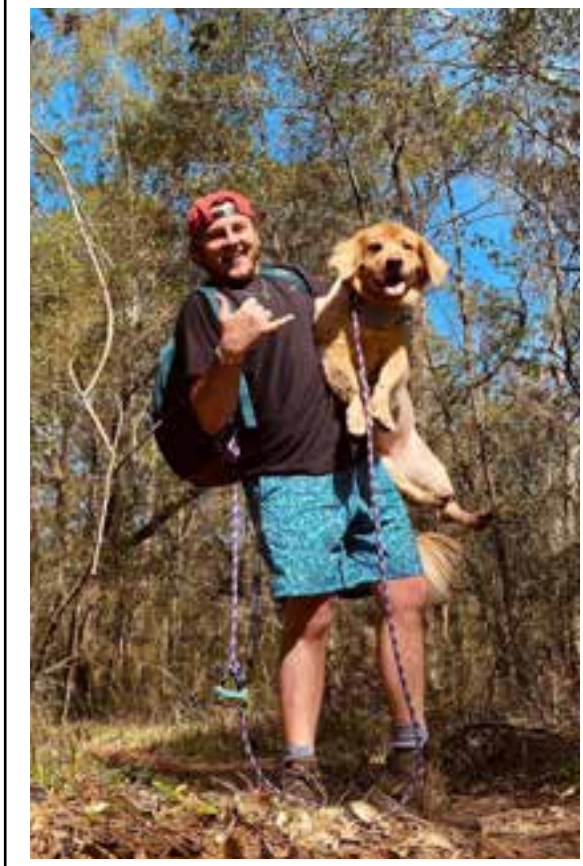
SYDNA MARSHALL AND HER DAD, DICK MARSHALL, AT THE ROOSEVELT BAR IN DOWNTOWN DENVER, JULY 2021.

KATIE, ARDEN AND ROSE LOCKWOOD ON A VINTAGE GREEN TRUCK GETTING THEIR CHRISTMAS CARD PHOTO TAKEN.

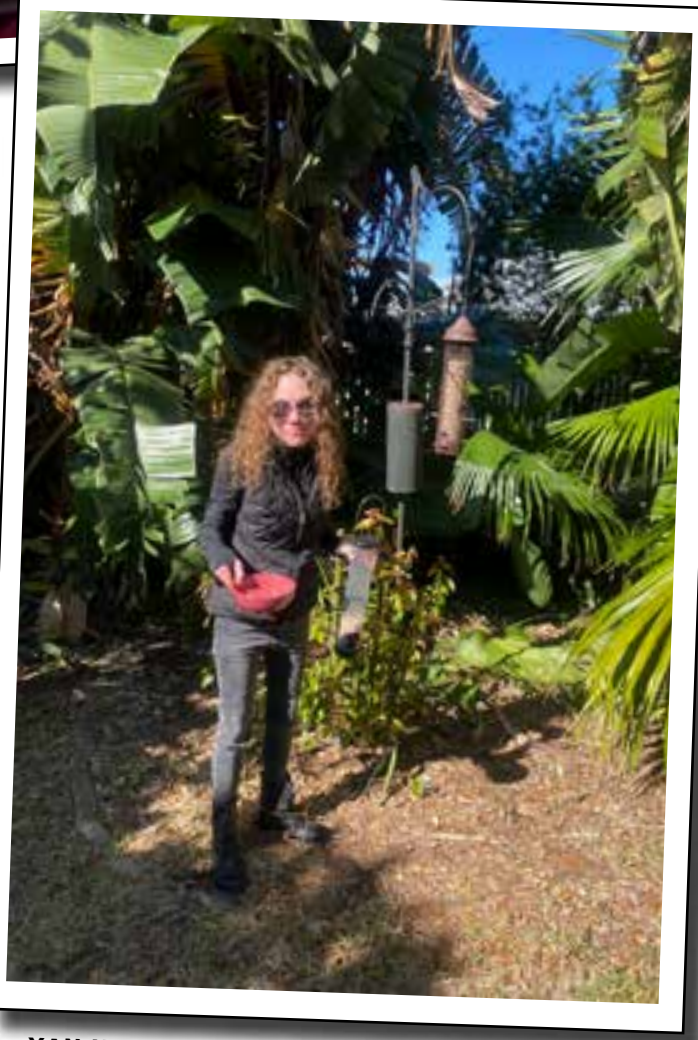
JULIE DESCH, GRIFFIN (JULIE'S SON), AND BARB ONLY UNMASKED IN ORDER TO EAT.



RAVEN ARAGON PLAYS THE GUITAR.



TRE LAROSA AND DUNCAN ON A HIKE.



XAN NOWAKOWSKI IN THEIR BACK YARD GETTING READY TO REFILL A SEED TUBE FEEDER AND WATER DISH FOR THE LOCAL BIRDS.

Surviving The Odds And Ends

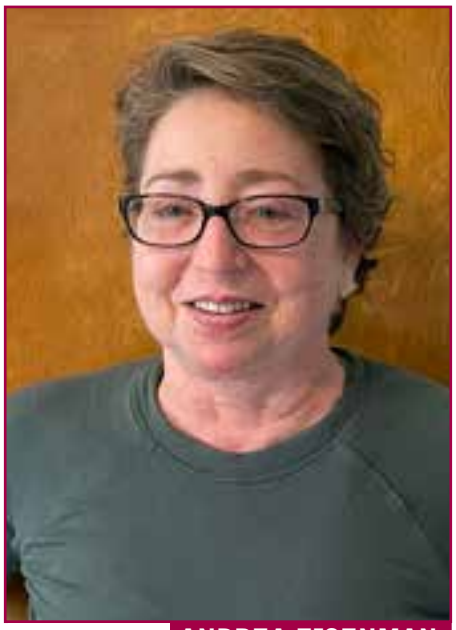
By Andrea Eisenman

Growing up, I was rather sickly. I was gaunt and had the "CF look" before newer medications made this obsolete. I was usually at the median age of survival, even as this slowly increased over time, but my quality of life was that of a person who coughed constantly, was confused with a two-pack-a-day smoker, and ate mounds of food while gaining nary an ounce of weight. My survival prospects were bleak at best. I frequently lived close to death. I was constantly on IV antibiotics, which seemed to no longer make a difference in the way I felt; they just kept the infections from further wreaking havoc and overwhelming my weakened body.

I was blissfully unaware of survivor's guilt as I didn't know anyone else with CF until I was well into my late 20s or early 30s. However, being culturally Jewish, I was familiar with other forms of guilt. Everything made me feel guilty—if I got a new pair of pants or something good happened, I had feelings of discomfort. It was hard for me to enjoy the things I was given without feeling twinges of guilt if those around me were not getting something, too. This may have come from being bullied and teased mercilessly as a kid or because my mom was overprotective of me—I'll never know for sure. Regardless, those feelings stuck with me.

I wouldn't truly know and experience survivor's guilt until May 2000, when I had my double-lung transplant at Columbia Presbyterian. A mere eight days after receiving this new gift of life, I awoke from a dream about my donor in the wee morning hours, just as the sun was starting to crest. At the time, I was in the step-down unit on the transplant floor. I had so many incongruous feelings, everything ranging from gratitude for this life-saving gift to gut-wrenching sadness from the underlying loss. I was

> "Many of the people whom I waited with for our transplants are now gone, but it is even sadder to think about the deaths of people who were transplanted after me."



ANDREA EISENMAN

incredibly grateful I was alive but also deeply saddened that the gift of a second chance came at the cost of my donor's own life, not to mention how hard that was for my donor's family. At the time, I didn't yet know that these feelings of guilt would be with me for some time—not feeling worthy of this gift of life when so many people do not even make it to the transplant surgery simply from a lack of suitable donors.

In some ways, it felt normal having myriad emotions and feelings about this wonderful gift of a new life because I imagined that my donor's family was simultaneously devastated by their loss. Thankfully, meeting my donor's family about 18 months after my surgery tempered these discordant thoughts. My donor's family said meeting the various recipients of their mother's organs helped them in processing their own grief—in the end, something positive resulted from her death. While they still missed their mom horribly, somehow this gave them some sense of peace in the end.

A year or so later, I was at an event honoring donor families and my donor's daughter invited me and my mom to go with her. It was in November near Thanksgiving and my donor's daughter was saying how her and her mom's birthday were coming up. Mine was, too, so I asked what days they were. She said that she was November 27. Her mom's was November 29. Eventually, they hoped to have someone fill in the gap. With tears in my eyes, I took a gulp of air and croaked, "mine is the 28." It almost felt like it was kismet. She said she got chills and, from then on, we had another bond that held us together. And thankfully, it helped me to feel less guilty over my renewed ability to live and breathe freely.

Prior to transplant, I was pretty sick and was living a very limited life. I had moved in with my mom as I could no longer live on my own. Post-lung transplant, as my health continued to improve, I met many new people with

CF, both pre- and post-transplant. This was thrilling to me. Getting to finally know and socialize with others who share this disease has been a blessing.

Having CF feels like knowing a shorthand or a secret language. I feel we learn to have empathy at a younger age than most. We understand what the stakes are with our disease. We get how all-consuming it can be to stay well. Very little needs to be said in order to convey what is happening. Our empathy for one another just reflexively kicks in. I treasure all my CF peeps and am always so happy to be in their realm. As I mourn those who are no longer here, it is not so much that they have died but seeing them suffer has been the hardest. Why them, why not me? Hardest were those who were younger than I.

It was against all odds that I recently turned 57. At times I cannot believe I am still here. (My friends remind me that it is because I am so compliant, but I am never sure.) While I am honored, in April, to celebrate 22 years with my donor's lungs, I do carry a lot of guilt at still being around when so many of my peers are not. Being in the CF and transplanted community can be a mixed blessing. Getting to know so many great people is fulfilling, but losing them gets to be soul-crushing over time. Many of the people whom I waited with for our transplants are now gone, but it is even sadder to think about the deaths of people who were transplanted after me.

While the survivor's guilt hovers, it is clear—life is not fair. Why do some people suffer so much while others do not? When I ask myself why them and not me, I mean, am I still worthy to be here? What can I take away from this guilt?

After much agonizing and therapy, I understand how lucky I am. Possibly this guilt pushes me to remember to be appreciative. I remain thankful to still be able to do most things most days. I continue to do what I can to keep living in my donor's memory. My days are still treatment-filled—exercise, naps, cooking healthy-ish food, going to myriad medical appointments *ad nauseum*. I feel I am still here through the grace of many people's support. It truly takes a village of wonderful people whom I have been fortunate to know. Some no longer here, sadly. But their memory resonates with me and is part of what keeps me going.

It has taken me years of thinking about this. Loss is painful. We miss our friends and people we have come to admire in our communities who pass away. We cannot explain it. The best we can say is, we are all fighting this disease—or, in the case of transplant, the complications that arise—the best way we know how. I try to be there for them when they are alive and then for their families when they no longer are. I mourn them. I try to process the emotions that come up and let the sadness come. When I can, I try to remember the good times we had or funny stories they told and the wisdom they imparted. Their spirit to be alive continues to inspire me to push myself to keep striving. Their lives enriched mine. And for that, I am forever grateful. ▲

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

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included 2 safety measures, 6 demonstrated differences with the intervention vs usual care.
https://tinyurl.com/4mxaj9e4

More Support Urged for 'Stressed' Patients With Anxiety

People with cystic fibrosis (CF) who have mild depression or anxiety report heightened stress, with significantly lower scores on 10 of 12 measures of health-related life quality as compared with the general population of CF patients. According to investigators, these patients may require more support, and specifically, possible individual interventions, such as talk therapy, than the current standard of care recommends. These findings indicate that monitoring, psychoeducation, and support may not be sufficient for adults with CF with mild depression or anxiety, suggesting the potential value of interventions. Living with a chronic disease like CF can be stressful, and many people with CF experience mental health problems such as anxiety and depression. Current guidelines suggest that CF patients should be screened yearly for such problems, beginning at age 12. For those with mild scores for anxiety or depression, the guidelines suggest offering preventive education and support, with follow-up screening a few months later. However, people with milder mental health symptoms have generally not been the focus of CF mental health research, despite the potential for symptoms to persist and progress in severity. A CF-specific form of cognitive behavioral therapy — called CBT — in patients who have mild anxiety or depression is being conducted. CBT is a form of talk therapy that works by targeting emotions by changing thoughts and behaviors. The CBT program being tested was developed by the researchers in collaboration with clinicians and adults living with CF. It

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Peer Engagement Groups From Attain Health

ADULT GROUPS

Facilitator: Brian Devine, adult with CF
Meeting #1: Tuesday 6MT/8ET
Meeting #2 Wednesday 6MT/8ET
Weekly meetings for adults with CF to connect and support one another through the trials, tribulations and joys of life, as well as current events!

YOUTH, TEEN, AND MINECRAFT GROUPS

Facilitator: Quinn Porco, teen with CF
Youth Group: Tuesday 4:30MT/6:30ET
Teen Group: Tuesday 5:30MT/7:30ET
Minecraft Group (siblings welcome): Friday 6MT/8ET
Weekly group meeting during which Quinn encourages peers to live their best life; includes chat groups and gaming fun!

INHALE MELANINE, EXHALE POWER

Facilitator: Raeshaun Jones, adult with CF Wednesday 5MT/7ET
This group was created and geared toward individuals in the Black Community who are living with cystic fibrosis. In this group, we will discuss our personal experiences within our communities as well as uplift and support one another on this trying journey in the fight against cystic fibrosis. "Living, one breath at a time"

CF FIGHTERS FOR RECOVERY & FREEDOM

Facilitator: Mark Tremblay, psychologist and adult living with CF Thursday 5MT/7ET
Mark Tremblay, age 51, with 32 years recovery from alcohol and drug addiction. Over the past 30 years, he has helped thousands of people with alcohol, drug, pornography, sex, relationship, and nicotine addictions. Mark has also managed addiction programs and ministries. He is opening this space to join you on your journey to find freedom.

MOMS WITH CF

Facilitator: Gillian Mocek, M.S.W., adult with CF and mother to sweet Simon
Monday 6MT/8ET
She has incredible insights and wisdom for balancing CF and motherhood, and uses this platform group for other mothers who have CF to get together, share, discuss victories and struggles with motherhood.

SOLID GROUND–BIBLICAL FAITH-BASED

Facilitator: Alma Martinez Svarthumle, adult with CF
Friday 1MT/3ET
Solid Ground—"A place to encourage one another, love on one another, and grow our faith in Christ together!"

Young Adult Group

Facilitator: Emma Chenier, college student living with CF
Monday 5MT/7ET
All about the transition into being a young adult—whether college or working—and being dedicated to your health.

LGBTQAI+

Facilitator: Dr. Xan Nowakowski, openly queer, agender, and polyamorous person living with cystic fibrosis
Times vary. Meets monthly. Accepting people exactly as they are; encouraging them to embrace their inner fire.

For more information:

http://attainhealth.org/peer-engagement-groups
https://www.jotform.com/attainhealth/support-group-intake-form

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covers key topics and skills for living with CF, including relaxation, depression in CF, adaptive thinking, health-related goals, and anxiety. The study found elevated symptoms at two to three times more frequently than expected in the community, with rates increasing from adolescence into adulthood. Most of the participants had received some form of mental health treatment in the past. However, the majority had never received CBT or any similar individual therapy. Half of the patients were actively taking medications for anxiety and/or depression, and another 25% had taken such medications in the past. These data suggest that the patients' mild scores for anxiety/depression could reflect a partial remission from a prior episode that was more severe, rather than a first episode of mental health problems. Similarly, on measures of health-related quality of life scores in 10 of a dozen areas were significantly worse among these CF patients compared with the values found in the general population of people with CF. The two exceptions were respiratory symptoms and weight. Further analyses of these scores indicated that patients with advanced disease tended to report their care as significantly less burdensome. Also of note, males tended to report significantly worse quality of life

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

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

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

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

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

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

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

To apply for this scholarship, please specify what type of creative degree you are pursuing (e.g., fine arts, graphics, music, singing, photography, filmmaking, writing, poetry, dance, theatre, and other performance arts). Candidates should have a minimum GPA of 3.0. Please submit essay answers, electronic samples of your work for consideration, transcripts, and a letter from your physician confirming CF diagnosis. The application deadline is April 15, 2022. ▲

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

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relative to weight than did females. These initial data suggest the potential value of interventions, such as CBT aimed at secondary prevention and better long-term health.
https://tinyurl.com/y2y6ksm6

Simple Intervention Improved Spirometry Adherence For Outpatient Cystic Fibrosis Clinic

Pre-clinic phone call reminders improved the number of patients with cystic fibrosis who submitted home spirometry results. Initial provider communication involved emailing or calling patients before their scheduled telehealth visit to remind them to submit home spirometry data prior to the visit. If spirometry results were not submitted, providers would follow-up by email or phone after the telehealth visit. Several barriers observed during this project included: (1) Some patients did not have their devices on hand for the telehealth visit. (2) Many patients did not prep their devices or install the appropriate app for the telehealth visit. (3) Many patients waited to perform their at-home spirometry testing during the telehealth visit. Overall,

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PEARLS OF WISDOM

Survival Is Serious Business: Reframing Guilt As Motivation For Self-Care

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

It feels very appropriate to write about survivor's guilt for this issue of *CF Roundtable* after reflecting on caregiving in the last one. For me, the two topics have become closely linked. Historically I have used survivor's guilt as a reason to neglect my own self-care in favor of working endlessly to help fellow people with CF and other chronic conditions. But recently, I have realized in earnest that surviving means more than just existing.

I got into a pattern for many years where I set no boundaries with myself, despite improving steadily at setting needed boundaries with others. I still struggle with unrealistic expectations of myself, especially when it comes to work. Given that I work full time as a medical school professor, in addition to volunteering full time in the CF community, and have not yet managed to overcome the essential human need for sleep, none of this even made sense mathematically.

During the summer of 2021, I wound up so overwhelmed by commitments I had readily agreed to that I felt as if I could not breathe intellectually. Although that would have been a familiar sensation physically, it disturbed me deeply when I felt as if I had no space at all inside my own mind. I had put myself in that position partly because I thought I owed my boundless energies to other people who had not gotten to live as long with their CF as I had with mine.

Of course, I had also survived other things that doubtless played a role in my sense that my only value in life came from working tirelessly to meet the needs of others. I fight daily against a deep, insidious fear of being expendable—or perhaps more accurately, a sense that I already *am* expendable but have managed to convince people otherwise through sheer magnitude of effort. This may be my version of the "impostor syndrome" many academics experience.

As an abuse survivor who engages these lived experiences in all areas of my professional practice, I have long since imparted to my students that consent cannot be given when a person feels afraid to refuse. Yet I wound up reproducing those same circumstances in my own life as a survivor. When I was given an earnest compliment by a colleague that "people always ask you for things because you never say no," it disturbed me profoundly. It was not the kind or earnest tenor of the feedback that chilled me, but rather the logical implications in my own mind.

So it seems fitting that what ultimately helped me change my behavior was thinking about the lessons I pass along to my students. I admitted readily, both to myself and to colleagues, that I could not knowingly model behavior for my students that was obviously hurting me. I knew my students deserved education from faculty who modeled the same compassion for ourselves that we nurture in our students for others. I leaned into this with the same zeal I have always brought to education: by sharing openly about my own relevant experiences as case examples.

> *"As I embrace survivorship as motivation to do better for myself, I feel as if I am just getting started."*

XAN NOWAKOWSKI

I have learned that I can best cope with survivor's guilt by doing everything I can to get out of situations that I know are harming my health. This is what honoring survivorship means to me now. I have begun to reap some of the benefits of this approach by getting settled in a new home in a different community with better air quality and fewer environmental stressors. After exploring relocating to a smaller city roughly halfway between the campuses where my partner and I have taught for over two years, we decided to move forward.

With working from home some days each week becoming almost expected after changes introduced by the COVID-19 pandemic, we felt confident that moving would improve our quality of life both individually and

jointly. I also suspected it would require me to continue practicing setting boundaries with myself and also facilitate getting more of the rest I have often denied myself.

So far, our projections have proven wholly accurate. I am understandably unsure whether my colleagues even believed me when I said that I would finally take the surplus annual leave I had accumulated during the pandemic to allow me to spend time packing up the old house, moving our possessions, supervising the crew that moved our furniture, and setting up the new house while still getting adequate sleep and completing my daily CF care tasks. But as both an experienced educator and an aging researcher, I know no one is ever too old to learn new skills.

At 38, I have survived a great deal. I have gotten second chances that others did not. I have walked out of hospital beds and intimate relationships that I was never guaranteed to leave alive. I have left homes, literally and metaphorically. Yet as I embrace survivorship as motivation to do better for myself, I feel as if I am just getting started. I told my parents that even amid the chaos of moving all our possessions ourselves, it felt like we were "finally getting to the good part" of our lives. That still feels true today.

The story of our housing search—which wound up being uncharacteristically short and easy—mirrors these broader changes in our lives and what we have learned along the way. I knew when I saw our house in person for the first time that it would be perfect for the next chapter of our lives. I could also tell from touring the home that one of the people who had the house custom-built 21 years ago had firsthand experience with disabling chronic illness.

I would not learn the full story of the house until after moving in. When I did, it resonated deeply. As I tended to the yard and landscaping plots, I found a decorative piece of stonework beside a palm tree near the front portico. It memorializes someone who passed away from cancer and also welcomes others to the property. I already knew from my research on the property that the surviving owner had moved away and sold the house to an investor. I wonder if they dealt with survivor's guilt of their own, or if the grief of staying in the home they shared with their partner became too much—or both.

I washed the stone carefully until its message shone clearly in the sunlight. Later that day, I showed it to my wife and told her what I had learned. I hope the people who lived their dreams in this house before death intervened would feel happy to know who lives here now. After all, I have come to know that feeling firsthand myself.

Watching my peers in the CF community thrive as they gain access to innovative therapies, like CFTR protein modulators, has balanced the feelings of guilt I have sometimes experienced surrounding my own survival. Although I cannot take those specific drugs myself due to genetic ineligibility, I embrace the deeper wisdom of this moment in our community story. Survival requires not only fighting to ensure our fellow patients get those same chances, but also intentionally doing the best we can for ourselves each day. ▲

Dr. Alexandra "Xan" Nowakowski is 38 years old and has CF. Xan is a director of CF Roundtable, *in addition to being a medical sociologist and public health program evaluator. They currently serve as an Assistant Professor in the Geriatrics and Behavioral Sciences and Social Medicine departments at Florida State University College of Medicine. They also founded the Write Where It Hurts project (www.writewhereithurts.net) on scholarship engaging lessons from lived experience of illness and trauma with their spouse, Dr. J Sumerau. You can find their contact information on page 2.*

LIVE OUT LOUD

What Brings You Joy? Do More of That.

By Lara Govendo, M.S.Ed.

We all could use a little more joy this season. The winter months are long; often dark and cold (well, it is here in the Northeast, anyway). The importance of mental health is elevated. As humans, we need a glimmer of hope to keep going. Some sort of light in the darkness that shows us that there are better days ahead. Perhaps joy is the solution.

In the CF world, we aren't strangers to dark times. When CF threatens to take us out, we are enveloped by darkness. Respiratory infections, gastrointestinal blockages, diabetic crises—you name it, we fight it. In the moment of fighting for our lives, it feels so heavy and dark. We often wonder if we'll make it out alive. That's not being dramatic; it's real life. That's the reality that we live with every single day.

While the world has grown darker in some ways amid the pandemic, we've also seen similar reactions in "normal" people who have been forced to live outside of their comfort zones, often beyond their control. People are disheartened, frustrated, overwhelmed, scared, and many entertain hopelessness.

The emotional cycle has parallels to our frequent exacerbations and dances with death. The irony isn't lost on our community that the rest of the world is getting a small dose of how we have to live our lives on the daily. Rarely are our lives comfortable. And a big part of our lives—our health—is outside of our control.

Something shifts in us when we are faced with our own mortality. It shakes us awake to what's truly important. And it tends to strip away the things that don't really matter.

What I've witnessed in our community is the steadfast resilience that overtakes those who are fighting for their lives. The ability to rally in times of life and death is one of our greatest attributes. We are the kings and queens of never giving up. And regardless of whether we realize it, the world is watching and is inspired by our valiance. We also have this incredible innate drive to focus on what brings us joy. It's often the focal point that keeps us going in seemingly dark times. Our light radiates when we can find joy despite our circumstances. When we share that with those around us, the days become lighter in every sense of the word.

LARA GOVENDO

As a community, we are built on care, compassion, and camaraderie. We have risen up together both in times of tragedy and in times of triumph. My gratitude for the connections I have in the CF and transplant community runs deep. We are connected in a way that only we understand and that is such a beautiful gift. I know that we will continue to rally around one another; together apart.

On an individual level, we need to take care of ourselves mentally, emotionally, spiritually, and physically. Whatever it takes, we have to do it. We cannot apologize for our decisions, actions, and responses. While we do have to be intentional and think beforehand, we must follow through with what makes sense for each of our individual paths in this life. Part of that is determining what the best course is. Only we know what that means for each of us.

So my challenge for you: decide this year to do the things that spark joy. Get curious. Chase what is in your heart to do. Don't stop believing that better days are coming. Chances are they're right around the corner. Be present always, cultivate peace, and share your light with those around you.

This will be my last column here with *CF Roundtable*. I have so enjoyed sharing my heart with all of you. Please come join me on my journey into new beginnings: @Lungs4Lovey (Facebook & Instagram). Lots of love sent for a peaceful season and a joy-filled 2022! ▲

Lara Govendo is 35 years old and has CF. She lives in Vermont as a wild adventure enthusiast who holds a Master's Degree in Mental Health Counseling. She currently works as a mental health counselor for middle schoolers. She also writes about living life beyond chronic illness and develops educational programs to restore hope to those in need. Thanks to her double-lung transplant in 2017, you can now find Lara traveling on the regular, exploring the glorious outdoors, and belly laughing with her loves. You can find her online at www.laragovendo.com (and on Facebook and Instagram) at "Lungs4Lovey." You can email her at lgovendo@usacfa.org.

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researchers observed a 16% higher rate of spirometry result submissions before or on the day of a patient's telehealth visit when simple interventions such as reminder phone calls and emails were implemented.
https://tinyurl.com/37ss5ssk

Cystic Fibrosis Triple Therapy Likely Contributed To Exacerbation Decrease During Pandemic

A new study highlighted a decrease in pulmonary exacerbations during the COVID-19 pandemic among patients with cystic fibrosis receiving triple therapy with elexacaftor/tezacaftor/ivacaftor. Researchers reviewed clinic and telehealth visit patient encounters and pulmonary exacerbations among patients who were enrolled in the Cystic Fibrosis Foundation Patient Registry from 2019 and 2020. From 2019 to 2020, there was a 24% decrease in the number of patient encounters overall, a 55.9% decrease in in-person clinic visits and a 60.6% decrease in reported pulmonary exacerbations. The number of patient encounters in 2020 was similar among patients who started Trikafta before or within the first quarter of 2020 compared with those who were receiving other modulator therapies or who initiated triple therapy after the first quarter 2020. Researchers also observed a greater reduction in overall pulmonary exacerbation incidence rates

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CF: THE MIND GAME

Living In The Valley Of The Shadow: Coping With Survivor's Guilt

By Mark Tremblay, M.A., M.P.A.

Like many CF patients growing up in the 1970s, I watched too many tiny black-and-white coffins rolling down church aisles amidst the anguished sounds of family and friends trying to hold it together while their lives were breaking apart. In fact, it was after attending one too many of these memorials that I first experienced survivor's guilt. The last one I attended was for a 12-year-old girl named Jenx who was my best and last surviving friend with CF at the time. The survivor's guilt from her passing cast a shadow over the rest of my life.

She and I were opposites in many ways. She reached a level of acceptance in her life that made her ooze virtue, wisdom, and grace; whereas I "made a mess," at least according to the baby book where my first words were recorded. For example, when some bully at school picked on her for her sunken eyes, unkempt hair (caused by the mist tent she slept in), and never coming to school, she walked away only to go home and pray for her bullies. In the meantime, I waited until the bully in the front turned his head and hit him in the ear with my full lunch box. Later, when she was too sick to ride Apple, the broken-down old horse in the pasture down the street, she would ask me to wheel her to the edge of the fence so she could watch me. Often, even though I didn't want to ride Apple (nor did he want me to ride him, I imagine) in circles in that undersized paddock, I would do it just to see the ear-to-ear smile tucked under her tattered Mickey Mouse cap from the Make-a-Wish trip to Disney she took when she was five.

For those who are interested, survivor's guilt was first observed in Holocaust survivors. Their disruptive and obtrusive thoughts were documented and studied. The overarching question was always why did they survive when others, whom they deemed more worthy and deserving of survival, perish instead? Unfortunately, all too many survived the hellscape of the camps only to be imprisoned by their own guilt, which drove them to drink themselves to death or commit suicide in surprisingly large numbers. Typically, when we think of survivor's guilt, we think of cases in which individuals survive war, accidents, natural disasters, or violence. However, surviving an illness deadly to others is a well documented and not uncommon cause of survivor's guilt as well. In the latter situation, survivor's guilt is completely normal; so much so that some theorists consider it abnormal if you do not feel survivor's guilt in such circumstances.

So what is survivor's guilt and how can we live rich, full lives despite the guilt of outliving our fellow warriors? Most psychologists agree that survivor's guilt is a particular type of grief in which you cannot move past the question "why them and not me?" Just like other types of grief, which are natural psychological and emotional adaptations to loss, survivor's guilt takes time to process and heal and this varies greatly from person to person in terms of acuity and length of time. Some of the symptoms of survivor's guilt include obtrusive and/or obsessive negative thoughts, emotional numbness, amotivational syndrome (e.g., not wanting to get out of bed), anhedonia (e.g., not wanting to do things you used to enjoy), intense fear, urges to engage in self-harm (e.g., cutting), suicidal thoughts, or substance abuse.

> *"Surviving an illness deadly to others is a well documented and not uncommon cause of survivor's guilt."*

MARK TREMBLAY

Survivor's guilt is fairly common in the general population, but it's particularly prevalent in the CF community where so many of us have siblings, family members, and close friends who have been taken from us too soon by this disease. Further, the more immersed you are in the community and the more your social support network relies on the community, the more likely you are to experience grief

and/or survivor's guilt because you connected to more people with short life spans. Over the course of my life, the methods I have employed to process survivor's guilt have ranged from drinking myself into a stupor to bringing it to my therapist so it can be used as fodder to spur my emotional growth.

The following are a few ideas for coping with survivor's guilt that I have found to be particularly helpful:

- **Focus on the deceased person's best qualities and try to emulate them.** Since Jenx's passing I have often thought about how she deeply accepted her CF and the way she let it mold her into a better, more patient, kind, compassionate, empathic, and graceful person. When I find myself thinking of her, I try to channel those thoughts into striving to embody her best qualities.
- **Let the waves of grief and guilt kindle your generosity.** One of the most powerful things I learned in sobriety is that connecting with and sharing my experience with others, providing a helping hand to someone in need, and offering my time and/or resources to assist others are all positive actions that I can take to disrupt my negative thought cycles and accelerate my emotional healing.
- **Acknowledge the survivor's guilt, label the emotions, and affirm your capacity.** When I first experienced survivor's guilt, it felt like a tsunami that drowned out all of my thoughts. When I got sober, I realized that what I thought was a tsunami was merely a set of emotions and thoughts which, individually, I had the strength and tools to manage.
- **Focus on your current relationships in the here and now.** When I'm struggling with loss, and survivor's guilt in particular, one of the most effective things I can do is lean more heavily on the love of friends, family, and fellow warriors in the moment. When I fight the urge to turn away and isolate and instead lean into others, I feel better faster.
- **Cherish your life by loving yourself and fighting your disease.** In my heart, I know what I need to do to live my best life despite CF, including taking my meds, doing treatments, attending clinic regularly, praying, working out, consuming the micro and macronutrients my body needs, and staying connected with my family, friends, and therapist. I strive to let my negative, intrusive thoughts be catalysts to engage in positive, productive actions.

In the CF community, we are constantly living in the valley of the shadow of death of loved ones and dear friends who have gone before us. We can choose to let these experiences mire us down in cycles of self-destructive behavior and corrosive thoughts or we can strive to embody the best of those who have gone before so that they can live in perpetuity and we can live more fully. My prayer for all you CF warriors is that you find the strength to keep fighting and find ways to live well even as you walk in the valley of the shadow. ▲

Mark Tremblay is 52 years old and has CF. He lives in Albany, NY, with his wife, MaryGrace. He has a Master of Arts in Psychology from Marywood University and a Master of Public Administration from Syracuse University. Mark has worked in the New York Governor's Division of Budget for six years and presently works full time at the Department of Health. He is the President of "CF Vests for Life," which collects donated therapy vests, nebulizers, and oxygen saturators for distribution to CF patients around the world. Additionally, he is the leader of the Attain Health group, "CF Warriors for Recovery and Freedom." Mark is also a director of USACFA. His contact information is on page 2.

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among those who received triple therapy compared with those who did not. Results showed a greater reduction in severe exacerbations from 2019 to 2020, compared with mild and moderate exacerbations, among both those who received triple therapy and those who did not. The exacerbation incidence rate within the elexacaftor/tezacaftor/ivacaftor population is more significantly reduced, indicating that highly effective elexacaftor/tezacaftor/ivacaftor therapy has positive therapeutic impacts for this population.
tinyurl.com/mr38s2ux

AND

https://tinyurl.com/yckjvs7v

Safety, Improved Outcomes With Cystic Fibrosis Triple Therapy Maintained Through 96 Weeks

Safety and improvements in lung function, respiratory symptoms and CFTR function with elexacaftor/tezacaftor/ivacaftor were maintained through 96 weeks of treatment in patients with cystic fibrosis and at least one F508del allele. Analyses will continue through 192 weeks. This open-label extension study included 506 participants with cystic fibrosis aged 12 years and older with F508del and a minimal function mutation or who were homozygous for F508del from the phase 3 pivotal parent studies. All participants received at least one dose of elexacaftor/tezacaftor/

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IN THE SPOTLIGHT
With Raven Aragon

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

Raven is 38 years old and lives in Phoenix, AZ. I first met Raven as she was starting to share openly online about her experiences as a transsex woman with CF. But we wound up having plenty more in common besides being part of the trans and queer CF communities! Over time we bonded about our shared love of music, from listening to heavy metal and hard rock to playing guitar ourselves and talking about our favorite equipment. We've hung out on her Twitch streams, talked about how gender-affirming healthcare is evolving as a field, recommended books to each other, supported each other in our journeys with CF and mental health, and shared many cute memes about bats. Spotlight, please!

RAVEN ARAGON

What would you like our readers to know about your journey? What do you wish healthcare providers knew about working with transsex folks who have CF?

I think I've been pretty lucky because my doctor was really accepting and open to it. I don't think even doctors realize how few resources there are for trans people to get any care. Every time I go to CF clinic, my doctor always asks me how the transition-related surgeries are going, whether I'm able to undergo them...and I'm like, "yeah, I just don't have the money," so the surgeries are pretty much on hold indefinitely. Aside from that, he's been very good. So I don't know if there's anything that I would change as far as my doctor is concerned, but I know that's not going to be the entirety of the community.

You've also met several other folks from the trans CF community who are interested in taking hormones and/or having surgery. What have you learned from connecting with others who are walking a similar path?

I've only met a few people with CF who are transitioning. It seems pretty simple for all of the female to male ones—they haven't really had many issues. However, of the two people with CF whom I've met who are transitioning from male to female, one was still in the closet. The other person, whom I just met, seems to be moving forward with everything and she's going to be talking to her CF doctors about whether she can get on hormones. I shared my experiences with her so who knows how that goes with her, but I have yet to meet another trans woman with CF who is on hormones.

What are your biggest challenges with CF? What is your unique presentation of the disease like and how has that changed in your experiences with CFTR modulators?

I always had really bad pancreatic issues. Since starting the modulators I haven't really noticed a lot of difference with my pancreas. My lungs, on the other hand, are doing fantastic. I think I had a sinus infection or something last week or two weeks ago, and even with that I just bounced back quickly. It wasn't like I was sick for a month because of it, as I would have been pre-modulators. So the CFTR modulators have really done a lot for my lungs. A few years ago, I was like, "okay, well I'm gonna die soon," and now I'm like, "wow, it's done so much for me."

You've had a lot of changes with your health since starting on Trikafta. How have your expectations and experiences with Trikafta evolved and differed from previous modulators you were taking?

When I was on Symdeko, it didn't seem like it really did anything for me. Although it may have, considering that there were a lot of other things going on when I started it. I quit smoking and a lot of other things happened around the time I went on Symdeko, but I didn't really notice a whole lot of differences in much of anything. I mean, I was still on constant oxygen—I was still having to pace myself to get through the day and errands like grocery shopping were very challenging. I would have to stop and take breathers for even the most basic tasks. With Trikafta, occasionally I'll get out of breath if I exert myself too much, but the amount that I can exert myself before I get out of breath and have to take a breather is way higher. And even at that, I just get winded; I don't start coughing mercilessly for 15 minutes, whereas most of my life before that I did.

Have you had any concerns about interactions between your hormone therapy and Trikafta? What

kinds of things did you talk about with your care team before switching from Symdeko?

I really didn't talk about much before switching to Trikafta. They pretty much just told me, "we're going to switch you; watch out for these things" and that was about it. As for interactions with my hormones, again there wasn't really any conversation about it. I told my CF doctor what I was doing and I asked him what he thought. He said I should be fine. That's kind of how my doctor is generally. I also told him about the surgeries and he wasn't particularly concerned about either that or hormone interactions.

What community resources have you found helpful in both coping with and living with CF?

I haven't found many helpful resources. I just disassociate when things get too rough. I bury myself in whatever I'm doing at the time, whether it be gaming or playing guitar or what have you. And that seems to be better for me than a lot of the other outreach things—because for me some of those just hammer the point of how I'm feeling. And, rather than alleviate them, they make them fester. Like when I was part of that CF group on Facebook, the constancy of people dying and people getting sick and everything just really hurt me, so I had to step away from it. And I find it really difficult to have friends who have CF because of that. My friend with CF who is transitioning is not in the best of health. Her lung function is really low. She's on Trikafta and she's hoping that it can keep her going until she's able to get a lung transplant.

It's just really difficult seeing what's going on in the community. So I pay attention to the news of what's going on with CF research, but as far as the community itself...I kind of stay away from it. Obviously, there's a few people I've met who have been very awesome and I would hate to not have them in my life now. But, for the most part, I kind of stay away from other people with CF just because it's really hard to get invested in somebody and then to lose that. I don't keep that from letting me get invested; I just don't often give myself the opportunity to be close to others.

You also have an adorable dog, Lilith ("Lily"). Does having her by your side help you stay well mentally? Were you concerned about allergies or other issues that could impact your health from having a dog at home?

I don't remember why exactly I picked her, but I do remember when we went to the pound. My roommate and I went to the pound just to look and we found two dogs that we liked. They liked the other dog a little bit more and I liked Lilith. There were three dogs in the kennel. She was sitting in the back, just kind of staring at me with this pitiful little face and I'm like, "oh she's so sweet." Occasionally she would nip at the other dogs because they would get too close or in her face. And I knew she was the dog for me. She just had a vibe about her. If I'm sad or upset or something, just having her to cuddle with and be a goofball is really great; she kind of opened my heart to dogs again. After my ex and I split and she took our dog Tia with her, I just felt like there was something missing. Getting Lily has been really amazing; she's such a good dog. She has her moments but she's a great dog. She has also gotten me up and moving. I think that's important—I have to take her out and I take her for walks because she won't do her business unless she's walking. So she gets me up and moving rather than just wasting away in my bed all the time. Her name comes from the TV show *Supernatural*...she's a Lilith, and she has turned out to be a little demon!

I know one of your biggest hobbies is gaming and you're working on going professional with it. How did you get into gaming? How do you want to be known in the gaming world–how would you like to stand out as you build your brand?

I started gaming when I was little. I remember being like four or five and staying with my mom playing *Legend of Zelda* and *Final Fantasy*. Ever since then I've been big into gaming. When I was in high school I didn't play as much, but I still had my computer and I would still play on that. My mom and I used to play *Pokémon* together. We would trade Pokémon online because I got *Pokémon Yellow* and then she played it for a little bit and then eventually bought herself a Gameboy. Gaming has always been a thing we share. Even now my mom has an Xbox and plays *Skyrim* and other similar role-playing games. So really gaming has just influenced me throughout my entire life.

You're also very skilled at guitar. Can you share about your history as a musician and what you're working on right now?

I've always wanted to play guitar. Always—ever since I was little. My mom has a photo of me when I was around five or six, playing with a toy guitar and microphone on Christmas morning. Music has been a huge part of my life ever since. When some kids were listening to children's music, I was listening to hard rock, heavy metal, and glam rock because that's what was popular at the time. My mom used to turn MTV on to soothe me when I was little. As I got older, I constantly asked her for a guitar. She was concerned that I would get frustrated and quit because my hands are so small. However, she met a guy at work who apparently had really small hands but he was a really good guitar player. Seeing him play was what pushed her to give it a chance. I dabbled

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MILESTONES

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

ANNIVERSARIES

Birthday

Andrea Eisenman
November 28, 2021
57 years old

Sydna Marshall
September 19, 2021
41 years old

Xan Nowakowski
December 29, 2021
38 years old

Michael Schnitzer
June 4, 2021
64 years old

Wedding

Sydna Marshall and Adam Keys
Austin, TX
8 years on October 26, 2021

Transplant

Michael Schnitzer
Double-lung Transplant
2 years on November 29, 2021

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in guitar a little bit, but not a whole lot, until I was 16, when my mom bought me my real guitar. I've had several different guitars since then and I have had to sell some of them to pay bills. But my current guitar is a PRS SE Paul Allender model—the purple one with bat inlays. I saw it in Guitar Center (https://www.guitarcenter.com) and really wanted it. So I went home and sold all of my guitars and my PlayStation 4 and bought it and I love it. It's the nicest I've ever had. This guitar—just the action on it and how it feels—it's just so perfect.

What musicians and bands inspire you? How has music shaped your journey with CF and your self-discovery as a woman? What are you listening to recently that moves you deeply?

I love Icon for Hire. Ariel Bloomer—just her strength and everything that she's dealt with in her past—has really inspired me. Her look and everything with the way that she creates her own clothes, her hair dye, and just everything about her screams "empowerment" to me. Looking at her and the way she held herself and the way she was singing is one of the many things that pushed me to finally transition as well. It was almost like stepping away from the dark gloominess that I had been in for so long. It was like a light there that I saw and thought, "yeah, I want that." For so long, I had seen that light in me and it was always stifled by so many things. I was never allowed to express myself in that way because I was "a boy," so there were a lot of things that caused me to repress it more and more as the years went on, and I got the feeling more strongly for me to transition.

Do you have a funny CF story you'd feel comfortable sharing? Is there an experience you look back on now that makes you laugh?

I have a story from when I was really little. My mom has told me was that I used to always ride this big wheel when I was in the hospital. And I would just ride it through the hospital and run over people's toes, apparently. But as far as growing up, I don't really have any funny CF stories because I was always so self-conscious of my coughing and other things.

Living with CF constantly requires us to change our plans and accept difficult things. What would a perfect day look like for you? Would your answer be different if the COVID-19 pandemic were under better control?

The pandemic wouldn't affect my perfect day. I think the perfect day for me would just be to have breakfast and then go to the lake and wakeboard for hours and hours. And then come home, eat dinner, and game until I go to sleep. I used to wakeboard a lot when I was younger. My mom's ex-husband had a boat, so we used to go every weekend during the summer. It was a lot of fun and it was the one space where I didn't have to worry about anything. ▲

Dr. Alexandra "Xan" Nowakowski is 38 years old and has CF. Xan is a director of CF Roundtable, *in addition to being a medical sociologist and public health program evaluator. They currently serve as an Assistant Professor in the Geriatrics and Behavioral Sciences and Social Medicine departments at Florida State University College of Medicine. They also founded the Write Where It Hurts project (www.writewhereithurts.net) on scholarship engaging lessons from lived experience of illness and trauma with their spouse, Dr. J Sumerau.*

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

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ivacaftor and enrolled in the open-label extension study. Nearly 500 participants experienced at least one adverse event. Most reported adverse events were mild or moderate. Exposure-adjusted adverse event rates and serious adverse events were lower in the extension study compared with the placebo group in the parent study that included participants who were heterozygous for F508del and a minimal function mutation. There were no new safety findings observed in the 96-week analysis. Improvements in percent predicted FEV1, CFTR function as assessed by decreases in sweat chloride concentrations, Cystic Fibrosis Questionnaire-Revised respiratory domain scores and BMI were maintained at 96 weeks. Thus, week 96 interim results of this open-label extension study were consistent with the previously established safety profile of elexacaftor/tezacaftor/ivacaftor in people with cystic fibrosis greater than or equal to 12 years of age and at least one F508del allele. Clinically meaningful improvements in lung function, respiratory symptoms and CFTR function assessed as decreases in sweat chloride concentration observed in the parent studies were maintained through an additional 96 weeks of elexacaftor/tezacaftor/ivacaftor treatment.

https://tinyurl.com/fzcpatae

Trikafta Found To Quickly Ease Sinus Symptoms In CF

Treatment with Trikafta rapidly eases symptoms of chronic nose and sinus inflammation in people with cystic fibrosis (CF). Trikafta improves symptoms of sinonasal disease within seven days of therapy, and the improvement persists up to at least six months. The study question was how does Trikafta affect the clinical manifestations of CF sinus disease? Prior to starting on Trikafta, and after about nine months on the medication, the patients underwent comprehensive assessments of sinus health. They also regularly completed a questionnaire,

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called SNOT-22 (sinonasal outcome test-22), measuring chronic rhinosinusitis symptoms. Higher scores on this test indicate more severe symptoms; of note, a change in score of 8.9 points or more is considered clinically significant. Prior to starting treatment, the mean SNOT-22 score was nearly 25 points. The scores rapidly decreased after patients started on Trikafta. One month after treatment, mean scores were less than 15 points, well below the minimum change needed to indicate a clinically significant improvement. Improvements were seen across all subscales of the SNOT-22 assessment. SNOT-22 scores remained low out to six months. Statistical analyses showed that people with more sinus and nasal inflammation prior to treatment were significantly more likely to have a reduction in chronic rhinosinusitis symptoms. Imaging of the patients' sinuses via CT scans suggested that treatment with Trikafta reduced mucus levels and mucosal thickening, which likely were key to the overall improvements seen in the study. Assessments of patients' nasal tissue, done via endoscopy, suggested that treatment with Trikafta decreased abnormalities such as polyps and crusting, with crusting "entirely resolved" with Trikafta treatment. Statistical analyses suggested that, like SNOT-22 scores, endoscopy improvements were more common in people with more sinus and nasal inflammation before starting Trikafta. Treatment with Trikafta did not improve patients' ability to smell, called olfaction, other analyses found.

https://tinyurl.com/2tdt4bny

Trikafta Shows Little Effect On GI Symptoms Over Six Months

Six months of treatment with Trikafta does not substantially alter digestive symptoms in people with cystic fibrosis. Analyses were presented that compared digestive symptoms before starting treatment and after six months, looking for trends. These analyses were broken down by age and sex. Overall there was not an enormous change in terms of upper digestive symptoms, such as nausea and abdominal pain. The most obvious changes were for bloating, but the change varied by sex: females tended to have less severe bloating after six months on Trikafta, whereas males tended to report worsening bloating. These differences were more pronounced in older patients. Similar results were found for lower abdominal symptoms, such as constipation. In terms of digestive health-associated life quality, overall trends showed improvement after six months of therapy. The most substantial effect noted was reduced dissatisfaction in males younger than 18. From baseline to six months, the number of patients reporting constipa-

ENGAGING PATIENTS ON RESEARCH TEAMS: THE CF PCOR TRAINING MANUAL

SEATTLE, WA (January 5, 2022)—Researchers from the University of Washington (UW) announce the release of a new Patient-Centered Outcomes Research (PCOR) Training Manual for the cystic fibrosis (CF) community. This step-by-step guide explains how CF researchers can include and authentically collaborate with patients on their teams. The complete manual includes four guides detailing each stage of creating and maintaining a PCOR team. The goal of the training manual is to provide a practical resource for CF researchers engaging with patients and caregivers on their teams.

The four guides are Prepare, Launch, Implement, and Monitor & Sustain. The manual comprises 57 pages including 28 pages of supplements providing examples of how to create essential documents such as team charters and feedback surveys. There are 64 references to peer-reviewed publications, PCORI documents, and tools from other outside groups.

The supplemental information includes vignettes with examples of the Development Team's own experiences and their solutions working with patient and caregiver partners on teams. Also included are educational resources such as introductory PCOR training webinars, project management tools, and a user guide website that supports PCOR teams who meet virtually.

About the CF PCOR Community Advisory Board

The University of Washington-based team included researchers and a community advisory board. The CF PCOR CAB consisted of 16 stakeholders that met six times between March and August 2021. The CAB included CF patients/caregivers, researchers, and stakeholders from the CF community. Stakeholders on the CAB were located across the country and were of various ages.

The CF PCOR Training Manual is available now on the Cystic Fibrosis Foundation website at this link: https://www.cff.org/researchers/patient-centered-outcomes-research-training-manual.

University of Washington
Department of Family Medicine
1959 NE Pacific St.
Seattle, WA 98195
Contact: Emily Godfrey
godfreye@uw.edu

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tion did not change substantially, while the proportion with diarrhea increased. This increase is not statistically significant, but it does line up with anecdotal reports of diarrhea in people starting on Trikafta and other CFTR modulators. Levels of fecal calprotectin – a marker of intestinal inflammation – decreased significantly after six months on Trikafta. In addition, there were no changes in levels of fecal elastase (a pancreatic enzyme needed for digestion) or steatocrit, a marker of fat, after six months of Trikafta treatment.

https://tinyurl.com/yh7xke69 ▲

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

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

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

IMPORTANT RESOURCES

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

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

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

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

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