

## Why Vaccinate?

By Julie Desch, M.D.

There is a rapidly growing group of people who now believe a concept of infectious disease that is reminiscent of the model of the flat earth. It is called the “terrain model” and its proponents are germ theory denialists, a/k/a people who don’t believe that microbes such as viruses and bacteria cause disease. The terrain folks actually believe that disease spreads from one person to another not due to an external agent—a pathogenic bug—but because the internal environment of the receiving person is “weak” or “toxic” and therefore they cannot fend off disease. Yes, this is a “victim is at fault” theory that can be infuriating to those of us who understand science and psychology, especially those of us who live with CF. Certainly, it is true that in order to stay healthy it helps to have a healthy/strong internal environment. Another phrase for this internal environment is “immune system,” so the theory isn’t completely off base. It is a bit like



JULIE DESCH

the old adage: the rich get richer. In this case, the idea is that the healthy stay healthy. The corollary is that the sick tend to get sicker, which is sort of true. But to blame the sick for this is heartless. And to actually deny that pathogens exist and cause disease is hard to swallow.

Now I’m sure that readers of this particular publication are well versed in the scientific theory/evidence that bacteria and viruses do, in fact, cause illness, so I don’t need to dispel terrain theory here. I’m simply mentioning it because there are people out there who likely have no significant scientific background, nor personal history of the effects of bacteria and viruses quite like we do. It is easy to get angry at these people and add to the polarity that we see every time we watch the news or walk into a Walmart. But anger doesn’t help. It just entrenches everyone further into their stance of “I am right, and you are an idiot.”

Before I knew anything about science or immune systems or pathogens, I got a flu shot every year. I did this because I was forced to do so. My parents gave me no choice. It was my least favorite day of the year, but they indoctrinated me into the wisdom of doing this by telling me over and over why it was important and by doing it themselves. They did such a good job that, by the time I was old enough to do so, I rode my bike voluntarily to Dr. Brugh’s office myself, without a

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## EDITOR'S NOTES

**F**all heralds a season of change—the leaves change color and the days grow shorter. In that spirit, we're talking about balancing caregiving with CF. We're also addressing the science surrounding the COVID-19 vaccine and various reasons why individuals in the CF community have chosen to get vaccinated. **Dr. Jeanie Hanley** asserts that getting vaccinated is very much a part of her equation when it comes to balancing caregiving with her own self-care. **Andrea Eisenman** writes about her dad's motto—take care of #1 first—and what that looks like in practice. **Gillian Mocek** shares her lessons along the way in balancing her own self-care all while raising two young kids. As part of our additional focus topic, former director **Colleen Adamson**, a high-risk PWCF, discusses her choice to get vaccinated after talking to her PCP, transplant team, and CF care team about what was best for her and what they recommended. **Dr. Richard Moss**, former Director of the Cystic Fibrosis Center at Stanford and site principal investigator for the Cystic Fibrosis Therapeutics Development Network, explains the science behind the safety and effectiveness of the COVID-19 vaccine, both for PWCF and post-transplant individuals in his article. Across the pond, **Maggie Williamson** writes about her experience as a transplant recipient waiting to get the vaccination in the U.K., as well as what her “new normal” looks like post-vaccination. **Elyse Goldberg** recounts her vaccine story as well—why she chose to get it and what side effects she experienced. **Anna Modlin** tells us about her COVID-19 vaccine experience in Australia, a country with national healthcare and some of the harshest and longest lockdown policies enacted.

You can read about the many accomplishments and accolades of Julia Kennedy and Emily Schutz, the two individuals selected from the pool of applicants for our Higher Education Scholarship. Be sure to read the latest CF research in **Laura Tillman's** expertly collated “From the Internet” column this issue. Additionally, **Leah Sands** opens up about the side effects of Trikafta in this issue.

In this issue's “In The Spotlight,” **Dr. Jeanie Hanley** talks with Sonya Haggett about her post-transplant life and what led her to become a social worker. **Dr. Xan Nowakowski**, in a nod to *The Matrix*, writes about spoon theory and sharing each other's realities in their “Pearls of Wisdom” column this issue. In our “Ask the Attorney” column, **Beth Sufian** answers readers' questions about working remotely after many employers have ended remote-working arrangements. **Isabel Stenzel Byrnes** talks about armor—the various types we employ and how this helps us navigate life—in her column. **Lara Govendo** lobbies for treats at adult CF clinics in her column this summer. Hindsight is 20/20, says **David Tarnow**, in his “Savoring Serendipity” column. You can read about the advice he wished he'd known when he was in the trenches with end-stage CF pre-Trikafta.

I hope you enjoy reading this autumn issue as much as I did! In the words of Effie Trinket from *Hunger Games*, may the odds be ever in your favor, Sydna.

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# Information From The Internet...

Compiled by Laura Tillman

## Partnership Aims To Lower Out-Of-Pocket Costs For Rare Disease Meds

AllianceRx Walgreens Prime, a specialty and home delivery pharmacy, is partnering with TailorMed, a health-care technology company, to help lower out-of-pocket prescription costs for specialty pharmacy patients. Medications attained through specialty pharmacies are those used to treat rare and chronic conditions in the U.S., and are often extremely costly. For this reason, Alliance Rx noted that it has staff dedicated to helping link eligible rare and chronic disease patients with financial aid programs. There was a 5% rate of non-adherence to prescribed treatment



LAURA TILLMAN

when out-of-pocket costs were zero, and a 45% non-adherence rate when these costs surpassed \$125. A majority of patients, 60%, stopped using treatments

as prescribed when out-of-pocket costs exceeded \$500. TailorMed's platform identifies people at risk of being unable to afford their specialty prescriptions, and automatically connects them to financial assistance programs. Its comprehensive database of financial assistance programs is updated in real-time. AllianceRx Walgreens Prime will have access to more than 5,000 aid programs via this platform. They include co-pay assistance, manufacturer voucher and bridge programs, government subsidies, community and state resources, and assistance from disease-specific foundations.

<https://tinyurl.com/yhjpvvpe>

## Relizorb Will Be Covered By Medicare

The Centers for Medicare and Medicaid Services (CMS) has published a local coverage determination confirming that Relizorb—an enzyme cartridge to help in digestion for cystic fibrosis (CF) patients receiving their nutrition

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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](mailto:articles@usacfa.org). Or go to our website: [www.cfroundtable.com/newsletter](http://www.cfroundtable.com/newsletter).

**Autumn (November) 2021: Caregiving with CF.** (Current issue)

**Winter (February) 2022: CF and Survivor's Guilt.** How do you manage your survivor's guilt? How has Trikafta affected your survivor's guilt? What triggers, if any, exacerbate your guilt? If you have had a transplant, how has that impacted any survivor's guilt?

**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 triaging and early prevention of common issues with aging?

**Summer (August) 2022: Dealing With Pain.**



# ASK THE ATTORNEY

## Remote Work For People With CF

By Beth Sufian, J.D.

**M**any employers have ended remote working arrangements, requiring all employees to return to in-person work. However, the COVID-19 pandemic continues. Many people with cystic fibrosis continue to be concerned about exposure to and infection with COVID-19 given the prevalence of the highly transmissible Delta variant. Many people with CF have successfully worked remotely during the past 18 months and want to remain working remotely (often called tele-work in federal guidance).

*Citations to published guidance from the United States EEOC, OSHA, and the CDC can be found at the end of this article.*

*Nothing in this article is meant to be legal advice and is only meant to be information. If you have questions about employment-related issues, Social Security benefits, education, or health insurance, you can contact the CF Legal Information Hotline and set up a time for a no-cost confidential call by emailing [CFLegal@sufianpassamano.com](mailto:CFLegal@sufianpassamano.com).*

**Question No. 1:** My employer says that a person with CF is not at risk of a bad outcome from a COVID-19 infection and therefore I cannot continue to work remotely.

The CDC has acknowledged in its COVID-19 guidance that people with CF (either with or without solid organ transplant) are more likely to become seriously ill if they contract COVID-19.<sup>1</sup>

By “seriously ill,” the CDC means that a person with CF (or one of the other underlying conditions it identifies) has a greater likelihood of needing hospitalization, intensive care, requiring ventilator-supported breathing, or

dying with COVID-19 compared to a person without health issues.<sup>1</sup>

Exposure to COVID-19 in the workplace is also a serious concern. The CDC and public health authorities have found that COVID-19 is highly contagious, potentially fatal, and spreads readily in the community. Based on these findings by the CDC, the U.S. Equal Employment Opportunity Commission (EEOC) concluded that COVID-19 meets the definition of “direct threat” because it presents a significant risk of substantial harm to the health or safety of others in the workplace.<sup>2</sup>

The U.S. Department of Labor Occupational Safety and Health Administration (OSHA) has also recognized that the SARS-CoV-2 virus (which causes COVID-19) is a significant health and safety risk in the workplace to workers with underlying conditions regardless of their vaccination status.<sup>3</sup> OSHA affirms that workers with disabilities may be legally entitled

to reasonable accommodations that protect them from the risk of contracting COVID-19.<sup>3</sup>

**Question No. 2:** When does an employer have to provide a reasonable accommodation under the Americans with Disabilities Act (ADA)?

Under the ADA, a covered employer must provide a reasonable accommodation to a qualified individual with a disability when an accommodation is necessary for the disabled employee to perform the essential functions of the job, or otherwise receive equal terms and conditions of employment. Reasonable accommodations are simply adjustments or modifications in the manner or circumstances in which a job is usually performed.<sup>4</sup>

Remote work is within the scope of potential reasonable accommodations, but whether remote work should be permitted as a reasonable accommodation depends on the circumstances of the individual employment situation. The duty to provide the accommodation arises when it is requested, or when the need is clearly apparent.<sup>4</sup>

However, employers covered by the ADA and Rehabilitation Act are not obligated to provide an accommodation that is unduly burdensome.<sup>5</sup> “Undue burden” is an objective standard based on the actual costs or administrative burdens of providing the accommodation.

Not all employers are covered by the ADA or the Rehabilitation Act. The ADA covers employers with 15 or more employees. The Rehabilitation Act has similar protections for disabled employees, and the Rehabilitation Act applies to employees who work for the federal government or who work for an employ-



BETH SUFIAN



er who receives financial assistance from the federal government, often known as a federal contractor. There is no employee minimum number.<sup>4</sup>

**Question No. 3:** All employees at my workplace performed remote work for the past 12 months or more. My employer now says everyone must work in-person. Can I remain working remotely?

A person with CF is a person with a disability under the ADA if they have a physical impairment that substantially limits a major life activity. Typically, a person with CF has a limitation in breathing or digestion. As an individual with a disability, an employee with CF can request to continue remote work as a reasonable accommodation even if an employer has said that all employees must work in-person. Whether an employee can work remotely as a reasonable accommodation depends on several factors. The increased risk of serious illness from COVID-19 for people with CF is a disability-related limitation that weighs in favor of allowing remote work.

However, the need for the reasonable accommodation must be communicated to the employer and every accommodation must be individually assessed. If an employer recalls employees from remote work to in-person work, a worker with CF will need to request remote work as an accommodation. The employer must then engage in discussions with the employee to evaluate whether a disability-related reason requires remote work. If there is a disability-related reason, then remote work can be provided as a reasonable accommodation.<sup>4</sup>

**Question No. 4:** I asked my employer for a reasonable accommodation, but my employer says that the ADA does not apply during a pandemic. Is my

employer correct?

The employer is not correct. Laws protecting the rights of workers including the ADA have not been suspended and continue to apply during the COVID-19 pandemic.

The EEOC specifically stated that the United States equal employment opportunity laws, including the ADA and Rehabilitation Act, continue to apply during the time of the COVID-19 pandemic, and they do not interfere with or prevent employers from following the guidelines and suggestions made by the CDC or state and local public health authorities about health and safety precautions.<sup>5</sup>

**Question No. 5:** My employer says that the COVID-19 pandemic is over, so I cannot have a reasonable accommodation. Is my employer correct?

The pandemic has not ended. A covered employer's obligation to provide a reasonable accommodation to a qualified individual with a disability exists without regard to whether a national emergency is declared. In addition, the COVID-19 national emergency was extended on February 24, 2021, and remains in effect nationwide.

**Question No. 6:** I do not have CF or another disability, but my adult child has CF and lives with me. I work in a place that has many people from the public visit each day. Is my employer obligated to provide me with the reasonable accommodation of remote work to protect my adult child?

An ADA or Rehabilitation Act covered employer's obligation to provide a reasonable accommodation is owed to an employee with a disability. An employer is not obligated to provide a reasonable accommodation to a non-disabled employee, even if the accommodation is needed to protect the health and safety of a disabled child or spouse.<sup>5</sup> However, an employer may

always voluntarily provide remote work to an employee even without the legal obligation to do so.

The ADA and Rehabilitation Act protect non-disabled employees from discrimination based on their association with a disabled person. If an employer took an adverse employment action against a non-disabled employee because that employee had a disabled child, spouse, or other disabled associate, then the non-disabled employee may have a claim for discrimination under the ADA or the Rehabilitation Act based on the association with a disabled person.

**Question No. 7:** I have not disclosed my CF diagnosis to my employer. Is it true that when I request a reasonable accommodation, I do not have to disclose my condition to my employer and they cannot ask what health condition causes my disability?

It is correct that an employer cannot make disability-based inquiries of a **job applicant** if the employer is covered by the ADA or the Rehabilitation Act of 1973. However, an employer may ask disability-related questions and conduct medical exams after an offer of employment is made and before work begins, as long all entering employees in the same job category are subjected to the same questions or examination.<sup>5</sup>

After an employee begins work, an employer may make disability-related inquiries and require medical examinations only if such questions and examinations are job related and consistent with business necessity. Any medical or disability-related information collected by an employer, regardless of when it is obtained, must be treated confidentially and, if retained, must be kept separately from other employment information.<sup>5</sup>

However, when an employee requests a reasonable accommodation,

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## What is the Boomer Esiason Foundation?

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

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

## What does BEF do?

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

- Educational Scholarships
- Lung Transplant Grant Program
- CF Patient Disaster Relief Fund
- Covid-19: CF Patient Assistance Program
- Team Boomer
- You Cannot Fail
- You Cannot Fail Hospital Bags & Survival Kits
- Gunnar Esiason's Blog
- Gunnar Esiason's The State of Health Podcast
- Jerry Cahill's Cystic Fibrosis Podcast
- Did You Know Video Series
- CF Education Days & CF Speaking Engagements

[www.esiason.org](http://www.esiason.org)

an employer **may** ask questions or request medical documentation to determine whether the employee's disability necessitates an accommodation. Obtaining this information is permitted because it is necessary for the employer to know the nature of the disability, how it affects performance of job functions, and how the disability can be accommodated.<sup>5</sup>

Typically, a letter from the treating physician is good evidence. Sometimes physician offices use a template letter that they use when a patient is filing an application for Social Security Disability benefits, which talks about all the complications CF may cause. This type of letter is not effective when requesting an accommodation under the ADA. If a letter makes it sound like the employee is too sick to do their job, the employer may conclude that the employee is not an employee who is protected by the ADA. The employee with CF should make sure the treating physician's letter provides information about why the person **CAN** perform the essential function of the job with a reasonable accommodation.

The ADA allows the employer to ask: (1) how the disability creates a limitation; (2) how the requested accommodation will effectively address the limitation; (3) whether another form of accommodation could be effective; and (4) how a proposed accommodation will enable the employee to continue performing the essential functions of the job.

If an employer fails to provide a reasonable accommodation, the employee can file a Charge of Discrimination with the EEOC within 300 days of the date the discrimination occurred. The EEOC now allows employees to file a Charge of Discrimination online at [www.eeoc.gov](http://www.eeoc.gov). Most states have state disability laws that provide a similar right to a reasonable accommodation. A person can also file a charge of discrimination with a state agency that will also inves-

tigate the charge of discrimination but often the time limit to file a charge is 180 days. A person should check their state law to determine the time limit for filing a complaint of disability related to disability discrimination.

## Endnotes:

1. People with Certain Medical Conditions (CDC Update August 20, 2021). <https://www.cdc.gov/coronavirus/2019-ncov/need-extra-precautions/people-with-medical-conditions.html>
2. Pandemic Preparedness in the Workplace and the Americans with Disabilities Act, ¶ II.B.; 29 C.F.R. § 1630. <https://www.eeoc.gov/laws/guidance/pandemic-preparedness-workplace-and-americans-disabilities-act>
3. Protecting Workers: Guidance on Mitigating and Preventing the Spread of COVID-19 in the Workplace. <https://www.osha.gov/coronavirus/safework>.
4. Pandemic Preparedness in the Workplace and the Americans with Disabilities Act, § C. <https://www.eeoc.gov/laws/guidance/pandemic-preparedness-workplace-and-americans-disabilities-act#23>.
5. What You Should Know About COVID-19 and the ADA, the Rehabilitation Act, and Other EEO Laws § D.13. <https://www.eeoc.gov/wysk/what-you-should-know-about-covid-19-and-ada-rehabilitation-act-and-other-eeo-laws>; and 29 C.F.R. pt. 1630 app. § 1630.2(o). ▲

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](mailto:CFLegal@sufianpassamano.com).

hint of objection, to take this unwelcome poke in my arm.

Most people aren't raised like this. Most people in our world haven't learned from a very young age that they routinely have to tolerate uncomfortable procedures to stay alive. Most people also haven't been conditioned to learn the names of the bacteria that colonize their lungs when they are sick. They don't have the direct lived experience of noticing that feeling like crap correlates perfectly with the green color of their sputum, which indicates that *pseudomonas* is on another rampage inside their body. To a person with CF, terrain theory is absolutely, mind-numbingly crazy.

But we are a minority. Fortunately, most people who don't have CF also believe in science, specifically in microbiology and virology. As I type this, there is a slim majority of people in the U.S. who are fully vaccinated against COVID-19. Today, the rate is 53.6%. Hopefully, by the time you read this, it will be higher, but reading about the terrain theory people has wiped out my hope that the vaccination rate will be the thing that ends this pandemic. Since access to the vaccine isn't actually a problem now, I fear the 46.4% who are not vaccinated have made a conscious decision not to do so.

Decisions like this are very personal, of course. The mRNA vaccinations are newer technology, and many people are very wary about subjecting their bodies to what they consider "unproven" science. They also likely have no idea how clinical trials are run, and therefore don't trust that adequate scrutiny of this newer technology was conducted at such a fast pace. Even I was amazed at how fast the vaccines were rolled out, being very aware from a young age at how frustratingly slow it can be for a new drug to be approved.

For me, this lightning speed was evidence of how much science has progressed. For the anti-vax people, I suspect this speed was evidence that the books were cooked, that we are all being

deceived, and that this is all a ploy to take away our freedom and allow the rich to get richer. We all come from different backgrounds, with more or less exposure to science, and with more or less trust in corporate America. Hence, the divide.

When I think about my decision process regarding the vaccine, it didn't take very long. The decision was automatic. Of course, I would get the vaccine. In fact, my main concern was how fast could I get it? What led to this automaticity of the decision? Well, it started with

act the way they do because of what they have experienced in life and what they have been told and believe. After all, when we enter into this world, we have no beliefs about anything. They are all acquired at some point. People who don't believe vaccinations work have clearly lived lives filled with very different experiences from mine. To call them crazy or idiotic, while it may feel good in the moment, is not seeing the big picture.

I have good friends who are in this

“Now I'm sure that readers of this particular publication are well versed in the scientific theory/evidence that bacteria and viruses do, in fact, cause illness, so I don't need to dispel terrain theory here.”

those mandatory flu shots. That conditioned me to understand by direct experience that vaccinations work. I rarely got the flu. Then I learned the science. Even before medical school, I loved science. I loved the scientific method and I trusted that using the scientific method got us as close to "the truth" as humans can get. Then I learned, through my own life with CF, as well as from professors of microbiology, about bacteria and viruses and how they do damage. I viewed them under the microscope (bacteria) and electron microscope (viruses). I learned that they exist. At some point, I did autopsies on people who had died from infections with bacteria and viruses. Talk about real-life experience!

So I had no doubts and the vaccination decision was automatic, like a reflex hammer causing a leg to kick. But I'm not your average bear. It helps me, when I read about terrain theory or see anti-vax or anti-mask people asserting their rights, to realize that people only

category. They won't get the vaccine. Some don't trust it, and others don't even believe that COVID-19 is real. A man I look up to in many ways believes in terrain theory. He is a good man with a good heart (he also happens to live in a very healthy body). At the end of the day, as I lie on my deathbed (hopefully not with COVID-19), I wonder what decision would sit best with me? Calling these other people stupid and shutting them completely out of my life or trying to understand their decision? And by "understanding" I don't mean "agreeing with" their decision. After all, we all want to be healthy. We all have an inborn instinct to do whatever we can to stay alive. We just have, at this moment in history, very different ideas of how that actually works. ▲

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*Dr. Julie Desch is 60 years old and has CF. She lives in San Rafael, CA, with her partner and their three dogs. She enjoys biking, meditating, and filling her days with joy.*



# SPIRIT MEDICINE

## Spiritual Armor

By Isabel Stenzel Byrnes

In the corner of my house is a new prized possession. It is an imposing, life-sized Japanese samurai with my mother's Arima family crest on the chest. The metal armor is intricately put together with leather and fabric ties. The helmet and facemask are daunting and will likely ward off an intruder who mistakenly thinks it is a person standing there. This was a gift from my Uncle Juichi, who boasts of coming from samurai blood.

Armor like this was worn for centuries when men went to battle. After several trips to European museums, I was always in awe of the heavy, bulky steel armor. I couldn't imagine anyone moving in these outfits. And yet, going into battle without armor meant almost certain death.

Regardless of whether we are into samurai or medieval armor, we all carry our own armor to protect ourselves in this world. We wear this armor in our battles and we choose what kind of armor we need for particular fights. There is so much hurt and pain in life that we can be weighed down by it all. Armor provides a barrier between us and that hurt. We would be too naked and vulnerable without it. What types of spiritual and emotional armor do you use? How much does your armor weigh you down?

When you have cystic fibrosis, character armor "forms in response to chronically stressed and traumatic events," according to alternative medicine practitioner, Ben Oofana. Along with our innate temperaments, how we cope with CF and the stresses it impos-

es creates character defenses and habitual patterns and attitudes. This armor can serve us and help us adapt or it can become problematic.

I'm pondering common types of armor we might use to deal with cystic fibrosis. *Achievement* can help us feel worthy and accomplished despite being sick. *Self-confidence* helps us feel good about ourselves in spite of, or because of, CF. *Passivity*, in the form of non-adherence or resignation, protects us

from the energy of fighting or the fear of failure or self-blame if we try to face a challenge and lose. *Hedonism* protects us from the harshness of life and helps us enjoy being alive no matter how much time we have. *Hypervigilance* gives us safety to help us feel we can catch illnesses or problems early, but it can also make us stuck in fear and what-ifs. *Boundaries* protect us from dysfunctional relationships that can deplete our energy and hurt us. Even *caregiving*

protects us from feelings of abandonment and gives us purpose and worth. *Stoicism* can protect us from feeling. Sometimes we have to suppress and inhibit our feelings to just get things done or keep

our cool during a medical procedure. Regulating our emotions is the heaviest armor we have to carry. *Numbing* or *avoidance* protect us from pain and give us an escape from unpleasant feelings. *Vulnerability* protects us from the exhausting energy of stoicism; we release our sorrow and pain when we cry alone or to a loved one, and we can acknowledge our suffering. Sometimes unprocessed emotion can lead to tightness and tension, creating a hardening around the body, much like the exoskeleton around a lobster. This kind of emotional armor is not good for the body. Just pause right now and take note of your physical sensations. Do you notice any armor around your body?

Real armor is made of metal, the strongest material. Emotional and spiritual armor has to be made of a strong material: control. Control helps us with fear and anxiety, gives us some

*"No matter what happens to our physical bodies, our spiritual armor can still lead to a happy life."*



ISABEL STENZEL BYRNES  
WITH HER UNCLE JUICHI'S  
SAMURAI ARMOR.



predictability, and gives us power. It builds our confidence and stretches our potential. Yet this kind of armor can backfire if it turns into rigidity, micromanaging, and criticism. In fact, judgment is a type of control armor that protects the ego by making us seem better than the other. With CF, controlling our healthcare demands can be empowering, yet it can lead to over-identifying with illness. Control is maladaptive when it is fueled by excessive fear (or power). I think the pandemic gives us ample evidence of that.

Faith is a significant type of armor as well; one where we put trust in the general of the army fighting the battle we are in. Faith means believing we are protected by a higher force, and that that force has our best interest and survival in mind. Reciting scripture that inspires and uplifts us serves as protection from pain for some. Spiritual armor helps us feel we are on

the right side of the perceived battle between good and evil and life and death. In some cases, spiritual armor can backfire if extreme views make it harder to empathize with others' perspectives.

In CF, faith as armor can be life sustaining. It might manifest itself in a belief that you can become a parent or will reach old age or that a modifier drug will come out for your mutation. Believing in life after death is armor to reduce our fear of death and diminish our grief over separation from our departed loved ones. Spiritual armor also means wearing things that will bolster your wellbeing, like self-compassion, self-care behaviors, mindfulness, or affirmations. No matter what happens to our physical bodies, our spiritual armor can still lead to a happy life. I think that's what Claire Wineland, CF advocate, meant by, "We teach sick people that when they are sick, somehow, some-

way, they cannot be as happy as normal, healthy people. We teach them that their happiness, their contentment in life, their joy in life, is tied to how healthy they are. We *can* be happy."

As I said before, armor can be heavy and bulky. In life we need to be able to move freely and nimbly. Can we be stealthy in armor? We have to learn how to wear it right and how to move in a certain way. We have to shed the pieces that weigh us down and impede us. Then we can face our battles with the greatest chance of winning. This is our great lesson—to be able to carry all the types of character armor we possess that promote our freedom, inner peace, and harmony with others. This is our true protection, and our most prized possession. ▲

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*Isa Stenzel Byrnes is 49 years old and has CF. She lives in Redwood City, California. She is 17 years post-lung transplant.*

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## TILLMAN continued from page 3

through enteral (tube) feeding—will be covered by Medicare. Relizorb is a first-of-its-kind digestive enzyme cartridge that can be attached to the end of a feeding tube. The cartridge mimics the function of pancreatic fat-digesting enzymes (lipase), so that as feeding formula passes the cartridge, fat molecules are broken down into components that are easier for the body to absorb. Relizorb can break down up to 90% of fat molecules found in most enteral feeding tube formulas, including molecules that are comparatively difficult to digest but are critical for growth and development, such as certain omega-3 fatty acids. With the newly issued LCD, the CMS has basically stated that Relizorb is both reasonable and necessary as a treatment for eligible patients. As such, Relizorb will be covered by Medicare.

<https://tinyurl.com/yfjm8o9h>

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## Functional Respiratory Imaging Offers Value In Cystic Fibrosis

Air trapping and pulmonary blood distribution were found to be clinically relevant functional respiratory imaging (FRI) parameters associated with spirometry and the 6-minute walk test (6MWT) in patients with CF, suggesting these FRI variables could offer value in the assessment of functional characteristics of the CF respiratory system. The FRI modality comprises a combination of high-resolution computed tomography (CT) images and computational fluid dynamics to gain objective and quantitative insights into lung structure and function. Investigators conducted a cross-sectional analysis in which the FRI outcomes were regional airway volume, airway wall volume, airway resistance, lobar volume, air trapping, and pulmonary blood distribution. The CF-CT score was used by 2

investigators to independently evaluate structural abnormalities on the CT scans. Patients also underwent spirometry and the 6MWT. A potential limitation of this study included the lack of controls without CF. Additionally, the researchers noted that additional investigation is needed to evaluate the association between longitudinal changes of FRI parameters and other relevant clinical outcomes. However, the investigators concluded that the set of structural components of FRI providing quantitative, objective and regional information have the potential to complement results derived from conventional outcome measures in future CF research as an alternative to visual CT scores.

<https://tinyurl.com/yjduljk8>

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## Joint Disease More Common With *P. aeruginosa* Infection, Female Sex

Continued on page 14



## PEARLS OF WISDOM

# There Are No Spoons: Caregiving As Reality Sharing

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

**M**y life's most important lesson about caregiving began with a spoon—or more accurately, with the lack of one.

Many years ago, before my wife and I were married, we commuted back and forth between our respective apartments in Tampa and Tallahassee. When I would come back to Tampa after being away for a few weeks, I would often find small things changed in our home: new art on the walls, new records or books on the shelves, new food in the cabinets and refrigerator. That weekend though, I felt more surprised by what I did not find: a single spoon in the entire place. I was making tea in the small kitchen space, looking for something to stir in the sweetener I had added.

I felt baffled by this. I also felt certain that the complete absence of spoons in the apartment had a story behind it. So, I walked over to J where she sat on the couch and said, “You know, I could have sworn we had some spoons last time I was here.” My wife immediately looked sheepish. She cast a sidelong look at the floor and explained, “I had to get rid of them. They were mocking me.”

I should back up a bit. Just as I live with chronic physical health issues because of my CF, my wife lives with chronic mental health conditions. One of the specific things J experiences is depersonalization, a process in which people simultaneously become alienated from their own sense of self and

project that sense of self onto inanimate objects. Depersonalization often stems from early life trauma, such as abuse experienced in childhood. It can become less profound over time as people experience healing and feelings of safety with those they love. This has happened for my wife, who these days will say with a grin that it has been a while since any cutlery spoke to her. But at the time, the spoons were a threat and had to be eliminated.

I could have said many things in that moment. Only one seemed appro-

priate, though. After all, it did not matter whether I thought the talking spoons were “real.” All that mattered at the time was the fact that J was authentically experiencing hostile communication from the spoons and took action to improve her own quality of life by removing them from the premises. My wife discarding the spoons seemed quite reasonable to me given how they were tormenting her. So I looked thoughtfully at J, then down at my cup, and then back at her. I nodded and said “okay.” And then I went back to the kitchen and stirred my tea with a fork.

That day, as on so many others, I was in the Matrix. I find the Wachowski sisters’ iconic film as good an allegory as any for the process of understanding and negotiating the socially (and sometimes technologically) constructed nature of reality.

On this occasion, there quite literally *was no spoon*. But in a sense, all things in our worlds are spoons—because our ability to interact meaningfully with them hinges on us not only constructing reality, but also sharing that reality with others. It thus seemed unsurprising to me as I progressed in my own graduate studies, and later in my work as a faculty member, that research clearly demonstrates acceptance of another person’s reality as one of the most powerful forms of caregiving.

And paradoxically—to reference another theory of “spoons” coined by fellow chronic illness patient Christine Miserandino—allowing ourselves to embrace with open arms the lack of spoons in a concrete sense can often

*With others, reality sharing likewise starts with acceptance. It means being fully present in the moment with another person, accepting how the world looks and feels to them.*



XAN NOWAKOWSKI

increase the “spoons” we have in an abstract one. In Miserandino’s Spoon Theory model, “spoons” represent the internal resources we bring to each day in living with our respective chronic conditions. These resources can be cognitive, emotional, social, etc. The amount and diversity of “spoons” with which we start each day shape how that day will go—and how exhausted we will become in trying to get through it.

My entire philosophy of caregiving thus begins and ends with the fact that I do not ever want to take spoons away from my wife in an abstract sense. And if that means she must sometimes take spoons away from me in a literal one, so be it.

I have been thinking a lot about spoons lately. I am just now recovering from a positively nasty bacterial sinus infection in which some 100+ teaspoons of neon orange muck emerged from my face. And yes, I measured! If I am to produce such ghastly secretions, I can at least find solace in quantifying them for later discussion with my clinic team. Collecting data increases my own “spoons” by distracting me from the physical pain of an acute infection. I have had people tell me that I am too data-driven, too clinical, too analytical in how I cope with my CF from day to day. I have told most of those people to stop talking and start listening, because caregiving looks different from person to person.

The care I give to myself reflects the same things I value in the care I receive from others—and the care I give to them in kind. I value that complete openness to my own unique personality as someone navigating lifelong chronic illness, and to the unique things I experience along the way. I value observation and affirmation over inference and judgment. And being a scientist, I also value the robust evidence basis for these practices.

Reality sharing has long been uplifted in the dementia care literature

as a transformational practice for helping people with memory loss and their loved ones enjoy time together and experience mutual joy. Rachael Wonderlin, one of my favorite authors in this field, describes how abandoning the paternalistic practice of attempting to convert others to our own experiences of reality liberates ample time and energy for sharing in the reality of our loved ones and making the most of quality time.

I have seen the value of this approach consistently in my own work as a medical sociologist and public health program evaluator focused on health equity in aging with chronic disease. For example, I have served for several years as the qualitative methodologist for the ACTS 2 skill building and support project centering Black dementia caregivers. The model for all the services and resources we provide to family caregivers focuses centrally on finding community and serenity with loved ones with dementia. This includes sharing in loved ones’ realities from moment to moment as caregivers also navigate their own realities with appropriate support. I have also served for several years as the evaluator for the REACH geriatrics workforce enhancement program at FSU. We offer several different trainings and resources to help family caregivers of all backgrounds and experiences live their best lives while providing impactful support for their loved ones. Our courses and materials consistently nurture skills and practices of reality sharing.

And I have likewise seen the value of reality sharing in how my wife cares for me as someone living and aging with CF. Unlike many of the people I have been close with in the past, J never expects me to perform health or hope in particular ways. Rather, she wants to share in whatever I am authentically experiencing on a given day—be it wellness or sickness, hope or despair. Her ability to be present with

me emotionally in a struggle without either centering or sidelining herself has taught me endless lessons about the meaning of love and caring. I often reflect that love is a verb more so than a noun; our actions are ultimately what communicate our affection for others.

Living with CF means spending ample time negotiating others’ expectations. We are bombarded with copious and often conflicting expectations from the people who care for us both formally and informally. We face complex expectations about how we will care for ourselves on a daily basis—what treatments we will choose, how we will monitor our health for signs of change, what interventions we will open ourselves to in times of crisis, how we will feel about all of it, and how we will behave as a result. We often spend so much time trying to manage these expectations and the feelings of others surrounding them that we can become lost to ourselves in the process. CF is an all-consuming disease socially as well as physically and mentally.

So, reality sharing can be absolutely transformational in how we care for ourselves and each other. Within ourselves, this can take the form of intentionally cultivating acceptance about changes in our health and our bodies. I often think about how my friend Dominic, who published some of his art in the last issue of *Roundtable*, nurtures this acceptance by exploring the full artistic potential of his post-transplant body. With others, reality sharing likewise starts with acceptance. It means being fully present in the moment with another person, accepting how the world looks and feels to them. I did this with my wife when the spoons disappeared, to give one example from a decade of shared history. And she does this with me every single day, whether I am coughing out bloody discharge and struggling to breathe or

Continued on page 14



# SAVORING SERENDIPITY

## Walk With Care When Grace Comes Running

By David Tarnow

Admittedly, I do not have a lot of experience as a caregiver. I am not yet a parent and my parents are not yet at the point of needing care. Like many individuals with CF, most of my experience with caregiving has been on the receiving end. I will not pretend as if I have anything to offer in the way of advice to anyone else who is currently in a caregiving role. God bless you if you are. What I will do, however, is take this opportunity to share some insights into my own shortcomings and lessons learned from my experience in the trenches of severe lung disease and end-stage CF pre-Trikafta.

The first thing you should know is that it wasn't pretty. In fact, most of the time it was downright ugly. My health declined fast and furiously, and none of us was ready for what was to come. My mom became my fulltime caregiver when I was no longer capable of taking care of myself. There was an odd tension between us before I acquiesced to the fact that, yes, I needed help. I'm sure pride and stubbornness played a role, along with a healthy dose of denial. My mother hesitated taking on certain responsibilities of mine after so much of my independence had already been ceded as a result of my ailing health.

The following months, during which I found myself in need of a significant amount of caregiving, were some of the most challenging times I have ever been through—I had never mentally prepared for finding myself in

a place where I was utterly reliant on others in order to fulfill many of my most basic needs.

If I could go back and give myself some advice, it would be this:

- Take it easy. Take it easy on yourself—what you are going through is hard. Take it easy on your mom, too—what she is going through is even harder.
- Your suffering affects everyone else around you, even though you may not realize it at the time. It is easy to take the myopic view that you are the only one going through

this. Don't allow yourself to be deceived by this false narrative.

- You are fortunate to have someone who loves you unconditionally and is willing to put up with you on your worst days. Not everyone is so fortunate. Be grateful, and don't take this for granted.
- No one expects to lose their independence and functional ability at such a young age, and, when you do, you won't be prepared. Don't be afraid to ask for help.
- This process is full of difficult emotions and experiences, many of

which will remain unspoken or unacknowledged. Don't keep them all to yourself. Life is better and richer when shared.

- There will be days when you feel like giving up. Don't.
- Be adaptable. Just as your caregiver will learn to adapt to your changing

*I had never mentally prepared for finding myself in a place where I was utterly reliant on others in order to fulfill many of my most basic needs.*



DAVID TARNOW

needs, you must be cognizant of the needs of your caregiver. We are all human.

- Find some meaning and purpose if you can. Reaching a point in life where you are no longer able to act of your own accord and volition requires that something fill this void. Try not to let it be bitterness and anger. Living for something outside of yourself is the best solution.
- Try not to live in your head so much. Living from the heart is the best way forward. A rigid intellect is an unwelcome companion when trying to rationalize and justify questions of the existential variety.

You hear the word dignity thrown around a lot when it comes to end-of-



life care or caregiving. What I can tell you, having watched people reach the end of their lives, is there is absolutely no dignity to be found in facing the stark realization that everything they once identified with or prided themselves in their ability to do is about to be lost. The best you can hope for is grace. The type of quiet and humble strength that is required to surrender totally and fully to love. Love in the form of someone else showing up and being there for you when your body no longer shows up for you. When the pain of daily living is too heavy a burden to bear and you question why it is that you are still here, grace is the answer.

My mother and other family members who took care of me when I fell to my lowest state are the epitome of grace. Although I did not arrive at this point in my life with any preparation for how to handle it, my mother sure did. She was born for it. I made a lot of mistakes my first go round, and I hope that I don't next time. Caregiving is hard on everyone. I can only hope that one day I will have the opportunity to return the favor in a meaningful way.

For me, being on the receiving end of caregiving has opened my eyes—we are all just doing the best we can. I am not sure anyone is ever quite ready for a truly demanding caregiving relationship, but if we can approach the experience with open minds and open hearts, maybe in some small way we can alleviate the collective weight of human suffering.

After all, the truth of the matter is that, in the words of Ram Dass, “[w]e’re all just walking each other home.” ▲

*Dave Tarnow is 32 years old and has CF. He lives in Erie, Colorado. Dave is the founder of “Dave’s Cycle for a Cure,” which inspired the national Cystic Fibrosis Foundation event now known as “CF Cycle for Life.” You may contact him at dtarnow@usacfa.org.*

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jumping up on her back and laughing so hard my sides hurt.

It has been years since the spoons disappeared from our Tampa apartment. Yet just as we both enjoy revisiting that story with people who are new in our lives, I continue to learn more from J every single day about what effective caregiving looks like. I try to reflect that spirit of openness and acceptance back to her in full, and to practice these approaches in kind with others in my

life, both within and beyond the CF community. We often learn at young ages that “sharing is caring” but leave behind these important lessons as we age. Indeed, sharing in the reality of those we love rather than trying to impose our own is the very essence of caring. ▲

*Dr. Alexandra “Xan” Nowakowski is 37 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.write-whereithurts.net](http://www.write-whereithurts.net)) on scholarship engaging lessons from lived experience of illness and trauma with their spouse, Dr. J Sumerau. You can find their contact information on page 2.*

Chronic *Pseudomonas aeruginosa* infection in adults with cystic fibrosis increases their likelihood of developing CF arthropathy. A significant risk for CF arthropathy was found with increasing age, a higher number of hospitalizations, CF-related diabetes, and the presence of comorbidities. Women were more than twice as likely to have CF arthropathy, as were patients with pancreatic insufficiency. Those with sinusitis or nasal polyps were also at higher risk for CF arthropathy. Treatment with anti-fungal medications linked with an almost three times greater risk of CF arthropathy in adults without *P. aeruginosa*. Arthropathy, a collective term for joint diseases, is associated with CF when patients have pain in the joints with signs of inflammation, but without evidence of periostitis (inflammation of the tissue that surrounds the bone) and with no other causes for joint inflammation.

<https://tinyurl.com/4nzxjuas>

### **Cystic Fibrosis-Related Diabetes (CFRD) And Cognitive Function In Adults With Cystic Fibrosis**

This study was carried out to assess cystic fibrosis-related diabetes (CFRD) as a mechanism for cognitive impairment in people with cystic fibrosis (CF). It was hypothesised that cognition would be poorer in adults with CFRD

than in those with CF without diabetes (CFND) or in healthy controls. Researchers evaluated cognitive performance using the Cambridge Neuropsychological Test Automated Battery which provides a comprehensive cognitive assessment with tests mapping onto specific brain regions. They recorded CF specific clinical variables for the two CF groups. The results showed that managing CF requires higher order executive function. It has been considered that impairments may be sufficient to interfere with self-care and the ability to conduct everyday tasks efficiently. Moreover, at which point in the CF disease trajectory these difficulties begin, and what may attenuate them, has yet to be ascertained.

<https://tinyurl.com/4x6ayw53>

### **Study: Use Lung Clearance Index To Measure Function**

An assessment called the lung clearance index could be used to help identify people with cystic fibrosis (CF) who are at high risk for worsening lung function. The study reported on factors associated with a worsening lung clearance index (LCI) over time in CF patients, which included initial LCI measurements and bacterial infections. LCI is a measurement of how well the lungs are able to take gas in and out. LCI represents the number of lung vol-

ume “turnovers” needed for clearance of a tracer gas; a higher value is indicative of worse lung function. LCI may provide useful clinical information, it is not used routinely in many places, in part because the assessment can be complex to administer. Another challenge is that questions remain about how this measurement tends to change over time. In this study, a research team reported data from a prospective study in which LCI was incorporated into routine clinical practice at three specialty centers. Because they were interested in how measurements might change over time—in the absence of noteworthy clinical changes—the researchers specifically looked at patients with relatively stable CF. The majority of patients with predominantly mild CF fell into a group with stable LCI throughout the course of the study.

Still, other trends were seen in a minority of patients. One in 10 started with LCI measurements about normal, and then the values increased over time (indicating worsening lung function). A similar proportion started with higher-than-normal measurements, which then worsened over time. In statistical models, the factor most powerfully associated with changes over time was LCI at baseline (at the start of the study). Other significantly associated factors

Continued on page 19

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

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

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

Nancy Wech established this scholarship in honor of her daughter, Lauren Melissa Kelly. This semester's winners demonstrated outstanding potential, just like Lauren years ago. Lauren was an inspiration to all who knew her. An incredible leader and scholar, her drive and success are the foundation of her memory. She was transformative in every aspect of her life. She had distinguished herself as a member of the Golden Key Honor Society, Mortar Board, Phi Upsilon Omicron, Gamma Beta Phi, Delta Gamma sorority, and was chosen as one of ten Senior Leads at the University of Georgia. She acted as one of the re-founding members of the Phi Kappa Literary Society and was significant in the metamorphosis of the Z Club into the William Tate Society. Although Lauren lost her battle with cystic fibrosis late in her senior year, her hard work and memory continue to live on through her inspiring involvement.

We are pleased to announce **Emily Schutz** and **Julia Kennedy** as the recipi-

ents of the scholarships for this calendar year. They will each be awarded \$2,500. Congratulations to both!

Emily Schutz is an accomplished young woman pursuing her bachelor's degree in English and Media Communication Studies at Florida State



**EMILY SCHUTZ**

University. Her writing has been featured in several publications in the last two years. Additionally, Emily was the Editorial Shoot Director for the *DWF Magazine* in Tallahassee for one semester in addition to serving as Graphic Designer for Her Campus FSU. Currently, she serves as both Secretary and Social Media Coordinator for Sigma Tau Delta in Tallahassee. Outside of her schoolwork, Emily is busy volunteering her time both within the CF community and within her wider campus community. Emily dreams of sharing stories with the world, especially the stories from those who need a voice.

Julia Kennedy is a motivated young woman who holds both a Bachelor of Arts in Social Studies and a Masters of Art in Educational Leadership from Michigan State University. She is currently complet-

ing the Special Education Supervisor Program offered through Wayne State University. Julia has worked as a resource room teacher in several elementary schools in Michigan for the past several years. In addition to her educational accomplishments, Julia has advocated tire-



**JULIA KENNEDY**

lessly for persons with CF in her community. In April of 2019, she and her husband met with Michigan representatives and senators to advocate for the continuation of medical benefits under the Children's Special Health Care Services of Michigan Medicaid programs.

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

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



## FOCUS TOPIC

### CAREGIVING WITH CF

# Self-Care While Caring For Others

By Gillian Mocek

**W**hen I saw two lines on the pregnancy test, I knew I was starting a journey that was going to change my life. I always imagined that moment to be one filled with unbridled joy. However, in reality, I was as scared as I was happy. I deeply wanted to raise children and have a family to call my own. I also didn't know how exactly I was going to do that well while also managing my CF.

I'm grateful to say that, since that day six years ago, I've figured some of that out. My son Simon, who is five, helped me learn many lessons along the way. He, along with my ten-month-old daughter Eloise and husband, Christian, together with our dog, Teddy, and cat, Rosa, make up my village of supporters. Among them are also my CF care team, our family, and our friends. The many recent medical advances in CF care have made this possible for me, too.

We have an active, hopefully intentional household and one in which I'm the manager and primary caregiver. I also work part time as a Community Support Specialist for the Cystic Fibrosis Foundation. Two kids, two animals, my husband, my job, and CF are a lot to balance. And as hard as I work to maintain a healthy balance of all those things, I don't always get it right.

But I do know I have come a long way from where I started and I am happy to share a few lessons I've learned about caregiving and balancing CF.

- **Self-care plan:**

Being a caregiver for others means I first need to care for myself. When I was in graduate school for my master's

degree in social work, every semester we had to develop a self-care plan. At first, these exercises felt redundant. However, as the semesters went on, I began routinely checking in with myself so much that it became a habit. I would review what I was doing to take care of myself and maintain my boundaries.

These plans had to include specific actions, a timeline for completing them, and a person to hold us accountable. Through that experience, I learned to hold myself accountable and



GILLIAN MOCEK

to routinely check in with myself. When I am feeling off, I know there is an unmet need. These self-care plans have also been useful to me as a caregiver. When my child is having a rough day, it's not because they want to make my life miserable, it's because they have an unmet need.

- **Therapy:**

My therapist is at the foundation of my self-care. She supports me

through living with CF and it is something we talk about often. I'm able to bring my thoughts and worries to her in a safe space where I know I am supported and can work toward healing. I think it's important to normalize this as an aspect of our CF care. Parts of living with CF can be traumatic. It has been wonderful to have more tools in my toolkit to care for my mental health with therapy.

- **My people:**

Another key piece of balancing caregiving with CF is knowing the people whom I can rely on in my life. This includes my close friends and family, as well as my CF friends and coworkers, with whom I communicate in a virtual capacity. Some days my spouse knows it's just a tough day for me and comes home for lunch or takes an item off my ever-growing to-do list. Some days it means going for a walk with a friend when our kids are restless. We live about six hours from our family so I don't have as much physical help as I'd like to, but it makes the other pieces that much more important. It means communicating with my spouse about what I need—accepting and humbly *asking* for help and/or saying no when I need to. I cannot and should not do it all, even if I didn't have CF. Empowering others is a gift and a learning experience for everyone.

Particularly important in this group of people are my CF friends. Their presence in my life is critical because they understand what it's like to wake up and not feel great, to calculate risks in the pandemic, to be working hard to take care of your body, and the daily ins and outs of CF. Having their support as I am a caregiver helps normalize my particular needs and



unique situation. They help remind me to take care of my needs—physically, mentally, and emotionally.

- **Exercise:**

Exercise has been critical for me this past year and a half, during which I have been living through the pandemic, working remotely, and experiencing pregnancy and postpartum. Until I used Beam, which is an exercise platform for people with CF, I never worked out as consistently. I especially like the live classes because accountability is important to me. Most days, Simon gets his tin cans and joins in on the classes with me. He, too, has made fast friends with the people who attend the classes and shares his love for dinosaurs with them willingly. I tried out Beam for physical and mental wellness, but it was the community of people that was a gift (to both me and my kids!) and kept us coming back.

- **Routine:**

On a very practical level, routines are very helpful and they ground us in our daily life. Routine does not need to be soul sucking. I find it gives me freedom to just have those anchors in my day to care for my needs (like sleep, meals, exercise, meds, treatments, insulin, etc.) and the needs of others in my life. My days now revolve around meals, Simon's school drop-off and pick-up times, and Eloise's nap times. I fit what I can for myself around those anchors in the day.

- **Involving my kids:**

“Two kids, two animals, my husband, my job, and CF are a lot to balance.”

Another important piece of my caregiving experience is involving my kids in my care. They will always know I have CF, and I strive to be open and honest (as is appropriate for their age and understanding.) Simon is quite skilled at giving me insulin shots, helping me organize my medicine, and he is sensitive and compassionate with my needs, just as I hope he will be with others throughout his life.

- **Practicing gratitude:**

The act of caregiving is not something I knew I would get the chance to do. So many people have cared for me in my life and now I have the chance to show up for my family tenderly and consistently. That means a lot to me, and it fills me up with joy and gratitude. It is tempting to be afraid because CF could take that away.

I remember when Simon was born a little over five years ago, when we came home from the hospital I remember very distinctly wondering “how long will I be here? What will I get to see?” and I decided then to make the best of the days I do get.

This quote by Mary Oliver from a poem called “Don’t Hesitate” speaks to my heart in this season of life that requires more intense caregiving for my

children. Mary Oliver says at the end of this poem that “joy is not meant to be a crumb.” This speaks to me as a person who has CF and is also a caregiver. It reminds me to be present in the moments of joy I get to experience and not let these moments be diminished by my worries for the future.

My kids show me best about the very present and tangible joy that can be found in any moment. That joy is meant to be immense! Caring for them gives me a front-row seat to their contagious wonder and awe.

Some days when my body is feeling more CF symptoms, it is harder to see the joy. I try to accept my body as it is that day and remember it is still good. Other days, CF is a lens that amplifies the joy in my life. Each night, I tuck in Simon and Eloise and tell them that I am so glad to be their mom. Every day, I wake up to the fact that it's a rare gift to be alive and care for my family. ▲

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Gillian Mocek is 30 years old and has CF. She lives in Evansville, IN. Gillian enjoys trying new recipes in the kitchen, hiking with her family, and fueling her caregiving tasks with walks to the local coffee shop. You can email her at [gillian.moczek@gmail.com](mailto:gillian.moczek@gmail.com).



## Encourage Family and Friends to Donate!

Give the gift of life that lives after you. United Network For Organ Sharing  
<https://unos.org/register-to-be-an-organ-donor/>

Make life possible. You can give hope to 106,764 people on the national organ transplant waiting list. Commit now to giving life to others when you die.



# The Conundrum Of Caring For Number One And Then Others

By *Andrea Eisenman*

One of my dad's mottos, and there were many (eyeroll emoji), was: take care of number one first! Which he had no problem doing. I always felt this was very selfish of him, and, if I were to adopt this behavior, I would also be selfish. I never wanted to be like my dad. But, in some ways, he was right. I cannot take care of others if I am not well or on top of my own treatments. But I think there are gradations in this motto—it's not all black or white. I eventually came to see that making sure I am well before I offer to care for or help others has been useful. I learned the hard way—as is mostly the case—by burning myself out for others or for work and then winding up in the hospital with pneumonia (pre-transplant). I am now better at this balancing act.

When I was younger, in my 20s, and my health was starting to decline with frequent IV antibiotics and hospitalizations, my dad constantly accused me of “not taking care of number one!” I later realized this was born out of frustration about my disease rather than about what I was doing, or supposedly not doing. That said, he was not wrong. I was mainly prioritizing fun or socializing over treatments as I wanted to be like my peers.

If I used a scale to balance my care with the care of others, I would say I usually lost that bargain. But things have changed in my favor in the last couple of decades. While in the past I tried to prioritize my health first, there were times where I sacrificed it in order to take care of my mom or my dog or my job (when I still worked). In retrospect, it was good I didn't have chil-



ANDREA EISENMAN

dren because I would have chosen their care over mine and probably would not have lived long enough to see them grow to adulthood.

One of the reasons I felt so strong about putting myself last might have had to do with low self-esteem—everyone was more important than me. And most likely, because my mom did so much for me, sacrificing so much time and most of life to keep me alive, I felt this is what everyone did for everyone else, right?

But learning how to balance my resources was important. I tried to balance my care with that of others around me. I sought to be mindful of my needs, which were many, and that of my friends, spouse, and elderly parents. My parents are now all deceased. My mom and stepmom died about a year apart and then my dad died eight months later. The losses were sad and shocking, but it was also clear to me

that my parents' care was no longer my obligation. Now I didn't have an excuse to not fully take care of my health. Sometimes what kept me so compliant in my own care was worrying about who would care for my mom if I died. Toward the end of her life, there were times when I needed to stay with her to ensure she was being taken care of—going to doctor appointments with her, cooking her food, etc. After she died, I no longer wanted to live. I had lost my reason to care for myself (forget that I am married and had a Boston terrier who was forever needing vet visits). It was a friend of mine who rightly made me see that I had to keep on living for my mom's sake. She pointed out how upset my mom would be if I gave up and stopped caring for myself. The real reality check for me was being diagnosed with post-transplant lymphoproliferative disease (PTLD), a lymphoma that can happen to people who are immune suppressed, post-transplant. It just made me angry and want to keep on fighting to live.

I've realized I am more fatigued and my energy is limited. I am currently in a clinical trial for the recurrence of PTLD. It has been extremely tiring. Instead of expending my energy on shopping for food and cooking, I realized I have a neighborhood friend who cooks for people for a reasonable fee. She is a chef, so we both benefit! I enjoy and take pride in my ability to transform my groceries into yummy delights, but I knew I needed a break to save my energy for other things. I would never have considered this option years ago. I might have ordered takeout more frequently, but I tend to get sick to my stomach if I eat too much restaurant food. I felt this was a good solution while I'm undergoing

“I eventually came to see that making sure I am well before I offer to care for or help others has been useful.”

treatment for my PTLT. I have also prioritized my exercise regimen over that of cooking. I realized cooking took a lot of my time away from things I needed to do each day like napping and exercising. Usually, I had to choose one over the other. I still do cook a few things on the weekends, which I can freeze for future meals. It is lovely to have fresh food that just needs minimal heating up.

While I feel it has taken me all of my life to get to a place where I am uber compliant in all of my treatments,

I see people respect me for it. They may think I am simply and sensibly spending hours a day on my health; however, the real reason I follow all my doctors' instructions is because I do not want to second-guess myself when I am dying!

Did I do enough? Could I have done more? And, at end-stage disease, it is too late to change anything. I try to take responsibility for my care and get creative with optimizing my health. I seek out helpful modalities like acupuncture and Reiki or other forms of healing to complement my western

medicine regimen and to help maintain a healthy balance.

All this prioritizing of my health has not stopped me from wanting to help others. I still want to, and, when I feel I can, I put myself out there. People in my life have been so supportive to me in my many times of need, I do want to give back, just more mindfully this time, with my father in mind, after I take care of number one. ▲

*Andrea is 56 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.*

**TILLMAN** continued from page 14

included age, infection with the bacteria *Pseudomonas aeruginosa*, use of antibiotics, and baseline forced expiratory volume in one second—a measure of lung function based on the amount of air a person can exhale in one second. Overall, the procedure was well-tolerated, with most patients saying it was easy to complete. The most common suggested improvement was shortening the time it took to complete the measurement (median of about 20 minutes). The findings support the use of LCI in clinical practice in identifying patients at risk of lung function decline. The measurement, however, is challenging to deliver in routine practice so may be better suited to annual assessments.  
<https://tinyurl.com/5265hdkd>

#### **Low VIP Hormone May Contribute To Early CF-Related Diabetes**

Reduced levels of the VIP hormone in the pancreas due to low nerve supply may contribute to the development of early diabetes in people with cystic

fibrosis. Cystic fibrosis-related diabetes (CFRD) is characterized by decreased production of insulin, a hormone secreted by the pancreas that lowers glucose in the bloodstream. VIP (vasoactive intestinal peptide) is a hormone known to modulate insulin secretion in the presence of glucose. VIP is produced in the nervous system and acts as a signaling molecule between nerve cells (neurotransmitter). It also is found in the respiratory, digestive, reproductive, and cardiovascular systems. VIP also has anti-inflammatory and immunomodulatory properties, and plays a crucial role in maintaining clean airways. Researchers demonstrated VIP was 50% lower in the lungs, sweat glands, and small intestines of mice that carry the F508del-CFTR mutation. The study found VIP deficiency originated from a reduction in nerve supply (innervation) starting at a young age before signs of CF-like disease were seen. The team also examined whether there were VIP changes in the pancreas, affected by

nerve supply, at different stages of CF in the same mouse model to determine whether changes in VIP levels would contribute to CFRD development. Based on their previous work, these mice showed minimal disease by eight weeks of age, whereas 17-week-old mice display well-developed CF-like disease. The data also showed that the amount of VIP is strongly reduced in the pancreas of CF mice at both early and late stages of disease progression. Finally, assessing the effect of altered insulin and glucagon production, the team found significantly higher levels of glucose in the bloodstream of CF mice. The results show a reduction in insulin and an upregulation of glucagon in the pancreas of CF mice, indicating that a CFRD phenotype can develop at an early stage (as early as 8-week-old) of disease progression.

<https://tinyurl.com/5x3bycu3>

#### **Hormonal Fluctuations Linked To**

Continued on page 25



# Familiar Is Unfamiliar

By Leah Sands

Constant anxiety runs through my mind. I try to fill my head with peaceful thoughts so that I can be present in the moments that make pleasurable memories. Some days it is easy to focus on the good, but some days are consumed by fighting through the aches in my body and in my soul. I was born with cystic fibrosis, and the life I have had so far is the only one I have known. My appetite is filled with handfuls of pills and my hours are consumed with breathing treatments and therapies. These aggressive attempts are to ease the difficulties that come from the mucus that has cemented itself inside my organs. No matter how hard I will the poison to exit from my body, it becomes something that I must adapt to and accept. The ability to do so many things has been stripped from my existence. My inability to breathe well has limited my involvement in activities. My stomach pains have brought on emergency procedures. My fears of being too far from a medical facility that can handle my health has kept me from traveling. My life came with boundaries and a strict routine, and I did not dare to deviate too far from my safety net.

Those struggling with the disease were given the possibility of relief, as scientists had developed a medication that helps the defective CFTR protein work more effectively inside our bodies. The first magic pill was only available for a small portion of those afflicted, and I was not a part of that group. To feel some hope that our disease was treatable, I hovered around those lucky enough to take the new modulators, asking for updates, hoping they were experiencing a new reality that the rest of us could only see in our dreams. After a few modifications of the drug, and several phases of research later, a miracle

drug was available for most of those suffering from CF. I had tried some of the earlier pills without much benefit, so I was looking forward to trying the latest medication, which was considered a serious breakthrough in the medical world. Our community of sick and dying was celebrating across the globe.

I never thought twice about taking the medication, as anything that might ease my tired, heaving lungs was worth it. Since most people with CF perish



LEAH SANDS

from respiratory failure, even the idea of wiping that fate from my future was almost unfathomable. However, within hours of taking my first dose, the vile mucus began fleeing from me. My sinuses began to open, and I was realizing everything had a smell. I was coughing intensely like I had been a drowning victim saved from the lake. My obstructed bowels were clearing, and the pains were flowing away. I could feel things in my body that I didn't even know could be felt. By the time I went to bed that night, I was able to fill my lungs with air beyond anything they had before. I could not pro-

cess the quietness of my breathing, so I silently slipped into a deep slumber.

Our community of desperately sick people was diminishing and our frequent pep talks were disappearing. The need for support from those that understood wasn't needed anymore because everyone was living. People were doing things they didn't think would ever be possible. Even though I was also experiencing amazing things, I longed for the relationships that used to keep me grounded. I used to walk up and down the hallways of the CF ward and chit chat with my fellow fighters from a distance, always learning a new way to deal with the disease. But instead of seeing each other in the hospital, we were seeing each other on social media living life to the fullest—images of fellow fighters having babies, traveling to extreme destinations, and being in all sorts of places other than a hospital bed. I even began to do things that I didn't think I'd have enough energy or years to accomplish—writing a book, starting a business, and traveling abroad.

I was also starting to see things from a whole different perspective. I felt like I didn't have to rush to fit everything in. I didn't have to maneuver my activities around how I was feeling. I didn't have to fit my hospitalizations into my work schedule. I wasn't tethered down to tubes and lines for hours each day. My life no longer had an early expiration date. I had a hard time figuring out what to do with all my spare time. The feelings of suffocation and impending doom that used to overtake most of my thoughts had been replaced with emptiness, and I had to find new thoughts to fill the void. I had to restructure my day and fill it with things that made me happy and filled my heart.

But being able to fill up your life where there used to be tedious

Continued on page 28



## THROUGH THE LOOKING GLASS

### For My Wife

Morning Comes  
My eyes open  
I feel the warmth of your body  
I breathe in  
No movement yet  
I watch oh so silently  
I can feel your presence  
I know, you know  
I'm watching you  
Movement  
Your hand slowly moves upward  
I watch Your hand slowly reaches  
for air  
Searching  
I lie still  
Waiting  
Anticipation overwhelms me  
Movement  
Morning sunlight shines off your  
hand  
My body feels my blood  
Still waiting  
The warmth of your hand  
so close  
Movement  
The fine hairs on my body tingle  
Then The touch  
The transfer of knowing love  
Complete  
No words  
No sound  
Movement  
With one touch  
Acknowledgement  
You know  
In my soul  
You will always know  
I love you  
In my mind, I will always find the time for you  
In my heart, we will never be apart!  
Movement

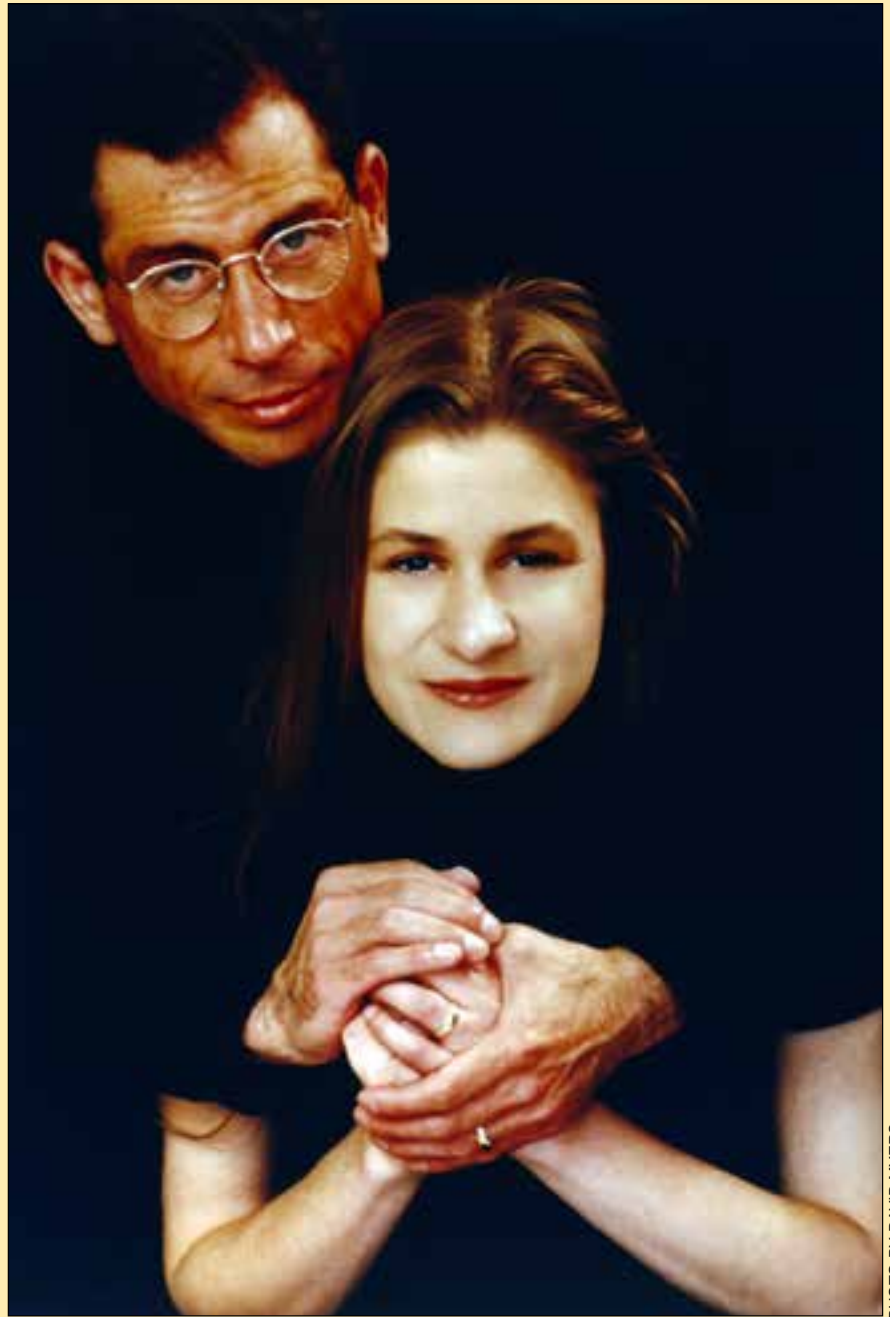


PHOTO BY DAVID MYERS

You roll over  
No words  
No sounds  
Movement  
Says it all  
Eyes touch.

-P. Howell, 1998

## FROM OUR FAMILY PHOTO ALBUM...



**ELYSE ELCONIN GOLDBERG IN KAUAI, HI.**



**GILLIAN AND CHRISTIAN MOECK WITH THEIR CHILDREN, SIMON AND ELOISE.**



**JEANIE HANLEY HOLDING HER GRAND-DAUGHTER, AMELIA HENDRICKSON.**

**LEAH SANDS  
KAYAKING AT  
PICTURED  
ROCKS ON  
LAKE  
SUPERIOR,  
AUGUST 2020.**







**ANNA MODLIN, ZOE HOLYOAK, TERRY HOLYOAK.**



**MAGGIE WILLIAMSON DRESSED UP HERE AS A UNICORN ON HER BIRTHDAY SPREADING CHEER TO HER TOWN IN SEVENOAKS, U.K. SHE PARADED AROUND HOPING TO GET SMILES FROM LOCAL RESIDENTS DURING THE LAST AND LONGEST LOCKDOWN OF COVID.**



**COLLEEN AND SCOTT ADAMSON.**



**SONYA HAGGETT IN ELDORADO NATIONAL FOREST IN 2019.**



# Caregiving And Vaccines—For Me, For You, For Everyone

By Jeanie Hanley, M.D.

Throughout my life, I've been a caregiver for both patients and families. When my own family member has a serious and/or prolonged illness that requires extended support, it can be the most daunting situation, emotionally and physically. When one of my sisters with CF was hospitalized and subsequently intubated for months, two different times, due to the flu, and when my aging parents could no longer manage for themselves, I learned quickly that, in order to advocate for them, I had to properly care for myself.

I needed to prioritize and maximize my health to have the ability to medically support others. In order to do that, it was important to keep other family and/or friends (a/k/a the "village") abreast of the medical needs and enlist their support. This opened up extra time for my own CF maintenance.

I realize I'm preaching to the choir here, but I found that our routine CF therapies are easy to throw by the wayside when advocating daily for someone else. For instance: missing one or more of what's optimal on a daily basis such as inhaled treatments, not getting enough rest, staying hydrated, and eating! It's also easy to skip on exercise, but I've found that regular exercise, at least three days per week, is necessary. I must also regularly keep appointments with my CF center and other doctors. And I make sure to get my annual flu vaccine and other vaccines as they are due—pneumococcal, shingles, etc. To have time for these visits and vaccines, the "village" of caregivers helps considerably.

Getting vaccinated protects me and those whom I treasure from preventable illnesses. Immunizations include the

“When I see others masked, I feel a kinship with them and quietly give gratitude for their considerate behavior of others while protecting themselves, too.”



**JEANIE HANLEY**

COVID-19 vaccine, which I received as soon as I possibly could. Why did I get the COVID-19 vaccine series? First, because it's effective. (See Dr. Moss's article on page 30 of this issue for the science behind the vaccine.) Second, to protect myself, you, anyone I look after (like my three-month-old granddaughter, whose immune system is not fully developed), and for everyone I know and don't know with whom I may come in contact.

The last year without the COVID-19 vaccine was a tragic time for many families who were unable to visit their

loved ones in the hospital, in hospice, or nursing homes. It was no different for me. Adding the risk of COVID-19 infection to the mix caused such heart-break during my father's last five months of life. I was unable to visit him face to face or give one last hug. Today, it is only because of effective COVID-19 vaccines that visiting restrictions can be relaxed, enabling a more peaceful end of life with loved ones.

I've already received my yearly flu vaccine, which is part of my usual autumn routine. Why? To boost my annual protection for myself, for you, for those under my care, for those who are immunocompromised, and for everyone I know and don't know. Very importantly, it's well known to be effective at reducing symptoms. Even when the flu vaccine is mismatched to the seasonal strain, there is significant protection against other influenza strains contained within the vaccine (Belongia, Expert Review of Vaccines 2017<sup>1</sup>) which, similar to COVID-19 vaccines, prevents severe disease and hospitalization.

My aforementioned sister skirted

<sup>1</sup> Edward A. Belongia, Danuta M. Skowronski, Huong Q. McLean, Catharine Chambers, Maria E. Sundaram & Gaston De Serres (2017) Repeated annual influenza vaccination and vaccine effectiveness: review of evidence, Expert Review of Vaccines, 16:7, 723-736, DOI: 10.1080/14760584.2017.1334554. <https://www.tandfonline.com/doi/full/10.1080/14760584.2017.1334554>



death two years in a row after catching the flu during winter. She was unvaccinated both times. Each hospitalization necessitated months-long intubation, tracheostomy, and pulmonary rehabilitation. It's been three and a half years since her last ICU hospitalization and her memory loss has never recovered. It took two near-fatal bouts of flu for her to be on board with all immunizations including influenza and COVID-19. Don't let that happen to you.

After maximizing my COVID-19 and seasonal flu protection, I will continue to mask up as much as possible.

Why? To protect myself, especially my bronchiectatic lungs, and others by not bringing home the multitude of other viruses out there that can wreak havoc. I used to get such strange looks pre-pandemic whenever I wore a mask in public. I love how much more commonplace masking has become recently. When I see others masked, I feel a kinship with them and quietly give gratitude for their considerate behavior of others while protecting themselves, too.

Caregiving is challenging for those of us with CF. It's difficult to stay on top of your health while urgent and

emergent situations arise when advocating for a loved one. But it's even harder when priority isn't given to your health on a daily and long-term basis. As is announced on planes: "please place the mask over your own mouth and nose before assisting others." In that same vein, ultimately, to be an effective caregiver to others, you must first be a good caregiver to yourself. ▲

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*Dr. Jeanie Hanley is 59 years old and has CF. She is a director and former President of USACFA. She is the founder of Planning Health, a patient advocacy non-profit. Her contact information is on page 2.*

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**TILLMAN** continued from page 19

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### **Cystic Fibrosis Exacerbations In Women: Study**

Epidemiologic studies demonstrate worse outcomes in women with cystic fibrosis (CF) than men. Women are colonized earlier with respiratory pathogens and have increased rates of pulmonary exacerbations after puberty and near ovulation. There are potentially important fluctuations in inflammatory biomarkers in the lungs that correlate with changes in lung function in women with CF. Researchers sought to explore whether natural hormonal fluctuations and hormonal contraception associate with changes in lung function, respiratory symptoms, or inflammatory markers. The researchers prospectively followed women with CF who were not on hormonal contraceptives and reported regular menstrual cycles. A subset of subjects were subsequently placed on a standard oral estrogen/progesterone combination contraceptive pill, ethinyl estradiol/norethindrone (loestrin), and reevaluated. Measurements included lung function, symptom questionnaires, sweat tests, blood for hormone concentrations, and sputum for inflammatory markers, bacterial density, and cytology. Hormone concentrations were as

expected on and off hormonal contraception. At times of peak estrogen (ovulation), there was a significant increase in sputum proinflammatory cytokines (neutrophil-free elastase) and a corresponding pattern of decrease in lung function. Furthermore, proinflammatory cytokines (IL-8, TNF- $\alpha$ , and neutrophil-free elastase) improved when placed on hormone contraception. As a result, the authors concluded that there are potentially important fluctuations in inflammatory biomarkers in the lungs that correlate with changes in lung function in women with CF.

<https://tinyurl.com/228svm6h>

AND

<https://tinyurl.com/2f2bdzr6>

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### **Pregnancy For Women With Cystic Fibrosis**

The Mayflowers study, which will begin in July and end in December 2025, will enroll 285 women with cystic fibrosis at 40 sites across the United States. The women will be followed through pregnancy and 2 years after giving birth to document their health changes. The study team expects that about 25% of the women will stop taking their cystic fibrosis transmembrane

conductance regulator (CFTR) modulator during part or all of their pregnancy. There is no published data in human women whether all 3 components of the newest CFTR modulator are transferred to the baby through the placenta or breast milk. Additionally, only small retrospective surveys have reported use of CFTR modulators in pregnancy, limiting understanding of the impact of modulators on outcomes for infants exposed in utero and during lactation. A substudy will evaluate the pharmacokinetics of modulators in pregnant women and in the infants exposed to modulators for women who choose to continue them during pregnancy and breastfeeding.

The study expects to show that it is safe for a woman to stay on her medications, especially one of the new modulators, and not experience a significant health decline during pregnancy, particularly in the first year after her child is born. The study will evaluate diabetic women with cystic fibrosis as well. A substudy of diabetic women will use continuous glucose monitoring. This substudy will provide evidence-based data for managing diabetes during preg-

Continued on page 34



# FOCUS TOPIC

## GETTING VACCINATED WITH CF

### A Jab Well Done

By Anna Modlin

**T**he past 18 months have been a journey. I turned 40 this year! It is such an achievement being both a person with CF and a lung transplant recipient of almost 11 years. I am so grateful to be alive even during a worldwide pandemic. For the past four and a half years I have lived in Melbourne, Australia. I immigrated here from the U.S. after meeting my Aussie husband at the Transplant Games of America. I am waiting for my dual citizenship to be processed as I write this. My life during the pandemic has been so different than my life prior to the pandemic. The biggest change is that I became a mother to my daughter, Zoe. Zoe was born through surrogacy in August 2020, at the height of our most restrictive lockdown.

Life in Melbourne, Australia, has been a rollercoaster for the past 18 months. We have been locked down with the longest and harshest lockdown in the world, for a total of 227 days and counting. We have national healthcare and, to protect our system, we must protect the community health. Australia has taken an elimination strategy to the virus. This means we have kept the lid on the virus for a good part of the pandemic, until now. We are once again in lockdown—our sixth. We have strict rules, with only essential retail, such as pharmacies, groceries, and gas stations staying open. Food is only for takeaway; no restaurants are open. We are not allowed to visit with others at home or outside. Masks are required inside and outside. We cannot go further than three miles from our homes and we have a nightly curfew of 9 p.m. to 5 a.m. It is strict here. If you break these

“I have always been someone who happily will take a vaccination to prevent illness.”



ANNA MODLIN

rules you can get as much as a \$5,000 fine. Yet, even with these restrictions, our numbers of COVID-19 cases of the Delta variant are continuing to grow. We have also been stuck in a very slow vaccination rollout wrapped up in a bad PR disaster with the AstraZeneca vaccine. We only had 4% of the population vaccinated when I was able to receive my second dose on June 20, 2021. My experience with both my first and second jab were virtually free of side effects—just a sore arm and a little fatigue. As I write this,

we have finally gotten more people vaccinated and have 34% fully vaccinated. We finally have more supply of the Pfizer vaccine and will soon be receiving Moderna as an additional choice. Right now, we are finally having a big campaign push for people 12 and up to be vaccinated. You see, we will continue to stay in our extreme lockdowns until we have reached an 80% vaccination rate. It is also important to note that, since the pandemic started, we have had our international borders shut to the entire world. With the travel ban we are not allowed to come and go. For myself, I am an American who married an Aussie and my whole family is back home in the States. I miss them so very much. I would do anything to see them again, and for them to meet my daughter, Zoe. We hope that once we have 80% of the population vaccinated, we will have some normalcy again and I will finally reunite with my family.

Living through these lockdowns has been a bizarre introduction to the world for our Zoe, and for us as parents. She doesn't know a world without masks and has never played with other children. She has kept me going through all this. She is the reason we work so hard to stay healthy and well. She is one of my reasons for being vaccinated. I hope that this world will return to a normal place for her to grow up in and I hope that I am able to survive this pandemic despite being immune suppressed. The vaccine is my hope; it is all of our hopes. Minimizing the disease that SARS-CoV-2 causes is the most important thing. Making this illness less lethal and less contagious is so important.

I look forward to one day finally having my booster shot. As we are so

slow in our countrywide rollout, the authorities have not yet considered approving the booster vaccination. I am just holding on for that third shot and however many after that to keep me alive so I can live and thrive alongside my darling daughter. As a one-year-old, she doesn't even have the option to be vaccinated, so we are even more careful to make sure she stays well. The first year of a child's life includes a plethora of vaccinations. We just finished her 12-month shots, and we feel proud as parents to be protecting her against all those crazy viruses out there that we are so fortunate not to experience as a society anymore. Until I became a parent, I didn't even realize that chicken pox in childhood had been virtually eradicated here\*!

I have read and discussed the pros and cons about vaccinations with my family and my healthcare providers. I have always been someone who happily will take a vaccination to prevent illness. I think a quick needle over a serious illness is a no-brainer. I love getting my flu shots. In fact, here in Australia, we get two flu vaccinations, six weeks apart from each other, for extra protection. To say that I was excited to get my COVID-19 vaccine is an understatement. I was in the second group to be allowed to get vaccinated here. I made my appointment as soon as I found a place I could get the vaccine. We have struggled with shortages of supply. I was scheduled back in April for an AstraZeneca jab (jab is the word they love to use here for a shot). The day before my appointment we were given disappointing news—the AstraZeneca vaccine was being recalled for anyone under 60 because of the risk of blood clots. Until further studies surfaced, it was all on hold. I was beyond disappointed. Even with the approval of my transplant team, I had to wait. It was then determined that if you were under 60 you could only receive the Pfizer

vaccination. Again, more waiting, as we didn't have the supply. Finally, on the day they had supply available, I was scheduled two weeks out to get my first Pfizer jab. The day I got the jab, May 28, was also my fourth wedding anniversary and the day we went back into lockdown for the fourth time! On May 29, they finally opened up the rollout for individuals 40+ to get vaccinated, so my husband went on the first day and waited in line starting at 7 a.m. to get his jab. After about four and half hours of waiting, he came home with his first shot completed. Similar to me, he experienced just a sore arm and a little fatigue after the jab. He did it for me and he did it for our baby Zoe. Our family is vaccinated to keep us together as long as possible. The minute that a COVID-19 vaccine is approved for our little girl, we will be signing her up!

I have confidence in the vaccinations being given to the world right now. Why? Because I trust medicine. I have put my life in the hands of doctors so many times. I have seen the benefits of all the vaccines I have taken and the ones I have watched my daughter receive. Prior to SARS-CoV-2, the mRNA vaccine has been studied for outbreaks such as flu and rabies. Once scientists had more information about the virus behind COVID-19, they worked to develop an mRNA vaccine specific to SARS-CoV-2. The science is there and I trust the science.

Since the pandemic began, I have received monthly updates from my transplant team at the Alfred Hospital in Melbourne. I want to note that, in Australia, we have only had one or two lung transplant patients who have caught COVID-19, so we are relying on data from other countries. One of the most reassuring things I have heard from my medical team is their support of the COVID-19 vaccines. They reported to us that in a government report from the U.K. using AstraZeneca

and Pfizer with about 50% of the included patients receiving each vaccine, 40% of the unvaccinated transplant patients died from COVID-19. Mortality with one COVID-19 vaccination decreased to 10% and the death rate decreased to 8% with two vaccinations. For me, this was a *huge* reassurance. I really don't want to die from COVID-19. I have much better things to do. So, until we reach an 80% vaccination rate here and until I can get my booster, I will just hang tight. I hope to see my family within the next year. I hope to swim in a pool again. I hope to be able to go shopping again. I hope to breathe fresh ocean air without a mask again. Until then I will consider I have a jab well done!

*\*Editorial Comment: It is only due to vaccinations that childhood chickenpox has been dramatically reduced (nearly eliminated) as have been hospitalizations and death (99.99% eradicated—only 1 in 100,000 reported deaths in children last year) in the U.S. and other countries like Australia. However, the same chickenpox virus still attacks 20% of adults in the form of shingles; therefore, it has not been eliminated in adults, nor will it be. Similarly, chickenpox has not been eliminated in many developing countries where vaccination or infections have not achieved herd immunity. ▲*

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*Anna Modlin is 40 years old and has CF. She is a double lung transplant recipient of almost 11 years. Anna is an American who lives in Melbourne, Australia, with her husband, Terry and one-year-old daughter, Zoe. Pre-pandemic, Anna and Terry competed in ballroom and Latin dancing together. Anna has always had a passion for swimming and bringing Zoe to the pool is a highlight in life for her. Anna is passionate about volunteering and helping where she can within the CF and transplant communities.*



# Post-Transplant/Post-Vaccine!

By Elyse Elconin Goldberg

**W**hat a year and a half this has been! When the COVID-19 pandemic hit in March 2020, I truly had the fear that this insidious virus might wipe out a great percentage of our patient population. For those of us living with cystic fibrosis, and those who have also had bilateral double lung transplants, the future seemed very uncertain.

As I reflect back on those months of shelter-in-place, Instacart, lack of toilet paper, and flour and yeast shortages, of all things, I must congratulate our CF community on doing an excellent job of caring for ourselves. We have trained our entire lives in excellent hand hygiene, social distancing (our six-feet-apart rule in CF group functions) and mask wearing. Gratefully, these practices literally saved our extended CF family. We have pivoted from in-person gatherings to virtual platforms for CF meetings,



ELYSE ELCONIN GOLDBERG

retreats, and conferences with such excellence. I am not sure if we will ever return to in-person functions.

We have been fortunate to have access to life-saving vaccines almost 18 months into this pandemic. My transplant team encouraged patients to receive the COVID-19 vaccine. I knew that being on immunosuppressant medications to prevent the rejection of my transplanted organs might also impact my immune system's ability to create antibodies against COVID-19. Interestingly, this most likely applies to other necessary routine health maintenance vaccines such as the flu shot, shingles vaccine, etc. There are limited studies with regards to these other vaccines that have been done on solid organ recipients<sup>1</sup>.

<sup>1</sup> Hum Vaccin Immunother. 2018; 14(6): 1311–1322. Published online 2018 Mar 21. doi: 10.1080/21645515.2018.1445446

**SANDS** continued from page 20

requirements was almost a chore. I had to find a new way to live with my body. I didn't have to rush to fit it all in. I didn't have to stop and smell the roses. My passion for photography and nature dwindled. The gorgeous sunsets didn't look so amazing anymore. I felt like I had so much time and so little to do. What was my purpose? I now had the ability to do amazing things and I was not sure how to apply it to my new life. I missed my people. The medical team who used to lift me up and delicately take care of me was no longer part of my family. The excitement of being able to breathe soon spiraled into a pit of depression.

I tried to reach out to people whom I had previously been neglecting because of my health and tried to organize get-togethers and reunions. I joined softball leagues

and put passion into my gardens. But just as my mind started to blossom into accepting this newfound freedom, my body started to feel worse. I developed a rash on 50 percent of my body that resembled an allergic response. I itched and scratched throughout the day and night, turning my inflamed skin to scabs. My elbows, knees, and shoulders began to give out, causing me pain with every movement. I had all this energy and breath but couldn't use it because of the crippling pains and rashes. I tried treatment after treatment, even considering surgeries, trying to sort out these new challenges.

One morning after I took my miracle drug, my airway started to tighten. Even though I was used to the feeling of suffocation, this sensation was different. I could feel my body rejecting the medication. I

decided to stop taking it immediately for fear that it would do more harm than good. My doctor agreed that it was too risky to continue. I was just beginning to figure out how to manage my new existence, when I was spun back into my old life. Almost immediately, my cough returned, my intestines plugged, and my weight dropped. My body was finding its way back to the way it was. Mentally though, it was almost exciting to be back to my old self; a reality that was familiar.

As I was returning to a state of health that was dire, I was finding that others were experiencing some of the same disappointments with our life-saving medicine. The side effects so severe that they also had to make the gut-wrenching choice to stop taking the medication. How can something so



I was eager to get the vaccine to avoid experiencing the very serious pulmonary complications and/or possible death as an outcome of COVID-19. I have received the standard protocol of two Moderna shots and followed the recommendation of my transplant team to get a third booster vaccine. In an effort to get a better understanding of how the vaccine is working in solid organ participants, I volunteered to participate in the Johns Hopkins COVID-19 Vaccine Transplant Research Study. After strictly following their post-vaccine lab draw schedule for the SARS-CoV-2 antibodies, I am seeing firsthand that being on immunosuppressant medications is inhibiting my immune system from creating the necessary level of antibodies to be protected from COVID-19. Further studies are underway studying solid organ recipients' T-cells and memory cells, which, beyond just antibodies, are also important defense mechanisms in our complex, multifaceted immune system. With this research, scientists may find that solid organ recipients have more

protection in order to fight COVID-19 than the initial antibody tests reveal.

Upon receiving a negative antibodies result following my third vaccine, I realized that I need to rethink this "new normal" and figure out a way to live a full life with healthy and safe boundaries. In the CF and transplant community, we all know how precious time is. There was no way I was going to let a year or more pass by just existing. I was optimistic for a miracle vaccine and to be able to return to life as normal like everyone else. For now, this doesn't seem to be the case. I have found meaningful ways to stay engaged and connected with family, friends, and my CF/post-transplant community through virtual volunteering, Zoom meetups, developing new interests and hobbies, and exercising! From the beginning of the COVID-19 pandemic, I made the decision to mask up and continue to walk/hike outdoors while social distancing with friends. The human connection is so important for our overall wellbeing. Caring for our

mental health is just as important as caring for our physical health!

While we wait for more information on how the COVID-19 vaccines ultimately perform in solid organ recipients, I encourage others to get vaccinated. The immune response system is exceedingly complex. Research is ongoing and I am optimistic that there is a commitment, by the medical establishment, to find a solution to protect our transplant community.

In honor of my donor and their gift of life, I feel a responsibility to take care of these precious lungs. As my daughter so beautifully said to me, "Mom, it is better to miss a few holidays, social gatherings, and travel experiences now than to miss them all in the future." ▲

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*Elyse Elconin Goldberg is 62 years old and has CF. She lives in Los Gatos, California with her husband, Craig, and their puppy Teddy. She is the mother of two grown children and one of her greatest post-transplant joys has been becoming a grandmother! You can email her at eeglilac@msn.com.*

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good make us feel so bad? Our freedom to live with health was being stripped from us. I noticed that even though my joints were feeling better, my lungs were filling at a quick rate. Breathing became tiresome again. I found myself in the emergency room on two occasions for lung issues, the latter ending in an admission for IV antibiotics. It was like my previous life came flying in at full force like it never left. Even though it was disheartening to be sick, I felt a sense of comfort as I was being treated by my old pals at the hospital.

It became clear that I cannot live without the drug, but I am not sure I can live with it either. Under medical supervision, I made the choice to try taking the drug again. At least I would be able to make an informed decision as to whether

taking the drug was even an option. As the nurses set up my hospital room with life-saving equipment in the event of me having an allergic response, I mentally prepared myself for the test. I needed to be okay if I was able to start the drug again, and I needed to be okay if I was not able to. The memories of the past year were swirling through my head as I lay the pill on my lips. I opened my mouth quickly and swallowed the pill before I could change my mind. Anxieties overran my thoughts as I intently focused on my airway. Was I feeling an adverse reaction? I calmed myself with a few deep breaths and found that my worries were causing me more difficulties than anything else. As the time passed, my vitals remained stable, and I was cleared to take the drug again. A new sense of relief filled my soul.

The tug of war with my self is currently at ease. I now feel that I have a second chance to fill my heart and soul with the things that make me happy. My life's time is not ticking so fast. Although I do not know what the future holds, I need to realize that my life-shortening disease is still a part of me and will always be. Whether my days are filled with medications and therapies, or whether they're filled with experiences, I must be able to find peace in that reality. I must continue to grow and learn about my body and my mind, finding a balance between the familiar and the unfamiliar. ▲

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*Leah Sands is 40 years old and has CF. She lives in Detroit, Michigan. She loves photography, team sports, and traveling. She can be contacted by email at leah.sands@gmail.com.*

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# Cystic Fibrosis And COVID-19 Vaccination

By Richard Moss, M.D.

As of September 2, 2021, CF Centers reporting to the CFF Patient Registry find 8,183 people with CF [PWCF] have received at least one COVID-19 vaccination. As about 30,000 PWCF are included in the Registry, this would suggest less than 30% of PWCF have been vaccinated. But when one considers that the PWCF Registry total also includes children <12 years not eligible for any COVID-19 vaccine, as well as underreporting of actual vaccinations (almost all of which are not given at a CF Center, but rather outside the CF care network, so Registry reporting is gathered later and at least secondhand), it seems vaccination, as well as public health measures of masks, social distancing, avoiding indoor gatherings, and hand hygiene, have substantially contributed to the CF community in the USA doing relatively well during the pandemic, with only 14 deaths and 268 hospitalizations.

How safe and effective are COVID-19 vaccines in PWCF? Studies specific to CF are not available, so we rely on data obtained in much larger populations. Two of these are relevant—first, the general population (which demographically skews older and therefore more susceptible to COVID-19 compared to CF) and second, the solid organ transplant population. Note that studies in immunocompromised populations are probably not relevant to non-transplant CF, as PWCF do not have an impaired adaptive (B & T cell) immune system (i.e., ability to make functional antibodies and develop cellular immunity), which is the body's defense system activated by vaccination.

The safety of COVID-19 vaccines has been established by intensive monitoring starting with large clinical trials in spring 2020 and continuing in real-world settings internationally, making these vaccines by far the most extensively studied vaccines in history. As of this writing, over 5.4 billion doses have been given, with 2.16 billion people



RICHARD MOSS

fully vaccinated—nearly 30% of the entire human race. Perhaps the best study of safety has come from Israel (Barda et al, NEJM 8/25/21), a nationwide study of nearly 900,000 Pfizer vaccine recipients, who were compared about six weeks after completing vaccination to matched unvaccinated controls. Vaccine safety data were presented along with medical records of roughly 200,000 cases of diagnosed COVID-19 compared to healthy controls. Probably the most worrisome vaccine-adverse event, mild and transient inflammation of the heart (myocarditis), occurred at 2.7 more events per

100,000 than unvaccinated; importantly, this compared to myocarditis occurring from COVID-19 itself in 11 more events per 100,000 than controls. So even the most concerning reported vaccine-adverse event—which occurs very rarely—occurred far less after vaccination than from catching COVID-19. Additionally, COVID-19 was associated with a wide range of other serious complications not seen with vaccination. This is but one of a large number of studies showing excellent vaccine safety. The vaccines have also been found to be very safe in the very old, in pregnant and breast-feeding women, and in people with many chronic conditions, including immunocompromised individuals and transplant recipients. Studies in children under 12 are ongoing with results expected soon.

When evaluating efficacy, the worldwide sweep of the Delta variant has made a difference. The Delta variant of SARS-CoV-2 is one of the most infectious, easily transmitted viruses ever discovered. It is also, we now know from a study in the UK, more virulent and inherently dangerous than prior COVID-19 strains (Twohig et al, Lancet Infect Dis 8/27/21). The three vaccines licensed under Emergency Use Authorization in the USA—Pfizer, Moderna and J&J, with Pfizer now fully approved after extensive safety analysis by the FDA—all continue to show superb efficacy in protection against serious illness, hospitalization and death, with only a slight fall from >95% to 85-90% in the new Delta era.

What confuses many people is the more dramatic falling efficacy against any infection as Delta has taken over. This endpoint includes asymptomatic carriage of the virus as well as mild, cold-like symptoms and more serious

potential manifestations of infection. With this endpoint, studies such as a recent large UK study comparing Delta to Alpha vaccine protection (Lopez Bernal et al, NEJM 8/12/21) have shown that while protection by an mRNA vaccine against any type of infection, including asymptomatic carriage (positive PCR nasal test), has fallen from >90% with earlier variants to perhaps ~50% with Delta, retention of protection against any *symptomatic* infection remains very good (88%). There is also some evidence that the mRNA vaccines may be more protective against Delta infection than other types of COVID-19 vaccines (Lopez Bernal et al, NEJM 8/12/21). The drop in protection against *any* infection, including asymptomatic nasal carriage, is what has led to the recommendations to resume public health measures to prevent transmission. Given the ongoing low numbers of hospitalizations and deaths in PWCF, there is no reason or evidence to believe that vaccination is losing its major protective effect. Concerns about waning immunity as time passes are being addressed by ongoing studies, but currently there is little evidence this is happening independently of variant effects.

The drop off in protection against any infection, including asymptomatic or very mild infection as opposed to the ongoing great protection against serious illness and death, has driven huge interest in giving a third, booster shot to further augment immunity and hopefully reduce *any* infection, and thereby cut transmission and thus help end the pandemic—here again, new data from Israel strongly suggests a booster can do so, reducing *any* infection by >70% in the short term (Patalon et al, medRxiv 8/31/21). Other research has shown that boosters work better when given after at least several months' interval following the initial jabs. The current trend suggests boosters will become

widely available and recommended, but there is a countervailing worry about the inequity of boosters in developed countries like the USA when the great majority of the world's population has yet to see a first dose.

What about PWCF who are transplant recipients? Studies in CF have clearly, if unsurprisingly, shown this is a very vulnerable population with disproportionate rates of hospitalization, ICU care, and mortality. Immunosuppressive drugs needed to maintain organ grafts target the same immune system components that vaccines stimulate. Here, studies show that the COVID-19 vaccines are much less effective. Less than half of solid organ transplant recipients had an antibody response after the standard two-dose mRNA vaccination (Boyarsky et al, JAMA 5/5/21); it may be even lower specifically in lung transplant recipients (Havlin et al, J Heart Lung Transplant 5/21/21). Breakthrough COVID-19 infections occurred 82 times more often (serious illness 485 times more often) than in nontransplant controls (Qin et al, Transplantation 7/23/21). Again, no studies focus solely on CF, but several include CF lung or liver transplant recipients along with those suffering many other diseases resulting in transplantation. There is some partially good news even here, though: a booster shot can result in a much better immune response, resulting in antibodies in 70% of solid organ transplant recipients (Kamar et al, NEJM 8/12/21), so transplant recipients join a list of those with other causes of immunocompromise currently encouraged to get a booster at least a month after the second mRNA vaccine shot. The CDC has an excellent website on this topic, which I encourage anyone who has received, or is listed to receive, a transplant, to review (<https://www.cdc.gov/coronavirus/2019-ncov/vaccines/>

[recommendations/immuno.html](https://www.cdc.gov/coronavirus/2019-ncov/vaccines/recommendations/immuno.html)).

Finally, a few words on influenza. It hasn't gone away, it's here to stay, mutating constantly so that annual flu shots to keep current (and occasionally additional shots for worrisome variants like 2009's H1N1 pandemic strain) have long been part of routine CF care. The 2020-2021 winter flu season was practically nonexistent, courtesy, it is thought, of COVID-19 public health measures. That doesn't mean it will stay subdued as the country has opened up and flu season approaches. People with CF have good antibody responses to flu shots (Dharmaraj & Smyth, Cochrane Database of Systemic Reviews 2014). Over 90% of PWCF get them (Ortiz et al, Chest 2010), and flu vaccines significantly reduce likelihood of PWCF getting the flu (Wat et al, JCF 2008). You should continue to get your annual flu vaccine. ▲

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*Dr. Richard B. Moss is former chief of the Pediatric Pulmonary and Allergy Divisions, and allergyimmunology and pulmonary fellowship training programs director, at Stanford University. He was educated at Columbia (BA), SUNY Downstate (MD), Northwestern/Children's Memorial Hospital (pediatrics residency) and Stanford (allergy-immunology, pulmonology fellowships). He was Director of the Cystic Fibrosis Center at Stanford (1991-2009) and site principal investigator for the Cystic Fibrosis Therapeutics Development Network (1999-2009), where he was also inaugural Chair of the Protocol Review Committee. Dr. Moss has reviewed and consulted for the NIH, CFF, national and international foundations, peer-review journals and biopharmaceutical companies. His research interests include immunopathogenesis, outcome measures, and treatment of chronic airway diseases of childhood. Recent work has focused on allergic fungal lung disease. If you have questions for Dr. Moss, you can email CF Roundtable at [cfoundtable@usacfa.org](mailto:cfoundtable@usacfa.org).*



# My COVID-19 Vaccine Story

By Maggie Williamson

**I**n January 2021, the first COVID-19 vaccines were beginning to arrive here in the U.K. The government announced that both Pfizer and AstraZeneca vaccines would be available and would initially be offered to those most in need. They did this by assigning every person in the country to one of ten priority groups and I discovered that I was in priority group four—"all those 70 years of age and over and clinically extremely vulnerable individuals (not including pregnant women and those under 16 years of age)."

In 2020, shortly after the pandemic began, I was designated as a "clinically extremely vulnerable individual" on account of having received a double lung transplant and having a suppressed immune system. I was asked to spend much of 2020 sheltering at home and was unable to leave for my own protection. At first, the government arranged for food supplies to be delivered to our homes because the supermarkets didn't have enough delivery slots. It was fantastic having a friendly person arrive at your door with canned goods, bread, pasta, cookies, jams, tea, instant coffee, and shelf-stable milk. Later on in the pandemic, the supermarkets started to offer priority delivery slots to people based on the same priority groups.

I was delighted that I would be receiving my vaccine soon and recall jumping up and down around my apartment yelling, "I'm getting the vaccine!" I was six years post-transplant and knew all too well the fragility of my immune system. I heard horror stories from both the U.S. and the U.K. about how serious COVID-19 was proving to be for transplant patients and how many of them were unfortunately dying. This vaccine would also be a ticket out of my

“Why did I get my vaccines? I wanted to protect myself given that I’ve had a transplant and knowing I have a compromised immune system.”



MAGGIE WILLIAMSON

apartment where, frankly, I had been stuck for far too long.

As the weeks went by and I patiently waited for an appointment, I spent a lot of time watching and reading the news. I gathered all the information I could on the vaccines. My transplant hospital was brilliant with getting information to us and I attended several of their virtual meetings and Q&A sessions. They explained the studies and calmed any fears I had that these vaccines were safe enough for both the cystic fibrosis and transplant communities.

It was late in February 2021 when the phone rang from my doctor's office, "Margaret, it is your turn for the jab. Would you like me to schedule that for you?" The poor woman on the phone must have thought I was insane because I was so giddy with excitement and my voice reflected that sentiment. The English already think Americans are too much and I was proving them right on this phone call! Thankfully, she seemed unfazed by my enthusiasm and the appointment was booked for a few days later.

I arrived at my appointment and was definitely the youngest person in the waiting area. I waited patiently to be called into the room. I was greeted by a very nice male nurse who asked a few questions, gave me a leaflet on the side effects of the vaccine and then proceeded to draw up the liquid from a vial. I was getting the AstraZeneca vaccine, which was what I wanted as I had heard of potential issues with the Pfizer vaccine if you've ever had severe allergies, as I have had in the past. It was a quick poke to the arm and I hardly felt a thing, far less than my flu shot every year! As I left, I felt like a weight had been lifted off my shoulders. I wasn't out of the woods but had taken the first step to regaining some normality. I didn't feel great for a few days after my first vaccine, but it was nothing too major—just a little tired, dizzy, and generally lethargic.



At the time, the U.K. government's strategy was to give the first dose of the vaccine to as many people as possible before giving out second doses of the vaccines. This meant I would have to wait 12 weeks for my second vaccine, which I wasn't too happy about as I watched some of my fellow Americans, who had gotten the Pfizer vaccine, receive their second shot after only three weeks. I waited 12 weeks for my second shot and just repeated the process. Since having my second shot, the advice has changed to only wait eight weeks between shots in order to receive the most immunity with the AstraZeneca vaccine.

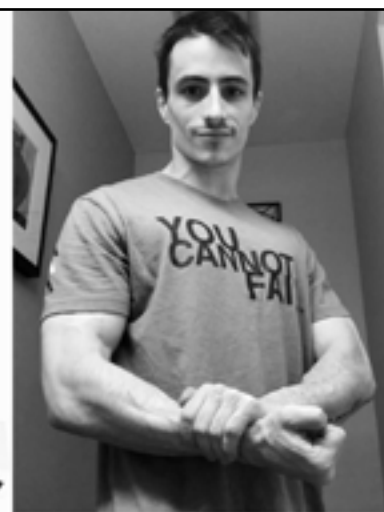
Why did I get my vaccines? I wanted to protect myself given that I've had a transplant and knowing I have a compromised immune system. I'd also spent a year stuck at home in my apartment and frankly, I wanted to be able to go out again.

How do I feel about two doses of vaccine causing a weak immune response in some post-transplant? I'm sad and angry that the vaccines have not produced the antibodies so many of us needed to feel safe. I'm hoping that the third booster vaccine that I'm getting in the coming weeks will give me some added protection.

What do I think about all the misinformation surrounding vaccines? It is fascinating to be living in another country outside of the U.S. and to see the differences between the U.K. and the U.S. The U.K. doesn't seem to have as much misinformation surrounding the vaccine. It never became political and most people wanted the vaccines to protect themselves and their loved ones. We still have people who are opposed to the vaccines, but we are already at a 90% vaccination rate in England. I was also fortunate to

receive information from my transplant team, which helped me feel safer in my choice to get the vaccines. ▲

*Maggie Williamson is 33 years old and has CF. Maggie received a double lung transplant in 2014 at Stanford Hospital in California. She now lives in the U.K. with her British husband. She is passionate about cooking and food in general and has built a small catering business called Just A Sprig. She is a peer connect volunteer with the Cystic Fibrosis Foundation, which involves helping those with cystic fibrosis who need support in coping with their disease. She is also a committee member for her transplant hospital club, The Harefield Hamster Club. Maggie is looking forward to traveling the world once it is safe and is particularly looking forward to seeing her family in Chicago. You can connect with Maggie through her @justasprig page on Instagram.*



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# I Am Vaccinated!

By Colleen Adamson

I have CF. I am a lung and kidney transplant recipient. I have diabetes and I have recurring bouts of skin cancer. I have a multitude of other health issues that are too long to list here. Do any of these things qualify me to be a doctor? Of course not. That is why I talked to my primary care provider (PCP), my CF doctor, and my lung transplant doctor about getting COVID-19 vaccinations. They all agreed that I should get them, so I did. I also got the third dose, as recommended by the Centers for Disease Control and Prevention (CDC) and my doctors. Because I am at high risk for getting all sorts of things due to my compromised immune system, I also get my yearly flu shot in addition to vaccinations for pneumonia, shingles, and Tdap (Tetanus, Diphtheria, and Pertussis), all with approval and endorsement from my doctors. I have had no health issues from any of these vaccines, and I am protected from these illnesses as much as I can be. That is what vaccines are for.

I think of getting the COVID-19 vaccines as me protecting myself and protecting everyone around me. This



**COLLEEN ADAMSON**

is not just about one person. This is not about personal liberty. This is about putting you, your family, and your community first. This is about caring for other people. This is a group effort, which means everyone needs to participate if we are ever going to get past this pandemic. There are breakthrough cases of COVID-19 (people getting COVID-19 even though they are fully vaccinated). Scientists tell us

that the vaccines are not 100% effective, which means it is no surprise that there are some breakthrough cases. This does not mean that people should not get vaccinated; COVID-19 vaccines are effective at preventing infection, serious illness, and death. We need the COVID-19 vaccines for protection, especially with the variants being more contagious and deadly than the original strains.

Since writing this, I have received good news: I still have COVID-19 antibodies! I am so thankful about this because I was told by my lung transplant clinic nurses that only a few of their patients developed COVID-19 antibodies after being vaccinated.

Yes, people with CF are probably smarter about medical stuff than the general population, but please do not be lulled into a false sense of security because you think you know what you are doing in this realm. COVID-19 is a force to be reckoned with, even for us. ▲

*Colleen Adamson is 52 years old and has CF. She lives in Alexandria, VA, with her husband, Scott, and their dog Penny. She can be reached at [scott.adamson@cox.net](mailto:scott.adamson@cox.net).*

**TILLMAN** continued from page 25

nancy that is specific to the unique needs of women with cystic fibrosis. The Mayflowers study will provide definitive data to guide women and their partners in their decision-making. <https://tinyurl.com/ve4u74e8>

## Fertility And Pregnancy In Cystic Fibrosis

Sexual and reproductive health is increasingly important for pwCF as many are considering parenthood. Most

men and some women with CF (wwCF) will have reduced fertility, which in both sexes is multifactorial. However, unplanned pregnancies in women are not rare, and contraception and its interaction with CF complications need to be addressed by the CF team. Reduced fertility may be overcome in most pwCF through use of assisted reproductive technologies. Most wwCF will have normal pregnancies, but premature birth is common especially in the setting of

reduced lung function and CF related diabetes (CFRD); optimization of treatment is recommended during pregnancy planning. Parenting imposes an increased burden on pwCF, with the challenges of caring for the newborn, postpartum physiologic changes and maintaining CF treatments. Most drugs used to treat CF are considered safe in pregnancy and lactation, but exceptions need to be acknowledged, including the limited data regarding safety of CF trans-



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## ENDOWMENT PARTNER

Carroll Groeger (in memory of Paul Schnackenberg)

membrane conductance regulator (CFTR) modulators during conception, pregnancy, and lactation. Prospective studies regarding these issues in people treated with CFTR modulators are paramount to provide evidence-based guidance for management in the current era of CF care.

<https://tinyurl.com/4xsxft2u>

### What To Know About Cystic Fibrosis And Pregnancy

This is an in-depth article that covers the following areas: Planning Your Pregnancy, Cystic Fibrosis Drugs and Pregnancy, Post-Transplant Considerations, Cystic Fibrosis Doctor Discussion Guide, Cystic Fibrosis and Fertility, Genetic Counseling, Cystic Fibrosis and Gestation, Pulmonary Exacerbations, Gestational Diabetes, Nutritional Deficiency, Constipation, Hypertension, Cystic Fibrosis and Postpartum, Impact on Recovery, and Breastfeeding. If you're planning on starting a family, you should definitely read this article.

<https://tinyurl.com/yecm2sy3>

### Depression, Anxiety Common Among CF Patients

Depression and anxiety are common among people with cystic fibrosis. Evidence suggests that adults with CF, as well as parents of children with CF, have an increased risk to experience depression compared to the general population. Depression and anxiety are known contributors to poorer quality of life, and linked with increased health-care costs. An investigative team conducted a systematic review to gather and summarize evidence from studies regarding rates of the conditions among CF patients worldwide. The results obtained after specific data analysis revealed that the overall global prevalence of anxiety was 24.91%, and depression 14.13% across CF patients. Researchers then conducted subgroup analyses and compared both anxiety and depression prevalence according to the geographical location. Anxiety prevalence showed a marked variation across continents: the lowest prevalence was seen in North America, and the highest

seen in Europe. The opposite was seen for depression, with the highest prevalence seen in North America and the lowest in Europe. Overall, these results support previous evidence and show that both anxiety and depression are common among CF patients. These findings highlight the need for close monitoring of the patient, regular screening for anxiety and depression and appropriate prevention techniques. <https://tinyurl.com/ytmnfc2d>

### Relatives With CF Can Be Source Of MRSA Transmission, Study Finds

Among people with cystic fibrosis (CF), antibiotic-resistant *Staphylococcus aureus* is sometimes transmitted among relatives, but rarely in healthcare settings. Findings also indicate that distinct subsets of *S. aureus* may be associated with differences in disease progression. A team of researchers reported an increase in MRSA infections among CF patients at their institution. They suspected that rising MRSA prevalence over the past decade may have been

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## LIVE OUT LOUD

# I Miss When I Was Rewarded For Being Alive: We Should Get Treats At Adult CF Clinic

By Lara Govendo

Life is so hard as an adult. I miss the days when I got lollipops and stickers when I went to the doctor or I got a prize if I didn't (and even if I did) cry during a blood draw. What happened to celebrating our life with CF like the good ole days? That may be the pick-me-up I'm craving.

I think we could all benefit from going back to easier days. In reality, those days were never easy, really, but snacks make *all* the things seem easier. It's less daunting going to multiple doctor appointments, suffering through procedures, and having tests done when there's a reward at the end.

Maybe it does mirror Pavlov's dog a bit; I don't see anything wrong with that, though. We could all benefit from a little classical conditioning—after all, our paycheck is the reward that gets us out of bed in the morning to go to work, and breathing clearly is the impetus for doing our breathing treatments. And the absolute bliss of being on a beach or a mountaintop is what gets us out of bed at 4:00 a.m. to leave for a trip, whether via car or airplane.

As a therapist, I believe we all need to find our individual motivator. Without it, it can be challenging to show up for the things we *have* to—the things we don't really have a choice in doing. And why wouldn't we make difficult matters more fun? It only makes sense given the amount of mutilation our bodies endure.

I am sure I am not alone within the CF or transplant communities in thinking of food as the only motivator for

going through the many bodily injustices we have to endure. We should have something to look forward to that will get us through the next thing we *have* to do. Since the majority of us have rocking metabolisms (or just terrible digestion), we have the glorious reward of being able to eat just about anything we want. In this instance, it's the epitome of Pavlov's dog.

At my pediatric hospital, they gave out bags to “get me through” the hospi-

tal stays. I miss that, too. I lived for those bags. I got my first (and only) iPod mini, beach towel (that I still use), and tons of games, candy, books, and more. It was like hitting the hospital lottery every inpatient stay (thank you Donna Crandall Foundation)!

I can't deny that growing up has been hard. Granted, it's a privilege to be in my 30s writing this as a person with CF who has also survived a double-lung transplant. I'm not negating that fact.

But why did everything change once we got older? It feels like it no longer matters that we need to be seen and validated just as much as when we were younger. We probably would benefit from that

level of validation now more than ever.

We CFers are living longer now, so let's change the adult culture! We don't become numb to life's assaults just because we grow up. Why, therefore, can't we have rewards just as we did when we were children? In fact, I would suggest we need more comfort, more awards, and more high-fives for still being alive! It's a huge feat.

As we know, clinic isn't always a happy place to attend. For me, this was especially true prior to my transplant. Having that pit of dread in my belly, otherwise known as anxiety, would hit me as I rode up the elevator. Usually it was a “you should really come into the hospital” talk that left a sour taste in my mouth. And mostly the disruption of my life that threw me into the cycle of anger, tears, and acceptance. But none of it was accompanied by a lollipop, which was drastically detrimental to my well-being.

Nowadays, clinic is a breeze with

*“We should have something to look forward to that will get us through the next thing we have to do.”*



LARA GOVENDO



my new lungs. Gosh I never thought I'd say that. And, although I don't get rewarded, the cafeteria's coffee is definitely my motivator... as well as the homemade chicken salad sandwiches on gluten-free rolls.

Who knew I'd be living so fully with little inhibition? Cruising into clinic devoid of oxygen tanks and heavy breathing is a miracle just as much as it is to leave clinic with a smile on my face. Maybe that's the sweet difference.

Don't underestimate the gravity of a sweet treat (unless you're salty) in stressful situations. It can be our greatest motivator, no matter how temporary it is. But it's worth it every time. And, hey, you only live once (or twice, for us transplantees). ▲

*Lara Govendo, M.S.Ed. is 34 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](http://www.laragovendo.com) (and on Facebook and Instagram) at "Lungs4Lovey." You can email her at [lgovendo@usacfa.org](mailto:lgovendo@usacfa.org).*

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## TILLMAN continued from page 35

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hastened by either person-to-person or healthcare worker-to-patient transmission.

To test this idea, the scientists sequenced the genomes (genetic code) of 97 MRSA isolates collected from 74 patients. By comparing genetic sequences among the different isolates, the researchers could look for clues as to how the bacteria were transmitted. In general, bacteria with relatively similar sequences are more closely related to each other. As such, if two people have MRSA isolates with very similar sequences, it is probable that one person acquired the infection from the other. Analysis showed evidence of MRSA transmission between relatives. However, instances of related individuals with distinctly different MRSA strains were also noted. In contrast, results showed minimal evidence suggesting transmission within hospitals, which indicates that these infections probably are not being acquired in a healthcare setting. Understanding the origins of MRSA in patients with CF could help limit acquisition of these resistant bacteria and improve patient outcomes.

<https://tinyurl.com/4nsbahkr>

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## Enterprise Therapeutics Doses First Subjects In Phase I Trial For Novel Cystic Fibrosis Therapy ETD001

CF Roundtable ■ Autumn 2021

Enterprise Therapeutics Ltd announced it has successfully dosed the first subjects in a Phase 1 trial for its novel inhaled cystic fibrosis therapy, ETD001. The first-in-man safety study is being conducted in healthy participants. ETD001 is an ENaC ion channel inhibitor with best-in-class potential aimed at treating all people with CF. ETD001 has previously been shown to have long duration of action in the lung and is therefore expected to provide a superior efficacy and safety profile compared to other ENaC drug candidates. ETD001 targets the epithelial sodium ion channel ENaC, which controls fluid volume and mucus clearance from the airways. By increasing the amount of airway fluid available to hydrate mucus, ETD001 addresses the underlying mechanisms of mucus congestion, and is expected to restore lung function, reduce the frequency of lung infections and improve patient quality of life. As ENaC inhibition is independent of the mutational status of CFTR, this makes the approach applicable to all people with CF. Additionally, ETD001 is expected to deliver benefit as a monotherapy and in combination with other therapies, including CFTR repair.

<https://tinyurl.com/yzx4k5rz>

AND  
<https://tinyurl.com/yhunz6bp>

AND  
<https://tinyurl.com/dxpdmrky>

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## RNA-Based Therapies Show Promise In Early Studies, ReCode Reports

RCT223 and RTX0001, ReCode Therapeutics' experimental RNA-based therapies for cystic fibrosis (CF), safely restored function to CFTR in patient-derived lung cells. Delivered through the company's non-viral platform—called the selective organ targeting (SORT) lipid nanoparticle (LNP) platform—the therapies were also found to be well-tolerated in lung cells from patients and in mice. ReCode plans to apply to the U.S. Food and Drug Administration in 2022, seeking the agency's clearance to start clinical trials of these therapies in the U.S. ReCode's CF program is focused on treating the 10–13% of CF patients who have non-sense mutations and limited therapeutic options. The CF program currently comprises two RNA-based therapies delivered through the company's SORT LNP platform, which uses tiny non-viral fat (lipid) particles to transport and deliver their cargo to targeted organs. The company's lead candidate, RCT223, uses a transfer RNA (tRNA) molecule—which plays a role in the process of protein production—to replace the pre-

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# IN THE SPOTLIGHT

## With Sonya Haggett

*By Jeanie Hanley, M.D., and Andrea Eisenman*

**R**eader, you are in for a treat. I (Jeanie) had the pleasure of interviewing Sonya Haggett, who is 46 years young and five years post-double-lung transplant. She lives in Oakland, California, and is a licensed clinical social worker. I first met Sonya six years ago at the CFRI Adult Retreat, which she attended while requiring oxygen 24/7 and awaiting new lungs. She was well loved and appreciated by others and her insightful nature, openness, and wit during the group rap sessions led to more profound discussions. When I saw her again a few years later, she was free of the oxygen tether, having received her transplant, and was radiant. Since then, Sonya has contributed to many organizations, like CFReSHC and CFRI, with her newfound energy, passing along her experiences of dealing with the issues surrounding transplant, women's health and CF overall. Please welcome, Sonya, our newest star. Spotlight, please!

### **What was your CF like prior to your lung transplant?**

Being extremely out of breath and fatigued. I asked my doctor why I had recently started coughing so much in the shower, and she explained it was because I was desaturating. In retrospect, I had been living without getting enough oxygen for about a year before starting supplemental oxygen, but only while sleeping, and about one and a half years before requiring supplemental oxygen 24/7. A lot of time was spent in the hospital on IV antibiotics between 2012 and 2016. On December 10, 2014, I was admitted for three weeks for an exacerbation—I celebrated my 40th birthday, Christmas, and the New Year in the hospital.



**SONYA HAGGETT**

### **What influenced your decision to go ahead with the transplant?**

Transplant was my best option to stay alive. Watching peers with CF on the path to transplant and getting to know them and their stories over the years at the CF retreat influenced me. I also cannot overstate the importance of the long and trusted relationship I had with my CF center and hospital. I cannot imagine going through the process without that level of trust and feeling the medical professionals were invested and working hard to keep me healthy and alive. Through hospitalizations I got to spend time with and know the pulmonologist on the transplant team and see the crossover between the transplant and CF teams at my hospital.

### **How long did you wait for new lungs and what was most challenging?**

I was on the transplant list for two years and one month. It was challenging: although emotionally healthy, I

had to energetically let go of so much because I needed all of my mental and physical energy focused on staying alive with peace of mind. I had to have faith things were going to work out for the best and let go of fear and doubt as much as I possibly could. It has been an incredible lesson I have tried to continue living.

I also learned the importance of leaning on others. For years, my coping mechanism was striving to live an independent life and working to meet life goals similar to peers without chronic illness. End-stage CF brought me to my knees, teaching me I cannot cling to delusions of independence and normalcy; it is not a healthy way to live. My family and friends and larger community showed up for me in ways I could never have imagined. Now that I am healthier and not so dependent on others, I find myself getting back into the independent mindset and have to work on keeping interdependence in mind.

### **What could you do after lung transplant that you couldn't do before?**

Travel, day trips, going to the beach, dancing, and enjoying exercise and yoga again. Travel highlights include going to New Mexico to meet a fellow individual with CF who also was a lung transplant recipient; visiting Italy and Thailand; as well as seeing Ireland with two of my nieces. Spending good, quality time with my nieces and nephews is a tremendously important part of my second chance at life. It is also important to mention I can now complete life's mundane tasks, such as showering and taking out trash, independently. I oddly feel a small joy taking trash out of my apartment and down the stairs to the outside garbage.

My body feels light and at relative ease as this task always takes my mind to a state of gratitude.

### **Is there anything you could do before that you can't do now?**

Healthcare decisions feel like a collaboration with my health professionals more so than life before transplant. They orchestrated and directly connected to me to my second chance at life and I feel accountable. In the past, I knew my body extremely well and felt like the expert; however, post-transplant I am no longer the expert.

I miss eating raw oysters.

Perhaps it is age, but I really feel I cannot push my body as much as I have in the past to meet a deadline, finish a project, or hang out enjoying myself. My body requires sleep and rest.

### **Can you take CFTR modulators?**

I took Trikafta for six months. My ENT proposed it to treat my sinus disease. My eyes teared up instantly. I had so many strong emotions and was hopeful that the latest modulator would help my sinus disease—clearing them of the *Pseudomonas* infection they have cultured for years—and ultimately keep my transplanted lungs as healthy as possible. Both my transplant and CF teams were in agreement that Trikafta was safe and held promise. Within the first week of taking Trikafta, I felt air in my sinuses like I never had. Not to mention all of the *Pseudomonas* infection I saw swirl down the drain with my daily sinus rinses. Graphic and unpleasant, I know, but such is life with CF. Then came the disappearance of the ubiquitous CF throat clearing.

I also noticed a pattern of waking earlier in the morning. I was averaging five to six hours of sleep a night despite feeling exhausted. Anxiety and extreme negative thinking were at a feverish pitch during this first month. I was trying to “tough it out” in hopes the dif-

ficult side effects would subside as my body acclimated to the profound changes spurred by Trikafta. At the end of the first month, I started experiencing worsening constipation which proved to be too much to manage. At day 30, already on a full dose of Trikafta, I was desperate for relief and followed my doctor's recommendation to lower the dose to one orange pill daily, rather than taking both the orange and blue pills.

Sleep deprivation, extreme negative thinking, and anxiety decreased significantly once the dose was lowered. My sinuses felt less clear, though, and the throat clearing returned. I felt disappointed and sad to have lost all the improvements to my sinuses I had gained over the first month. Over four months ago, I stopped taking Trikafta and I still have GI issues, which are worse than before I started.

### **What benefits have you had from counseling?**

I am fortunate enough to have weekly, long-term therapy. It has served as a place to process my hopes and fears, formulate goals for my life and my health, and peel back the layers of the onion to focus in on what is truly important and energetically worthy. There is no way I could have done this work with my friends and family. I need an unbiased third party.

### **Why did you become a social worker?**

My decision to go into social work was influenced by my time spent in support groups at CF Adult Retreat and how helpful that process was to me in my CF journey. So many ideas of how to take care of one's health and pursue life goals were sparked in these settings.

It also stemmed from recognizing the importance of self-advocacy in getting medical care. Some people need

support in developing self-advocacy skills or an empathetic party advocating on their behalf.

### **How did retreats with other CF adults impact your life?**

My peers with CF are the largest influence on my life and decision to get a transplant. I watched friends in varying stages of the disease process—before and after lung transplant—over the years at the CF Adult Retreat. Seeing and getting to know others who were going through the same pre-transplant struggles was scary. At a certain point I knew I was headed toward the transplant journey. Some seemed to live and soar post-transplant as they recounted the year since being together in person. People also talked about their imminent death related to complications post-lung transplant and deeply understood I would not see them next retreat. These are opposite ends of the post-transplant experience spectrum. So many nuances to the new purpose of life post-transplant along with the changed definition of good health and general feeling of satisfaction lie in-between. Time spent in support groups and informal settings at CF Adult Retreat—the empathizing and learning from my peers—is precious, sacred, invaluable, and helped me to feel more prepared as I became a patient with my lung transplant team.

### **What have you been doing to cope during the COVID-19 pandemic?**

The CF community has been more of a support than ever amidst the pandemic shutdown. The majority of my time spent on Zoom has been with my community participating in educational, emotionally supportive, and exercise programming provided by CF-related organizations.

There is an inherent connection

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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](mailto:cfroundtable@usacfa.org)**

## ANNIVERSARIES

### **Birthday**

**Cara Brahm**

Lebanon, KY

45 on September 10, 2021

### **Wedding**

**John & Jeanie Hanley**

Los Angeles, CA

35 years on June 28, 2021

### **Transplant**

**Cara Brahm, 45**

Lebanon, KY

Bilateral lung transplant

7 years on September 8, 2021

**Lara Govendo, 34**

Milton, Vermont

Bilateral lung transplant

4 years on August 18, 2021

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mature stop signal with a “go” sequence in the CFTR gene, so that the full CFTR protein can be produced. Data showed that both single and repeated dosing of RCT223 restored CFTR function in lung cells derived from CF patients and grown in the lab. This effect lasted for at least 72 hours (three days) after a single administration, and CFTR functional levels continued to rise with twice-weekly doses. ReCode’s mRNA replacement agent for CF, RTX0001, is designed to deliver to cells a working version of CFTR’s messenger RNA molecule (mRNA), the intermediate molecule derived from DNA that guides protein production. As such, RTX0001 is expected to increase the production of a working CFTR protein. Preclinical data showed that RTX0001, delivered through a commercially available mesh nebulizer, successfully delivered CFTR’s healthy mRNA into patient-derived lung cells and into the lungs of mice. The delivered mRNA was found to effectively and significantly increase the production of a working CFTR after a single, low dose, and CFTR’s activity was sustained for at least 72 hours after twice-weekly admin-

istration. Both RNA-based therapies were generally well-tolerated in the evaluated preclinical models.

<https://tinyurl.com/uf6z54e3>

### **Study Describes New Compound For Nonsense Mutations**

A novel compound that may hold promise for treating the roughly 11% of cases of cystic fibrosis (CF) that are caused by nonsense mutations has been described in a new study.

A nonsense mutation results in a premature stop codon introduced in the DNA sequence. This causes the cell to produce a truncated form of the CFTR protein, which is quickly degraded within the cell. In the new study, researchers set out to identify novel readthrough compounds that might be useful for treating CF and other diseases caused by nonsense mutations. As the name suggests, the goal of a readthrough compound is to let ribosomes “read through” the erroneous stop codon to produce a full-length protein. The team used a cell model that basically consisted of cells engineered with a gene encoding a detectable protein that had a nonsense mutation in it. The researchers used this

model to screen 771,345 small molecules, and found that 180 of them had readthrough activity. Among these molecules, the most active was called SRI-37240. Further testing using cellular models of CF with nonsense mutations demonstrated that treatment with this small molecule could allow the cells to produce functional, full-length CFTR protein. The researchers next synthesized dozens of derivatives of SRI-37240, and they found one—called SRI-41315—that had even more potent readthrough activity. The increased readthrough efficacy of SRI-41315 was confirmed in lung cells from people with CF, particularly when used with G418, a type of antibiotic known to aid readthrough of premature codon mutations. Further investigation revealed these readthrough small molecules work by reducing levels of eRF1, a protein used to recognize stop codons. The team suggested that targeting eRF1 may be a useful strategy in CF.

<https://tinyurl.com/4rwawphe>

### **Scientists Demonstrate Promising New Approach For Treating Cystic Fibrosis**

Researchers demonstrated a potentially powerful new strategy for treating



among my peers through the deep understanding of our need to follow guidelines set forth for people with CF and/or transplants to stay healthy. Developments in medical science have allowed me to live this long and now is not the time for me to stop following the guidance of medical professionals who care for me.

Peers who continue to believe in medical science validate the measures I take to decrease COVID-19 risk. The counter messages—those who don't believe in the science behind vaccines,

masking, and taking contact precautions—wear on me. It has been difficult to stay the path of best practices to not catch COVID-19 as healthy family members, peers, and the community around me return to something resembling life before March 2020.

Friends with transplants who are immunocompromised like me have been supportive in a special way, making me feel less isolated.

**What is your motto or favorite saying?**

Make a plan and God laughs. Nothing is perfect but everything is ok. ▲

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*Jeanie Hanley is 59 and has CF. She is a Director and the past President of USACFA. Andrea Eisenman is 56 and has CF. She is a Director of USACFA and is both the Webmaster and Executive Editor of CF Roundtable. Their contact information is on page 2.*

*If you would like to be interviewed for "In The Spotlight," please contact either Andrea or Jeanie.*

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cystic fibrosis (CF). It involves small, nucleic acid molecules called oligonucleotides that can correct some of the gene defects that underlie CF but are not addressed by existing modulator therapies. The researchers used a new delivery method that overcomes traditional obstacles of getting oligonucleotides into lung cells. The scientists reported that they were able to restore the activity of the protein that does not work normally in CF and saw a prolonged effect with just one modest dose. Treatments for CF now include CFTR modulator drugs, which effectively restore partial CFTR function in many cases. However, CFTR modulators cannot help roughly ten percent of CF patients, often because the underlying gene defect is of the type known as a splicing defect. In principle, properly designed oligonucleotides can correct some kinds of splicing defects. In practice, getting oligonucleotides into cells, and to the locations within cells where they can correct RNA splicing defects, has been extremely challenging, especially into the lungs. Therapeutic oligonucleotides, when injected into the blood, have to run a long gauntlet of biological systems that are designed to keep the body safe from viruses and other unwanted molecules. Even when oligonucleotides get into cells, the most

usually are trapped within vesicles called endosomes, and are sent back outside the cell or degraded by enzymes before they can ever do their work. This new strategy overcomes these obstacles by adding two new features to splice switching oligonucleotides: Firstly, the oligonucleotides are connected to short, protein-like molecules called peptides that are designed to help them to distribute in the body and get into cells. Secondly, there is a separate treatment with small molecules called OECs, which help the therapeutic oligonucleotides escape their entrapment within endosomes. The researchers demonstrated this combined approach in cultured airway cells from a human CF patient with a common splicing-defect mutation.

<https://tinyurl.com/ygutw7hf>

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### **Inhaled Drug Could Treat Rare Cystic Fibrosis Mutations**

Trikafta treatment combining three drugs – elexacaftor, tezacaftor and ivacaftor – reduces symptoms for those with cystic fibrosis. The new medication, although not a cure, works wonders for the 80 percent of cystic fibrosis patients with the predominant mutation causing the disease - F508del.

But what about the 20% of CF patients who have a different genetic

mutation? SpliSense manipulates and “fixes” defective messenger RNA that generates a non-functioning cystic fibrosis transmembrane conductance regulator (CFTR) protein. Rather than trying to repair defective proteins, SpliSense’s technology generates a new fully functioning protein from RNA. SpliSense has demonstrated in cells derived from patients that it can completely restore CFTR function. The company is now moving on to animal models and the first human clinical trials are planned for 2022. Since CF primarily affects the trachea, bronchi, bronchioles and alveoli, SpliSense’s treatment is meant to be inhaled so that it reaches the lungs quickly without any uptake by other organs or the bloodstream. If approved by regulators, the treatment would be administered weekly for 10 minutes at home throughout the patient’s life. Ideally, the cost of this expensive treatment would be picked up by health insurance providers. SpliSense’s lead product is based on “Anti Sense Oligonucleotide” (ASO), a synthetic nucleic acid molecule that can bind to specific sequences within target RNA molecules.

The ASO sequences are specific to the target mutation region in the RNA, so the treatment won’t affect (or damage) nearby organs and tissues. That

Continued on page 42

should reduce potential side effects.

<https://tinyurl.com/yhf7awht>

### **CF Foundation Launches New Collaboration With Deep Science Ventures To Overcome Challenges To Developing Genetic Therapies For CF**

The CF Foundation announced a new collaboration with venture creator Deep Science Ventures, focused on uncovering and designing new technologies with the potential to overcome challenges to developing genetic therapies for cystic fibrosis. Deep Science Ventures and the CF Foundation will work together to assess pressing barriers to genetic therapies in CF, explore the feasibility of potential solutions, and design proof-of-concept studies. By aligning the

Foundation's CF expertise and scientific capabilities with Deep Science Ventures' track record of uncovering innovative technologies, this agreement has the potential to push the CF field forward and significantly accelerate progress toward future therapies for CF.

<https://tinyurl.com/4ufcf6>

### **Vertex To Initiate Phase 3 Program For Once-Daily Triple Combination Regimen In Cystic Fibrosis**

Vertex Pharmaceuticals Inc. will initiate a phase 3 development program for the new once-daily investigational triple combination of VX-121/tezacaftor/VX-561 (deutivacaftor) in people with Cystic Fibrosis. Phase 2 data suggested the triple combination

holds the potential to restore cystic fibrosis transmembrane conductance regulator (CFTR) function in people with cystic fibrosis to even higher levels than seen with other Vertex CFTR modulators and thereby provide enhanced clinical benefit. The combination of VX-121/tezacaftor/VX-561 was first identified as having potential for increased efficacy based on its ability to drive higher levels of chloride transport compared to TRIKAFTA in human bronchial epithelial cells in vitro. The Phase 3 program consists of two randomized, double-blind, active-controlled 48-week trials, which will evaluate the safety and efficacy of VX-121 (20 mg)/tezacaftor (100 mg)/VX-561 (250 mg) in comparison to TRIKAFTA.

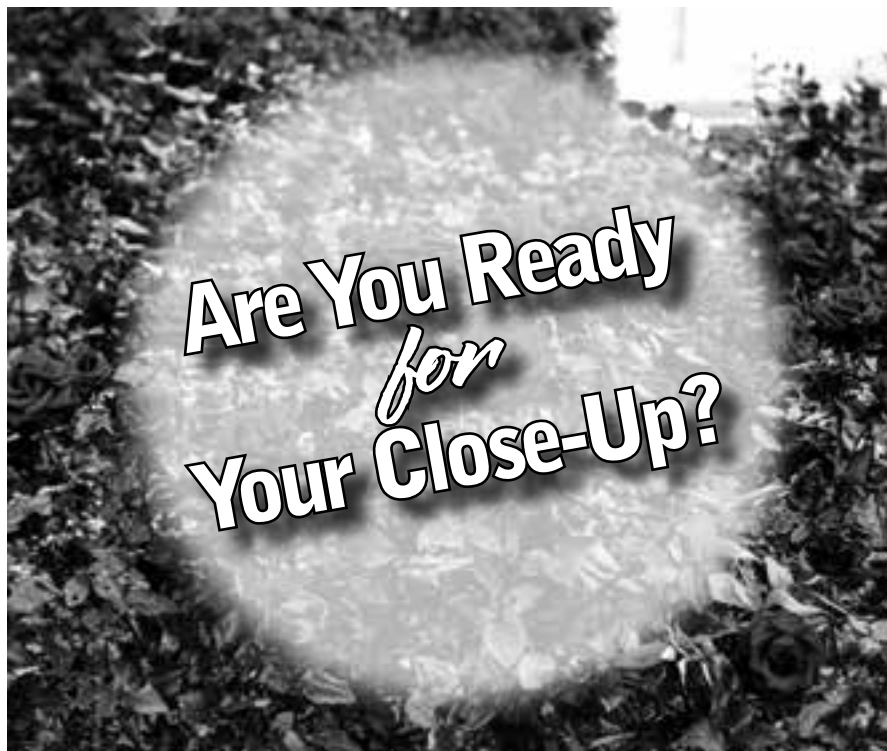
<https://tinyurl.com/32t3m398>

AND

<https://tinyurl.com/yz5ad85e>

### **Kaftrio Treated Severe CF Without Confirmed Secondary CFTR Mutation**

Kaftrio successfully improved lung function and quality of life in three cystic fibrosis patients who have had severe lung disease with an unknown mutation in the CFTR gene in addition to the common F508del mutation, a case series reported. Results showed that over 24 weeks of Kaftrio treatment (almost six months), the sweat chloride levels in all participants progressively lowered, eventually reaching the normal range. The treatment also led to clinically relevant improvements in lung function in all patients. CF Questionnaire-Revised (CFQ-R) respiratory domain scores significantly increased for all participants by eight weeks and were sustained for the six months of treatment. Other CFQ-R domains showed an enhanced quality of life. No pulmonary exacerbations were reported during the treatment. No abnormal adverse events were reported in terms of vital signs, clinical laboratory tests, or physical examinations. Kaftrio combines three CF medicines: elexacaftor and tezacaftor, designed to help faulty CFTR get to the



**D**o you have CF and you're 18 years or older? Want to share your story and CF journey? If so, consider being interviewed for our "In The Spotlight" column. Send an email to Andrea and Jeanie at [ITSinterviews@usacfa.org](mailto:ITSinterviews@usacfa.org) to set up an interview about your health, struggles, achievements, education, career, family, or other significant experiences while dealing with CF. Inspire others who have CF and similar experiences. The interview will be published in a future *CF Roundtable* issue.

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**You can also subscribe online!** Go to [www.cfroundtable.com](http://www.cfroundtable.com). You can now scan this QR Code with your smart phone to go directly to our online registration page. By subscribing online you have the ability to download a PDF of the latest newsletter, receive the mailed version or you can receive both. The online version will be available two weeks prior to the mailed version.



### KEEP YOUR INFORMATION CURRENT

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

**Thank you for helping us with this.**

cell membrane, and ivacaftor, a so-called CFTR potentiator that aims to open blocked channels allowing water and ion transport. Kaftrio, sold as Trikafta in the U.S. is available only for CF patients with at least one CFTR F508del mutation or another known secondary mutation. Patients whose DNA analysis is

inconclusive typically are excluded from Kaftrio clinical trials and treatment. These findings support expanding Kaftrio treatment in CF patients without a confirmed secondary CFTR mutation, especially in those with advanced lung disease.

<https://tinyurl.com/8wd2vbk7>

AND

<https://tinyurl.com/4ckac52t>

AND

<https://tinyurl.com/4x6ktkp> ▲

*Laura Tillman is 73 years old and has CF. She is a former director and President of USACFA. She and her husband, Lew, live in Northville, MI.*

## REMINDERS

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



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## IMPORTANT RESOURCES

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

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

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

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

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