CF Roundtable®
A NEWSLETTER FOR ADULTS WHO HAVE CYSTIC FIBROSIS
AUTUMN 2020

The Juggling Act: CF, Parenting, And Work

By Cindy Baldwin

When people ask me what I do, I usually answer that I work part time as an author and also stay home with my daughter. What I often *want* to add, but don't, is that I feel like I have a third job, too: taking care of my health.

Most people with CF are well acquainted with the way that can seem like a full-time job in itself. Scheduling daily breathing treatments, complex medication regimens, exercise, and adequate rest and nutrition are difficult enough. When you add in the other realities of a CF life, like doctor appointments and what feels like an infinite number of calls to pharmacies, insurance companies, and provider's offices to make sure everything is paid for, the delicate dance of CF care often feels all-consuming. Adding parenting into the mix? *Instant* chaos.

Although I've dreamed of being a published author for as long as I can remember, I spent many years doubting my own ability to balance a writing career, parenting, and keeping on top of caring for my CF and other health conditions. Because my energy is extremely limited, I wasn't sure I'd ever be able to divide myself in so many different directions.

> "I knew I had to figure out new ways to write that allowed me to get more done in less time."

CINDY BALDWIN

Still, writing was extremely important to me—so, as my daughter grew older, I found myself frequently reinventing the way I worked in order to make everything fit. I wrote my first published novel, *Where the Watermelons Grow*, when my daughter was two and had recently stopped napping. Since selling *Where the Watermelons Grow* to HarperCollins in 2016, I've juggled deadlines for other books while homeschooling preschool,

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INSIDE THIS ISSUE

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EDITOR'S NOTES

Autumn brings sweeping change in both the foliage and the temperatures. This issue we're focusing on being an entrepreneur in the CF community and what that looks like. We're featuring **Brian Callanan**'s inspiration from *The Tao of Pooh* for starting the Cystic Fibrosis Lifestyle Foundation. **Mandy Anderson** of RAYMA Team details her tips for building and growing your own business. Our focus topic section also features **Nicole Kohr**, who writes about using her CF journey to create *Fall Risk*, the musical. We've also got a story from **Shaun Clarke** about using art to help kids process and normalize their chronic health struggles. **Klyn Elsbury** retells her story of going from social security benefits to bestselling author and motivational speaker.

In the "Voices From The Roundtable" section you can read about **Devin Wakefield**'s reflections on hiking the Pacific Northwest throughout the years. **Amy Gutierrez** talks about the importance of both reviewing your medical notes from doctor visits and having access to these electronic medical records via OpenNotes. Our "Voices" section also features a story about **Jeremy Moore**'s efforts to write a book for others needing tips and techniques for seeing results in the gym. We've got a wonderful poem from **Rob De La Noval** in this issue whose written shape reflects the topic: shots! **Dr. Xan Nowakowski** discusses the importance of mental health in the age of new innovated modulator therapies for CF patients. We're also very excited to welcome **Xan** to our growing board of directors! You can read their story on page 31.

Laura Tillman expertly collates all the latest CF research from the internet in this issue. For our "In The Spotlight" interview this issue **Andrea Eisenman** and **Jeanie Hanley** talk with Amy Shroyer Branham, a CF patient who also has both MS and thyroid cancer. We recently awarded two $2,500 scholarships through our LMK scholarship. You can read about the winners, Daniel Gonzalez Davila and Ashley Wilson, in this issue. Be on the lookout for education updates from our winners in a future newsletter.

Beth Sufian answers questions from our readers in her column "Ask The Attorney." **Lara Govendo** writes about the importance of maintaining authenticity and vulnerability in a world that constantly pulls us many directions. **Mark Tremblay** discusses the importance of using various methods, such as HALT, to keep stress, loneliness, anger, and feelings of being alone at bay. Speaking of loneliness, **Isabel Stenzel Byrnes** writes about the difference between loneliness and solitude, especially in the time of COVID-19. **Kathy Russell** reflects on the origins of *CF Roundtable* and how much the newsletter and organization have evolved over time. She also reminisces about the early days of in-person conferences for CF patients in her column this issue.

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.

Publication of *CF Roundtable* is made possible by donations from our readers and grants from Sustaining Partners - AbbVie, Gilead Sciences, Maxor Specialty Pharmacy, Mylan, The Smith Foundation in memory of Jeannine Ricci, and Two Hawks Foundation in Memory of Dr. Lisa Marino; Pearl Sustaining Partners - Boomer Esiason Foundation, Cystic Fibrosis Foundation, John & Susan McMurry Fund in memory of Peter McMurry; Diamond Sustaining Partners - Marina Day, Trustee of the McComb Foundation, Nancy Wech (in memory of daughter, Lauren Melissa Kelly & in honor of son, Scott Kelly).

Information From The Internet...

Compiled by Laura Tillman

PRESS RELEASES

Drying Does Not Clear Nebulizers of NTM Bacteria, Study Finds

Nontuberculous mycobacteria (NTM) is able to survive being dried for a full day, suggesting that drying is not a sufficient way to disinfect nebulizers used by people with cystic fibrosis (CF). It is commonly recommended for nebulizers to be washed and then dried between each use. Previous research has demonstrated that drying can kill *Pseudomonas aeruginosa*. Researchers studied *Mycobacterium abscessus*, subspecies massiliense; *M. abscessus*, subspecies bolletii; and *M. abscessus*, subspecies abscessus. For each type, two clinical isolates were tested. Bacteria were placed on plastic surfaces mimicking a nebulizer, and dried for a day. Both drying at room temperature and drying in an incubator (at about 99 F) were tested. After drying, the team tested whether bacteria were able to grow in a culture in the lab. In all experiments, no evidence suggested that drying killed these bacteria. All NTM isolates tested were viable after drying for 24 hours, regardless of the *M. abscessus* species tested or the drying method used. Within the context of nebulizer hygiene, this means that these NTM organisms would be able to survive on a washed nebulizer following drying for 24 h, which has not undergone any formal disinfection protocol, which can eradicate NTM organisms. Based on the results, the team suggested that CF patients seek an effective alternative control to drying for NTM eradication such as *steam or heat disinfection*.
https://tinyurl.com/y555mz6b

LAURA TILLMAN

Clinical Characteristics And Outcomes Associated With Inquilinus Infection In Cystic Fibrosis

Researchers conducted a retrospective, case-control study of persons with

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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 photo of yourself as well as your name, address, and telephone number. Photos will be returned. Email all submissions to: cfroundtable@usacfa.org. Or go to our website: www.cfroundtable.com/newsletter.

Autumn (November) 2020: Entrepreneurs with CF (Current issue)

Winter (February) 2021: Late Diagnosis (Submissions due December 15, 2021.) How did your CF diagnosis help you with existing, untreated health issues? What acceptance or resistance have you experienced within the CF community? What resources have helped you in processing your diagnosis?

Spring (May) 2021: Sexuality and Sexual Health in CF (Submissions due March 2021.) How was your experience of puberty and adolescence influenced by having CF? In what ways has your sexual health been affected by CF? Do medications or treatments affect your sexual health? When and how do you bring up CF when dating?

Summer (August) 2021: Alternative Therapies and CF (Submissions due June 15, 2021.)

ASK THE ATTORNEY

Questions From Our Readers; Answers From The Attorney

By Beth S. Sufian, J.D.

Readers have submitted a variety of questions this month. People with CF continue to struggle with access to health insurance coverage if they have lost their job due to COVID-19. Some people receive Social Security benefits and have found remote work they can do part time and need to know the rules regarding working and receiving benefits. Others are in need of information regarding their legal right to Public Health Emergency Leave ("PHEL") because their school-age child is attending school virtually or their child's day care is closed. Others are finding that being laid off from their job has given them the time they never had to do their daily medical treatments, rest, and take better care of themselves. These individuals have questions about applying for Social Security Disability or SSI benefits.

The information provided below is not legal advice about a specific situation and is only meant to be legal information. If you have questions, please contact the CF Legal Information Hotline at CFLegal@sufianpassamano.com or call 1-800-622-0385.

Question: I lost my job, and, subsequently, my health insurance, because my employer lost business due to COVID-19. How can I get health insurance?

Answer: There are a number of health insurance options. If a person with CF has a spouse who has employer-based health insurance coverage, then the person with CF has 60 days to enroll as a dependent on that health insurance plan. A person with CF can also purchase a plan through the Affordable Care Act health insurance marketplace, which can be found at www.healthcare.gov. The person can qualify for special enrollment within 60 days of the loss of their health insurance plan. Another option is to enroll in COBRA. Typically, the person would have 60 days to enroll in COBRA, but, under new federal guidance, a person can now enroll in COBRA after that 60 days has passed. The rule extends most COBRA deadlines beyond the "Outbreak Period," which is currently defined as March 1, 2020, to 60 days after the end of the declared COVID-19 national emergency or another date, if provided by the agencies in future guidance.

Question: I receive Social Security Disability Insurance (SSDI) and would like to work part time. What is the maximum amount I can make each month and what is the maximum number of hours I can work each week?

Answer: The 2020 Social Security Disability part-time work limits are as follows:

A person can work part time and make up to $1,260 before taxes are taken out of the monthly work check. If a person is self-employed, the maximum work earnings a person can make are $910 a month before taxes are taken out of the monthly work check.

BETH SUFIAN

The maximum amount of work hours for a SSDI or SSI recipient is no more than 20 hours a week.

A person who receives SSI benefits has the same monthly income limits and work hour limits if the person receives the full SSI amount offered in their state. The difference is that for every $2 made in work earnings, the SSI check is reduced by $1. So, if a person earns $400 in one month from work activity, the SSI check is reduced by $200 for that month. Because the Social Security Administration typically does not know about the work earnings until the month after the work income is received, the SSI recipient often has an overpayment for SSI benefits received when the person should have had the SSI check reduced.

Question: I was laid off from work and have found my health has improved because I have more time to take care of myself. How can I determine if I can apply for Social Security Disability or SSI benefits?

Answer: A person can check to see if they have enough work credits to possibly be eligible for Social Security Disability Insurance (SSDI) benefits by going to www.ssa.gov and setting up a personal account. Then a person can see what a possible SSDI benefit would be if a person also meets certain medical eligibility criteria. If a person has not worked enough to be eligible for SSD benefits, the person may be eligible for SSI benefits.

There is a lot of misinformation in the CF community about who can qualify for SSDI or SSI benefits. Social

Security reviews medical evidence for the 12 months prior to applying for benefits. A person can meet the SSA medical criteria by having a low FEV1 or hospitalization in the past year, plus breathing treatments or other medical treatments during the day. In addition, a person who must spend a good amount of time each day taking care of themselves may also be eligible for benefits even if their FEV1 is not low or the person has not had any hospital stays in the past year. For those who cannot work right now, Social Security benefits can provide a monthly cash benefit and access to health insurance coverage.

Question: I have CF and I have a child who is at home doing virtual school. I would like to take PHEL and take some time off from my job. I read the child's school has to be closed to be eligible for PHEL. Is my child's school closed if they offer virtual school?

Answer: Many schools have converted to full-time or part-time virtual learning models. This is sometimes called a hybrid model to describe a school that is partly open to in-person instruction and partly closed to in-person instruction, with the closed portion receiving online, virtual instruction.

As a result, school is still in session, but in-person instruction is either not provided or provided on a limited basis. This situation raises the question of whether such a school is closed for purposes of PHEL under Family First Corona Relief Act ("FFCRA").

The Department of Labor ("DOL") has issued guidance in a series of Frequently Asked Questions confirming that when the physical location of a school or child care facility is closed, regardless of remote options, employees who have children at home participating in virtual school may be eligible for PHEL leave as a result of such closure. Notably, if a school is partially open and an employee's child attends intermittent in-person instruction when it is available, the employee would not be eligible for leave on in-person instruction days. The FFCRA regulations and the DOL's FAQ make clear that PHEL can be provided on an intermittent basis with employer approval.

The guidance and regulations do not describe how much of the school must be closed for the school to be considered "closed" for the purposes of PHEL. There are at least two responses to an employer that denies the request for PHEL saying the employee's child's school is not "closed." First, if the school is closed to any portion of the student body due to COVID-19, it is closed for purposes of PHEL. A school operating at 50% in-person instruction and a school operating at 95% in-person instruction would both be considered "closed" for purposes of PHEL because the school is, in fact, closed for some part of the student body because of COVID-19. The regulations and guidance do not state this assertion specifically, but this assertion is based on the meaning and effect of what is stated in the regulations and guidance. The second analysis is that PHEL is intended to allow parental leave when the employee's child is unable to attend school because the school is closed due to COVID-19. If the DOL considers partially open/partially closed schools to be "closed," then it would seem to follow that even if the school were closed to one child because of COVID-19, the school would be partially open/partially closed (because the school is closed to that child), and the parent of the child would be eligible for leave so that the child is not unsupervised at home. Again, the regulations and guidance do not state this assertion specifically, but this assertion is based on the meaning and effect of what is stated in the regulations and guidance.

For more information and discussion on PHEL, see the CF Roundtable *Blog post "New Laws to Address COVID-19: Part III of IV" and "New Laws to Address COVID-19: Part IV of IV" posted on May 6, 2020.* ▲

Beth is 55 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.

Cystic Fibrosis Mothers

Cystic Fibrosis Mothers is a website dedicated to providing information on parenthood to women with cystic fibrosis around the world. Our aim is to provide a central online resource for the global cystic fibrosis community. It includes personal stories, research articles, advice and links to further sources of information built up over time.

We also provide a private support group on Facebook with more than 500 members worldwide. To visit our website go to: www.cfmothers.com.

If you would like to join our Facebook support group, please go to: https://cfmothers.com/cfmothers-forums/.

SPIRIT MEDICINE

The Spirit Of Solitude Versus Loneliness

By Isabel Stenzel Byrnes

In this time of COVID-19, many people with CF who live in hot spots have been spending months living indoors, avoiding their usual social activities. Thankfully, technology allows us to connect to each other. Some of us feel it's safe to socially distance while wearing a mask. However, COVID-19 can intensify feelings of loneliness, and loneliness is common in the CF experience, anyway. Some of us have depression or we may withdraw emotionally if we do not feel well or feel understood. If we are disabled, we've lost work or school contacts. As a transplant patient, I've needed to avoid potentially infectious gatherings in the winter—even before COVID-19. In this article, I'd like to ponder the experience of loneliness in hopes that some of these reflections can alter the way we feel our loneliness—especially during this pandemic.

The CF community is not alone. Our country has another epidemic going on right now: loneliness. The American emphasis on individualism and independence seems to have broken down the sense of community and social connection. Millions of people live with sparse human contact, and research tells us that lonely people are more likely to become ill, experience cognitive decline, and die early. As a grief counselor, I work with many lonely, bereaved elders and I see the impact of their loneliness. During COVID-19, some elders say that sheltering in place feels like a punishment—like solitary confinement.

Connecting to others is a biological need. It ties back to the idea that being part of a group is adaptive behavior. In ancient times, a person abandoned by his tribe was more likely to die; therefore, it makes sense that research shows that loneliness elicits a biological response in the form of a survival threat. Lonely people experience a chronic stress response. We also know that loneliness reduces our sense of self and self-confidence. People are our mirrors; if we don't have them, we can't always see the strength in ourselves. Loneliness makes it harder to cultivate courage—to pick up the phone, to go out and meet people. This cycle makes lonely people lonelier.

Some of us introverts don't mind being alone, but we can still crave connection. Being lonely as an extrovert is indeed harder. In addition, it is possible to feel lonely even if you are living with other people in your home. If we are in a relationship that makes us feel alone, we need to examine that and address it. We also cannot expect one person to fulfill all of our needs. Sometimes it takes a village to overcome loneliness!

Psychologist John Cacioppo said, "Loneliness is an aversive signal, much like thirst, hunger or pain." We have to respond to it. When alone, it's important to make an effort to reach out to others and push through the instincts to withdraw. Talking to the cashier at the store or the mailman, using social media, joining Zoom meetings, or picking up the phone are some examples. Just now a new neighbor knocked on my door. My initial response was to be taken aback or imposed upon, but this neighbor models what we can all do to spark connection. My husband has started regular Karaoke sessions with his friends. I often have Zoom dinners with friends. I joined the CFRI Retreat by Zoom in July and loved connecting to others. We can also focus on helping others during times of loneliness. It takes a little creativity to adapt to volunteering during these COVID-19 times.

ISABEL STENZEL BYRNES

Another way to respond to our loneliness is by thinking about it in different ways. Sometimes it is a matter of getting used to being alone. It is a process that takes time and we can accept loneliness as a part of the human experience, rather than resist it. We can tell ourselves there's nothing wrong with us for being alone. We can also challenge thoughts that we are truly alone because, spiritually, we never are. For those who believe, we have a Higher Power that is always present within us, guiding and supporting us. Reading spiritual texts can inspire us to feel that presence.

And there is Solitude. Mae Sarton wrote, "Loneliness is the poverty of self; solitude is the richness of self." Loneliness is a yearning, an incompleteness. It is a desire to escape from oneself. Our fears and thoughts take over. Our responses are automatic reactions. Paul Tillich said, "Language has created the word 'loneliness' to express the pain of being alone. And it has created the word 'soli-

tude' to express the glory of being alone." When we are solitary, there is no yearning. We are at ease and at leisure with ourselves. In other words, we are at home with ourselves. We are less reactive and more emotionally intelligent with the present moment.

Sometimes lonely people tell me they like to "keep busy." This is a way of keeping the mind preoccupied and thereby distracting oneself from *feeling*. Yet, there is often tension in this way of being. If we can relax around our loneliness and pursue things that don't activate restlessness or agitation but rather calm us or bring us positive feelings, then we experience a very different way of being alone. This is the richness of being alone. An example of this is when I create art, play music or write, or read a good book. I get fully immersed in the activity and I don't notice feeling alone. This is a good kind of aloneness.

Ideas, creativity, and imagination are germinated in solitude. Nelson Mandela lived alone in prison for 27 years. Rather than feeling frustrated and limited in his loneliness, he wrote. He discovered the power of words. He transformed imprisonment into a deeper resource in himself. Isolation created his maturity and depth. Through his words, something inside him became enriched and nurtured. He is a shining example of transforming loneliness into solitude.

When I've been lonely, and subsequently met people, I felt a subtle desperation to connect. It is a profound need—a hunger so fierce that it can often prevent us from recognizing what the other person actually has to offer. Yet solitude helps us connect to each other in a richer way.

Our society wants us to be individualistic and ironically doesn't treat solitude very well. If we are dining alone in a restaurant, people may pity or judge us. Taking a trip alone is becoming more popular, but many still feel awkward or unsafe doing so. Solitude does not really mean a walk in a forest or sitting in meditation by ourselves. It is deeper than behavior. It is found by dwelling in your body and in your senses. And *being with* and *being okay with* our own thoughts and feelings.

In the CF experience, I believe loneliness leads to depression, and loneliness leads to decreased motivation to do our treatments or exercise. This creates a spiral of negative consequences. Loneliness is simply bad for our health. We all need a "who" to live for—someone who cares about us and loves us. Claude Anshin Thomas, author of *At Hell's Gate: A Soldier's Journey from War to Peace*, suffered from addiction and PTSD. He wrote, "I can feel very alone in this journey, but I don't have to take this journey in isolation. We are conditioned by our society and culture not to talk about our pain. But if we don't talk [or write], if we don't create a language to express our feelings, healing will not take place. We will continue to store up and re-create the cycles of suffering." These words apply to those of us in the CF community. The greatest antidote to loneliness with CF, to me, is to reach out and express this life experience to others. We can express it online, in writing, or verbally to anyone who will listen compassionately. Though a certain amount of loneliness is natural during these trying times, each of us has our own level of tolerance of loneliness. With the abundance of online CF resources, I hope we can all reach out for support as well as support each other and reduce loneliness. We can all actively prevent lonely people with CF from becoming a statistic—one where they'll have more health and cognitive issues and premature death. May our own walk through lonely times help us be more kind to those who are alone. ▲

Isa is 48 years old, 16 years post-transplant, and lives in Redwood City, CA. She works at Mission Hospice in San Mateo.

SPEEDING PAST 50

From Bad Air To A Bright Outlook

By Kathy Russell

As I write this, it is September and most of the western states are suffering wild fires. Here the smoke is thick and dense. Pilots have reported that it rises all the way up to 12,000 feet. It goes out 1,000 miles over the Pacific Ocean and obscures the sun much like a solar eclipse. The air has a foggy yellowish tint. We are staying inside and keeping all our windows closed with the air purifier going full blast. There are fires on all sides of us, but, so far, they have stayed away. We are hoping for some rain in the next week or so, but there are no guarantees—of course.

We have been staying at home most of the time because of COVID-19. I have gone out once each week to grocery shop. I'm still wearing a mask and gloves when I'm out as I really don't want to be exposed to this virus.

I am rooting for the researchers who are trying to create a vaccine that will be effective against COVID-19. I doubt that I will take the first version that is presented, but I will take it when I feel that it has been proven to be both safe and effective. I remember, all too well, epidemics of Asian flu, swine flu, avian flu, H1N1, and illnesses such as polio, measles, chicken pox, whooping cough, and others that caused such widespread death and destruction. I am not taking this lightly and I feel that others should be serious about it, too.

As I have been having a lot of time to think, I have realized that this issue of *CF Roundtable* marks the conclusion of our first 30 years of publishing. When we started out, we had no way of knowing if the publication would thrive and survive for 30 years. Of course, we did hope that it would. We all put in as much effort as we could to see it get off the ground and to establish it as a viable and important source of information for all adults who have CF.

I believe that the founders—Joe, Connie, Ken, Larry, and Melinda—would be happy with what the newsletter has become. It took many years and a lot of hard work to reach a position where we are thought of as reliable and informative. I take great pride in all that we have done to get there.

Over these years, many companies, foundations, and charities have learned of USACFA and *CF Roundtable* and have been moved to help out with generous contributions, as have many individuals who have an interest in CF. These donations, coupled with the great writing of our readers, have kept the newsletter running.

I am delighted to see so many new writers participating in the newsletter. Each new writer brings another perspective and new insights into living with CF. This is important to maintaining the relevance and worth of the newsletter. I hope that, by reading the stories from all of these people, we all learn and broaden our understanding of life with CF.

> *"It has always been a goal of USACFA to be inclusive and to provide a place where adults who have CF can find community."*

KATHY RUSSELL

It has always been a goal of USACFA to be inclusive and to provide a place where adults who have CF can find community—a place where they can feel that they are a part of something. When it still was possible, we held national conferences. We utilized safe practices so that we could lessen the chances of anyone getting ill because of their attendance at one of our conferences. We were able to have speakers present information that we might not get anywhere else, and that was tailored to our interests and needs.

I remember after our first conference, in 1999 in Houston, TX, one woman, who had never met another adult with CF, told me that it meant so much to her to be around other adults who have CF that she wore her name badge home on the plane. She was proud to be a part of something that not just anyone could join. It made her feel important and a part of a special community. Remember, this was before the internet was a big part of our lives

and many people felt very isolated. I am grateful that we didn't have to endure a pandemic such as this one back then. It would have been so much more isolating than it is now.

I found it very informative to be able to meet people from all over the country at our conferences. During those earlier years, I learned that there were many differences in how CF was regarded and how it was treated. I hope that there are not such wide variances in treatment now. It seems to me that there is much better communication between medical centers and between doctors and other caregivers than there used to be. I feel that this is definitely to our benefit. I miss being able to gather with others who have CF and I miss the feeling of camaraderie and shared knowledge that I get from such gatherings. It means a lot to me to see people doing so well—those that I have known for many years and those who are newer acquaintances as well.

I remember I received a phone call somewhere circa 1991. The caller was the mother of a newly diagnosed infant daughter. We talked for quite a while and I was able to answer many questions and to help allay some of her fears about her daughter's diagnosis. We kept in touch, periodically, over the intervening years. I learned of the daughter entering school, moving on to high school, going to college, graduating, getting married, and eventually having a baby of her own. This really made me happy to hear of her near-normal life. I believe that many more people, who were diagnosed at birth or soon after, will have near-normal lives thanks to the new personalized medicines. I am so happy about this.

When I was a nurse in a children's hospital back in the 1960s, the outlook for babies diagnosed with CF wasn't so rosy. Our kids coughed and hacked and choked all the time. One could always recognize the cough of a person who had CF. I have great hopes that in the coming years that will no longer be the case.

The past 30 years have seen a lot of changes in the world of CF. I think that there will be many more wonderful changes in this century. The outlook is bright. Who knows, maybe it will become unnecessary to have a newsletter that is especially for adults who have CF. Wouldn't that be amazing?

Until next time, please stay healthy and happy.

Kathy ▲

Kathy is 76 years old and has CF. She and her husband of 55 years, Paul, live in Gresham, OR. She is a past director of USACFA and served as President and Treasurer and was Managing Editor of CF Roundtable *for several years. You may contact her at krussell@usacfa.org.*

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CF from two CF centers with at least one respiratory culture positive for Inquilinus spp. comparing with age-matched CF controls with chronic *Pseudomonas aeruginosa*. They compared rates of pulmonary exacerbations. Inquilinus isolates were genotyped to assess strain diversity. They distinguished 17 patients with Inquilinus infection with a mean age of 13 years at first positive culture. In childhood, Inquilinus can present and is often correlated with chronic infection in CF. It was reported that lung function and nutrition status, lung function decline at the time of detection, and pulmonary exacerbation rates in Inquilinus cases were similar to those with chronic *P. aeruginosa*.

https://tinyurl.com/y645v46g

Higher Prevalence Of Oral Candida Fungus Found In CF Patients

People with *cystic fibrosis* (CF) are more frequently infected with oral Candida fungus and carry a higher number of fungi compared to healthy individuals. All fungal species identified in the study's CF patients were susceptible to currently available anti-fungal medications. Fungal infections, either alone or in combination with bacteria, have been linked to decreased lung function. In particular, mixed infections with the bacteria *Pseudomonas aeruginosa* and the fungus *Candida albicans* have been correlated with the progression of CF lung disease. 80% of CF patients with severe disease tested positive for Candida, whereas 68% of those with low disease severity tested positive, compared to 44% of control subjects. Two Candida species—*Candida dubliniensis* and *Candida tropicalis*—were detected only in the CF groups, whereas *Trichosporon asahii* was found exclusively in patients with severe CF, and *Saccharomyces cerevisiae* (baker's yeast) was found exclusively in the control group.

https://tinyurl.com/y64jgrck

1 In 3 Adults With Cystic Fibrosis Also Living With Diabetes, Cutting Lives Even Shorter

The signs and symptoms of CFRD share similarities to both Type 1 and Type 2 diabetes, but CFRD is a distinct condition with a different underlying cause. People with CFRD have worse lung function than people with just CF, and, ultimately, likely to have shorter lives. The condition requires daily careful dietary monitoring, regular monitoring of blood sugar levels, and insulin injections multiple times a day. Due to the launch of a new research program, the end of painful insulin injections

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LIVE OUT LOUD

Death Of Expectations:
The Journey Back To Your Truest Self

By Lara Govendo

As CFers, we're all too familiar with how the cha-cha dance between life and death changes a person; however, it's our choices that truly affect who we are as individuals—specifically our most authentic selves. Staying true to ourselves is often our biggest battle. It's tragic how our authentic selves die slowly. It starts with the tiny sacrifices made to fit in with others. We compromise our standards to *earn* love and respect. The death of our truest selves happens so slowly, we don't even notice. By the time we do recognize a difference within us, we're so far away from who we once were that we don't know if we will ever come back to ourselves, or how.

I was the closest to physical death I've ever been when I was on the highest oxygen flow possible before intubation. It was a truly magical time for me. Something changed within me during that season of near death. The matters of the world faded away; drama and discourse became more trivial. It didn't matter what was trending, politics didn't cross my mind, and there was no room for nonsense. People dropped out of my life faster than quicksand, but I barely noticed. I did, however, notice when people came out of the woodwork—they showed up in my hospital room, sent letters of encouragement, and kept up with text messages even when I was too exhausted to respond.

During that season of life, I felt the most like *me* than ever before. It surprised me that my authenticity shone the brightest when I was on my way out physically. My communication during that season was genuine. I boldly expressed my feelings to others. My language was clear and direct. I was amazed at the bravery within me to have such sincere conversations. I didn't have the time to mince words, nor did I have the breath to waste. I became even more deliberate in what and on whom I spent my breath as I didn't have the luxury of focusing on things or people that weren't adding to my life.

Life calls us to be intentional on every level, in all areas of our existence, especially for those of us with CF—we don't have the time, energy, or breath to waste on matters or people that suck the life out of us. Every moment feels more urgent when there are odds stacked against us. Getting caught up in the matters of the world only zaps us of energy that could be channeled in positive directions. Each inhale is precious when you're fighting for every breath.

Like everything else in life, vulnerability and authenticity aren't "one and done" matters. Both are an ongoing process of inner work that requires accountability, personal responsibility, and continuous growth. These matters often become pertinent in dire situations.

I've made the unfortunate mistakes of sacrificing my values, time, and energy on unimportant matters. My biggest regret is investing in relationships in all capacities, which ultimately required me to change who I was in order to fit into a box defined by other people's expectations. Learning that it is my responsibility to teach others how I want to be treated was an invaluable lesson. We accept people's lack of boundaries when we don't set or enforce them.

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LARA GOVENDO

As much as we would like to blame others, taking this as an opportunity to look within is crucial. As a society, we are quick to point fingers at others and play the victim (or passive) card. Being stuck in these habitual patterns of toxicity is grounds for a renewed vision. We *get* to choose how we live our lives—depending on others to alter our life course is irresponsible. This goes for people in our immediate circles as well as those on the peripheries of our social life.

Focusing on what we can do to spark change is more important than ever. And that comes from being our most authentic selves and loving who we were created to be. Abiding by these two life mantras evokes empathy within

us. It also helps us refrain from seeing the world through a judgmental lens. Security in ourselves and the ability to be true to our core invites others to do the same.

The desire to meet others' expectations comes from a place of insecurity. If we are constantly striving to please everyone, it's a telltale sign that we aren't being true to ourselves. Staying in alignment with who we are attracts the people meant to be in our lives. It also paves the way for which path we are meant to be on. This is impossible without knowing who you truly are.

At the end of the day, who feels comfortable being themselves around those who are judgmental, self-righteous, or noticeably fake? Right, nobody does. But we can't expect it from others if we aren't willing to take the risk and wholeheartedly be ourselves.

We can't afford to lose our individuality for fear it will cost someone else's comfort. We were created differently so that we could all do our part in the tapestry of the human race. The point isn't to be all alike; the point is to be ourselves and fit our unique gifts into the piece of the bigger puzzle.

Are you willing to take the risk of sacrificing the fake persona you've built and trade it in for the one you are when you are the most *you*? What would happen if we all did this together and promised to welcome each other as we are?

In a world that wants to tell us who we are, how to feel, and what to believe, fighting harder to stay true to ourselves is imperative. We don't have the luxury of caring what others think of us if we want to make an impact. We have to rise up and dare to be different. Choose to be true to ourselves with the driving force of love. Let us lead by example—and begin anew as our truest selves each and every day. ▲

Lara Govendo is 33 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 writes about living out loud and develops educational programs to restore hope to those in need. Thanks to her double lung transplant in 2017, you can now find Lara traveling on the regular, exploring the glorious outdoors, and belly laughing with her loves. You can find her online at www.laragovendo.com (and on Facebook and Instagram) at "Lungs4Lovey." Her email is lgovendo@usacfa.org.

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could be in sight as researchers are set to explore how to avoid the need for daily injections, eventually leading to cutting-edge treatment. The scientists will look at the way signals move from the digestive-juice-producing parts of the pancreas to the insulin-producing cells, which signals cause the most damage and whether these signals can be measured in the blood of people with cystic fibrosis to help researchers understand more about CFRD and how it develops. Researchers believe that CFRD is caused by signals from damage to the digestive-juice-producing part of the pancreas, which stops insulin-producing cells from working properly. Understanding more about these signals could lead to entirely new approaches to treating diabetes, avoiding the need for insulin injections.
https://tinyurl.com/y45wgep8

Prolonged Heat Exposure In Cystic Fibrosis Can Lead To Metabolic Alkalosis

In patients with cystic fibrosis, prolonged exposure to heat can lead to chloride-deficient metabolic alkalosis as a result of a loss of electrolytes through sweat. Patients with *cystic fibrosis* can have massive losses of sodium chloride that can lead to extracellular volume contraction and produce metabolic alkalosis. Furthermore, extracellular volume contraction causes secondary hyperaldosteronism, which leads to hypokalemia from potassium wasting in both sweat and urine.
https://tinyurl.com/y56teey3

Hearing Loss Screening In Patients With Cystic Fibrosis

Evidence indicates that treatment for cystic fibrosis (CF) often includes antimicrobial therapy with such agents as aminoglycosides; unfortunately, research suggests such therapy can lead to ototoxicity. "Hearing loss is an under-recognized problem among individuals with CF and can cause significant effects on quality of life. Audiology units are rarely co-located with respiratory centers, requiring further outpatient visits to identify ototoxicity and greater expense for patients. To decrease inconvenience and patient expenses, researchers tested the performance of a tablet-based audiology system that could be administered by non-audiologists, as well as a Web-based test that could be completed at home. The tablet-based test accurately screened for hearing loss with 93% sensitivity and a 94% negative predictive value. While the Web-based test had high specificity, it only had 14% sensitivity and 58% negative predictive value. Early ototoxicity occurs at high frequencies, which are often not noticed by the patient. Web-based testing didn't test extended high frequencies, which explains the poor sensitivity. Significant impact on quality

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while battling a months-long case of pneumonia when my daughter was three…and now, during a global pandemic that closed my daughter's school in March and led me and my husband to the decision to homeschool full time for the 2020/2021 school year.

I confess that when people ask me how I balance work, parenting, and CF, my usual reaction is a slightly hysterical laugh and the response that *I don't think anything gets balanced!* When you're trying to run a work-from-home career, raise a kid, and handle health concerns, it can feel like you're barely staying afloat. Still, even though life is often chaotic, I wanted to share some of the tips and strategies that *have* helped me get a little better at this difficult juggling act. While most of my tips will apply particularly to the work-at-home parent, I hope that any working parents reading this will find something helpful!

1. I had to learn to work as efficiently as possible.

Writing is a bit of a different career than many; you spend months or years writing, rewriting, and rewriting again before you ever sell a book to a publisher and make money. Before writing *Where the Watermelons Grow*, I had written other books, and each one had taken a very long and arduous time. When my toddler stopped napping right as I began *Watermelons*, I knew I had to figure out new ways to write that allowed me to get more done in less time. I attended classes, read books, and challenged myself to try writing techniques I'd never been willing to try before, in an effort to find ways to write more quickly and create drafts that needed less rewriting later. Ultimately, I was able to greatly increase my efficiency, which allowed me to finish the book and then balance the early years of my publishing career while raising a very young child.

My specific productivity tips don't translate well across different jobs, but the piece of advice that does is to make *sure you're working as efficiently as possible.*

> "I had to figure out how to work in the moments I had, rather than the ones I wished I had."

Have you examined the way you're structuring your work day, the distractions you have around you, and the methods you use to get your work done? If not, that's an excellent place to start.

2. Learn to work in small bursts.

For those of us with the ability to work at home, flexible scheduling is often a must. Before my daughter came along, I had a real tendency to be precious about my process. I liked to write at certain times of day, for certain lengths of time, and I was pretty sure I couldn't do it any differently. With a toddler, though, I had to admit that my preferred schedule simply wasn't realistic; I had to figure out how to work in the moments I had, rather than the ones I wished I had, which meant structuring my work so that I could pick it up and get a few straightforward tasks done any time my daughter was occupied. Learning how to break my work down into small, flexible chunks like this was *hard*, but it really helped increase my output, especially during the years when my daughter was a very high-needs toddler and preschooler.

3. Learn to work in times that don't feel natural.

I am NOT an evening person. Most days, I feel like my brain turns off around two or three in the afternoon and doesn't fully click on again until midmorning of the next day. By the time my daughter is in bed, I'm *wiped!* But when she was very young, I hit a point where I realized that I would never be able to make my publishing dreams come true if I didn't learn how to work in the evenings after she was asleep. Instead of pulling out a book to read or selecting a Netflix show to watch, I started opening my laptop after getting her to bed and making sure I worked for at least an hour or so each night. At first, my efforts felt laughable—I hardly got any real work done, and what I did get done didn't feel like it was very good quality. But the longer I practiced, the more my brain started to get used to working in the evenings. Eventually, I was able to work smoothly and efficiently during those hours that used to feel completely worthless.

4. Get help.

No working parent is an island, and I highly recommend that, if it's at all possible, CF working parents ask for help—whether that means hiring a house cleaner if you have the financial means, getting grandma to take care of your child(ren) one day a week, or asking if your spouse can adjust his/her schedule to give you some child-free time during the day. Things are a little different in the middle of a global pandemic, but, in regular times, I often hire a local teen to help with basic cleaning jobs or get childcare help when I'm on a deadline and have to work longer hours than normal.

5. Learn which balls are plastic and which are glass.

Early in 2020, I heard a story about advice that the prolific author Nora Roberts gave on balancing a writing career while raising children. She said that every working parent has a lot of balls to keep in the air—usually, more than we physically or mentally *can* keep in the air all the time. Sometimes, no matter how hard we try to hone our juggling skills, some of those balls are going to fall and hit the ground.

The trick, Roberts explained, is to

figure out which balls are plastic and which are glass. In other words, we have to decide which balls are going to hit the floor and bounce (not cause irreparable damage) and which balls are going to hit the floor and permanently shatter. Dropping a plastic ball might still cause problems, but they will be problems that we can ultimately fix. Dropping a glass ball, on the other hand, is a much more serious thing. The makeup of the balls you're trying to juggle isn't static—sometimes, your glass ball might be an important work project that is due soon. Other times, the glass ball might be a child who is going through a difficult time. Your glass ball might also be a CF exacerbation or other health challenge that needs focus, time, and rest to heal. But by continually checking in with ourselves and our responsibilities to figure out which of the balls we're juggling are glass and which are plastic, we can mitigate lasting fallout as much as possible. No working parent—and especially not a working parent with CF!—is going to be able to do it all, all the time; part of successfully juggling so many different things is to recognize that there will be times when balls get dropped, and we should give ourselves grace in those times.

Balancing a career, parenting, and CF is tough work. It takes time, diligence, a frankly exhausting amount of creativity, and a willingness to be flexible and think outside the box. For me, it has definitely come with concessions: I'm not nearly as prolific an author as I would like to be, for instance! But I've found true joy in both raising my daughter and building my career.

I hope you do, too. ▲

Cindy is 32, has CF, and lives just outside Portland, OR, with her husband and seven-year-old daughter. She is the author of Where the Watermelons Grow, Beginners Welcome, *and the forthcoming* The Stars of Whistling Ridge, *all with HarperCollins Children's Books.*

Read Your Doctors' Notes!

By Amy Gutierrez

Like most people with cystic fibrosis, I see multiple care providers every year, and the burden to coordinate my care with all these different physicians falls on me. Coordinating this care means keeping track of a myriad medications, appointment dates, care instructions, and treatment plans. Caring for my CF is a full-time job and I would bet you feel the same way.

As a 39-year old with CF, I see: a pulmonologist; an endocrinologist for my CFRD; a gynecologist for my hormones and lady bits; a gastroenterologist for my CF tummy issues; a dermatologist for skin cancer screenings; a cardiologist because of my history of supra ventricular tachycardia; a hematologist for treating von Willebrand disease; a psychologist; an internist for vaccines and wellness visits outside my CF care; a podiatrist for diabetic foot exams; an ophthalmologist for diabetic eye exams; an otolaryngologist for CF-related sinus issues; a dentist, and a hepatologist to make sure my liver is in good shape yearly. Sometimes I see all these specialists several times a year.

In order to adhere to treatment plans and coordinate care between specialists, I must have real-time access to my medical notes. Not all of my providers are at the same medical institution, much less the same state, so I have to make sure I am able to share visit notes in a timely manner.

Having access to electronic medical notes (OpenNotes) is a game changer, not only for people with CF, but the chronic disease community as a whole. Sadly, not every medical institution and/or doctor's office gives patients electronic access to their chart notes. Some institutions allow providers the choice of whether to share their notes. For example, I have to ask my internist to "release" my electronic notes following every appointment with her. Why doesn't she do it automatically? She's afraid that patients will be offended or will have too many questions. She's an amazing doctor, and I am really grateful for her fantastic care, but I don't agree with her philosophy on sharing medical notes with patients.

AMY GUTIERREZ

Thankfully, the federal government agrees with me. Beginning November 2, 2020, all medical providers in the United States must provide all patients electronic access to their medical notes, with a few exceptions. This is a huge victory for the chronic disease community.

Medical notes are different than the After-Visit Summary. My friend, Liz Salmi, at OpenNotes (twitter @TheLizArmy) does an outstanding job illustrating the difference between clinical notes and the After-Visit Summary. The After-Visit Summary isn't very helpful and leaves out quite a bit of detailed information from the care visit. People with CF want electronic access to what is on the left in the graphic below, not on the right.

Above and beyond convenience, patients' access to their notes allows patients to more effectively coordinate their care, adhere to treatment plans, identify and fix errors more quickly, and secure and/or maintain disability benefits where applicable.

Studies show that "most patients remember less than half of what they discuss with their medical profession-

COMPARISON OF OPENNOTES (LEFT) AND AFTER-VIST SUMMARY (RIGHT).

als. To make sure you don't miss important information, OpenNotes gives you the opportunity to review the details of your visit at any time. Above all, OpenNotes helps you make sure that you and your healthcare team are always on the same page." That's a win for both patient and provider. https://www.opennotes.org/notes-you/

When notes from a visit are available, I email them over to my other care providers so that everyone is concordant. This provides continuity of care, education across my various specialists, and ensures that my care plan is documented with each provider. God forbid, if I'm ever incapacitated or unable to advocate for myself, my husband or extended family can have access to my medical history.

In 2019, I traveled five states away to have sinus surgery. I have von Willebrand, a bleeding disorder characterized by missing the Von Willebrand factor clotting protein. Any time I have surgery, I need DDAVP, a medication to control potential excessive bleeding. My hematologist is an expert in my bleeding disorder, so electronically sharing my hematologist's surgery treatment plan medical notes with my ENT prior to my sinus surgery (as opposed to verbally communicating my medical need, which can be easily forgotten or misunderstood), was the best way to ensure I was properly cared for during the surgery. This either isn't possible or is extremely difficult if I have to request analog medical records.

According to a study published in the *British Medical Journal*, "OpenNotes evidence has shown that transparent medical records can increase patient engagement—patients who read the clinical notes written by their doctors report feeling more in control of their care and being better able to adhere to the treatment plan." That sounds like what the Cystic Fibrosis Foundation advocates—coproduction of care.

> "Patients who read the clinical notes written by their doctors report feeling more in control of their care and being better able to adhere to the treatment plan."

According to a study published in the *Permanente Journal* in 2018, "[m]edication nonadherence for patients with chronic diseases is extremely common, affecting as many as 40% to 50% of patients who are prescribed medications for management of chronic conditions such as diabetes or hypertension. This nonadherence to prescribed treatment is thought to cause at least 100,000 preventable deaths and $100 billion in preventable medical costs per year."

So how can OpenNotes help our community? In a study published in 2012, "up to 78% of patients reported that OpenNotes helped them take their medications as prescribed." Additional evidence can be found in a study by the Geisinger Center for Health Research published in 2015 that found patients offered access to notes were more likely to fill their prescriptions for blood pressure medication. Treatment adherence and persistence in CF is a huge burden, so anything and everything we can do to improve that is a win for our community. We are all eager for the next, new CF treatment, but imagine what our health would be like if we regularly complied with the treatments available?

A recent Johns Hopkins study claims more than 250,000 people in the U.S. die every year from medical errors. I'm not sure about you, but I work too dang hard on my health to die from a medical error. You might be surprised at what you read in your chart notes when you get access to them after each visit. I've found several errors in my medical record that I likely would not have seen if I didn't have access to OpenNotes. According to my medical record: I'm adopted (I'm not), I have Type 2 diabetes (I don't, I have CFRD, which is very different), and I have Factor V Liden (nope, I don't have that). These are all big errors that could have a negative impact on my health. The right to access my records electronically, in real time, empowers me to make sure my record is accurate. *JAMA* found that one in five patients finds an error in their medical notes. That's way too common! 42% of these characterized a mistake as serious. How can we fix errors in our medical records if we don't have access to them?

Lastly, chart notes are essential for disability benefits, both public (like SSDI) and private. Anyone who has had the privilege of working with Beth Sufian on their disability case knows that what your physician does or doesn't write in your chart notes can make or break your disability case. Even if you aren't on disability now, making sure your medical notes fully capture your health is essential. OpenNotes is fundamental to this process.

So, what can we expect beginning November 2, 2020? "The program rule on Interoperability, Information Blocking, and ONC Health IT Certification, which implements the 21st Century Cures Act and requires patients be provided access to all the health information in their electronic medical records without charge by their healthcare provider **beginning November 2, 2020**. Clinical records must be shared with a patient's 3rd party application (e.g., downloaded to a smart phone) by November 2, 2022. (See this rule posted in the Federal Register.)"

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FOCUS TOPIC

ENTREPRENEURS WITH CF

How I Went From Social Security To Business Owner (The Untold Story)

By Klyn Elsbury

The doctors walked out of the ER room and the nurse walked in. It was my fifth hospitalization that year. My lung function hovered in the high 30s to low 40s. I had a six-figure career as a corporate recruiter with a corner office and a house I just paid mostly cash for with my own money.

And I couldn't keep up. My office at work was cluttered with IV polls and medical supplies. I had to be moved to a part of the building that wasn't as populated so my nebulizer wouldn't disturb my coworkers.

Every cold call, I loved. Every LinkedIn post, I loved. I felt a surge of purpose every time the employer said yes to a candidate I found and recruited to work for my employer.

My health, however, had bigger plans.

The nurse and I had become friends over the years so she didn't just see the numbers and progressions; she knew my inner desires and ambitions. And she said with bated breath, "Klyn, it may be time you file for social security. It's common in your 20s. You get a steady paycheck and can focus on your health." I nodded and cried myself to sleep that night, alone in the ER.

After I filed, I took odd jobs for cash because living in San Diego with diabetes and cystic fibrosis is so tough. I didn't want to move in with my parents again (who were living in Texas). I didn't have a significant other. I was alone. And to tell you I was positive that it would work out would be a blatant lie.

I felt every emotion and sometimes all at once: disgust, angst, hatred, loss of my faith in God, and loss of my identity. Who was I if I wasn't a corporate career climber?

I traded groceries for recruiting consultations. I put myself through Zumba training on an IV drip because it cost $150 to get certified and gyms paid $15 for cash instructions. I was so sick, I couldn't ever finish the class I taught but I could get hired for the next gig because I knew how to interview. I slept on couches. I dog sat for food for the millionaires who lived downtown. For three years, I took my government money and feelings of inadequacy and told myself this was not my final destination.

I learned everything I could about mindset and neuroscience. How are some people able to accomplish incredible feats no matter the circumstances while I, talented and sick, am apparently utterly worthless?

Eventually, people started asking me to speak. Nobody paid me. I met my now fiancé through these speaking engagements. He belonged to a group of entrepreneurs who are socially conscious and incredibly helpful.

During another hospitalization, I was talking on Instagram to a guy friend with cystic fibrosis. I asked him what he did all day, two rooms down from me. His response: "I play video games." I then asked a few more patients what they did to occupy the time. One by one, I heard answers like watch TV, read, and other fun, but mindless, activities.

I knew that wasn't my path. Why did we all suddenly know so much about the Kardashians but not how to market ourselves digitally? Why can we do math when it comes to calculating our Amazon purchases and how much insulin we need with 42.1g of carbs,

KLYN ELSBURY PRESENTING AT A CONFERENCE.

but we can't figure out how to create an LLC? There's got to be someone out there who had a tough life and built an empire? Who capitalized on their ambitions? Right?

So, I got on LinkedIn and searched relentlessly for Inc. 5000 winners. I didn't know what that was. I just knew if a business made it on that list, they must be rich. And, I found a group of successful entrepreneurs who agreed to talk to me.

I found many of them had really tough stories. One person couldn't decide if he should pay his light bill or buy food for his dog when he started out. Another person grew up in unimaginable poverty. Story after story of survival and strength enabled each of them to, in turn, become successful entrepreneurs.

During that same hospitalization, I wrote my book, *I AM ___: The Untold Story of Success*. It gave me the confidence to get off social security benefits and build a recruiting company of my own.

Within one year, I made more money than I could comprehend. And in year two, I hit rock bottom. Person after person after person whom I cold called could not have cared less about recruiting. They wanted to hear about mindset, entrepreneurship, and cystic fibrosis. I even had the idea of starting online courses to teach others with CF what I was learning about business, digital marketing, social media, and basic sales skills. The CF Foundation turned my application down.

I was told confidentially that cystic fibrosis patients aren't ready and have no desire to learn about business.

One night, at a party, the president of a local chapter of an international group and I were fortunate enough to be having dirty martinis on a beach. I told him about recruiting and how I knew this was not my life's purpose. I shared about my book, which had by then become an international bestseller, and he asked if I wanted to be the keynote speaker for a business conference of over 1,500 guests.

I said yes.

I leveraged that speaking opportunity to book a few other engagements for practice before I took the stage. None of them paid me. I was voted the top keynote speaker for that event, beating out Jocko Willink, Shep Gordon, and Tony Hawk, among other famous titans of industry.

One guy represented a bureau. The day the bureau called with news they wanted to sign me, I cried so hard. Finally, the message that you don't have to accept your medical limitations became my life purpose.

I immersed myself in self-help books, personal growth, neuroscience, and countless business trainings. I had a rule for every hospitalization: no TV; only online business courses. I found that the more I helped myself, the more others wanted to help me.

Suddenly, friends with million-dollar company valuations were gifting me courses and teaching me what I didn't know. They offered feedback. They even bought a few copies of my book. They told their friends, bureau connections, anyone and everyone about this wacko girl with 50% lung function who was building a business around changing lives.

The speaking gigs started coming in. The first time I was paid five figures for a 45-minute keynote, I was absolutely blown away. I couldn't believe that my name was listed up there among Mel Robbins, Rachel Hollis, and females I had been looking up to for years.

To this day, I still don't watch TV—with the exception of *Naked and Afraid* at night with my fiancé. Instead, I'm listening to Audible books, taking business courses, or attending neuroscience seminars. The more I learned, the more I developed courses on overcoming fear, embracing change, and creating your own headline for your life.

In time, my social media hit 60,000 followers as people started to rally behind this mindset. So, I needed a podcast, "The Motivated Mind," which has been a slow start but is now at 10,000 downloads.

I can't advise anyone on whether they should start a business. Heck, sometimes, I think I just got lucky. There have been months of zero income and thinking I'm still a mess up. There have been crushing blows to my heart as well. I work more now than I ever have. The first time I got hate mail and had to file a police report because I told someone online that they don't have to stay mediocre, that they can make small steps every day towards their goals, I questioned if I was tough enough. But, I remembered why it's worth it when the filed report was followed by a message of how I helped someone overcome their fear of failure, or repaired their divorce, or ended their nicotine addiction.

Business and entrepreneurship are not for everyone. But, for the person reading this who knows they aren't living up to their potential and believes they have what it takes to put themselves out there, to give up TV and luxuries, to struggle and ultimately figure out an IV drip on a conference call, to go months without knowing when the next paycheck is—my message is simple: trust your gut. ▲

Klyn Elsbury is 32 years old and has CF. She lives in San Diego, CA, with her fiancé and furbaby. She is an international best-selling author, business owner, and motivational speaker. Klyn is passionate about teaching others what is possible through her health journey and entrepreneurial skillset. You can listen to her podcast at "The Motivated Mind" and contact her on her website: www.missklyn.com.

The Business Of Stronger And Longer Lives With CF

By Brian Callanan

One of my favorite quotes is from the book *The Tao of Pooh* by Benjamin Hoff, which illustrates the concepts of Taoism through the characters of Winnie the Pooh. Hoff writes, "A saying from the area of Chinese medicine would be appropriate to mention here: 'one disease, long life; no disease, short life.' In other words, those who know what's wrong with them and take care of themselves accordingly will tend to live a lot longer than those who consider themselves perfectly healthy and neglect their weaknesses."

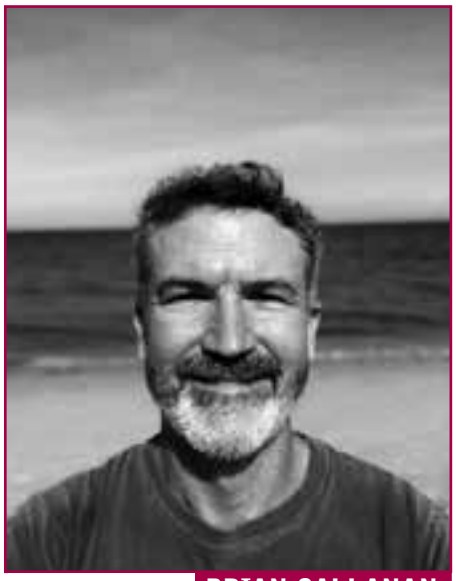
Late in high school, I experienced a deep depression and I learned how important, yet difficult, it could be to deal with unseeable forces such as anxiety and depression, forces that would impact my living with CF as much as not doing my chest therapy or exercising.

Studying psychology in high school, college, and later in my post-graduate studies taught me about the connection between mind and body. Living with CF has taught me to accept my physical and psychological weak points so that I can focus on taking care of them in order to follow Hoff's advice in *The Tao of Pooh*.

When I was 21, my older brother, who wasn't a CF patient, died at the young age of 39 from a heart condition left untreated. It was then that I realized what the Taoist concepts truly meant.

The connection of addressing both physical and psychological weak points (*i.e.*, lungs, digestion, diabetes, isolation, depression, and anxiety) for me became exercise.

Exercise is what I have come to consider "the bridge" between both physical and mental health and well-being. My desire to help others came from a combination of psychological, social, emotional, physical, and financial challenges all of which were concentrated by the presence of multiple rare diseases. All together, these challenges afforded me the opportunity to nurture wellbeing in accordance with the goal of living a long life.



BRIAN CALLANAN

This was the complete opposite of the statistics, prognosis, and empirically based mindset I had been raised with that guided my sights toward looking down instead of looking up.

In this vein, CF was never keeping me from my goal of helping others in life, but instead was my driving force. I never aspired to work for myself and, honestly, was scared of it at times—I felt like I didn't know what I was doing. This insecurity dovetailed with an ethical distinction between creating a mission focused not on helping me with my CF management, but on helping me help others who were interested in taking care of themselves to hopefully live stronger and longer lives.

My initial idea for the Cystic Fibrosis Lifestyle Foundation ("CFLF") was very different than what it looks like today. The idea of an outdoor therapeutic clinic for the traditional "two-week tune-ups" came to me during my first hospitalization when I was in my junior year of high school. I was fed up with the atrophy of lying in bed for two weeks, the buzzing and flickering of fluorescent lights, the disrupted sleep, the unpredictability of treatment times and, even more so, the feeling of missing out on life as my peers continued to enjoy it.

I believed there could be alternative, more holistic ways of providing IV treatments, nutritional supplementation, and intensive airway clearance. I felt that a setting that offered traditional tune-up therapy in conjunction with outdoor activities aimed at building confidence, stamina, and empowerment over CF, while also offering support for the psychological and social issues would be ideal. I needed to hammer out the logistics of making that a reality. Building relationships with the individuals, families, and care providers within the CF community was the prerequisite for the CFLF to thrive.

In 2003, after extensive internet research, I took the first step and incorporated the CFLF. Over the past 17 years, I have learned that there are pros and cons to running your own business versus working for a company:

- I could work from home, but at the same time would experience isolation from the social life of the office.
- I could avoid sick coworkers, but at the same time would not be exposed to bugs that would also gradually build a stronger immune system.
- As a CEO of my own business, I

could have the flexibility to scale back on work when I was exhausted, sick, or hospitalized, but also did not have a team of colleagues that could help cover for me in my absence since many of my job responsibilities aren't something that can passed down to company employees. If I didn't do the work, it just wouldn't get done.

- If the business wasn't profitable, my employees and I would be without a paycheck.
- Running a nonprofit organization meant a limited earning potential whereas, in a traditional company environment, the physical demands of travel, stressful deadlines, office politics, etc., affected my CF management, thereby limiting my earning potential in a traditional corporate environment.

I evaluated these risks constantly throughout the journey of starting CFLF, whose mission has a substantial impact on people's lives. The feeling of this mission being more of a vocation than a job was a significant element in my decision making.

Initially, after college, I wanted to pursue a Ph.D. in kinesiology with the goal of publishing research about the importance of exercise in CF but never found a program that recognized the intersection of mental health and physical health as a holistic approach to wellness. I discussed my interests with my physician at the time, Dr. Patricia Walker, who suggested starting a nonprofit program to help patients, as opposed to just studying the impacts of exercise on life with CF. That was the epiphany that changed my mindset from observing to doing.

So, I began the CFLF as a 501(c)3 nonprofit organization specifically focused on teaching people with CF the importance of exercise as one of many critical factors in surviving CF. By helping people with CF overcome financial barriers to exercise activities, we were providing opportunities for people with CF to address any of their physical or psychosocial weaknesses, while also enjoying an activity of interest.

Part of my choosing this business was an interest in having a professional life that would require me to be physically active. I knew that I did not want to pursue a field that would mostly confine me to a desk nor compromise my health in pursuit of financial ascent.

Initially I relied on building relationships within the community and coordinating with CF clinics with whom I already had a relationship. I transitioned through two pediatric doctors at the University of Vermont into adult care, and my physicians saw that my consistently high lung function and quality of life was tied in with the active lifestyle I was leading. Pediatric doctors Tom Lahiri and Laurie Whittaker-LeClaire were huge supporters in my initial outreach for starting the CFLF after seeing my consistently high lung functions and quality of life during clinic visits. I also felt it was important to demonstrate on a larger scale that people with CF could defy statistics. In 2006, I rode my bicycle roughly 2,000 miles from Canada to Key West, Florida, in order to bring awareness and fundraise in support of the CFLF.

> "Part of my choosing this business was an interest in having a professional life that would require me to be physically active."

Although I achieved national coverage, awareness within the CF community did not happen until we secured a marketing partnership with a pharmaceutical company that promoted the CFLF Recreation Grant program through their sales team in all CF clinics around the country. In 2013, we had more people asking for help than we were financially able to support. As a result, we had to scale back our marketing efforts as the program became increasingly competitive.

Fundraising is an extremely challenging element of running a nonprofit organization. It's critical to build a donor base that both gives support and spreads awareness of the nonprofit mission in their respective networks. I still have a lot to learn, especially with the changes resulting from COVID-19 this year. I strive to always focus on my desire to help others and shine the spotlight on others living the CFLF mission of *stronger* and *longer* (#STROLO) lives.

In starting the CFLF, help came in many different forms: consultations; evaluation of need, transferability, and viability; driving a car hundreds of miles to support a cycling campaign; and jumping in to help organize and plan new events, some of which still exist 13 years later. The occasional fear and uncertainty of not knowing if we will make it as an organization requires a tremendous amount of emotional support from others. Fortunately, in both good times and bad, the CFLF Board of Directors has provided unwavering guidance in line with our mission. It goes without saying that building relationships in the community has proved invaluable for financial support in all shapes and sizes. All of this together means that the CFLF is changing lives with CF for the better; we're

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Three Tips To Building A Business For People With Cystic Fibrosis

By Mandy B. Anderson

RAYMA Team. The name dropped in my head while completing a five-mile run on the treadmill in the spring of 2019. It's a combination of my name and my bestie's name, and it became the new name of our organization when we rebranded from a company that focused on life coaching for women to a company that gives leaders and entrepreneurs the skills to grow their influence and hope to stay the course.

My name is Mandy B. Anderson and I own my company with my best friend. That fact alone brings with it a set of unique challenges. However, overcoming challenges is not a new adventure for me because I also have cystic fibrosis. My best friend and I started our first company back in 2014—we rose up and took the leap of faith into being business owners with a bottle of wine and determination. I'd like to share with you a few of the lessons I've learned since then on how to handle being an entrepreneur with CF.

> "Years before I jumped into owning a company, I spent time learning and applying basic skills like time management and intentional rest days."

MANDY B. ANDERSON

Tip 1: Schedule Your Energy

Years before I jumped into owning a company, I spent time learning and applying basic skills like time management and intentional rest days. I'm so glad I learned those skills because the journey of entrepreneurship has been known to burn out many people.

My health is non-negotiable. This means I need to schedule my energy accordingly. That looks like making sure I have one day a week where I can work from home in my jammies and not have meetings so if I need extra rest, I can get it. It also means that I

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https://www.opennotes.org/onc-federal-rule-interoperabilty-information-blocking-and-open-notes/

OpenNotes provides us with further information about this law: "[t]he eight (8) types of clinical notes that must be shared are outlined in the United States Core Data for Interoperability (USCDI), and include: consultation notes, discharge summary notes, history & physical, imaging narratives, laboratory report narratives, pathology report narratives, procedure notes and progress note. Clinical notes to which the rules do not apply: (1) Psychotherapy notes recorded (in any medium) by a health care provider who is a mental health professional documenting or analyzing the contents of conversation during a private counseling session or a group, joint, or family counseling session and that are separated from the rest of the individual's medical record. (2) Information compiled in reasonable anticipation of, or use in, a civil, criminal or administrative action or proceeding." However, due to COVID-19, the national coordinator for health information technology at the Department of Health and Human Services says it may not enforce the November 2, 2020 rule until February 1, 2021.

Whatever the enforcement date ends up being, this law is a big win for people with cystic fibrosis and those with chronic illnesses. If you have access to your data electronically, make sure you're always checking your latest visit notes. If you don't have access, make sure your institutions know that you are aware of this federal law and you will be expecting their compliance. This is our data, and in the amazing words of ePatient Dave, "give me my damn data!" If you haven't seen his TED talk, The year of patients rising, I highly recommend watching it—he is an amazing leader in our community. ▲

Amy Gutierrez is 39 years old and has CF. She lives in Los Angeles, CA, with her husband Joel. She loves traveling internationally (when there isn't a pandemic), standup comedy, and college football.

practice having healthy boundaries around work time v. home time.

When you know yourself well, you can schedule your time better. This has been one of the reasons why I can stay healthy and on top of things when it comes to cystic fibrosis. Being intentional with my rest days and stress management led to my creation of a course called, *The Art of the P.A.U.S.E.*, which is available on our website if you need help with scheduling healthy boundaries.

Tip 2: Have Your Own Health Insurance Plan

Starting a business can be exciting and scary all at the same time. There are so many unknowns! I made the decision to be on my own individual health insurance policy so that I didn't have the added stress of worrying about medical bills while building our business. I also suggest that you lean into the organizations that are available to help cover medical costs that insurance doesn't cover.

In my state, there is an organization called the Cystic Fibrosis Association of North Dakota. They reimburse CF members for a percentage of out-of-pocket medical expenses not covered by insurance. This is a *huge* help to families, and especially to those of us building businesses at the same time. Never be afraid to tap into those type of resources. They are there for *you*. Connect with them and become an advocate for them at the same time.

Tip 3: Keep Your Day Job

Most businesses take five to seven years before they become profitable, and many times the owner doesn't take a salary during that time. This fact is not talked about that often, and the daydreams of being your own boss and making your own money glamorize the entrepreneurial adventure.

Building a business takes work and it is wise to keep a part-time job so you have a steady income while building your business to provide the full-time income you want and deserve. Full disclosure: I used to think that I couldn't handle both building my business and working a part-time job because of CF. I was wrong; and I put our family through some lean financial times because of that limiting belief.

I now have a part-time job that allows me to work 10 hours per week and still have time to build my business. It gives our family the ability to have the income we need while my business is still growing to pay me a full-time income. And the funny thing is, I'm actually way more productive in my business because of the part-time job! My health has been thriving, too.

These three tips have helped me be confident as I build my business and still give me the ability to manage a life affected by CF. And when I hit a setback because of CF, I have the peace of mind that everyone involved will understand and work with me because I've already established the expectation that my health is nonnegotiable. ▲

Mandy B. Anderson is 38 years old and has CF. She's a leadership coach and the Chief Creative Officer (CCO) of RAYMA Team, where she helps leaders and entrepreneurs be healthy, stay creative, and have a dangerous hope. Mandy is the host of the "She Who Overcomes" podcast and is an upcoming TEDx Speaker. She lives in North Dakota with her husband, Nate. Connect with her on Instagram at @msmandybanderson. Find more information about her books, courses, and coaching at www.raymateam.com.

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helping people strive to live long enough to see a cure.

Relationships are everything. Building new relationships, and nurturing old relationships are the key to new ideas and opportunities.

Taking care of our team is as important as providing help to the CF community—without an effective team, none of this would happen. Because we are a mid-sized nonprofit, accessing health insurance for myself and employees is a challenge. Group policy rates often outweigh our employees' abilities to access private or marketplace insurance at significantly lower costs. Because we have employees in different states with different needs and family situations, CFLF provides team members with the ability to secure their own level of insurance with a certain percentage of the monthly premiums being paid by CFLF directly.

I still face challenges specific to both me and our Program Coordinator, who also has CF. I am a social person and would prefer a work environment that allows for person-to-person interaction and project collaboration; however, we must work remotely. So, in this era of COVID-19, we have actually been a more prepared than most as we already have remote capabilities for working from home.

While I know that CFLF is continually capable of achieving more, we are always welcoming committee members, volunteers, and board members who live and ascribe to the importance of shaping a lifestyle that promotes an active role in taking care of one's health through physical and mental wellbeing. ▲

Brian Callanan is 43 years old and has CF. He lives in Miami, FL, with his wife, two stepdaughters, and their furbabies. Brian is the founder and Executive Director of the Cystic Fibrosis Lifestyle Foundation (CFLF). He enjoys open-water swimming, distance cycling, high-altitude snowboarding, hiking, sailing, and rock-climbing. You may email him directly at brian@cflf.org or visit his website at www.cflf.org.

Creating While You Cough: How Nicole Made Her Journey With CF Profitable

By Nicole Kohr

NICOLE KOHR

I was always a business owner at heart. When I was eight years old, my mother bought me the laminator that I had been asking for, and I used it religiously. I was always that type A individual. As I grew older, however, my career path went down a windy road. I started out in the theater as an actor and worked my way up to community theater choreographer after several professional acting gigs in my youth. The overall profession became more and more taxing on my body, especially after I was diagnosed with *M. abscessus.* I knew I would have to take a more "realistic" path if I wanted to guarantee my own employment with insurance. So, I majored in public relations. This is where I found my second love: the creative aspect of the administration process. Logos, campaigns, and press...oh, my! I also minored in child psychology and became certified in the theory and practice of child life just in case my CF was cured so I could work in peds. Yes, CF created its obstacles, but overall forced me to be more creative and strategic with my career decisions. After working for a few years in a nonprofit organization, I published my first book and discovered my third love: writing and producing musicals about CF. Who knew all three loves would come together?

Things got off to a rocky start. I had an amazing musical sitting on my laptop, but no one, with the exception of my mother, was willing to listen. I started writing it when I was 12. I copyrighted it when I was 16, and it sat on my laptop covered in dust until just before my transplant in 2019. I knew early on that I would have to write something smaller to get my name out there—something that would identify me as a writer. So, I reached out to a few artists associated with my nonprofit work. Within six months I, with the help of one of my artist friends, published *Two Cents* and I started my own LLC (for writing). I loved writing the book, but it was just a stepping stone. With my name on the map, I started blogging for everything and everyone. The blogs were conveniently timed because my health was declining quickly, so every story made for a good read. Reaching a FEV1 of 20% is what gave me that final chip on my shoulder that I needed to get my play off my computer. I sent it to everyone I could think of, including the CF Foundation, and asked them to read it. I also applied for an Impact Grant Award, albeit late because I was in the middle of my transplant evaluation. A friend, who helped with some of the budgeting, and I were awarded the grant the day before my wedding, May 31, 2019. I was married on June 1 and received my new lungs on June 14. That didn't stop me from having a full staff of CF musicians ready to go by July 1. It was an uphill climb from there.

Most people underestimate how much work goes into writing a book or writing a musical. They're both overwhelming tasks. You have to find a composer who understands your story and the style of music that you're trying to convey. You need a strong team of people to help you market the show, apply for grants, ensure you stay on budget, recruit and cultivate participants, hold professional and semi-professional auditions and shows, send contracts, be a devil's advocate for different aspects of the show, lighting, costume, and design. The list goes on and on...

Two Cents catered to a wide variety of people. I did that on purpose. I

didn't want my first book to be specific to CF. My mother always encouraged me to write about CF. However, the arts were my escape from CF. I never wanted the two to collide and, up until the day of my transplant, only a handful of friends and family knew that I had CF. I came out of the closet, if you will, when I was being wheeled into my transplant surgery. "Post everything!" I shouted to my mom, and she did. This gave me the freedom and the platform to write about cystic fibrosis. The only thing that I would not budge on—it had to be a comedy.

Fall Risk is a musical comedy about chronic illness and transplant. The show shines a special light on cystic fibrosis by chronicling the life of Gabriella Michaels, a CF patient being evaluated for double lung transplant. Gabby has just received a callback as the leading lady for a regional theater show. Now her family, her doctor, and her director have to convince her which aspect of her life is more important. Described as "this generation's *Rent*," *Fall Risk* will pull on your heart strings and tickle your funny bone.

I chose this business because being my own boss allows me to be as type A as I want to be on the days that my body is able and to rest on the days that my body is not. I do most of my business through recruitment or word of mouth, but the amount of logistics that goes on behind the scenes is a team effort. I do my best to team up with as many CF patients and CF organizations as I can, despite the competitive nature of grants and the arts. You can't do it by yourself—healthy or otherwise, it's not physically possible. It's all about steadiness and endurance. We're all fighting for the same cause: awareness, funding, and a cure. So, you must have some kind of team supporting you.

I've learned a lot about myself throughout this experience of business ownership and leadership. I've learned that there are no rules. Yes, there are laws that you have to follow. But there's no one-size-fits-all when it comes to a disability, nor is there a one-size-fits-all for business. We're entering a new era of remote access and online accessibility. If there's anyone who knows how to multitask, cultivate constituents online, and handle the stress of isolation, it's a CF patient. I've learned how to balance work, life, *and* health. I've learned that it's not that hard to start a business if you do all of the research and have a couple hundred dollars. It's *incredibly* difficult to *upkeep* a business, especially when you're pushing yourself through your medication routine or a respiratory infection. I've learned that my laminator is priceless. I've learned that I'm even stronger and more resilient than I thought. And I've learned to believe in my own company. ▲

Nicole Kohr is 27 years old and has CF. Nicole received her double lung transplant in 2019. She resides in Somerset, NJ, with her husband Jared. Nicole is an author, lyricist, and owner/executive producer of her infamous Fall Risk *musical designed for the CF community. You can reach Nicole on her website: www.fallriskthemusical.com.*

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of life was found in 8% of participants. Evidence confirms previous recommendations for routine hearing evaluations at high frequencies (>8 kHz). Age is a known risk factor for hearing loss and, within this study cohort, also likely represents cumulative antimicrobial therapy. The authors suggest that annual screening including vestibular and tinnitus questionnaires could identify patients with unidentified hearing loss, but that further research is still required to validate the use of tablet audiology-based longitudinal monitoring.
https://tinyurl.com/yxro8pkw

Effect Of Concomitant Azithromycin And Tobramycin Use On Cystic Fibrosis Pulmonary Exacerbation Treatment

Azithromycin (AZM) is one of the most widely prescribed chronic medications for CF in the United States. Recent evidence has identified a potential antagonistic relationship between AZM and tobramycin. AZM use both at the most recent outpatient clinic encounter and during pulmonary exacerbation (PEx) treatment in combination with IV tobramycin was associated with a significantly lower absolute improvement in percent predicted forced expiratory volume in 1 second (ppFEV1), a lesser odds of returning to ≥90% of baseline ppFEV1, and a shorter time to next PEx requiring IV antibiotics compared to IV tobramycin use without concomitant AZM. These results support the hypothesis that an antagonistic relationship between these two medications might exist.
https://tinyurl.com/y3dnfgq8
AND
https://tinyurl.com/y37zz3tq

Azithromycin And Tezacaftor/Ivacaftor Is Associated With First-Degree Heart Block In An Adult With Cystic Fibrosis

The Cystic Fibrosis Transmembrane Conductance Regulator (CFTR) gene is expressed in the heart, but this is not known to cause myocardial dysfunction or abnormalities in the ECG in people with CF. CFTR modulators such as tezacaftor/ivacaftor improve lung func-

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Designing My Life

By Shaun Clarke

Owning my own business was always a thought in the back of my mind. I felt that, in doing so, I could cater to my CF/post-transplant lifestyle. As my work is all online, I knew it would allow me to take my work wherever I needed to, especially with the unpredictable hospital visits.

CF was the primary reason for wanting to run my own business. I believe COVID-19 solidified and validated that decision. The changes that the world is facing right now have become a bigger challenge for those of us with chronic illnesses. I felt the chances of me working directly with the public again were getting slim as it was all far too uncertain. While in quarantine, I was able to focus on where I should—and ultimately would want to—be with my work.

I am a graphic designer and illustrator. Starting a business in that field made perfect sense for me. Various aspects of my job will definitely cater to the CF community as well as the general population. My goal is to bring my knowledge and life experiences to the table and share that through my artistic abilities. As of now, I am in the early stages of building my business. Clients and advertising will come at a later date, but planning is in forward motion.

It has actually been easy for me to start building my business. There are very little overhead costs, and the only support I need as of now is emotional. I have not used any specific business model, so I am just winging it for now. I've never made a business plan. Right now, I am in the stages of trial and error. I got my idea for the business from thinking like a kid. I wanted to help kids process what they are going through and normalize chronic health struggles by means of art. Artistic creativity will be a helpful tool in a fun, but educational, way. The goal is to use my story to support others.

SHAUN CLARKE

> "I got my idea for the business from thinking like a kid."

My biggest challenge to date would be focus. I have learned that in order to succeed, there has to be a measure of self-discipline. Because I had not held a traditional job for a long time, I had developed some bad habits. I went back to school a few years ago and earned my degree in graphic design. This helped me regain the focus and energy that I needed to move forward. I took an online course, which kept me safe from any potential illnesses that circulate through college campuses. I was forced to create a schedule that worked for me. I had to make sure my work got done in a timely manner, which demanded time management and responsibility. Working at home has many benefits and downfalls. Without the skills I learned, I am unsure how I would have been able to achieve my goal.

I live in Canada and have health insurance through the government. I also have the added benefit from supplemental insurance through my wife's work. This has made a tremendous difference in having the flexibility to develop my own business.

As much as I would love to work with the public again, I don't think I could work under anyone again. I do miss meeting new people and having new experiences that can't happen from the comforts of my home. Not working a traditional job has made me very independent, however. The thought of being controlled by an employer at my age truthfully does not sit well with me. I am far too set in my ways, so having my own business caters to my personal needs. Besides, why would I want to make someone else money when I can make it for myself? ▲

Shaun Clarke is 40 years old and has CF. He lives in Niagara Falls/Ontario, Canada, with his wife Andrea and their two furbabies. Five years post-double lung transplant, Shaun has adjusted to the rollercoaster of his new life through the support of family and friends. You can find him and his wife on long-distance walks with their pups, experimenting in the kitchen with delicious recipes, and spending time with loved ones. You can reach Shaun on Instagram: @theartofshaun

PHOTO BY RAY LOCKETT

Augie's Clinic

She doesn't ask
About the productivity
of my cough
or what meds I'm taking.
My weight is meaningless to her.

But she willingly keeps me company
on those sleepless nights
full of coughing fits.
And extra snacks are just a bonus
to her foraging nature.

She takes no interest
in the results of blood work
or sputum cultures.
Readings from the peak flow
and glucose meters tell her nothing.

For, she already knows
the best way to counteract
unpleasant procedures
a game of tug-o-war,
a belly rub or squeaky toy
makes everything seem a little better.

She has her own methods
for counteracting bad days,
those of shortness of breath,
fatigue and no appetite.

And sometimes all I need
is what she has to offer
the silliness of a jester,
an extra nap on the couch,
soft fur soothing
skin that has felt
too much harshness for one life.

—C.Martinet, 2003

FROM OUR FAMILY PHOTO ALBUM...

SHAUN CLARKE AND HIS WIFE, ANDREA, AT A CHRISTMAS PARTY.

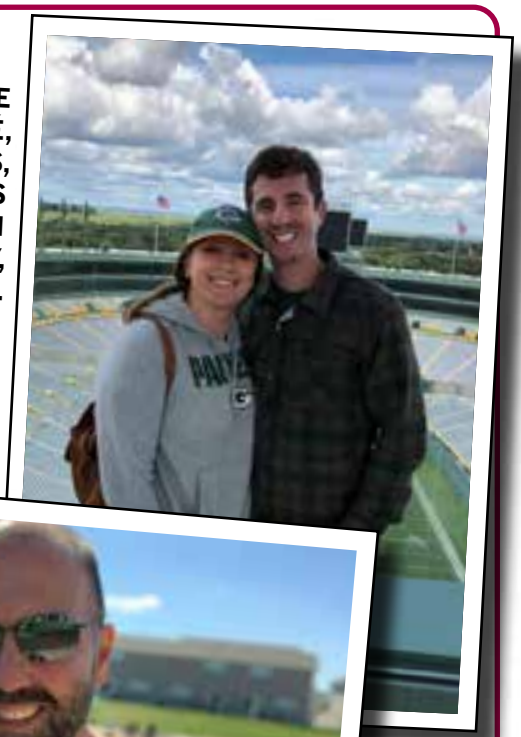
BRAIN CALLANAN AND HIS WIFE, MARISSA WONG, CYCLING ALONG LAKE GARDA, ITALY, ON THEIR HONEYMOON IN 2019.

HUSBAND, JARED, AND MOTHER, PATTY, GIVE NICOLE KOHR LOVE ON HER WAY INTO TRANSPLANT SURGERY.

KLYN ELSBURY

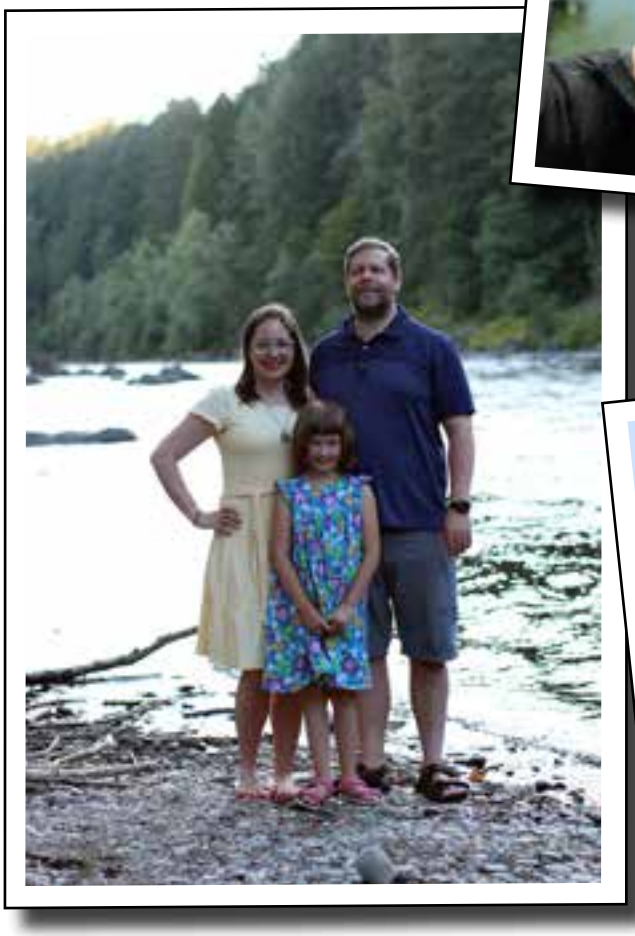
DEVIN WAKEFIELD MARCHES DOWN BUCK CREEK VALLEY NEAR GLACIER PEAK IN THE CASCADE MOUNTAINS OF WASHINGTON.

ARDEN AND KATIE LOCKWOOD WITH THEIR NEW DAUGHTER, ROSE MILLY LOCKWOOD.



JEREMY MOORE AND HIS FIANCÉ, SARAH PHILLIPS, AT THE PACKERS STADIUM IN GREEN BAY, WISCONSIN.

MANDY AND NATE ANDERSON.



THE BALDWINS: CINDY WITH HUSBAND, MAHON, AND DAUGHTER, KATE.

AMY SHROYER BRANHAM AND HUSBAND, SGT. ROBERT BRANHAM.

Recipients Of The Lauren Melissa Kelly Scholarship Announced

The U.S. Adult CF Association (USACFA) is pleased to announce the recipients of the Lauren Melissa Kelly ("LMK") 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 **Daniel Gonzalez Davila** and **Ashley Wilson** as the recipients of the scholarships for this calendar year. They will each be awarded $2,500. Congratulations to both!

Daniel Gonzalez Davila is an accomplished young man pursuing his Bachelor's Degree in Business at Pennsylvania State University, where he has been recognized on the Dean's List. He has been a volunteer with the *CF Roundtable* as an assistant to the treasurer for the last few months. As someone who immigrated from Mexico when he was young and who received a lifesaving transplant a few years ago, Daniel has faced tremendous health and financial struggles, yet he is fueled by his passion for helping others in his educational pursuits. One day he hopes to start a nonprofit that financially helps families of children and people who have a chronic illness so that they don't have to limit their lives to make ends meet. Daniel helps assist USACFA's treasurer behind the scenes. He also raises money for the CF Foundation and hopes to continue giving back to the chronic illness communities for as long as he can.

DANIEL GONZALEZ DAVILA

Ashley Wilson is a motivated young woman who is pursuing a Bachelor's Degree in Cinematic Arts at California State University of Monterey Bay. She has become fascinated with the art of creating videos and the world of being behind the camera. She also created a YouTube channel to share about her life with CF and how she has learned to live with a positive perspective. Through this, she strives to encourage the youth in the cystic fibrosis community. She has also been a videographer at local cystic fibrosis awareness and fundraising events like the Living Breath Foundation. She hopes to continue mentoring for cystic fibrosis youth and serving as a resource and support through her social media platforms. She has volunteered for many years with 4-H club, holding various leadership positions including president and secretary, as well as volunteered on various advocacy and awareness projects with the CF Foundation.

ASHLEY WILSON

Both Daniel and Ashley 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 offered in the fall semester 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. ▲

Benefactors

BRONZE

Anonymous (in memory of Rich DeNagel)
Kerry Bender (in memory of Jeannine Ricci)
Nancy Coleman
Daniel Crain (in memory of Jeannine Ricci)
Jonathan Fella (in memory of Jeannine Ricci)
Christine Greco (in memory of Jeannine Ricci)
Thomas Hatton (in memory of Jeannine Ricci)
Michael and Angela Hennessey (in memory of Jeannine Ricci)
Douglas Johnson
Kim Walcott King (in memory of Jeannine Ricci)
Ken and Peggy Martinet (in memory of Catharine Martinet)
Nancy Moseley
Angela Plummer (in memory of Jeannine Ricci)
Cantor S. Rabinowitz
Rosemary Randazzo
Charles Seiferman, Jr. (in memory of Jeannine Ricci)
Steven Tonkovich (in memory of Jeannine Ricci)
Charles Van Orden (in memory of Jeannine Ricci)
Donna Van Vlack (in memory of Jeannine Ricci)
Robert Zacharatos (in memory of Jeannine Ricci)

SILVER

Mary Jo and John Edelman (in memory of Jeannine Ricci)

PLATINUM

Brandywine Global (in memory of Jeannine Ricci)
Kroger Specialty Pharmacy
Michael Ricci (in memory of Jeannine Ricci)

SUSTAINING PARTNERS

Abbvie
Gilead Sciences
Maxor
Smith Foundation (in memory of Jeannine Ricci)

PEARL SUSTAINING PARTNERS

Boomer Esiason Foundation
Cystic Fibrosis Foundation

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tion and overall health in people with CF. Minor adverse events have been reported in the early clinical trials, but no consistent changes in the ECGs have been reported. This case highlights an unusual side effect of first-degree heart block that occurred after more than 8 months of azithromycin and tezacaftor/ivacaftor in combination. Drug withdrawal and reintroduction confirmed that neither drug alone, but only the combination, caused this change. As tezacaftor/ivacaftor is also present in elexacaftor/tezacaftor/ivacaftor, care may be needed to exclude this delayed interaction with azithromycin. This patient remained well but exhibited prolongation of PR interval to 334 ms. Evidence of drug/drug interaction appeared only after 8 months of treatment. PR interval was affected only by the combination but not by either of the drugs alone. They suggest a possible necessity of monitoring ECG when using azithromycin and tezacaftor/ivacaftor.
https://tinyurl.com/yyjsnzno
AND
https://tinyurl.com/yx9tshun
AND
https://tinyurl.com/y6s289kv

Scientists Discover Curious Clues In The War Between Cystic Fibrosis Bacteria

Several kinds of bacteria can cause lung infections in people with cystic fibrosis (CF). *Pseudomonas aeruginosa* typically infects infants or young children and persists for life, while *Burkholderia cepacia* complex species only infect teenagers and adults. Although Burkholderia infections are rare, when they do take hold, they are deadly. Researchers have discovered a reason for this pathogen's apparent age discrimination. The research shows that both Pseudomonas and Burkholderia use toxic weaponry, called Type VI Secretion Systems (T6SS), to compete with and establish dominance over each other. Pseudomonas bacteria isolated from infants and young children use their harpoon-like T6SS to fire toxins at, and kill, competitor bacteria, including Burkholderia. Although Burkholderia also produce T6SSs, they cannot effectively compete with Pseudomonas isolates from young CF patients. However, as those Pseudomonas bacteria adapt to living in the lungs of CF patients, they lose their ability to produce T6SSs and to fight with Burkholderia. The Burkholderia, using their own T6SSs, are then able to kill the Pseudomonas and establish *infection*. Some strains of Pseudomonas evolve to persist in the CF lung, but they also evolve to lose their T6SSs, and their competitive edge over Burkholderia, which are then free to colonize the respiratory tract.The scientists think the Burkholderia T6SS is an important fac-

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tor promoting the ability of these pathogens to infect CF patients. Therefore, researchers could potentially develop therapeutics to target these secretion systems to prevent infections. Assessing the T6SS potential of resident Pseudomonas populations within the CF *respiratory tract* may also predict susceptibility of patients to potentially fatal Burkholderia infections.
https://tinyurl.com/y43ru8p7

Bacteria-Eating Viruses Could Provide A Route To Stability In Cystic Fibrosis

Because phages can defeat bacteria when antibiotics fail, people with late-stage cystic fibrosis are increasingly seeking the treatment on a compassionate-use basis. A dedicated network of virologists, pulmonologists, clinicians and others has stepped up to deliver, hunting down the right strains of phage to treat patients' infections. By the end of 2020, Armata Pharmaceuticals expects to launch one of the first clinical trials of phage therapy for cystic fibrosis. Phages attach to bacterial cell surfaces and inject their genetic material, hijacking the bacterium's machinery and forcing it to churn out thousands of phages. The overstuffed bacterial cell then bursts, releasing the viruses. That is, the phages replicate at the site of the infection and continue to destroy bacteria. Because each phage attacks a specific strain of bacteria, the treatment does not damage surrounding tissue.
https://www.nature.com/articles/d41586-020-02109-7

UVA Researchers Studying Mucus To Make A Difference In Cystic Fibrosis Patients

Researchers are hoping to create a computer model to understand which genes cause cystic fibrosis and other diseases. They have been working on the research to determine if properties of mucus can affect whether or not an infection can occur in diseases like cystic fibrosis. One of the important outcomes from this will be tools that doctors and clinicians can use to improve their approaches for treatment and possibly for prevention of different health issues related to mucosa. By understanding how bacteria interact with these molecules, researchers can understand better how to stop these infections from settling in.
https://tinyurl.com/y32w9r9m

Beyond Air Publishes New Data On Nitric Oxide To Treat *M. Abscessus* In The Peer-Reviewed Journal Access Microbiology

Beyond Air, Inc., a clinical-stage medical device and biopharmaceutical company focused on developing inhaled nitric oxide (NO) for the treatment of patients with respiratory conditions, announced that the results from a compassionate use patient case study using NO to treat pulmonary *Mycobacterium abscessus* disease (*M. abscessus* disease) was published. The study evaluated the effect of inhaled NO therapy delivered via the LungFit™ System as compassionate treatment in a 24-year-old, female cystic

POETRY CORNER

IV

Stretch-tie-snap-pull
my skin taut
to accept the
floating dagger
threatening, gleaming, coming
plastic missile,
key fit for the
gates of life--
One, two, three
four jabs all
miserable mis--
haps, an unbloody
mess; no red or
green in sight--
only brown and
ow! stop poking
dammit; all right i
resign: here's the
other arm here's
another shot--
will this one
ultimately
be in
vain
?

Rob De La Noval is 31 and has CF. He lives in South Bend, IN, where he teaches theology and philosophy. He enjoys slasa dancing, working out, and is currently obsessed with the musical Hamilton.

Meet A New Director—Xan Nowakowski

Dr. Alexandra "Xan" Nowakowski is a medical sociologist, public health program evaluator, community advocate, and peer counselor living with cystic fibrosis. You may recognize them from *The CF Warrior Project Book* or various Cystic Fibrosis Foundation conferences. They also run support groups for LGBTQIA+ adults with CF in partnership with the Attain Health Foundation. Being openly queer and agender, as well as multiethnic and part of "the 10%" of patients without mutations qualifying for CFTR modulator drugs, gives them unique insight into questions of inclusion and justice in CF community. Born in December 1983 with very rare CFTR gene mutations, Xan had a long and complex road to a conclusive CF diagnosis despite being sick since birth and having their first sweat test in early childhood. They now receive care at the Orlando Health adult CF clinic in central Florida. Xan also works full-time as an Assistant Professor at the Florida State University College of Medicine regional campus in Orlando. In their work with FSU COM they spend a lot of time looking at connections between health and social life, and exploring how programs for people with different health conditions work. Their work focuses strongly on the experiences and needs of people aging with chronic diseases, which is basically studying what they know best! In addition to living with CF, Xan deals with PTSD from a long history of intimate partner abuse. Their experiences of chronic illness and complex trauma have helped them make an impact for others. Along with their spouse Dr. J Sumerau, Xan runs an outreach project called Write Where It Hurts for scholars who bring their lived experiences of hardship into their professional activities. Xan holds a Ph.D. and M.S. in Medical Sociology from Florida State University, an M.P.H. in Health Systems and Policy from Rutgers University, and a B.A. in Political Science from Columbia University. ▲

XAN NOWAKOWSKI

fibrosis (CF) patient with chronic and progressive pulmonary *M. abscessus* disease. The patient had an eight-year history of *M. abscessus* refractory to treatment with multiple drug combinations. The patient had progressive deterioration in lung function, functional status, and quality of life, and was denied lung transplantation consideration at multiple centers in the US and Canada due to chronic *M. abscessus* lung infection. In addition to treatment of the patient, the study examined the response of the patient's bacterial isolate to high-dose NO relative to other clinical *M. abscessus* isolates by performing in vitro susceptibility tests using an NO exposure chamber. In general, the patient noted improved respiratory symptoms and quality of life and had small improvements in her lung function, six-minute walk distance, and inflammatory markers but no significant change in tests and cultures for *M. abscessus*. A retreatment protocol was designed. However, dosing was stopped after day eight of the retreatment due to adverse symptoms. In vitro susceptibility tests showed a dose-dependent NO effect on *M. abscessus* susceptibility and significant heterogeneity in response among *M. abscessus* clinical isolates. The patient's isolate was found to be the least susceptible strain in vitro. The heterogeneity in *M. abscessus* susceptibility to NO suggests that longer treatment regimens could be required to see reduction or eradication of more resistant pulmonary strains. Nitric Oxide (NO) is a powerful molecule, naturally synthesized in the human body, proven to play a critical role in a broad array of biological functions. In the airways, NO targets the vascular smooth muscle cells that surround the small resistance arteries in the lungs. NO is also believed to play a key role in the innate immune system and in vitro studies suggest that NO possesses anti-microbial activity not only against common bacteria, including both gram-positive and gram-negative, but also against other diverse pathogens, including mycobacteria, viruses, fungi, yeast and parasites, and has the potential to eliminate multi-drug resistant strains.
https://tinyurl.com/y6nbz6hn

Urine Marker Could Help In Screening For CF Lung Infections, Study Finds

Measuring levels of a molecule called *lipoarabinomannan*, or LAM, in a person's urine could be an easier way of screening for airway infections due to nontuberculous mycobacteria (NTM) in people with *cystic fibrosis* (CF). Previous research indicates that about 1 in 5 CF

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Backpacking Through CF

By Devin Wakefield

> "Saying "no biggie" to hemoptysis on a backpacking trip tells me I've changed a lot since my earliest back-country adventures."

I dropped my Aerobika (similar to an Acapella or Flutter) off a cliff during my first treatment on my first backpacking trip without family. Later that same day, I also coughed up blood. I went on this trip with a fellow CF patient who had already received a lung transplant, her dad, and two mutual friends without CF whom I had known for several years. We all agreed this was an exciting start to the adventure. We planned to go 34 miles in three days and two nights from Castle Rock State Park (in California) through the Santa Cruz mountains to the Pacific Ocean—the trip was known as Skyline to the Sea.

This was back in 2017 when I was yet not eligible for any modulators. In civilization, I would nebulize at least three doses of Albuterol, two doses of Pulmozyme, and two or three doses of antibiotics daily. All would include rigorous airway clearance. I also suffered from bowel obstructions, necessitating both Amitiza and Miralax multiple times a day, along with plenty of hydration. All of this with the usual pills I popped during the day to maintain my CF. You know the drill. Outside civilization, surrounded by trees, hills, streams, and absolutely no electricity outlets, I had to simplify. I brought my ill-fated Aerobika; a portable, battery-powered nebulizer; extra Albuterol; Miralax in individually wrapped packets my GI doctor slipped me one day after complaining that my insurance wouldn't cover the drug; and my pills, sorted into small plastic baggies. I also brought the equipment everyone brings when they go backpacking: food, clothing, sleeping bag and pad, and on and on.

My post-transplant friend and her father are fondly known in my local CF community for going on long "death marches." She had been grieving the recent death of her twin sister, who also had CF. As you can imagine, twin sisters with cystic fibrosis had a bond forged in the great fires of bowel obstructions, end-stage CF, and lung transplant. In my confident enthusiasm, I suggested she take me on one of these famed journeys. I thought something fun might cheer her up.

DEVIN WAKEFIELD AT HIS CAMPSITE.

With all my equipment strapped snugly to my back and hips, I set out on the hike with my friends. Thankfully, my hemoptysis episode struck with camp less than a mile away. My friends carried my pack the last stretch, and I rested with no new symptoms that evening. We had hiked nine miles and had eight to go the next day. The following night, my lungs sufficiently recovered and, with no more Aerobika to loosen my mucus, my post-transplant friend's father dusted off his CPT skills to loosen my gunk. We braved a night in a dry riverbed and our water supply ran low. We lasted just until we found a wet creek from which to filter and refill. We finished the trip with a 17-mile march to the Pacific. At the ocean, we stuck our feet into the water to numb our aching soles. We celebrated at a diner, reminiscing about the gorgeous trees, diverse flora and fauna we saw, and rugged landscape. Stuffed and weary, we then left to pass out in a proper bed.

Saying "no biggie" to hemoptysis on a backpacking trip tells me I've changed a lot since my earliest back-country adventures. When I was growing up, my family loved camping and hiking. My mom took up a large part of the burden of organizing my meds. In a way, she shielded me from some part of my fraught reality—but not totally. In the 1990s and early 2000s, packing and taking medications on hikes was more complicated—I didn't have a portable battery-powered nebulizer that could last the whole trip, for one. I remember one absurd time when we had to hike to the ranger's cabin for electricity for my nebulizer. I think I was eight years old, and I

remember starting to feel self-conscious about my CF. Shielding took a toll on my mother. She tells me of a moment when, after boiling nebulizer cups in the camp stove, she set them out to dry. An unfortunate change of the wind blew ash from the campfire right onto the freshly sterilized cups. That moment hurt, and we went camping less often.

After that first backpacking trip, we traveled through new lands. In 2018, we hiked through the Lost Coast, near Eureka, California. We hiked along the coast, on the sand or unstable rocks at the coastline. We could only travel when the tides permitted, from 4 a.m. to 11 a.m. and then 4 p.m. to 11 p.m. This meant getting up before dawn, packing up with a headlamp and the stars twinkling above, then expectorating green gunk as the rising sun lit our path forward. One day we could not make it to our next campsite in time before the tide came in, so we snuck inland along a river to rest. In 2019, we explored the Hoh river valley, on the Olympic peninsula in Washington. We marched through an old-growth, temperate rain forest and marveled at the massive trees covered in moss. When the sun shone on the moss just right, it seemed the trees glowed, throwing an emerald sheen onto the trail. On the third day, we hiked up to the alpine beauty of the Olympics and our jaws dropped when we could finally see Blue Glacier. Thankfully, the clouds gave us a window to see the glacier's deep blue hue, just visible through the cracks in the white snow on top. Beyond the glacier, we could see the bottom of the north side of Mt. Olympus, but clouds hid its peak. Unfortunately, due to my error in planning, Blue Glacier was further than I thought, and we again hiked 17 miles that day. Worth it. It was all downhill after that.

On my latest backpacking trip I had the help of Trikafta. I traveled with a new friend through Spider Meadows and beyond Lyman Lakes, in the Glacier Peak wilderness of Washington. Trikafta simplified packing for me. I will still, obviously, have CF and need to bring and consume my medications, but I was freer to focus and plan against the usual things that kill people in the mountains. Bears, failing to bring enough food, or accidentally eating a hallucinogenic berry (which looks like a thimble berry) fueled my pre-trip disaster scenario brainstorming. Luckily, my friend on this trip knew which berries we could safely eat and I stuffed my bear cannister to keep me safe from my other worries. This adventure featured non-stop alpine meadows filled with wildflowers, jaw-dropping mountain views, an abandoned mining shack, old growth forest, and rain. On the second day, my friend's boot sported an ever-widening hole on the side. We fastened some clothes pins on it to keep it from ripping further and decided to cut short our journey by hiking 17 miles back to the car in one go on the third day. I keep swearing no more 17-mile hikes, gosh darn it!

On my backpacking journeys before Trikafta, health loomed larger in my mind. It heightened my anticipation for danger and kept me second-guessing myself while planning. A backcountry journey always needed backup plans, but I also made backups for my backups. I recall on my Lost Coast journey I had a mild sore throat—probably from coughing—but I prepared for the possibility of leaving early if it worsened. All in all, though, CF-induced worrying did not stop my appreciation for the backcountry. Once I got out into the field, the beauty of nature secured my attention.

I am still adjusting to my new confidence in my health, which Trikafta has given me. Fear still resides in my psyche and I fill it with other concerns, but I see it slowly healing and my anxieties loosening. Wrought from years of various medical traumas and an active imagination, this fear won't go away easily—I should note this global pandemic does not help. I have started to react differently to the aftermath of an increasingly rare hemoptysis episode now—an eyeroll and a quick note in my medical notebook—and to the few bowel slow-downs I face today I say, "water chugging contest time!" I got this.

> "I am still adjusting to my new confidence in my health, which Trikafta has given me."

Now I have come fully into adulthood and I call Seattle, WA, my home. I have rediscovered my childhood love of the outdoors and organize my own medications. Trikafta enables me to explore the backcountry further than pre-Trikafta days. Improved medical technology simplifies my packing as well. The gorgeous scenery of the West calls, and I'll be out there again soon. I won't be forgetting the remarkable resilience older CF patients and I could show as we climbed mountains with blood in our lungs. Standing on Trikafta, the mountains look shorter to me now, but they remain just as tantalizing to explore. ▲

Devin Wakefield is 29 years old and has CF. He lives in Seattle, WA. In his free time, he likes to run, hike, and backpack. His contact information is on page 2.

IN THE SPOTLIGHT

With Amy Shroyer Branham

By Andrea Eisenman and Jeanie Hanley

Amy considers herself an army wife and dog mom. And only after that, does she list her diseases—cystic fibrosis (CF), multiple sclerosis (MS), and thyroid cancer. This is not the best kind of trifecta to have, but she is not weighed down by any of this. Her focus is to stay well and enjoy life with her husband and puppy. See how she does it all with grace and joy. Please welcome our newest star. Spotlight please!

Age: 44

Location: Fairmont, West Virginia, born and raised.

What do you want our readers to know about?

I feel like I have unique issues having been diagnosed with CF when I was three months old and then MS when I was 37. I only know two other people who have both CF and MS. One is a man from Hawaii, whom I correspond with via Facebook, and the other person is a female about whom I know nothing else. While I am glad to have one other person to talk with about both diseases, I would really prefer to talk to another female as it's my understanding that women experience MS differently than men. This makes sense as my father is living with MS. Also, if you are female and have CF and MS, I would love to correspond with you. You can find me on Facebook—look for "Amy Shroyer Branham" and send me a friend request!

Even though my older brother doesn't have CF, I was fortunate that my aunt, who was unable to have a child of her own, adopted a baby with CF because the wait was much shorter than waiting for a child with no underlying conditions. My cousin and I grew up like siblings who were both treated for our disease. We went to clinic appointments together and each felt like we were not alone in battling CF. In 1994, when the cross-infection protocols were first instituted, we were no longer able to be so close. I still appreciated growing up feeling like I had someone who, having the same disease I had, understood me. It was comforting.

AMY SHROYER BRANAHM AND PUP, GIZMO.

How did you know you had MS?

In August 2013, I went to bed one night and woke up blind in one eye. I saw a doctor who said it was a complication from my CF. I knew that was nonsense and my CF doctor confirmed that. I consulted with my dad's neurologist, who eventually diagnosed me with MS. I have taken all sorts of medications for it and most stopped working eventually or had horrible side effects. Currently, I am getting Rituxan infusions every six months as a maintenance treatment and, so far, it is working. Currently, I walk with a cane, but can otherwise do everything just fine. I have Relapsing Remitting MS, which mainly causes muscle pain. I am right-handed but it mostly affects the left side of my body.

How did you meet your husband? Did you tell him about your CF right away?

I met my husband on a blind date my mom set up for us in 2006. She knew he was in the service and my family's long history of service would be a likely connection between the two of us. She was right—we married a year later in 2007. It was a third marriage for both of us and, as they say, the third time was indeed the charm! I told him about my CF right away. I explained what CF was and how it affected me. He had never heard of it. Luckily, he was OK with it and wanted me to educate him about it.

He had been in Iraq, in operation "Iraqi Freedom," so I was sure he had endured a lot in his life and could therefore handle my CF. He retired in 2016 from the military, specifically in the army as a Sergeant First Class. Now he works full time at the Army National Guard as a maintenance man for the armory. He likes to work.

Thyroid cancer?

I was told I had hypothyroidism (low thyroid function) most of my life, so I was used to seeing an endocrinologist. One day in 2012, I woke up with a baseball-sized tumor on my neck. I promptly went to see a surgeon to find out what it was. After three separate trips with subsequent waiting periods for needle biopsy results, I had confirmation of a thyroid cancer diagnosis. On Christmas Eve, I received news that the surgeon would have to remove the tumor. I scheduled the procedure for the first few days of the new year so I wouldn't ruin my Christmas. My surgeon removed my thyroid, tumor, and nearby lymph nodes on January 3, 2013. I was given radioactive iodine for

three years and have since been in remission.

How does CF affect you?

So far, I have endured 36 different surgeries in my life, including seven nasal polyp surgeries. Now, my sinuses are fine. Unlike many with CF, I have never had an issue with gaining or maintaining my weight. I do take enzymes, the highest dose of pancreas capsules possible. In the past, my lung function hovered around 100%, but has declined in the last three years. My lung function now is around 60%. It decreased further after a bout of pneumonia last year. My PFTs dipped to 40%. After starting Trikafta seven months ago, my PFTs are now back up to 87% as of my last clinic visit! I am so relieved and happy about that.

Are you religious or spiritual?

Religious. Being a Methodist, I read the Bible and "attend" church by watching it on television on Sundays. I remain grateful to God. I sometimes wonder why I have all of these diseases and I think, God just knew I could handle them. I believe in God and thank him every day when I wake up.

Have you ever worked?

My first job was babysitting at age 11. Then, later in life, I was a Certified Nursing Assistant (CNA), medical assistant, pharmacy tech, and a sleep tech for Bi-PAP and CPAP machines. I had on-the-job training through the company where I initially started working. The last job I had was as a CNA and I was always getting sick from being exposed to illness at work. I finally had to quit working as it was not good for my health. I was being written up for missing work due to the colds I got during my time working. It was for my health that I quit in 2008, at age 38.

What do you do for fun?

I like to browse through Facebook, take care of the house, and play with our new puppy Gizmo. I stay at home mostly, especially right now with COVID-19 and being higher risk. My husband does the shopping and I sometimes go to get my medications. I love listening to music and watching Netflix.

Favorite movies?

50 Shades of Grey, *Aquaman*, and basically anything with Jason Momoa.

Favorite TV series?

The Ranch, on Netflix.

Favorite music?

90s R&B, rap, dance, 50 Cent, Enigma, Eminem, and recently, Post Malone. A town near me had a Vodka promotion in their liquor store. The local radio had a contest to meet 50 Cent so I called in and I won! I could not believe it. I got to meet him briefly and pose for a photo with him. I brought my brother with me to help me navigate the crowd with my cane. I was able to cut in front of a huge line because I had won the contest. It was worth all the dirty looks I received—50 Cent is my favorite rapper.

What keeps you going?

My husband, family, and dog keep me going through all of my diseases and complications. Also, if something happens with my family, like they get sick, they immediately call me and I like to try to fix and help them. They call me the "fixer" and ask me for a first opinion before going to their doctor! I guess this is due to my CF and because I have worked in the medical field. ▲

Andrea Eisenman is 54 and has CF. She is a Director of USACFA and is both the Webmaster and Executive Editor of CF Roundtable. *Jeanie Hanley is 57 and is a physician who has CF. She is a Director of USACFA. Their contact information is on page 2. If you would like to be interviewed for "In The Spotlight," please contact either Andrea or Jeanie.*

TILLMAN continued from page 31

patients will test positive for NTM. The current gold standard for diagnosing NTM infection is a *bacterial culture test* of the airways. However, this method has notable drawbacks: it can be costly, take weeks to get results, its sensitivity is low, and it can be difficult to obtain samples from certain individuals. LAM is a molecule found in all mycobacteria species, and released by the bacteria when they are active. Additional analyses showed that LAM levels may also be linked with a particular type of NTM—higher urine LAM levels were significantly associated with positive culture for the bacterium M. *abscessus*. These results indicate that detecting LAM in urine can help to rule out individuals who probably do not need an NTM culture. That is, patients who test negative for urine LAM are likely to also test negative in a NTM sputum culture, so this test would not be necessary.

https://tinyurl.com/yys8mztu

New Technology Could Improve Detection Of Biomarkers For CF, Others

A new strategy for detecting very small concentrations of specific molecules could allow for better diagnosis and management of a variety of diseases, including *cystic fibrosis* (CF). Measuring the amount of molecules in the blood or other bodily tissue can provide important information related to health—for example, levels of *C-reactive protein* (CRP) in the blood are associated with inflammation. CRP measurements are important for the management of many conditions, including CF. Measuring the levels of certain

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CF: THE MIND GAME

Is It Time For You To HALT?

By Mark Tremblay

I was diagnosed with CF in 1970. After years of drinking and drugging to deal with the emotional fallout, I was admitted to an inpatient addiction treatment center shortly after my 18th birthday. Upon successful discharge, the situation I faced was daunting, to say the least. Not only was I in poor health from ignoring my CF for several years, but I was trapped in a small town with a suspended license, my father was dying of cancer, and all my closest friends were alcoholics, addicts, dealers, or other folks on the fringes of society. All those factors, plus my lackluster progress in treatment, earned me a big fat red check mark in the "high risk" box on my discharge report.

Such was the inauspicious beginning of my 32-year recovery journey. Not surprisingly, within a few days of discharge, after fighting the urge to drink for hours, I found myself peddling my beat-up, old BMX bike to the nearest AA meeting in a desperate attempt to stay sober. The AA meeting took place in the first engine bay at the local fire house and looked pretty much like some townies sipping coffee as they sat on mismatched chairs around a sagging, plastic table. After a shaky start (I ticked off a few of the old timers by having the gall to discuss my drug use at an Alcoholics Anonymous meeting), a couple of the group members warmed up to me a bit. One in particular, my buddy's mom, whom I remembered as a violent drunk, but who apparently turned her life around in AA, shot me a warm smile and said, "Honey, sounds like you better HALT before you slip." Curious about what she meant, I stuck around to hear more. Thousands of AA meetings and more than 30 years later, I credit her admonition and support with keeping me sober for those first few shaky days and many more tough weeks after that.

For those of you not familiar with recovery clichés, "Hungry, Angry, Lonely, Tired ('HALT')" is a warning used in 12-step programs to prevent relapse. It harkens back to the 50s but is still just as prescient today. In fact, I've used it countless times to assess my own status as well as that of others I've tried to help who were also struggling with addictions, depression, anxiety, and codependency. HALT is a simple reminder to assess the degree to which you are feeling stressed, angry, isolated, and/or alone. Additionally, it is a reminder to pay attention to your diet and sleep patterns. Admittedly, I view emotional wellness through the lens of my own recovery. Nevertheless, I have found, through years of helping others, that these factors play a key role in predicting, catalyzing, or compounding relapse, not to mention bouts of depression, as well as other emotional and mental exacerbations.

> *"HALT is a simple reminder to assess the degree to which you are feeling stressed, angry, isolated, and/or alone."*

MARK TREMBLAY

Most folks in recovery would heartily agree that stress and anger are the most serious of the aforementioned factors when predicting impending relapse. We're likely all experiencing some of these to one degree or another between being in the midst of a global pandemic, nationwide social unrest and violence, economic malaise, political turmoil, and historic unemployment. If you've spent any time on social media lately, you'd likely agree that everyone seems more stressed and angrier than ever. As CF patients, we have to deal not only with all that's going on in the world, but we are more profoundly impacted by healthcare access, drug prices, and coverage during the pandemic.

Personally, I have a long list of things that stoke my anger. To name a few: waking up in the hospital next to a roommate that had MRSA (which I've cultured ever since), or the time my boss told me during my evaluation that, "I was lucky to have a job with my health issues." However, every day I choose to live free of the dark cloud of anger and bitterness because the AA big book says, "...we are not skilled in separating justified from unjustified

anger...because our wrath is always justified. Anger, that occasional luxury of more balanced people often led us straight to the bottle." However, regardless of the reasons, studies show failing to effectively manage anger delays healing from injuries, lowers pulmonary capacity, dampens immune function, and decreases antibody levels.

The second most important factor in determining risk of an emotional/behavioral exacerbation is the degree to which you are feeling isolated and alone. Studies show that people are three times more likely to experience loneliness in the midst of this COVID-19 crisis than before it began. Several studies found social isolation contributes to elevated blood pressure, spikes in cortisol levels (stress hormones), delays in healing, disrupted sleep, and is just as harmful as smoking 15 cigarettes a day. Although many folks in our community try to ease these feelings by leaning heavily on social media and virtual relationships, studies show that time spent on social media is inversely related to happiness and people do not, in fact, feel as secure or satisfied with their virtual relationships compared to their in-person relationships.

Additional factors that warrant consideration when it comes to assessing emotional and mental wellness are sleep, diet, and exercise. Possibly you're oversleeping or experiencing greater sleep disruptions. Perhaps you're eating too much, eating too little, or experiencing greater fluctuations in your glucose levels (if you have CFRD). Maybe you're experiencing increased loneliness or anger. All of these scenarios may indicate the need to HALT and get help.

Sometimes, even when we sense we need help, certain personality traits, which aren't necessarily exclusive to having CF but often present in CF patients, can prevent us from seeking help: denial, perception of invincibility, and stigma. Denial is a mental device many patients use to cope with CF. One patient, in particular, stands out as a prime example of denial as she genuinely described her illness as "mild" even up to and after being listed for a transplant. While denial may be helpful in dealing with CF, it is never helpful when dealing with behavioral and mental health issues. Another CF-related personality trait often seen is invincibility, which outwardly presents as something akin to teenage indestructability but manifests in patients who know they are anything but invincible and are, indeed, often in dire need of more social, emotional, and relational support in their lives. The final prong of help-inhibiting personality traits often seen in CF patients is a strong aversion to talk to professionals (e.g., a counselor, social worker, or psychologist) about difficult emotions for fear it will bring up more problems. I had a friend who said, "It's bad enough I'm a CF patient, I'll be darned if I'm going to be a psych patient, too!" While visiting him in the psych hospital after his second suicide attempt, I thought he would have been better off admitting that he was both before his emotional problems threatened his life.

Even when a patient assesses their situation honestly and admits they need help, the healthcare system itself sometimes stands in the way. In 2008, when I finally admitted I needed mental healthcare, it took my clinic six months for them to refer me to a counselor who, it transpired, was no longer taking patients. Even after I found my own counselor, it took my clinic another six months before they found a nurse practitioner who could prescribe and manage my antidepressant medications. It's not uncommon for smaller CF centers to lack experienced and knowledgeable staff, sufficiently robust referral networks, and training to provide the necessary emotional, mental, and behavioral support CF patients often need. Many CF centers do not have licensed psychologists, psychiatrists, or Certified Alcoholism Substance Abuse Counselors ("CASAC") with CF population experience on staff or within their referral networks to properly assess, diagnose, as well as prescribe and manage psych meds for patients with mental/behavioral disorders. Another issue is that mental/behavioral health screening is not routinely provided in CF clinics and, even when intermittent screening attempts are made, clinicians don't always have the necessary training and experience to provide intervention techniques widely used with healthy adults, chronically ill, elderly, and disabled populations.

I hope by bringing awareness to the warning signs that you may need help, as well as to the personality traits that may make you more resistant to help, that you will be empowered to take the first step by reaching out to your CF clinic or local counseling center.

I am not a licensed clinician but I am leading a free, online open recovery group on Wednesdays at 7:00 p.m. EST—"CF Warriors for Freedom and Recovery," through Attain Health. The group provides an opportunity for CF patients to learn about and work through a 12-step recovery program to deal with their hurts, hang-ups, and addictions. ▲

Mark is 51 years old and lives in Albany, NY, with his wife MaryGrace and stepson. He holds a Master of Arts in Psychology from Marywood University and a Master of Public Administration from Syracuse University. Mark has worked for six years in the New York Governor's Division of Budget and currently works full time at the Department of Health. He is the President of CF Vests 4 Life Foundation. He and his wife love cycling, church ministry, and riding their motorcycles. You can follow them on their YouTube channel, "Breathing Grace."

Mental Health In The Age Of Innovative CF Therapies

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

Even amid a global pandemic, people with cystic fibrosis in the United States are living longer and in better health than we were a decade ago. Back in 2010, only a few people enrolled in clinical trials had access to early CFTR protein modulator drugs. When the first successful CFTR modulator, Kalydeco, hit the market in 2012, nobody knew quite what to expect. Because only about two percent of people with CF were genetically eligible to take Kalydeco, understanding the impact of this drug on the general CF community required some creativity. But one impact that immediately became apparent was that of hope, which extended far beyond the limited number of patients eligible to take Kalydeco in particular.

Modulators nurtured and transformed a spirit of hope that grew steadily in the U.S. CF community. Other contributing factors included new and improved airway clearance technologies, off-label use of drugs for other conditions, and promising research on infection mitigation treatments like phage therapy. In the process, new modulators also emerged—first Orkambi and Symdeko for people with two copies of the 508F deletion, and then the highly anticipated triple combination therapy, Trikafta, for people with any combination of 508F and residual function mutations.

As many as 90% of people with CF can now theoretically benefit from CFTR modulators tailored for their unique genetic profiles. Those of us who cannot—the so-called "10%er" group within the CF community—still benefit from a wide array of other advancements in treatment that have been developed within the last decade. So, we face a world in which the future looks not only brighter but also potentially much longer. And that means adjusting to changes.

I remember being in my 20s and struggling to breathe or having to cough out half a cup of neon green mucus in the middle of the night before being able to speak more than a couple of words at a time. Experimental treatment with inhaled corticosteroids rescued my lungs and my future alike, leaving me with more mental energy to manage other CF complications like kidney disease and cardiovascular problems, and, of course, the severe GI issues I got in exchange for relatively easier lung challenges. In the process, I was also trying to heal from the trauma of an abusive relationship and move forward in my relationship with my whole self—not just the body that had put me through so much.

XAN NOWAKOWSKI

In some ways CF seemed like the least of my worries, even though it was killing me, and I was clearly running out of time. I also did not know my CFTR mutations yet, just that I was very sick and getting sicker with what might be the same disease my childhood healthcare providers had tentatively identified when I was around five years old. Slowly suffocating from my own lung secretions certainly instilled a feeling of running out of time—one that would fuel me in finishing my Ph.D. just a few days past the two-year mark. But I was always running from something back then, whether the shadows that chased me in endless bad dreams or the thick deposits of infected mucus that clogged my airways.

In the middle of all this I met my wife, who knows plenty about dying young, even though she does not have CF herself. I consider her a cat of sorts, with seemingly endless extra lives. We did a lot of healing together. She helped me understand my bisexuality better; I helped her find greater peace with being transsex. And along the way, we realized they call it "post-traumatic stress disorder" for a reason. It felt easier to put our ghosts and fears into context when we were both so focused on surviving. As she strived to find some financial security after a lifetime of extreme hardship, she stayed awake to watch me breathe, shaking me gently when I would stop, clapping on my back to help bring up that inexhaustible supply of foul sputum.

Then my wife landed her first faculty position, a literal dream job that let her stay in Florida; I finished my

own Ph.D. and got prescribed beclomethasone inhalers as a final attempt to save my lungs. Things got easier physically after that but became exponentially harder mentally. I began to learn firsthand the meaning of the term "complex PTSD" and how it differs from post-traumatic stress induced by a single incident, such as surviving a terrible car accident or getting robbed at gunpoint. My nightmares intensified; any better sleep quality I got from inhaled steroids was offset by acting out my terrible dreams. I woke up exhausted and haunted.

I also had the good sense to go to counseling, something I had done a few times before with excellent results. I found a counselor who understood and resonated with my unique and complex ethnic and cultural background, and who supported people who were polyamorous and queer and nonbinary. I also talked a lot with my wife about what I was experiencing. Being somewhat further along in her own healing journey, she helped me stay focused on the light at the end of the tunnel that I often had difficulty seeing. Over time, I learned that complex PTSD requires thoughtful management with different types of cognitive reframing than those that tend to help with the symptoms of acute PTSD.

The fact that my own trauma from abuse intersected in a variety of ways with the challenges of living with CF and the medical trauma it created introduced challenges of its own. When I finally got the genetic testing in 2016 to identify my CFTR gene mutations and confirm my clinical CF diagnosis, it came as a relief. Even though my kidneys were showing signs of incipient failure and my digestive function had nearly disappeared, I felt grateful that something finally made sense.

Coordinated care at an accredited CF clinic—along with pancreatic enzymes, specially formulated vitamins, and better airway clearance methods—changed my health dramatically. But just as important was the enormous hug my CF doctor gave me at the end of my intake visit in early 2017. He held space then, and has continued to ever since, for a range of emotions: my anger, my disappointment, my fear, and all the trauma that created those things. I did not need to explain any of this to him, either the trauma itself or how various aspects of my personal biography intersected with it. This kind of awareness should be standard in the field of CF care, not an exception.

> "As we continue to push for new treatments for the various physical challenges of living with CF, we should also innovate in mental healthcare."

Who we are shapes how we process trauma, medical or otherwise. And when we spend our lives becoming slowly more accepting of how little time remains, it can be difficult to shift out of that mindset. For some, that means living fast and dying young. For others, it means isolating oneself and still dying young. For still others, it means being extremely cautious all the time and maybe dying a little less young. My old goal was not to die without my Ph.D. in hand. After my parents placed the hood over my shoulders at graduation, white noise crackled in my head. I had a wonderful relationship and now I had my doctorate...and lungs that course after course of antibiotics could not clear of infected mucus thick as rubber cement. Then everything changed, and the world suddenly seemed very large.

We are now learning how to live expansively—not only to dream of the future, but to plan for it and process all the changes that entails. What seems like a uniformly wonderful thing on the surface is complex and often fraught upon closer examination. For those of us with intersecting forms of trauma from queerphobia, transphobia, racism, misogyny, poverty, etc., it becomes much more difficult to ignore the need for healing and the painful work it involves. I might have taken my trauma to an early grave had those steroids not saved my lungs. And, in my early years of healing, I often wished that I had.

By contrast, when Trikafta finally came out I felt mostly excited for all the people who could take it rather than disappointed about my own continued exclusion from modulator eligibility. After all I had already endured, a life of being well managed with other types of drugs and devices seemed like a distinct upgrade. One of my first friends in the CF community, who was also one of the first people to take Kalydeco when it came out, said that his struggle had become mostly mental once his physical health became easier to manage. I still think about that a lot and see the wisdom in it.

Modulators and other innovative therapies are transforming the lives of many adults with CF in the U.S. These changes are long overdue and also riddled with challenges for mental health. Likewise, modulators and other new drugs can themselves have pronounced mental health effects. We continue to learn about these side effects as more patients experience therapeutic innovations for ourselves. And, in the process, our mental health needs continue to evolve. So, as we continue to push for

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Voices from the Roundtable

Mapping The Trail

By Jeremy Moore

After being diagnosed with CF at 15, I finally had a reason for why I was not developing like other boys my age. Now that I knew why, I had to decipher out how to catch up. I had to figure out how to improve my cardio capacity, build muscle, and make up for lost growth. Three years ago, I started writing a book documenting how I achieved success in my quest.

I knew I had to write this book to give others a direct trail map to follow, whereas I had been walking blindly for many years in my search for answers. I published *Trail Map to Muscle: How to Defeat Genetics, Disease, and Build a Confident Body* in July of 2020. In my book, I detail what to do to see real results for anyone looking to get into shape at any age. I know there are many people out there who walk into a gym, or want to work out at home, and have no idea where to begin. My book is not specifically meant for those with CF; rather, it is meant for anyone who may or may not have a genetic barrier or disease holding them back.

Mapping out the book outline was a real challenge—I had to revisit what did and did not work for me and what allowed me to make significant progress from 15 years prior. It took many long days of writing. I sought to cover everything from understanding what to eat and how it affected my workouts to how to get the most out of my time in the gym. I strove to not leave out any detail because I knew how much this book could help others, especially parents who had teenagers with CF who were looking for a way to improve their child's health.

> "In my book, I detail what to do to see real results for anyone looking to get into shape at any age."

JEREMY MOORE

My hope for the book is that it changes lives, plain and simple. It is the book I wish I had when I was starting my journey, and that it will serve as a clear trail map for others to follow for many years to come. ▲

Jeremy Moore is 33 years old and has CF. He lives in Chesapeake, VA. He is the author of Trail Map to Muscle: How to Defeat Genetics, Disease, and Build a Confident Body. *He enjoys fishing, playing drums, and lifting weights in his free time. He can be reached at trailmaptomuscle@gmail.com.*

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new treatments for the various physical challenges of living with CF, we should also innovate in mental healthcare.

Including social workers and other mental health professionals in CF clinic care helps a lot. But this approach must complement support systems outside the clinics themselves. Access to individual and family counseling can help to bridge those gaps. This requires both capacity in the professional workforce and the means to pay for care—something often lacking in the CF community, given the financial burdens many of us already face.

Perhaps most importantly, just having social spaces to talk openly about the mental health challenges of *not* dying young can do wonders for our ability to thrive. Peer support, modeled by programs like the Cystic Fibrosis Foundation's Peer Connect initiative or the Attain Health Foundation's weekly support groups, can make a vital impact for patients. Running a support group for Attain Health focused on LGBTQIA+ adults in the CF community has transformed my own life in wonderful ways, and given me an even deeper sense of purpose and meaning in my own journey with CF. ▲

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

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, **PO Box 1618, Gresham, OR 97030-0519. Or email to: cfroundtable@usacfa.org**

ANNIVERSARIES

Birthday

Scott Reece
Albuquerque, NM
50 on September 1, 2020

Transplant

Lara Govendo, 33
Milton, Vermont
Bilateral lung transplant
3 years on August 18, 2020

Scott Reece, 50
Albuquerque, NM
Bilateral lung transplant
19 years on June 9, 2020

Wedding

Scott Reece & Carrie Rust-Reece
Albuquerque, NM
20 years on April 1, 2020

NEW BEGINNINGS

Katherine Lockwood, 32
Sandwich, MA
Rose Milly Lockwood born
July 24, 2020

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molecules requires the use of specialized technology. However, there are inherent limitations as to how sensitively these technologies can detect their targets. Developing technologies that can more precisely perform these measurements is an ongoing effort in biomedical research – the "ideal" technology would, in theory, be able to detect single molecules in a sample. The researchers therefore developed a new technique based on two concepts: nanopores and DNA origami. *Nanopores* refer to very small holes. Specific adaptations can be designed so that the nanopore is a sensor for a range of specific molecules. DNA origami involves taking a strand of DNA and "folding" it to create a specific structure. These structures can be tailored such that they have a central cavity that will only bind to a specific molecular target. In the study, the researchers basically used DNA origami to create nanopores that could only be "filled" by a single, specific molecule. The captured biomarkers [target molecules] are then read with nanopores and can be done one molecule at a time. By coupling DNA origami and nanopores researchers are able to quantitatively detect disease biomarkers can be with single molecule sensitivity. One of the main advantages [of the technique] is the minimal sample needed. The process is very quick, and takes just minutes to provide results.

https://tinyurl.com/y265yvo3

AND

https://tinyurl.com/y3eomb6o

Bacterial 'Death Stars' Could Be Tricked Into Destroying Themselves

Researchers discovered a network of nutrient-transporting channels that are formed when bacteria grow in large communities, and this could be used to kill bacteria more quickly by tricking the bacteria into transporting drugs through the channels instead of food. The communities—called biofilms—are involved in up to 80% of persistent human infections and cannot be killed easily by antibiotics. It's like a bacterial Death Star, with the biofilms able to be blown up from the inside by targeting the channel systems with drugs. The channels take the nutrients from underneath to transport them through the biofilm, whereas traditionally antibiotic treatment would be from above the biofilm. The structure of large bacterial communities is complex and poorly understood but the discovery of these channel systems in E. coli biofilms is important because it tells us how bacteria in biofilms can move nutrients throughout their community. These biofilms are like domes, which antibiotics try to remove when applied to their surface, but the problem is that antibiotics don't penetrate into it. This then leads to persistence of the infection and then development of antimicrobial resistance, which is a major public health issue. Because a route into the biofilm from underneath has been discovered, potentially the drugs can get in under the dome to kill the *bacteria* quicker and more effectively.

https://tinyurl.com/y4fl4hhq

CF Foundation Gives Nearly $15M To Groups Researching Infections In CF

The *Cystic Fibrosis Foundation* (CFF) has awarded nearly $15 million to 33 organizations that are researching how

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to improve outcomes for *cystic fibrosis* (CF) patients who are battling infections. The awards fall under the CFF's *Infection Research Initiative*, a large-scale effort to provide $100 million in funding for research into CF and infections through 2023. The newly funded projects fall under three broad *categories*: detection and diagnosis of infection, new approaches to fight antibiotic resistance, and addressing pathogens that are difficult to treat. Three of the awarded research projects are centered around new diagnostic approaches that require less sputum and that can generate results more quickly. Two others aim to detect pathogens through either a blood or a urine sample. To address bacterial resistance to antibiotics, three of the awarded research projects are focused on developing an alternative approach to treat bacterial infection based on *phage therapy*. Phage therapy uses viruses, called *bacteriophages*, that specifically infect and destroy bacteria while not harming human cells. One of these studies is researching bacteriophages to target antibiotic-resistant *Pseudomonas* bacteria, another is engineering bacteriophages to target a wide range of *Burkholderia* bacteria. The third awarded group of phage therapy researchers is developing a library of bacteriophages to target each of the different strains of Burkholderia bacteria known. Other projects awarded by the CFF are focusing on specific challenges faced by CF patients, including infection with *nontuberculous mycobacteria*, fungi, and multi-organism infections.

https://tinyurl.com/yxufzgeb

CFF Funding To Further Contrafect's Research Into Alternative For Antibiotics

ContraFect will receive funding from the *Cystic Fibrosis Foundation* to conduct preclinical research about the potential of direct lytic agents, an alternative to conventional *antibiotics*. The first stage of the agreement will profile funding for the in vitro activity of ContraFect's next product candidate, CF-370, an engineered lysin targeting *Pseudomonas aeruginosa*, and amurin peptides, against bacterial specimens obtained from CF patients at different stages of disease. Direct lytic agents, (DLAs), are a next generation of antimicrobial therapeutics. DLAs include both *lysins* and *amurin peptides* and cause the rapid death of bacteria — much faster than conventional antibiotics. Both agents destroy the bacterial cell wall. Lysins are proteins derived from naturally occurring bacteriophages (viruses that infect bacteria). Lysins are lytic agents, as they can kill bacteria by breaking down a key component in the structure of the bacterial cell wall. According to ContraFect, once the cell wall is breached, the bacteria disintegrates rapidly. Amurin peptides, a small-class of peptides (short chains of amino acids, the building blocks of proteins), were shown to effectively kill several Gram-negative bacteria, including *Pseudomonas aeruginosa*, in lab studies. Moreover, these peptides strengthen the action of standard-of-care antibiotics, making them particularly suitable for CF patients infected with antibiotic-resistant bacteria.

https://tinyurl.com/yx9tshun
AND
https://tinyurl.com/y3aukc6n
AND
https://tinyurl.com/y26cjkrz

Orkambi Benefits CF Patients Across Levels Of Lung Health, Study Finds

Orkambi (*ivacaftor/lumacaftor*) can significantly reduce the number of days people with *cystic fibrosis* (CF) require intravenous *antibiotics* for flares and helps with weight gain—independently of a patient's level of lung function at the start of treatment. While the greatest lung improvements were seen in patients with suboptimal lung function at treatment start, these findings support *Orkambi*'s use by all eligible patients, regardless of their current lung status. The researchers' goal was to compare over one year of Orkambi's use, the magnitude of improvements in ppFEV1 and *body mass index* (a measure of body fat), and changes in exacerbation frequency in patients with higher and lower lung function scores. Changes noted were compared with those seen in study patients with intermediate range baseline ppFEV1. Patients with a low ppFEV1 had a nearly two times higher discontinuation rate compared with the others. The main reason given for stopping treatment by low ppFEV1 patients was adverse respiratory adverse events. Among continuous Orkambi users over the one-year study, a significant increase in ppFEV1 was seen both in patients with intermediate and low ppFEV1. Further lung function gains were not evident in those with high ppFEV1 scores at baseline. A sizable number of patients in the intermediate ppFEV1 group also experienced a 5% or greater improvement in lung function scores. Similar gains were seen in 22% of low ppFEV1 patients and in 27% of those with high ppFEV1. While Orkambi had the greatest lung functions benefits in the intermediate ppFEV1 group, the treatment significantly increased body mass index across all patients, regardless of their baseline lung function and age. Orkambi also significantly reduced exacerbations requiring intravenous (IV) antibiotics in intermediate ppFEV1 patients, and showed a similar trend in the high ppFEV1 group. The number of days each year with IV antibiotic treatment was significantly lower among all groups, compared with their number over the year prior to starting Orkambi. These results suggest that adolescents and adults across all ranges of ppFEV1 may benefit from Orkambi.

https://tinyurl.com/y3nmpaps

Real-World Impact Of Trikafta Will Be Evaluated In RECOVER Study

A new study will assess the real-world effects of *Vertex Pharmaceutical*'s *Trikafta*, a triple combo of *elexacaftor*, *tezacaftor*, and *ivacaftor*. The study, called RECOVER, will assess the impact

Peer Engagement Groups From Attain Health

ADULT GROUPS

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

YOUTH, TEEN, AND MINECRAFT GROUPS

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

INHALE MELANINE, EXHALE POWER

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

CF FIGHTERS FOR RECOVERY & FREEDOM

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

MOMS WITH CF

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

SOLID GROUND–BIBLICAL FAITH-BASED

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

Young Adult Group

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

LGBTQAI+

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

ATTAIN HEALTH

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

of *Trikafta* on the everyday lives of people with CF, rather than in controlled clinical trial conditions. This study will allow the researchers to discover in detail how this new treatment affects the health and everyday lives of people with cystic fibrosis, to understand why different people might respond differently to the drug and to gain insight into how this treatment might affect the very significant treatment burden that people with cystic fibrosis currently endure. Besides examining routine health parameters, researchers will evaluate parameters not included in the *AURORA clinical trials* that led to the approval of the therapy in the U.S., including imaging, functional, biological and quality of life measurements.
https://tinyurl.com/y5xktl7m

Vertex Seeks To Expand FDA Approval Of Its CFTR Modulators

The *U.S. Food and Drug Administration* (FDA) is reviewing applications from *Vertex Pharmaceuticals* to expand the approval for three of the company's *cystic fibrosis* (CF) therapies: *Trikafta* (elexacaftor/tezacaftor/*ivacaftor* and ivacaftor), *Symdeko* (tezacaftor/ivacaftor

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and ivacaftor), and *Kalydeco* (ivacaftor). The three novel *supplemental new drug applications* (sNDAs), if approved, would give access to these therapies to people with rare CF-causing mutations that are not covered by current FDA approvals. Vertex estimates that 600 people with such rare mutations live in the U.S. Approval of the applications would also allow some individuals access to more Vertex therapies. For example, patients currently eligible for *Kalydeco* treatment only could also become eligible for Trikafta and/or *Symdeko*. Vertex's new applications to the FDA are based on in vitro data (data acquired from experiments on cells), which showed that many rare CFTR mutations respond to one or more of these modulators.
https://tinyurl.com/y27tvtrb

Arrowhead Pharmaceuticals Initiates Dosing Phase 1/2 Study Of ARO-Enac For Treatment Of Cystic Fibrosis

Arrowhead Pharmaceuticals, Inc. announced that it has dosed the first subjects in AROENaC1001, a Phase 1/2 clinical study of ARO-ENaC, the company's investigational RNA interference (RNAi) therapeutic being developed as a treatment for patients with cystic fibrosis (CF). ARO-ENaC utilizes Arrowhead's proprietary Targeted RNAi Molecule (TRiM™) platform and is the company's first inhaled RNAi candidate to target pulmonary epithelium. The Phase 1/2 clinical study, AROENaC1001, is designed to assess safety, tolerability, and pharmacokinetics and potentially provide an early assessment of efficacy in patients with CF. ENaC (epithelial sodium channel) is a target with great potential for many CF patients that may not be eligible for existing therapies due to their specific genotypes, commonly called class I patients, and for those that have an inadequate response to therapy. ARO-ENaC is designed to reduce activity of the epithelial sodium channel alpha subunit in the airways of the lung. In patients with CF, dysfunction in the cystic fibrosis transmembrane conductance regulator (CFTR) causes increased ENaC activity which contributes to airway dehydration and reduced mucociliary transport. This predisposes patients to persistent lung infections, structural damage, and progressive loss of pulmonary function. ENaC has been extensively explored as a potential therapeutic target for CF, but the development of inhaled small molecule ENaC inhibitors has been limited by on-target renal toxicity and short duration of action in the lung.
https://tinyurl.com/y5dah4l2
AND
https://tinyurl.com/y2uj4lza

Dosing Begins In Phase 1 Trial Of ETD002, Potential CF Inhalation Therapy

Enterprise Therapeutics announced that dosing has begun in a Phase 1 clinical trial testing *ETD002*, a potential inhaled treatment for *cystic fibrosis* (CF). The study (*NCT04488705*) will assess the safety and tolerability of EDT002 in ascending single and multiple doses in healthy people as compared with a matching placebo. ETD002 is designed to boost—or potentiate — the activity of *TMEM16A*, a chloride channel found on the surfaces of cells in airway tissues, where it helps to regulate the amount of salt and fluids. ETD002 enhanced the activity of TMEM16A in preclinical studies, thereby increasing fluid flow into the airways, as well as promoting mucus thinning and clearance. TMEM16A potentiation works regardless of a patient's CFTR mutational status, making this approach applicable to all with CF, and potentially to patients with other lung diseases. ETD002 is expected to work both as a single therapy and in combination with other therapies, including those that repair the mutated CFTR.
https://tinyurl.com/y5e4psuu
AND
https://tinyurl.com/y3afxer3

4d Molecular Therapeutics Announces New Agreement With Cystic Fibrosis Foundation

4D Molecular Therapeutics (4DMT) announced a new agreement with Cystic Fibrosis Foundation (CF Foundation) to develop precision gene medicines for cystic fibrosis (CF). Under this agreement, the CF Foundation will support the completion of IND-enabling research and development activities, and the planned Phase 1/2 clinical study of 4D-710, 4DMT's wholly-owned product candidate for the aerosol treatment of CF lung disease. 4DMT's gene therapy approach holds promise for the treatment of CF by using a proprietary and optimized AAV vector to deliver a functional copy of the CFTR gene to the airway and lungs to restore function and alleviate disease symptoms. 4D-710 is comprised of a transgene insert encoding for the CFTR gene and 4DMT's proprietary vector 4D-A101, a vector that is designed for an efficient, single dose aerosol delivery to the lung airways and with resistance to pre-existing antibodies.
https://tinyurl.com/y4nbrnvh

Azurrx Biopharma Initiates Phase 2b Clinical Trial Of MS1819 In Cystic Fibrosis—With First Patient Screened And Three Clinical Trial Sites Activated

AzurRx BioPharma, Inc.announced that it has initiated its Phase 2b OPTION 2 clinical trial to investigate MS1819 in cystic fibrosis (CF) patients with exocrine pancreatic insufficiency (EPI) with the activation of three clinical sites and initial screening of the first patient. The Phase 2b multi-center study is designed to investigate the safety, tolerability, and efficacy of MS1819 in a head-to-head comparison against the current porcine enzyme replacement therapy (PERT) standard of care. The primary efficacy endpoint is the coefficient of fat absorption (CFA).

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Previous clinical trials have demonstrated that MS1819 is safe and well tolerated. With this trial, the optimal dose for MS1819 will be determined in preparation for a Phase 3 study. MS1819 will be administered in enteric capsules to provide gastric protection and allow optimal delivery of the enzyme to the duodenum. MS1819 is a recombinant lipase enzyme for the treatment of exocrine pancreatic insufficiency. MS1819, supplied as an oral non-systemic biologic capsule, is derived from the Yarrowia lipolytica yeast lipase and breaks up fat molecules in the digestive tract of EPI patients so that they can be absorbed as nutrients. Unlike the standard of care, the MS1819 synthetic lipase does not contain any animal products.

https://tinyurl.com/yxj8lzkz

AND

https://tinyurl.com/y2xr4l99

ELX-02, Potential Therapy For CF Nonsense Mutations, Named Orphan Drug

ELX-02, an investigational treatment of *cystic fibrosis* due to nonsense mutations, has been designated an *orphan drug* by the *U.S. Food and Drug Administration* (FDA). One *type of CF-causing mutation* is known as a *nonsense mutation*. These are mutations that cause a "stop" signal to be coded in the gene, somewhat analogous to having a period erroneously placed in the middle of a sentence. A premature stop codon results in the production of a shortened protein that is unable to work properly. ELX-02 is designed to restore the production of full-length and functional CFTR protein. The therapy *works* by binding to *ribosomes*—the cells's protein-making machinery—allowing them to "read through" the aberrant stop signal to generate fully working protein. *Phase 1 clinical trials* conducted in healthy volunteers showed that ELX-02 was generally well tolerated, and preclinical studies have supported its efficacy.

Eloxx is currently sponsoring two Phase 2 clinical trials—called EL-004 (*NCT04126473*) and EL-012 (*NCT04135495*)—to test ELX-02 in people with CF who have at least one *G542X mutation in CFTR*, the most common CF-causing nonsense mutation.

https://tinyurl.com/y3flxu9q

Aridis Reports AR-501 Clinical Data: Positive Safety Data In Healthy Subjects Of A Phase 1/2a Clinical Trial

Aridis Pharmaceuticals, Inc. announced positive results from the Phase 1 portion of its Phase 1/2a clinical trial of AR-501, an inhaled formulation of gallium citrate being developed for the treatment of chronic lung infections in patients with cystic fibrosis (CF). It is a non-antibiotic, broad acting antimicrobial with a mechanism of action involving interference with iron and disruption of microbial iron-dependent metabolic pathways distinct from current antibiotics. AR-501 acts as an iron analog and is believed to disrupt multiple iron dependent pathways in microbes, leading to growth inhibition. AR-501 has antimicrobial activities against a number of gram-negative and gram-positive bacteria, including antibiotic resistant strains. The Phase 1/2a clinical trial, which is being funded by the Cystic Fibrosis Foundation, is a randomized, double blinded, placebo-controlled study evaluating the safety and pharmacokinetics in healthy volunteers and *Pseudomonas aeruginosa* infected CF patients. AR-501 is being developed as a once-per-week dosing regimen that is self-administered using a hand-held nebulizer device. Preclinical efficacy and safety data have demonstrated that AR-501 works synergistically with multiple antibiotics, is effective against antibiotic resistant strains, and has a low intrinsic resistance profile. Separately, an intravenous formulation of gallium nitrate citrate has been evaluated in Phase 1 and Phase 2 clinical studies as a single, 5-day infusion in moderate and severe cystic fibrosis patients. Both clinical studies of IV gallium demonstrated safety and efficacy as measured by improvement in lung function.

https://tinyurl.com/y4h99mud

Renovion Raises $8.1M To Advance ARINA-1 Nebulizer Therapy

Renovion has secured $8.1 million in funding for the clinical development of *ARINA-1* (ascorbic acid), an experimental *nebulizer* therapy to clear mucus and reduce inflammation in people with non-cystic fibrosis *bronchiectasis* and *cystic fibrosis* (CF). People with a lung transplant or chronic lung disease such as CF or bronchiectasis experience high levels of mucus production and inflammation. Among possible causes are defects to the hair-like projections in the lungs, called cilia, that clear out mucus, and chronic respiratory infections that impair immune system responses. An inhaled formulation of *ascorbic acid*, or vitamin C, ARINA-1 is designed for twice daily use. According to Renovion, the therapy is intended to restore mucus clearance and reduce inflammation caused by immune system alterations and infections in the airway. ARINA-1 has been granted *orphan drug designation* by the *U.S. Food and Drug Administration* for the treatment of people with lung transplants and CF. The treatment is administered via the investigational *eFlow nebulizer* which enables short inhalation times. Initial studies investigating ARINA-1 in patients have indicated that the treatment has a strong safety profile. In people with CF, ARINA-1 showed greater efficiency in clearing mucus than *hypertonic saline*.

https://tinyurl.com/y5k6aoep

Corbus Pharmaceuticals Reports Last Subject Visit In Phase 2b Study Of Lenabasum For Treatment Of Cystic Fibrosis

Corbus Pharmaceuticals Holdings, Inc. announced that the last subject completed their final visit in the Company's Phase 2b JBT101-CF-002 trial of lenabasum for the treatment of cystic fibrosis. The Phase 2b trial is a multinational,

An Update On The Speakers Bureau

We relaunched the Speakers Bureau last fall. So far, in 2020, we have delivered five talks. We are proud of our ability to roll with the punches as we've had to do since COVID-19 has forced us to suspend in-person speeches for the calendar year. One of our directors gave a speech to Translate Bio, Tré LaRosa spoke to Integrated Genetics, Ella Balasa spoke to Corbus Pharmaceuticals and Translate Bio, and Isabel Stenzel Byrnes spoke to Walgreens.

Our Speakers Bureau is able to speak on a far-reaching range of topics, from daily living with cystic fibrosis, working with CF, going to college with CF, legal measures and CF, mental health, grief, struggling with clinics and/or adherence to treatments, and just about anything else you can think of. We are proud of our ability to sponsor our speeches for companies that are looking to host and better understand the patient experience. We are also thrilled with our ability to educate clinics, research organizations, or just about anybody else. It is our belief that we when we tell our stories, we improve the world for others with CF, and, hopefully, we encourage a bit more compassion in the world.

Please let us know if you would like to request a speaker—or, if you have CF, if you'd be interested in joining our Bureau!

https://www.cfroundtable.com/speakers-bureau

426-subject study evaluating the efficacy and safety of lenabasum in cystic fibrosis. The primary efficacy endpoint is the event rate of pulmonary exacerbation (PEx). Secondary efficacy outcomes include other measures of PEx, change in forced expiratory volume in 1 second (FEV1), % predicted, and change in Cystic Fibrosis Questionnaire-Revised respiratory domain score. Lenabasum is a rationally designed, oral, small molecule that selectively binds as an agonist to the cannabinoid receptor type 2 (CB2), resolves inflammation, and limits fibrosis. CB2 is preferentially expressed on activated immune cells and on fibroblasts, muscle cells, and endothelial cells. In both animal and human studies conducted to date, lenabasum has induced the production of pro-resolving lipid mediators that activate endogenous pathways which resolve inflammation and speed bacterial clearance without immunosuppression. Data suggest that lenabasum can reduce expression of genes and proteins involved in inflammation and fibrosis. Lenabasum is also active in animal models of lung infection and inflammation in cystic fibrosis. Lenabasum has demonstrated acceptable safety and tolerability profiles in clinical studies to date. Lenabasum treatment also was associated with a lower rate of and longer time to pulmonary exacerbations in a Phase 2 cystic fibrosis study.

https://tinyurl.com/y23j7dyr

New Technology Offers Potential For Faster, More Accurate CF Diagnosis

A new technology, called X-ray velocimetry (XV), could enable faster and more accurate *cystic fibrosis* (CF) diagnoses by non-invasively tracking the movement of air through even the fine structures of the lungs at high-resolution. This new technology measures how different layers of tissue diffract or absorb X-rays as they pass through the body. Capturing these images at high speed over the course of a patient's breathing cycle provides information about both lung structure and function. These measurements enable precise assessments of the expansion of even very small lung tissues, which makes it possible to pinpoint the origin of any functional change. According to the researchers, a useful byproduct of XV's heightened sensitivity is that it provides a quantitative way of classifying CF cases. Based on the presence and pattern of airflow obstructions, the researchers could assign different cases as "healthy," "heterogenous," or "clustered." Healthy lungs expanded in a uniform manner, consistent with both lungs being clear and unobstructed. Heterogenous disease was defined when CF-like obstructions occurred relatively evenly throughout the lungs. Cases were classified as clustered disease when lungs became partially obstructed with mucus, showing poor ventilation and trapped air in the obstructed regions. Understanding the differences between different types of CF can have important implications for treatment and outcomes. The combination of X-ray velocimetry and progressive automation of the data analysis is an important step in the development of more sophisticated methods of lung function testing, and should assist research internationally to improve the health and lives of people with cystic fibrosis, and a range of other lung diseases. The U.S. Food and Drug Administration *recently approved this technology* for all respiratory indications in adults.

https://tinyurl.com/y5xw6h57

AND

https://tinyurl.com/y3w4othr

Academy Of Nutrition And Dietetics Releases Practice Guideline: Nutritional Needs Of People With Cystic Fibrosis

The Academy of Nutrition and

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Dietetics has released a comprehensive guideline to help registered dietitian nutritionists address the needs of people with cystic fibrosis who are at risk of developing nutrition problems commonly associated with pulmonary disorders. This guideline is the first of its kind, addressing new medications and increased risk of obesity among some people with cystic fibrosis. The Academy conducted an evidence-based review of current scientific literature and created a guideline with 28 nutrition recommendations for RDNs. They include medical nutrition therapy, nutrition screening and assessments and individualized meal planning to improve health and stem the progression of the disease.
https://tinyurl.com/y5yemdxh

Age At Lung Transplant Impacts Long-Term Survival Of Patients With Cystic Fibrosis

Patients with cystic fibrosis aged 30 years and older at the time of lung transplant had superior long-term survival compared with younger patients. *Patients with cystic fibrosis* aged at least 30 years had significantly higher survival compared with those aged 18 to 29 years at the time of transplant. Median survival was 9.47 years in the older cohort vs. 5.21 years in the younger cohort, according to the data. Death due to allograft failure was significantly lower among patients aged at least 30 years vs. those aged 18 to 29 years. Researchers noted a higher incidence of malignancy in patients aged at least 30 years vs. those aged 18 to 29 years.
https://tinyurl.com/yykdssc2

Newly Funded Research Seeks To Improve CF Lung Transplant Outcomes

A series of new research projects will seek to better understand how the immune system responds to transplanted organs. The goal is to find ways to increase the viability of organs after transplant. Understanding how immune cells respond to transplanted organs sets the stage for developing novel therapeutic strategies to improve outcomes for transplant patients. There is a fairly high risk of organ failure, with only about half of transplanted lungs still functioning five years after the surgery. This is markedly lower than the five-year organ survival rates for heart, liver and kidney transplants, all of which are about 70%. Uncovering the basis for the poor survival of lung transplant recipients should also provide new insight into the causes of other inflammatory diseases that affect the lung. The goal behind studying tolerance in the context of transplants is to find ways to ensure that the transplanted organ is tolerated by the immune system. If the immune system attacks the transplanted organ, it can lead to *rejection*.
https://tinyurl.com/y53rrz8g

CF Patients With Poorer Lung Function At Greater Risk Of Premature Or Smaller Baby, Study Shows

Pregnancy is generally safe for both women with *cystic fibrosis* (CF) and their child, but those with poorer lung function are at greater risk of having a premature or smaller baby. Improvements in CF care standards have allowed more women with the disease to become pregnant. However, their chronic disease may pose a serious risk to their own health and to that of their baby; which is why determining the risk of pregnancy-associated complications in women with CF is key to helping them make informed decisions regarding pregnancy and family planning. Previous studies suggest that pre-pregnancy lung function is the stronger predictor of pregnancy outcomes for both mother and baby. Severely impaired lung function —less than 50% of predicted *FEV1* is *associated* with an increased risk of premature delivery (prior to 37 weeks of gestation), cesarean section, and low birth weight. While FEV1 of less than

50% *has been advised* as a contraindication to pregnancy in these women, successful pregnancies in those with 20–30% of predicted FEV1 have been reported. The data suggest that pregnancy is possible and that fetal outcomes are good in most women receiving prenatal care, even in those with a starting FEV1 [less than] 60%.
https://tinyurl.com/y22pazql

Baseline Cystic Fibrosis Disease Severity Has An Adverse Impact On Pregnancy And Infant Outcomes, But Does Not Impact Disease Progression

In this multicenter-retrospective cohort study, researchers sought to examine the correlation of baseline disease severity, pancreatic insufficiency (PI), and *Pseudomonas aeruginosa* (PA) infection with fertility, the pregnancy course, delivery, neonatal outcome, and subsequent disease progression. Baseline disease severity, PA infection, and PI have an adverse effect on infant outcomes but do not substantially affect the progression of the disease during and after pregnancy. Therefore, pregnancies may have a good prognosis in severe CF patients.
https://tinyurl.com/y27qapfj

The Outcome Of Pregnancy In Women With Cystic Fibrosis: A UK Population Based Descriptive Study

This population-based study was undertaken to estimate the incidence of cystic fibrosis in pregnancy, as well as to determine obstetric and neonatal outcomes. Participants were all pregnant women with cystic fibrosis who booked for prenatal care in a UK obstetric unit between March 2015 and February 2017. In total, 71 pregnancies were reported during a two-year span. No maternal deaths occurred. Findings revealed generally good pregnancy outcomes of women with cystic fibrosis. Even in those women with FEV1 < 60% predicted, successful pregnancy was achievable, however, a higher likelihood of preterm delivery and a smaller baby was noted in such women.
https://tinyurl.com/yx98jcow

CF ARTICLES

Outcomes of pregnancy in women with cystic fibrosis (CF) taking CFTR modulators—an international survey. Edward F Nash, Peter G Middleton, Jennifer L Taylor-Cousar, J Cyst Fibros, 2020 Jul;19(4):521-526

As their long-term prognosis improves, women with CF are increasingly choosing to have children, but the safety of CFTR modulators in pregnancy and breastfeeding is currently unknown.

Methods: A survey was sent to lead clinicians of adult CF centres in Europe, the United Kingdom (UK), United States of America (USA), Australia and Israel requesting anonymised data on pregnancy outcomes in women using CFTR modulators before and during pregnancy and lactation. 64 pregnancies in 61 women taking IVA (n = 31), LUM/IVA (n = 26) or TEZ/IVA (n = 7), resulting in 60 live births were identified. In 44 pregnancies, CFTR modulators were either continued throughout pregnancy or temporarily stopped and then restarted. Two maternal complications were deemed related to CFTR modulator therapy; cessation of modulator therapy resulted in clinical decline in 9 women prompting resumption of therapy during pregnancy. No modulator-related complications were reported in infants exposed in utero and/or during breastfeeding.
https://tinyurl.com/yy6svscu

Rapid lung function decline in adults with early-stage cystic fibrosis lung disease. Elliott C Dasenbrook, Aliza K Fink, Michael S Schechter, Don B Sanders, Stefanie J Millar, David J Pasta, Nicole Mayer-Hamblett. J Cyst Fibros, 2020 Jul;19(4):527-533

The prevalence of adults living with cystic fibrosis (CF) who have early-stage lung disease is increasing. The authors studied the prevalence and evaluated spirometric risk factors associated with the subgroup of patients with early-stage lung disease and FEV1 decline of ≥5% predicted/year. They found that even among adults with early-stage lung disease, approximately 15% are shown to progress and experience a large decline in lung function. This reinforces the concept that lung function in early-stage CF is not normal or mild. Rather, lung function decline may be delayed, but not avoided, in these individuals. Variability in FEV1% predicted and airway obstruction as measured by FEV1/FVC ratio may identify individuals at increased risk of decline. Adults with early-stage lung disease should be followed in clinic to monitor for onset of decline.
https://tinyurl.com/y2c53kak

Screening practices for nontuberculous mycobacteria at US cystic fibrosis centers. Derek Low, Dulaney A Wilson, Patrick A Flume. J Cyst Fibros, 2020 Jul;19(4):569-574

Current guidelines recommend at least once yearly screening for nontuberculous mycobacteria (NTM) in Cystic Fibrosis (CF), however screening practices remain widely variable. This study evaluates current practices among United States CF centers with specific focus on clinical predictive factors for NTM screening. The authors found that NTM screening practices vary widely among United States CF centers with many centers testing only on clinical changes. With higher rates of testing shown as successful in identifying more patients with NTM, routine screening should be emphasized in CF care going forward.
https://tinyurl.com/y24nhcfv

Identification of Mycobacterium porcinum in patients with cystic Fibrosis: Pathogen or contaminant? Grace R

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Paul, Amy Leber, Christopher J Nemastil, Kimberly J Novak, Michael Brady, Stephanie Stack-Simone, Alexander L Greninger, Stella Antonara. J Cyst Fibros, 2020 Jul;19(4):580-586

Mycobacterium porcinum is a non-tuberculous mycobacterium (NTM) identified in potable water. The identification and clinical impact of *M. porcinum* in patients with cystic fibrosis (CF) has not been described. In one institution, *M. porcinum* was isolated exclusively during hospitalization in a cluster of patients with CF. Patients with CF who were hospitalized between September 2016 and September 2018 and could expectorate sputum were included, and samples were processed per institutional guidelines. Post-hospitalization and one-year clinical outcomes on those who isolated *M. porcinum* in respiratory cultures were reviewed. Whole genome sequencing was performed on *M. porcinum* isolates obtained from patients and environmental sources to identify source of acquisition. Review of 14 CF patients with 16 *M. porcinum* isolates revealed rapid time to culture positivity after admission. *M. porcinum* was isolated in teenagers and adults irrespective of baseline pulmonary function, body mass index, or CF genotype. Whole genome sequencing suggested all isolates belong to the same *M. porcinum* strain and confirmed the source of acquisition to the ice machine. Review of patients' clinical course, including three patients who underwent lung transplantation, suggested a pseudo-outbreak with minimal clinical impact. NTM, including *M. porcinum*, are ubiquitous in potable water and institutional water reservoirs. Our findings suggest *M. porcinum* is a transient colonizer rather than a pathogen. Challenges exist in discerning the role of NTM as a contributor of pulmonary morbidity in patients with CF, and adherence to established guidelines regarding NTM related pulmonary disease remains important.
https://tinyurl.com/y588l66z

Clinical effect of lumacaftor/ivacaftor in F508del homozygous CF patients with FEV 1 ≥ 90% predicted at baseline. B L Aalbers, K M de Winter-de Groot, H G M Arets, R W Hofland, A C de Kiviet, M M M van Oirschot-van de Ven, M A Kruijswijk, S Schotman, S Michel, C K van der Ent, H G M Heijerman. J Cyst Fibros, 2020 Jul;19(4):654-658

The first available CFTR modulator combination for homozygous F508del patients, lumacaftor/ivacaftor, has not been tested in patients with percentage predicted (pp)FEV1 > 90 in the phase III trials. The objective of this study is to share real life experience about treatment results in this group. Researchers discovered that homozygous F508del patients starting lumacaftor/ivacaftor at ppFEV1 ≥ 90 improved significantly in nutritional status, sweat chloride levels and exacerbation rate, but did not respond in ppFEV1. Treatment is well tolerated in this patient group. These effects make it worth considering to treat this group of patients with lumacaftor/ivacaftor.
https://tinyurl.com/y63vpftr

Intraoperative extracorporeal membrane oxygenation for lung transplantation in cystic fibrosis patients: Predictors and impact on outcome. Vittorio Scaravilli, Letizia Corinna Morlacchi, Alessandra Merrino, Edoardo Piacentino, Davide Marasco, Alberto Zanella, Mario Nosotti, Lorenzo Rosso, Federico Polli, Francesco Blasi, Antonio Pesenti, Giacomo Grasselli. J Cyst Fibros, 2020 Jul;19(4):659-665

Predictors and outcomes of intraoperative extracorporeal membrane oxygenation (ECMO) during lung transplantation (LUTX) for cystic fibrosis (CF) are unknown. Thus, researchers collected data and found that while undeniably useful as a life-saving procedure, ECMO during LUTX for CF is associated with worsened short-term outcomes. ECMO should be implemented weighing its risk and benefits.
https://tinyurl.com/y48khn5r

Streptococcus pseudopneumoniae, an opportunistic pathogen in patients with cystic fibrosis. Chloé Dupont, Anne-Laure Michon, Marion Normandin, Guillaume Salom, Marie Latypov, Raphaël Chiron, Hélène Marchandin. J Cyst Fibros, 2020 Jul;19(4):e28-e31

S. pseudopneumoniae isolation has not been described before in CF patients. Identification of *S. pseudopneumoniae* remains challenging due to the high similarity level between species of the Streptococcus mitis group. *S. pseudopneumoniae* was associated with pulmonary exacerbation in 46% of the patients, either as the sole pathogen or as part of a polymicrobial infectious process. *S. pseudopneumoniae* has to be considered as an additional opportunistic pathogen in CF and additional studies are needed to increase knowledge of its epidemiology and clinical significance in CF.
https://tinyurl.com/y6m8jth3

Attitudes of pain and opioids prescription practices in US cystic fibrosis centers. Yaoli Y, Trina H, Zubin M, Laxova A, Decker C,Braun Andrew. Journal of Cystic Fibrosis, August 12, 2020

This research was carried out to compare responses between adult and pediatric cystic fibrosis (CF) centers in the US. Researchers conducted a questionnaire examining care team members' views on the prevalence and characteristics of pain, pain management, and opioid use. Via a CF Foundation (CFF) email listserve, the questionnaire was distributed to accredited programs throughout the US. This study's findings demonstrate that chronic pain is common in adult patients with CF and management presents a formidable challenge to providers. The data recognized that the development of guidelines and/or collaboration with pain specialists will seemingly benefit both individuals and providers.
https://tinyurl.com/y4xxl7zm

Aerosolized lancovutide in adolescents (≥12 years) and adults with cystic fibrosis – A randomized trial. Eber E, Trawinska-

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Bartnicka M, Sands D, Schoergenhofer C,Jilma B, Ratjen F. Journal of Cystic Fibrosis, September 2, 2020

Given that lancovutide, a polycyclic peptide derived from *Streptomyces cinnamoneous*, activates a chloride channel (TMEM-16A) other than the cystic fibrosis (CF) transmembrane conductance regulator protein and could benefit CF patients, researchers conducted this randomized, controlled trial to test the effectiveness and safety of three different lancovutide treatment regimens vs placebo in a larger trial over a longer treatment period (8 weeks). They found that compared with placebo, lancovutide did not improve forced expiratory volume in 1 second percent predicted. Overall, lancovutide's safety and tolerability were acceptable.
https://tinyurl.com/y5989gh8 ▲

Laura is 72 and has CF. She is a former director and President of USACFA. She and her husband, Lew, live in Northville, MI.

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IMPORTANT RESOURCES

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

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

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

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

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