

The Art Of Reinvention

By Jeanie Hanley, M.D.

All too often, when I hear the term “growing older,” usually it is accompanied by a litany of complaints, whether physical (wrinkles!), emotional (so tired!), or regrets (I wish I had...). Every once in a while, and more often in the CF community, I hear how amazing and beautiful it is that we’re aging, a sentiment to which I wholeheartedly subscribe. Growing older with CF, every decade conquered, is a victory. I overcame the poor odds of survival that accompanied the initial diagnosis and turned 60 (yes, 60!!) last June. Yet I feel that I’m a twenty-something at heart and hope to have many great years ahead of me.

To me, growing older means being more in tune with yourself—knowing what you can and can’t handle. This is why I continually adapt and adjust what I’m doing in order to pursue new interests and passions. It’s not always successful but hey, life is about learning, right?

In 2006, I had to leave my won-



JEANIE HANLEY

derful practice as an allergist, tending to others’ health, and go on disability due to rapidly declining lung function. During the next 16 years, I had to tend full time to someone else’s health—my own. Concurrently, I reinvented myself and my life and tried to figure out what I could accomplish while trying to heal and maintain my health. It didn’t come easily. Looking

back, I view my life so far as an artful jigsaw puzzle, each piece an experience while not knowing the full details of the final picture. Some experiences lock in place better than others. I’ve learned to fine tune my experiences to create my own masterpiece. I continue to experiment with new pieces, throwing out those that don’t fit and keeping the best ones.

Spending so much time on my health every day limited what I could do. However, I did find I was able to try out new interests and accomplish a lot through volunteering. Every year, I changed up my volunteering to test the waters in different roles. For example, I started a nonprofit patient advocacy organization to use my medical degree to help others receive the best medical care possible. I volunteered and joined the board of nonprofits like USACFA and CF Research Institute (CFRI). I became my neighborhood’s Block Captain and dove into genealogy, loving to research and investigate ancestral roots, not just for

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EDITOR'S NOTES

Summer is in full swing, and with that, a slower pace. This issue we're focusing on CF and pain. Down here in Texas, summer means extreme heat. Most years, summer is challenging for my sinus pain—the heat and humidity cause more inflammation, especially in my sinuses, resulting in an increase in headaches. Fortunately, I have fared better this summer—doing things outside in the heat doesn't end in debilitating pain. I suspect it's the addition of turmeric gummies in my daily pills. Whatever the reason, I'll take it.

Andrea Eisenman shares her story of abdominal pain so severe she ended up in the ER, multiple times. She writes about the long road to discovering the source of her pain and how she treats it now. Meanwhile, **Nicole Kowal** elaborates on the difference between the physical pain CF causes and the ever-present mental pain that comes with the territory. **Dr. Julie Desch** writes about chronic pain and the freedom that comes with experiencing but not owning pain. We're also featuring artwork exploring pain by **Roy Berkeley-Haddox**.

In this issue, you can also read about the latest CF research in **Laura Tillman's** expertly collated "From the Internet" column. In our "In The Spotlight" column this issue, **Dominic Quagliozzi** talks about his transplant journey, his art, being a first-time father, and moving across the country during a global pandemic. He also speaks about Trikafta and various community resources that have helped him cope with CF. **Dr. Nowakowski** discusses how we perceive pain and what appropriate pain management looks like, especially as the CF community ages. As a follow-up to the Spring 2022 issue, **Beth Sufian**, in our "Ask the Attorney" column, covers options for SSI and SSDI beneficiaries in part III of her III-part series of articles regarding working while receiving social security disability benefits. **Isabel Stenzel Byrnes** writes about the importance of wellbeing and incorporating more fun in our lives as a means for spiritual pain relief. **Mark Tremblay**, in his column, writes about his various sources of CF-related pain and the ensuing anger that arises because it prevents him from fully living the life of his choosing. In our "Family Matters" column this issue, **Cindy Baldwin** lays out three tips for parenting while working through pain.

Jerry Cahill, in the first of a series of interviews, talks about his lung, kidney, and liver transplants.

In our "Voices from the Roundtable" section, **Jennifer Kyle** details her adventures and mishaps in online dating again in her 50s.

On the cover, **Dr. Jeanie Hanley** writes about growing older and the artful jigsaw puzzle of her life. Growing older also means both trying new things and letting go of others. With that, we bid farewell to Jeanie as she starts anew her part-time medical practice. Jeanie has served on the USACFA board for many years as both a director and in various officer roles. Her wisdom and guidance will be missed by all of us at USACFA.

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

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Information From The Internet...

Compiled by Laura Tillman

Lupin Gets Tentative Approval For Cystic Fibrosis Drug

The drug company announced that it has received tentative approval from the US Food & Drug Administration (USFDA) for its Abbreviated New Drug Application (ANDA) Ivacaftor tablets, 150 mg. Ivacaftor tablet is a generic equivalent of Kalydeco tablets manufactured by Vertex Pharmaceuticals. These tablets will be manufactured at Lupin's Nagpur facility in India.

<https://tinyurl.com/vhmbfjup>



LAURA TILLMAN

New Research Could Prevent Hearing Loss For 50% Of People With Cystic Fibrosis

People with cystic fibrosis are prone to recurring lung infections which need to be treated with aminoglycoside antibiotics. Aminoglycoside antibiotics are

very effective against life threatening infections and are associated with low rates of antibiotic resistance; however, they can also cause hearing loss. Researchers estimate that it may be 50% of adults with the condition. Aminoglycosides enter and kill the sensory hair cells in the inner ear that are vital for hearing. Researchers are developing new aminoglycosides that aren't able to get into hair cells, but still retain the ability to kill bacteria, thus making them less toxic to hearing. By the end of three years the researchers hope to have at least three new aminoglycosides that can be moved towards clinical testing.

<https://tinyurl.com/yu37krkn>

AND

<https://tinyurl.com/yuaaex85>

AND

<https://tinyurl.com/yuaaex85>

Pilot RCT Of A Telehealth Intervention To Reduce Symptoms Of Depression And Anxiety In Adults

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LOOKING AHEAD

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

Summer (August) 2022: Dealing With Pain. (Current issue)

Autumn (November) 2022: Transitioning from Pediatric to Adult Care. What do you wish your providers had known when you were transitioning from pediatric to adult care? How did your family's involvement in your care change as you matured into adulthood, if at all? Which of your care needs were fulfilled during your transition to adult care? Which ones were left unmet? How did other developments in your life during the transition to adulthood impact your CF management? If you could change one thing about your transitional care experience, what would it be and why?

Winter (February) 2023: Accessibility and CF Care. What does accessibility look like for CF care in clinic? What steps can our health care providers take to make sure every patient can fully participate in their own care? What made that experience work so well? What does accessibility mean for you at home? Have you and your family modified things in your physical space, sensory environment, social interactions, etc. to help you or others live better?

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



ASK THE ATTORNEY

Working While Receiving Social Security Disability Benefits—Options For SSI And SSDI Beneficiaries:

Part III of III: Extended Period Of Eligibility And Extended Period Of Medicare Coverage After A Trial Work Period For SSDI Recipients.

By *Beth Sufian, J.D.*

I. Introduction.

In the previous issues of *CF Roundtable*, the first and second parts of this three-part series appeared. The first part addressed §1619(b) of the Social Security Act, which applies only to individuals receiving Supplemental Security Income benefits (SSI). The second part addressed Trial Work Periods (TWP) for Social Security Disability Insurance (SSDI) beneficiaries.

This third part addresses extended periods of eligibility and extended periods of Medicare coverage for beneficiaries after a TWP. The rules that apply to SSDI and Medicare coverage *do not* apply to SSI and Medicaid benefits. The rules regarding continuation of SSI benefits are different from the rules governing continuation of SSDI benefits. If you are a SSI beneficiary, see Part I of this series on continued Medicaid eligibility under §1619(b) in the Winter 2022 *CF Roundtable* issue. Part II of this series relates to SSDI and the TWP and appeared in the Spring 2022 *CF Roundtable* issue.

Three separate provisions in the Social Security Act allow a SSDI beneficiary to maintain Medicare benefits while he or she remains disabled and has work income. These three provisions are: (1) Trial Work Period; (2) Extended Period of Eligibility; and (3) Continued Extended Medicare Coverage. This article addresses the Extended Period of Eligibility and Continued Extended Medicare Coverage, which only apply to SSDI and Medicare recipients and do not apply to SSI beneficiaries. Some people

with CF think they are receiving SSI benefits but are actually receiving SSDI benefits, and some believe they are receiving SSDI benefits but are actually receiving SSI benefits.

Please make sure you know the benefit you are receiving as it makes a big difference in terms of non-medical eligibility criteria and the ability to work and keep benefits or keep Medicare or Medicaid benefits. However, medical eligibility criteria is the same for SSI and SSDI.

II. Extended Period Of Eligibility After A Trial Work Period.

A. Social Security Administration (SSA) Determination.

At the end of the nine-month TWP, some SSDI beneficiaries decide

their TWP is a failed work attempt. They discontinue work activity and want to continue receiving the monthly SSDI cash benefit and Medicare coverage. However, the beneficiary does not determine whether the TWP is successful or unsuccessful. SSA makes the determination. A person may continue to be eligible for SSDI only if SSA determines the person continues to be disabled under its rules.

B. Suspension Of Payments Versus Termination of Benefits.

At the end of a TWP, SSA may stop benefits. However, not all cessations of benefit payments are the same. A suspension of payments is different from a termination of benefits, even though both are a cessation of benefit payments.

Termination of benefits often occurs after a TWP when SSA concludes that the beneficiary has made a successful return to work and is no longer disabled. When SSA determines that a beneficiary is no longer disabled, SSA will terminate benefits because the beneficiary is no longer eligible to receive further benefits based on that claim. If a person wants to resume benefits after a termination, that person must submit a new claim by filing a new application for benefits and show that he or she meets all medical and non-medical criteria. However, the person may not be able to show insured status if the claimant's work credits have expired while receiving benefits.

Suspension of benefits is different. If SSA still regards the beneficiary as disabled under its rules (despite the beneficiary having earnings from work activity), SSA may suspend benefit pay-



BETH SUFIAN

ments. When benefits are suspended, the beneficiary remains eligible to resume benefit payments based on the claim or application for benefits. The beneficiary is still required to show that he or she remains disabled and meets the medical eligibility criteria, but their work credits and insured status will not have expired or lapsed. If benefits are suspended, SSA will extend the beneficiary's eligibility based on the prior work record. This is called an Extended Period of Eligibility.

C. Benefits During An Extended Period Of Eligibility.

The Extended Period of Eligibility may be up to 36 months, but the beneficiary may not continue to receive cash benefits for 36 months. Extended Period of Eligibility means that the eligibility to receive SSDI benefits on the prior work record continues for up to 36 months if the individual continues to be disabled under SSA rules. During the Extended Period of Eligibility, a beneficiary does not receive monthly cash benefit payments, but the beneficiary continues to be eligible to resume benefits based on their prior work record if the beneficiary stops working.

However, a beneficiary who wants to resume benefits based on their prior work record must still show evidence that he or she continues to be disabled under SSA Rules. The medical criteria for determining disability status are the same during an extended period of eligibility as during an initial application. The return to active benefit status is not automatic. SSA must still review the evidence to determine if the individual is disabled under the medical criteria set out in the law. SSA often describes this review as an "expedited review." "Expedited review" describes SSA's internal procedure on review, but there is little or no difference in the speed or ease of the procedure experienced by the claimant.

III. Extended Period Of Medicare

Coverage (EPMC).

A. EPMC Is An Additional Work Incentive.

SSDI beneficiaries often believe that Medicare entitlement stops when cash payments stop. This is true in most cases, but not all cases. It is possible under certain circumstances for a beneficiary to continue Medicare coverage, even when monthly cash benefits have ceased. In 2000, Congress extended Medicare coverage for people who receive SSDI benefits and who work while remaining disabled.

The EPMC provision makes it possible for beneficiaries who continue to be disabled to continue Medicare coverage after the TWP ends and after cash benefits stop due to substantial gainful activity. A SSDI beneficiary may keep Medicare coverage when the beneficiary is working as long as the disabling condition continues to meet SSA rules.

The EPMC is a work incentive for SSDI beneficiaries. EPMC is intended to benefit individuals who remain disabled and who have lost their monthly cash benefit due to work activity. EPMC is not intended to maintain Medicare coverage when SSA terminates benefits due to medical improvement that makes the beneficiary no longer disabled under the medical criteria for benefits or for other reasons resulting in a termination of eligibility. Individuals receiving the EPMC must still meet SSA disability requirements, even though these individuals may be working and are not due a cash payment.

B. How Long Is The EPMC?

The current rules allow EPMC to continue for at least eight and a half years (93 months) after a TWP ends. During the EPMC, the beneficiary will not receive a monthly cash benefit, but will continue to be covered by Medicare.

The EPMC period typically will not begin until a person's TWP is completed. This is because monthly cash

benefits are protected during a TWP, and Medicare enrollment continues while cash benefits continue. Determining exactly when EPMC begins and ends will depend on several factors. Making an exact calculation is often difficult for an individual because the information required to make an exact calculation is based on Social Security's records and determinations.

C. Obtaining EPMC.

EPMC is not automatic. EPMC is an extension of a person's current Medicare enrollment. EPMC cannot be obtained if SSDI benefits have already been terminated. Thus, it is important to confirm EPMC before work activity causes SSA to terminate benefits. The EPMC and the Extended Period of Eligibility run concurrently, not consecutively. It is best to confirm the EPMC with SSA near the end of any TWP if the beneficiary intends to continue work activity with income over the SSA allowable amount of earnings.

D. Continuation Of Medicare Coverage.

Individuals seeking continuation of Medicare have typically been eligible for SSDI benefits for more than 29 months. This is because a beneficiary does not begin to receive Medicare until 29 months after the date SSA determines the beneficiary is disabled.

The 29-month delay in Medicare enrollment is due to Social Security's waiting period before Medicare starts. Under SSA rules, a beneficiary must wait five full months after the date of disability determined by SSA before the beneficiary may begin to receive the SSDI monthly cash benefit. During the five-month initial waiting period, no cash benefits are paid and no Medicare coverage is provided to the SSDI beneficiary. After the initial five-month waiting period, the monthly cash benefits may begin. However, Medicare enrollment does not begin until an

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me, but for others, too. I enrolled in research studies to advance CF science that may benefit all those with CF. Everything is on the table and I'm open to considering all kinds of new professions and new avenues.

The beauty of volunteering is that it can be done from home, very often during nebulized breathing treatments. It allows the flexibility to take care of yourself and doesn't interfere with healthcare appointments. If it ever feels overwhelming, then cutting back is in order. The biggest benefit of volunteering is meeting and getting to know like-minded individuals throughout the U.S.—people who have enriched my life immeasurably.

I feel very fortunate that my health has improved on a grand scale in the past few years with the advent of better CFTR modulators. This led to my kicking around the idea of returning to practice as an allergist in direct patient care. Basically, rein-

venting a new permutation of what felt like a previous life in academia. I wrote down what this new permutation would entail. I knew I couldn't return full time, as in the past. It would have to be part time, a few days at most per week and I'd need my mornings free. Like that would ever happen! My mornings are the most important part of my day—necessary for treatments, exercise, and, if I can, even a power nap. As God is my witness, a position materialized, as if sending the universe an outline of the best job situation for me led to a meta-physical all-points-bulletin to local allergists. An allergy practice responded to the universe's request with my near-exact specifications. What a wonder! One thing led to another and, lo and behold, I've signed on the dotted line and will start soon. I couldn't be more excited to add this new beautiful jigsaw piece. It's taken oodles of preparation to catch up and get back up to snuff. It's been worth it.

While I'll have to put much of my volunteering aside for now, I hope to feel settled in my new position before eventually adding it back since it's such an emotional salve and extremely rewarding. I believe growing older with CF makes us value each and every unique experience, particularly the challenging times such as being on disability, a time when I learned so much about myself. I'm ready to tackle new challenges, regardless of physical age, on my journey to complete my jigsaw masterpiece. ▲

Jeanie is 60 years old and has CF. She is a doctor, lives in Los Angeles with her husband and has three grown children, a son-in-law, soon-to-be daughter-in-law, and a granddaughter, all who light up her life. Not too surprising, she did her share of jigsaw puzzles during the COVID-19 pandemic. She is also a director of USACFA and CFRI. Her contact information is on page 2.



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additional 24 months have passed. The initial five-month waiting period, plus the 24-month Medicare waiting period, combine for a total of 29 months after the date of disability.

A beneficiary cannot continue Medicare until after Medicare begins. Therefore, a person cannot seek a continuation of Medicare benefits until they are enrolled in Medicare, which occurs 29 months after the date of disability.

IV. Conclusion.

The rules discussed in this article are complicated. Nothing in this article is meant to be legal advice about your specific situation. If you have questions please contact the CF Legal Information Hotline at CFLegal@sufianpassamano.com or by calling 1-800-622-0385. All calls to the CF Legal Information Hotline (CFLIH) are confidential and free of charge to the caller. The CFLIH is spon-

sored by a grant from the CF Foundation but the CFLIH employees are not employed by the CF Foundation. ▲

Beth Sufian is 56 years old and has CF. She is an attorney who focuses her law practice on disability law and is the Vice President of USACFA. Her contact information is on page 2. You may contact her with your legal questions about CF-related issues at CFLegal@sufianpassamano.com.

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With Cystic Fibrosis

Adults with cystic fibrosis (awCF) have higher levels of depression and anxiety than community samples. The Coping and Learning to Manage Stress with CF (CALM) intervention was developed for awCF reporting elevated symptoms of depression or anxiety. In this pilot study, awCF were randomly assigned to either six telehealth sessions (CALM; n = 15) or treatment-as-usual (TAU; n = Primary outcomes were depression and anxiety. Secondary outcomes were coping self-efficacy and health-related quality of life (HrQOL). Tertiary outcomes were feasibility, acceptability, and satisfaction. Assessments were completed at baseline, post-intervention, and 3-month follow-up. At post-intervention, the CALM group had a lower mean score than the TAU group for depression and anxiety. The CALM group had higher (i.e., better) mean scores than the TAU group for coping and HrQOL domains of Social Functioning and Vitality. Most treatment gains were not sustained at 3-month follow-up. The researchers concluded that CALM shows promise as an intervention to reduce symptoms of depression and anxiety and improve coping and HrQOL.

<https://tinyurl.com/2a8cbbdp>

This review aimed to address the impact of CF on the occupational functioning of patients. A significant proportion of patients were reported to retain a job on a full- or part-time schedule. Less physically demanding occupations were most frequently performed, perhaps due to CF-related inability to sustain a heavy workload. Disease severity parameters (e.g., lung function measurements, or personal, psycho-social, or economic conditions) have been reported as determinant or co-determinant factors for the development of work-related disability. Although further research is necessary, these results may be useful to inform interdisciplinary CF healthcare management, including the assessment of work function, and to define career counseling plans and workplace risk assessment and management strategies to support the personal, social and professional lives of patients.

<https://tinyurl.com/2ad5okpt>

Flares, Poorer Lung Health Can Follow Becoming A Parent With CF

People with mild-to-moderate cystic fibrosis commonly experience a decline in lung function and an uptick in exacerbations shortly after becoming parents, a study reports. The use of CFTR modulator therapy can lessen the negative health impacts of newfound parenthood, findings also suggest. With available care, people with CF are living

longer than was once possible, and more and more patients are choosing to become parents. The researchers looked at changes in lung health by assessing percent predicted forced expiratory volume in one second (ppFEV1). They also assessed the frequency of exacerbations, or times when symptoms suddenly worsen, by looking at times when patients were treated with intravenous (IV) antibiotics. Changes in patients' body mass index (BMI) were also evaluated. Results indicated that lung function decreased significantly following the birth of a child. On average, ppFEV1 lowered by 3.19% from the year before to the year after a child's birth. BMI also decreased while the frequency of exacerbations significantly increased by about 30%. The researchers reported that among people with CF with mild to moderate disease, parenthood adversely impacted health outcomes in the year following the birth of a child compared to the year before birth. Analyses based on sex showed a significant decline in BMI among women but not men. Rates of ppFEV1 decline or exacerbations did not significantly differ based on sex. Older age was not associated with any of the outcomes analyzed. Declines in patients' health might be attributed to the distractions of parenthood, from "sleep deprivation" to less time for self-care. Statistical analyses showed that, after adjusting for age and sex, patients

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The Impact Of Cystic Fibrosis On The Working Life Of Patients: A Systematic Review



SPIRIT MEDICINE

Fun And Wellbeing As Spiritual Pain Relief

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

My spiritual life is upheld by a fundamental belief: to embrace sorrow and pain together with joy and wellbeing. This is the true meaning of yin and yang—that life has light and darkness swirling together in every experience, every moment, and every lifetime. In this issue focusing on pain, I'd like to ponder how the pains in life can coexist with fun and wellbeing.

The day after cancer surgery on my eye, I discovered Audible and randomly chose a book that popped up called *The Power of Fun*. Catherine Price, its author, who also wrote *How to Break Up with Your Phone*, argues that adults do not have enough fun.

We are so caught up in achievement, productivity, and capitalistic consumer culture that we devalue fun. Cell phones and laptops have also pulled us away from sources of creative fun, entertainment, and engagement. Price defines fun as anything that includes these three things: playfulness, connection, and flow. Flow is defined as a state of being fully immersed in an activity, with energized focus and full involvement, where time seems to stand still. It is actively engaging in fun rather than being passive, like watching TV. It is different from a “relaxing” activity (though sometimes those feel good), such as walking in nature, getting a massage, or lying on the beach. Fun is good for our physical, spiritual, and emotional wellbeing. Fun is not selfish, indulgent, or immature. We all need to enjoy life.

This book inspired me to pursue more fun in my life, intentionally and

deliberately. Because of this book, I've started a FunSquad with the CFRI Retreat monthly Zoom calls. As people with CF, we have plenty of pain and struggles, but incorporating fun is such a vital compensatory act. When working nearly full time, I was deeply deprived of fun. My “free” time was spent on healthcare and exercise. My father always used to say, “work hard and play hard.” I worked hard at a job

and healthcare and then went to sleep. That was pretty much it.

So what can we do to have more fun? I'm planning more. I plan to go to pickleball practice every week or two. I've planned to mosaic my garden wall with friends. I've planned a few trips with close friends. I make sure to schedule things with my nieces. Even if I feel physical pain, I still do these things, because they are healthy distractions

and inevitably I feel better afterwards. I have the kind of body where the more I move, the better I feel. Some people have a hard time feeling relief from pain much of the time. But fun can also be stationary: a board game,

comedy writing, karaoke, or even painting.

Distraction is not the same thing as denial or avoidance. It is a deliberate compartmentalization of our experiences. We create space for positive emotions and sensations and set aside the painful ones.

I do believe that fun is part of a healthy spiritual life. Some religions teach asceticism, saying that abstinence from sensual pleasures can lead to spiritual transformation. This has given fun a bad rap when it comes to spirituality.

I believe God wants us to be happy and celebrate life and laughter. After all, many of the times we've had the most fun have been done with love for the people around us. Love is the foundation of all spirituality, after all. Fun lightens us from the heaviness of our struggles. We need not overindulge, get intoxicated, or succumb to physical

“We are so caught up in achievement, productivity, and capitalistic consumer culture that we devalue fun.”



ISABEL STENZEL BYRNES

temptations to have fun. But we do need to wake up our life-force energy that makes us glad to be here and to feel grateful for life, no matter its challenges. Fun helps us balance the yin and yang, the dark and light, the negative and positive. When we endure pain, we have a choice to set it aside (when we can) and redirect our focus on the good, the joyful, and the fun side of life. And when our pain and fatigue are too much to have fun, we can assume the attitude, as my dear cousin did: “I’m learning to appreciate living through others’ experiences.” We can still have fun vicariously.

Pain is an inevitable part of living with CF, and sometimes it seems to get worse as we get older. If pain is going to be present, how can we live a full life, anyway? Besides having fun, I’d like to explore the idea of wellbeing. Positive psychologist Martin Seligman’s book, *Flourish*, summarizes his research to see what makes some people thrive in the midst of adversity. He studied happiness and life satisfaction but felt these themes did not fully capture what it is that helps us endure through struggles. He tried to see what makes life worth living. He decided that wellbeing is a state that helps us judge life positively and feel good. He uses the acronym PERMA to define wellbeing: positive emotions, engagement, relationships, meaning, and accomplishment. We need to feel some positive emotions to be well. We need to have some engagement in life activities—hobbies, work, volunteerism—to be well. We need to have positive relationships with others to be well. We need meaning—purpose, a why, a reason—to be here to be well. And finally, we need a sense of accomplishment to feel well—some achievement vocationally, academically, socially, or artistically that helps us feel good about ourselves.

In this CF life, I believe wellbeing is literally life-giving. Wellbeing gives us our drive to keep going. Our PFTs can

be dismally low, our futures bleak, and our bodies full of chronic pain, but we can still reach PERMA if the right variables are set in place. I’ve witnessed that being part of the CF community can definitely create PERMA. Being aware of our mortality can also push us to seek and find PERMA sooner in life compared to the pace for our healthy peers. Seligman emphasizes that wellbeing can be learned and calls this “learned optimism.” He has developed specific trainings to help teach PERMA and learned optimism.

Seligman’s work about psychological wellbeing has a direct connection to spiritual wellbeing as well. Chronic pain can make us feel abandoned by God. In my professional world of hospice, we define spiritual (or existential) pain as a time when a person is “unable to find sources of meaning, hope, love, peace, comfort, strength, and connection in life or when conflict occurs between their beliefs and what is happening in their life.” (Anandarajah and Hight, 2001). PERMA can help us find some relief from this kind of spiritual suffering. Feeling the effort of life is worthwhile is often augmented when we have connection, love, and meaning.

Fun and wellbeing help us embrace the gift of the miracle of life, no matter how painful it is. Rather than succumbing to pain, we have agency to decide how we carry it. Fun and PERMA remind us that we can still be well despite the challenges of our bodies. If you’d like to join CFRI Retreat’s FunSquad and share your fun, email me at the address below. You can google the Penn Resiliency Program and “PERMA Workshops” to learn more ways to boost your wellbeing. I wish you pain relief and wellness. ▲

Isa Stenzel Byrnes is 50 years old and has CF. She lives in Redwood City, California. She is 18 years post-lung transplant. You can contact her at isabear27@hotmail.com.



What is the Boomer Esiason Foundation?

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

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

What does BEF do?

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

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

www.esiason.org



PEARLS OF WISDOM

How Much Is Too Much? Understanding And Responding To Our Pain Signals

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

It is the middle of May and my face remains under construction. Right now, I have an even bigger gap in my bite thanks to getting mounts built for the permanent bridge that will get installed about two weeks from now. That work also included some surprise gum surgery—words that do not seem like they should go together in a sentence. But with CF, anything is possible. So the continuing saga with getting my jawbone repaired and filling the gap in my bite has me thinking anew about pain and how we perceive it as people living and aging with CF.

I still feel confused and frustrated about why it took so long to get adequate treatment for an infection and cyst that had eaten through the bone in my tooth socket when I was getting regular dental care. The short answer here is that neither x-rays nor CT scans—the only imaging that was available to both my dentist and my surgeon—showed the full extent of the problems with the root of that tooth. So even though I had let my dentist know periodically that the socket below that crown felt strange, which seemed especially concerning since the nerve in that root had long since been removed via root canal, she was not able to do much until the prosthetic in that spot broke apart.

This feels frustrating to me because I suspect I would have experienced much worse pain—and thus sounded a serious alarm about needing the root removed—

years earlier if I were not so accustomed to dealing with various kinds of pain each day as a result of CF-related infections. Of course, those same infections are likely what caused such severe dental issues in the first place. Understanding the origins and implications of the pain I feel in different parts of my body challenges me constantly. Figuring out how to respond to these experiences feels even more difficult.

It probably comes as no surprise that I have done some work on pain

management in my own career, dating back to my days in the Master of Public Health program at Rutgers, where I focused my capstone project on what hospitals are doing to offer patients alternatives to opioids for chronic pain management. About two years before starting my M.P.H., I had undergone an operation to remove scar tissue that had accumulated in my bladder from many years of recurrent infections and my body attacking the tissue with histamine as part of the immune response.

Over time, thick masses of scar tissue started pressing on the nerves in the wall of my bladder, causing constant sensations of someone squeezing my pelvis in a vise or stabbing it with a knife.

I first experienced that pain when I was 11 years old. I had just turned 22

when I underwent surgery to remove the scar tissue. Being in constant pain feels scary no matter your age. It was also incredibly disorienting when so much else about my body was changing. Because, at that point, nobody knew for sure that my health issues were caused by CF rather than some sort of primary autoimmune disease, there was little I could do to stop the vicious cycle of infection and scarring.

Even several years after identifying the genetic mutations that cause my CF, my ability to stop what remains of my natural dentition from being destroyed by virulent bacteria remains limited. I have already had most of my teeth completely replaced above the gumline; I have also undergone multiple operations to restore the gums themselves. Now my prosthetic teeth are breaking

“My ability to perceive something harmful as painful is already so compromised by nearly four decades of acclimating to chronic pain.”



XAN NOWAKOWSKI

apart as infections destroy the roots that support them. I expect that if I live long enough, I will wind up wearing full dentures. I would feel happy with this outcome because it would mean I survived long enough to lose my remaining roots. What I do not look forward to are the years of renewed pain that would likely precede that transition.

What frustrates me most is that even after 38 years of living with this disease and six years of knowing for sure that I have it, I still do not always know when the pain I feel is something usual versus something requiring urgent action. My ability to perceive something harmful as painful is already so compromised by nearly four decades of acclimating to chronic pain that I have to exercise caution when cooking so I do not accidentally put my hand on a hot pan. I burned the skin off one of my knuckles several years ago because I did not realize I was touching something hot until I smelled singed flesh.

I have voluntarily had deep fillings done on some of my teeth without any anesthesia because I know I will not feel anything from the drill except vibration. When I had my wisdom teeth extracted, I asked for only local anesthesia and then drove myself home afterwards. I did the same for the surgeries to remove my diseased root and install the bone graft. I took my antibiotics as prescribed afterward but declined pain medication. When I had my gumline reshaped a couple weeks ago around where the bridge will go, I again asked for the minimum possible amount of Novocain. I took an ibuprofen after the procedure to help control the swelling then forgot to take more later. My upper lip felt stiff and immobile for a few days afterwards. I could feel pressure and swelling around that part of my jaw, and some tingling in the nerves in the roots of the teeth that had been ground away, if I drank a cold beverage. But I cannot say I felt any real pain.

What do I even consider “real” when it comes to my own pain, though? Is that even a relevant term for people who experience pain similarly? The memory of spending an entire night vomiting from the combination of general anesthesia and morphine I was given during and after my bladder surgery still lingers. It makes me averse to pain medication unless I “absolutely need it.” For pain to distract me it has to be incredibly severe. I would rather deal with nuisance discomfort than become incapacitated in other ways. But does that mean my pain is not real or that it should not be managed?

I grew up in a neuroscience research lab, which gave me some good insight on why we feel pain and how not all types of pain are alike. Some pain originates in nerves outside of our brains and some originates within the brain itself. This means not all pain responds to the same types of therapies. It also means understanding our own pain can be incredibly difficult even when we have years of experience. And as other articles in this issue of *CF Roundtable* demonstrate, not everyone with CF experiences the same things when it comes to feeling pain. Some people experience heightened awareness and some, like me, barely register discomfort. It may be peak neurodivergence that I get more bothered by certain fabric or food textures than when I walked around with broken ribs for several months.

Then again, I may feel more bothered than I consciously know. My wife can often tell when I am in more pain than usual because of my body language and affect—even if I do not realize anything is amiss myself. The sociologist in me thinks there is probably more to this than just acclimation. In the distant past, I was punished for showing any evidence of being sick. I was also constantly reminded that my health issues made me less desirable

and less valuable than my peers. This has not been the case for a long time, but it certainly took a toll during the years I experienced it. I think I do prize being “tough” to a certain extent—or at least, I used to. Now I just feel lost a lot of the time, and sad about all the opportunities I have missed to advocate for myself because I could not voice that I was hurting.

I have no easy answers, either for us as patients or for those without CF who care for us. But I do feel strongly that we need focused attention to pain as an integral part of living and aging with CF. This remains a progressive disease even for people who benefit tremendously from therapies that keep our respiratory and digestive systems clear and functional. More of us will experience the breakdown of other body parts—our teeth, our bones, our joints—as we live longer without going into respiratory failure or starving to death. Pain is often part of aging even without underlying chronic conditions, simply because of wear and tear on the body. What does that mean for our community in the age of CFTR modulators and other innovations in treatment? We need to ask those questions early and often to ensure patients can receive pain management appropriate to our unique individual needs as we grow older. ▲

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



FAMILY MATTERS

Parenting Through Pain

By Cindy Baldwin

Like many CF patients, I've dealt with decades of chronic pain. Whether it's bouts of recurrent pleurisy, dislocated ribs from coughing, sinusitis triggering migraines, or abdominal pain from Distal Intestinal Obstruction Syndrome (DIOS) and gastroparesis, there's always *something* that hurts!

Chronic pain can be particularly challenging as a parent. The physicality of raising kids—carrying babies, being climbed on by toddlers, getting hit by your preschooler mid-tantrum—is often unrelenting, and it's easy for the everyday rigors of parenting to exacerbate existing chronic pain issues.

As a mama to a wonderful nine-year-old who has always been a highly physical kid, I've spent years trying to figure out how best to balance my body's needs with my daughter's needs. While some seasons are harder than others, here are a few of the things I've found most helpful when it comes to parenting through pain.

1. Honor Your Limits

Just like parents are reminded on



CINDY BALDWIN

airplanes to put on their own oxygen mask in case of emergency *before* they put on their child's, there are times when we, as parents with CF, have to get serious about respecting our physical needs, even if that means we can't do exactly what our child wants.

Because sleep is key for me in dealing with both chronic pain and pulmonary health, I have always needed to schedule time for an afternoon nap. Often, this means setting my daughter up with games, books, or a TV show that she can enjoy on her own, even if she'd *really* rather have my company. (When she was little, I shamelessly bribed my daughter to let me have some quiet time in my bedroom alone—we started with five minutes of quiet time and slowly worked up until she could play or read for an hour at a time.)

I also have mobility limitations and chronic muscle pain due to other conditions and have to be careful about how much walking I do in a day. Sometimes this means saying no when my daughter wants to take a walk or play an energetic game outside while other times it means finding ways to modify the activity she wants to do so that I can do it while sitting.

Although it can be hard to tell a pleading kid that you can't play because you need to sleep, or that you can't push them on the swing anymore, or that you can't go to the park—honoring

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using a CFTR modulator actually experienced a slight increase in ppFEV1 after becoming parents, in contrast to the significant decline seen in those not using these treatments. Modulator therapy did not significantly affect BMI or exacerbation rates.

<https://tinyurl.com/222ab5r5>

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

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

Test (SNOT-22) scores and lower scores in the respiratory domain of Cystic Fibrosis Questionnaire-Revised (CFQ-R) and physical health domains of Cystic Fibrosis Quality of Life Evaluative Self-administered Test (CF-QUEST) and Short Form Survey (SF-36). The physical and mental domains of SF-36 and CF-QUEST had a strong correlation with SNOT-22. Higher scores of SNOT-22 nasal subdomains correlated with lower scores of SF-36, CFQ-R and CF-QUEST. Thus, CRS is a prevalent

your limits and making sure you're respecting your body's needs ultimately gives you the tools you need to be a more present parent.

2. Set Firm Boundaries

Because I have fibromyalgia, I sometimes experience hypersensitivity to touch. During those times, I've found I have to be explicit with my daughter in setting boundaries about how she's allowed to touch or cuddle me. We've also had a lot of talks about how mama needs gentle hugs, because my ever-present abdominal pain from a lifetime of DIOS and dysmotility makes tight squeezes very unpleasant!

When setting boundaries with kids, it's important to be kind and caring, but firm. Talk to your kid about what your pain is like and help them to understand what kind of touch is okay and what isn't. Practice ways they can touch you that don't trigger pain, and, if they forget, give a gentle reminder.

3. Get Creative

Whether you can't join your kid

on a hike because of the altitude, or you can't carry them because of chest pain, get creative to find ways to work around your pain! Some of the workarounds I've used throughout my daughter's life include:

A. Learning about babywearing when she was a baby and experiment-

chair I'm sitting in becomes the soccer goal; the fact that I need to lay down on the couch is because I have a mysterious illness she needs to go on a fantasy quest to solve; I can't run around in the kiddie pool with her, but I can sit beside it and splash her with my feet.

E. Finding engaging, kid-friendly audiobooks and podcasts to listen to together when I don't have the stamina to do anything more active.

Chronic pain is definitely an extra layer of hard on top of the normal difficulties of parenting...but finding ways to honor our limits, set firm boundaries, and get creative about ways to play on bad pain days

can not only help us to manage our pain, but help us to raise children who are empathetic and compassionate. ▲

Cindy Baldwin is 34 years old and has CF. She is the author of several books with HarperCollins, including the upcoming *No Matter The Distance* (February 2023), which features a protagonist with cystic fibrosis. Cindy lives near Portland, OR, with her husband and daughter.

There are times when we, as parents with CF, have to get serious about respecting our physical needs, even if that means we can't do exactly what our child wants.

ing with several types of wraps and slings so that I could hold her without additional stress on my body.

B. Snuggling together on the couch instead of holding her when she got too big for me to lift safely.

C. Researching activities and games we could play with me sitting or lying down.

D. Incorporating my need to rest into the games she wants to play. The

co-morbidity of CF patients, which significantly reduces HRQoL. SNOT-22, CFQ-R, CF-QUEST and SF-36 were strongly correlated. Severity of sinonasal symptoms have a strong correlation with HRQoL in CF patients.

<https://tinyurl.com/53defjfp>

AND
<https://tinyurl.com/2h6x4wyt>

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

CF Roundtable ■ Summer 2022

Diabetes is known to cause changes in brain structure and long-term cognitive dysfunction. This work investigated cystic fibrosis-related diabetes (CFRD) as a mechanism for cognitive impairment in people with CF. It was hypothesised that cognition would be poorer in adults with CFRD than in those with CF without diabetes (CFND) or in healthy controls. Cognitive performance was assessed using the Cambridge Neuropsychological Test Automated Battery which provides a comprehensive

cognitive assessment with tests mapping onto specific brain regions. Demographic, clinical and self-reported health data were documented for all participants. CF specific clinical variables were recorded for the two CF groups. People with CF demonstrated deficits in aspects of verbal and spatial memory, processing speed and cognitive flexibility compared with healthy controls, with all areas of the brain implicated. Those with CFRD had additional

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FOCUS TOPIC

DEALING WITH PAIN

What Pain Is Teaching Me

By Julie Desch, M.D.

I didn't write about aging for the last edition of *CF Roundtable*, even though I am quite qualified to do so given my grey hairs, because I was in too much pain to think about writing. So, when I saw this time the job is to write about pain, I laughed...a tiny bit. Here is what comes through me regarding pain at this stage of my longer-than-expected life.

If you are in pain, first of all, have compassion for yourself, because physical pain sucks. No sane person wants to feel pain. In my body, there is pain in the lower back and down the back of both legs. It seems to "always" be there to some degree, and it has become the master of my activities. In other words, unless I prefer that it be excruciating rather than just super annoying, I cannot do the movements that up until now have kept my body healthy and my mind relatively sane. Walking is out. Weight training is out. Any intense form of movement that gets my heart-beat up seems to be a memory. I don't know if it will always be this way. But right now, it is.

I've learned through reading and watching videos and podcasts that chronic pain is understood way better than it was when I trained in medicine. In fact, pretty much everything I learned is useless, like a DVD player or something. We now know that with chronic pain (defined as pain that lasts longer than three months), the brain actually learns to send pain signals as a response to increasingly smaller stimuli. It develops supersensitivity to incoming signals over time. This creates a vicious cycle that, in my opinion, is frankly evil. The end result is that the pain is always there, even when the body is complete-



JULIE DESCH

ly safe and the brain could totally chill out if it just took a moment to look around. This is obviously a very brief sketch of a complicated subject. If you are interested, google "central sensitization." Get a snack, because you will be reading for a while.

So I watch. I watch muscles atrophy. I watch stiffness become the new norm. I am powerless over this. But I'm not special. All bodies decay. All bodies die, either slowly or suddenly. If I think of this through the lens of time, it is happening slowly in my body. It seems to me that looking through this lens of time is what makes it sad to watch. I see what is the case right now, in terms of what my body could do in the past. Then I grieve about what is lost and am scared shitless of what must certainly be my future.

But what is actually here now? A body is sitting in a chair typing. There is tingling and numbness down the legs, especially the left. Calf muscles are *rigor mortis* tight. There is a tiny bit of

pain in the left hamstring, which is relieved if I stretch it a bit. That's all. That is what I am aware of right now. My awareness also hears beautiful music and tastes morning coffee. Awareness looks out the window in front of this body and sees oak trees and a vineyard in the distance. Birds flitter by, evading a clear view needed for labeling with a name. Suddenly, there is silence in the mind and the fingers stop typing. That silence is so sweet, so free.

The message the pain gives is that it is not me. This sounds esoteric and unhelpful. It sounds like a really hard thing to grasp, but really it just takes a moment of reflection. If "I" can sit back and observe this uncomfortable sensation I call pain, then how can it be me? I am watching it. If I am watching it, I am not it. Is it there? Yes. Is it mine? Do I have to own it? No, I do that in a thought. There is a thought that says, "Hell, yes, it's my pain; nobody else feels it!" This very convincing thought is the source of all of my misery.

We have been trained to think this way. It is a conditioned pattern to think that what my body experiences is me. Sensations in my body experienced only by me must be mine, or so we believe, but only because it is what we have always believed. The same is true for thoughts and emotions. It flows like this: This fear of a future of incapacity is "my" fear. Nobody else feels this fear. Right there, I know that is not an accurate thought. Of course other people feel fear. All people feel fear at some point. I haven't been singled out. And yet my fear (or any emotion I feel) is personal and private and, because I am the one experiencing it so intensely, it singles me out. It is mine.

This claiming of things—sensa-

“This claiming of things—sensations, thoughts, feelings—this process of identifying these things as me or mine is why I suffer.”

tions, thoughts, feelings—this process of identifying these things as me or mine is why I suffer. But I can very firmly say, no, these things belong to nothing. They are simply here being experienced, but not owned. This is where freedom is found.

I can suffer if I want, but I don't have to. All I have to do is get better and better at catching this claiming process by the mind in action and inquire into it. Is it true that these sensations are me? There will be a lot of arguing in the mind by the various voices that I have listened to over the

years and believed to hold the truth. Some will be screaming, “F*%k you and your mindfulness, Julie. This hurts like hell, and if you don't f#*king do something about it, it is going to hurt forever!” Others will be shaking their imaginary heads, saying, “You have lost your mind. This makes no sense.”

I am not my body or my mind. What I really am is the witness—I can listen, observe these voices, and firmly understand that these thoughts are not me. I can say to the pain, “Prove to me that you are me.” Can a sensation

prove its identity? How would that work? In direct experience, how is it that this painful sensation is mine but the sound of music is not mine? The taste of coffee is mine but the sound of the woodpecker on the side of the house is not mine? How? Where is the proof? Without believing a thought, what is me or mine?

All of these sensations floating around in awareness are just what they are, sensations. Yes, they are experienced. Yes, they are uncomfortable and it would be nice if they go away. But not claiming them is much more comfortable than the alternative, and I don't have to do it anymore. It is entirely up to me. ▲

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

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difficulties with higher-level processes known collectively as ‘executive function’, which demand greater cognitive load and recruit the prefrontal cortex. Compared with healthy controls, those with CFND and CFRD had an estimated 20% and up to 40% reduction in processing speed respectively. Managing CF requires higher order executive function. Impairments may be sufficient to interfere with self-care and the ability to perform everyday tasks efficiently. <https://tinyurl.com/27j3mvsu>

Indoor Air Pollution Exposure Is Associated With Greater Morbidity In Cystic Fibrosis

Exposure to higher levels of outdoor air pollution is associated with worse lung function and greater rates of pulmonary exacerbations in CF, but limited data on exposure to indoor air pollution exists. Individuals with cystic

fibrosis who were enrolled in the Twin and Sibling Study self-reported exposure to four known sources of indoor air pollution (secondhand smoke, forced hot air, wood stove and fireplace). Change in lung function, rates of hospitalizations and pulmonary exacerbations were followed over 4 years to compare outcomes in those who were exposed to those who were not exposed. Adults exposed to secondhand smoke had 42% increased yearly risk of hospitalization compared to those adults who were not exposed. Questionnaire-based data suggest that exposure to sources of indoor air pollution increase morbidity in both the pediatric and adult cystic fibrosis populations. Future studies with qualitative indoor air pollution measurements are needed to further quantify exposure risks for the CF population.

<https://tinyurl.com/8bh2vf8k>

CFF Grants \$1.6M To Study Lung Transplant Complications

The Cystic Fibrosis Foundation (CFF) has awarded \$1.6 million to support research focused on identifying biomarkers of chronic lung allograft dysfunction (CLAD)—a complication of lung transplants—in people with cystic fibrosis. With this research, the CFF hopes that CLAD diagnoses can be made sooner, leading to improved prognoses in the nearly 50% of transplant recipients who experience the condition in the first five years after transplant. CLAD encompasses a range of complications that occur when one's body rejects a transplant, leading to an inability of the transplanted lungs to function normally. It is the most common complication leading to death in transplant recipients, and strategies to sooner detect it are greatly needed. One of the

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Pain In The You Know What

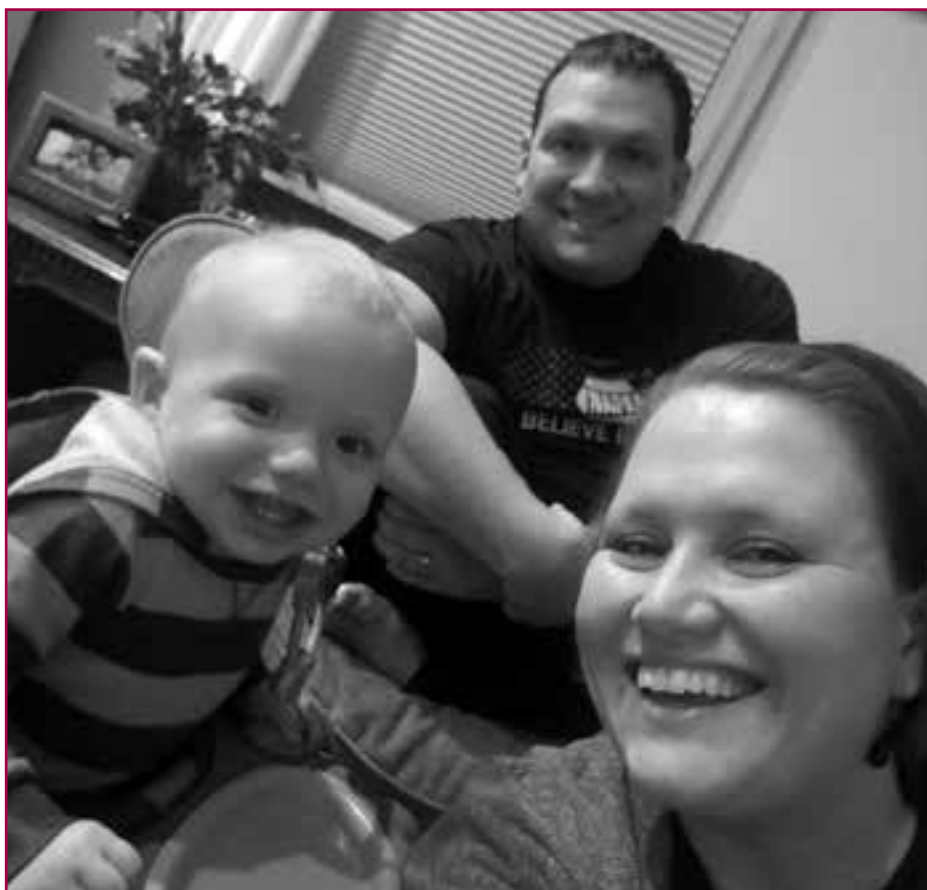
By Nicole Kowal

Pain is usually associated with physical pain, or at least most people I've asked associate it with physical pain—like in January, when I was folding laundry and decided to “crack” my own knee, but, in reality, I dislocated it and then popped it back in myself! The result was everything but my ACL being torn in my left knee (which is good). No surgery was needed, but the pain for four months was awful and still, to this day, the knee does not feel right. But the physical pain wasn't as bad as the mental pain. Mental pain always seems to hurt more to me than the physical. Mentally, I was shot. I hated not being able to take care of myself or my son the way I always have. I hated having to depend on others so much for simple tasks.

Having CF can physically be painful—I have coughed so hard that I have dislocated some of my ribs and needed a visit to my chiropractor to have them popped back in. Some amazing cryotherapy to soothe the resultant discomfort afterward was my only relief. I have had costochondritis—the inflammation of the cartilage adjoining the ribs to the sternum—more times than I can remember, and I have even gone to the ER for treatment on several occasions. My joints have the beginning signs of rheumatoid arthritis; living in Buffalo, NY, makes this challenging because of the constant weather changes. My sinuses, even after having had two sinus surgeries, still give me pain during the allergy season. Again, the frequent weather changes here don't help. Nonetheless, my mental pain is always there and always more pronounced than my physical pain.

I am in pain mentally. That may sound super confusing but let me explain. The fact that my mind never

“The constant worry I experience makes my heart and mind feel almost as if the pain were physical.”



MICHAEL, NICOLE, AND ERNIE KOWAL (THEIR MIRACLE TRIKAFTA BABY) AT HOME IN EDEN, NY.

stops gives me pain. The constant worry I experience makes my heart and mind feel almost as if the pain were physical. Pain management is important, but we cannot forget to address the pain of our mind and heart. Having a disease like CF can even inflict pain on others, and that also weighs heavily on my mind.

Our families have pain and worry because they watch us go to endless doctor appointments and undergo

endless treatments. And I worry about the pain that may come from watching me “suffer” or die. Now that I am a mom, I worry about my son potentially growing up without me, or having a mommy who is too sick to participate. Even after starting Trikafta and being the healthiest I have ever been, I still worry about all of this. In addition, I am sure we all feel the pain of survivor's guilt or the pain of the cost of

medications. I also worry about how much of a pain in the a\$\$ I can be toward others—my boss, my friends, or my family—with my random appointments or issues.

Ultimately, the mental pain of this disease is what hurts the most. I discuss my pain with my husband and my therapist. I take two anxiety medications to assist with this as well, and, my gosh, that has helped tremendously. I wish it were more “normal” to discuss

this type of inner suffering. Mental pain is a real thing, and I believe a lot of people overlook that. I tend to resort to sarcasm and humor to cover my pain. We all have different coping mechanisms for our pain and the main thing is to make sure you seek help for whatever pain you’re experiencing. If you are physically in pain, please do not just deal with it, and, if mentally in pain, please reach out to someone who will help you through it. Pain comes in

so many forms and we have so many sources nowadays to relieve our suffering and get the help we deserve. ▲

Nicole is 34 years old and has CF. She has been married to her husband Michael for over six years and they have been together almost 11 years. They were blessed with their son Ernest within six months of starting Trikafta. They live in a small farm town near Buffalo, NY. You can email her at abnormalnicole@gmail.com.

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funded projects will explore the relationship between transplant rejection and digestive reflux, a condition characterized by indigestion (so-called heartburn), chest pain, and swallowing difficulties. To do so, the researchers will examine the bile acids—digestive fluids made in the liver—found in the lungs of transplant recipients. In collaboration with the Cleveland Clinic, CFF launched the CF Lung Transplant Consortium (CFLTC) patient registry and biorepository last year. The project’s goal is to collect clinical data and biological samples to aid scientists in better understanding CLAD and other transplant outcomes.

<https://tinyurl.com/mrxx9utw>

Sustained Effectiveness Of Elexacaftor-Tezacaftor-Ivacaftor In Lung Transplant Candidates With Cystic Fibrosis

Elexacaftor-tezacaftor-ivacaftor induces rapid clinical improvement in patients with cystic fibrosis (CF) and advanced pulmonary disease, often leading to suspend the indication for lung transplantation. Yet no long-term data is available in lung transplant candidates. The authors found that in lung transplant candidates eligible for elexacaftor-tezacaftor-ivacaftor, the rapid improvement following initiation of

treatment persisted over one year with a reduction in treatment burden and lung transplantation could be safely deferred in most patients.

<https://tinyurl.com/2nhsr3ve>

Outcomes Following Lung Re-Transplantation In Patients With Cystic Fibrosis

Compared to their initial transplant, CF patients experience significant clinical decline in renal, cardiac, and pulmonary function at the time of lung retransplantation. This may indicate that an earlier evaluation and rehabilitation process may be necessary to identify patients earlier for lung retransplantation prior significant clinical decline.

<https://tinyurl.com/3vc5xcsm>

Low Body Mass Index As A Barrier To Lung Transplant In Cystic Fibrosis

Patients with advanced CF lung disease and BMI ≤ 17 kg/m² are less likely to be listed for lung transplant and have a higher risk of dying without listing, compared to those with higher BMI. Regional differences suggest access to transplant for malnourished CF patients may be limited by location.

<https://tinyurl.com/wtwxjh34>

Survival Improved For Patients With

Cystic Fibrosis With BMI Recovery Post-Lung Transplant

In patients with cystic fibrosis, lower preoperative BMI was linked to lower likelihood for BMI recovery within 1 year of lung transplant, but for those who achieved BMI recovery within 1 year after transplant survival improved. Poor nutritional status as measured by BMI is an independent risk factor for pre-transplant death in advanced cystic fibrosis lung disease and low BMI is an indication for early referral to a lung transplant center. Thus, having a low preoperative BMI is a risk factor for poor outcomes after lung transplant, but this study underscores the importance of attention to BMI recovery posttransplant. Future research should investigate whether approaches to augment weight gain posttransplant improve outcomes, particularly among recipients with very low BMI at the time of transplant.

<https://tinyurl.com/37v6f2x3>

CF Patients With CFRD Treated With Lung-Pancreatic Cell Transplant

A lung and pancreatic cell transplant from a single donor to people with end-stage cystic fibrosis and CF-related diabetes (CFRD) safely and effectively improved their lung func-

Continued on page 23



Voices from the Roundtable

Dating At An Older Age

By Jennifer Kyle

Here I was, 51 years old, living with CF, post-menopause and divorced. After going through a 13-year emotionally and mentally abusive marriage, I was happily on my own again. But who was I? Who did I want to be in my 50s? Was I ready to date again? How would someone take the news when I had to tell them I have CF? When I was dating almost 20 years ago, it was very different and I did not know I had CF. I was 37 years old when I was diagnosed. Going out and meeting someone then was the norm, but now I had to learn a whole new way of dating. Scary! After my divorce, it took four years to heal, learn, reflect, and grow until I felt I was ready for the dating scene. I had heard horror stories about online dating from friends and colleagues, and here I was about to start my own journey.

So the first thing I did was talk to all my friends about which sites I should use. The recommendations were overwhelming—go on Match.com, try eHarmony, or see if you like Bumble. Ugh! So many choices. I settled on Match.com and quickly had to learn about scamming. Thankfully, my closest friend was very familiar with this and taught me all the signs to look for when a “fake” person was texting me. I quickly learned how to block people. Some would think this factor alone would be enough to scare you away from dating, but I was determined to get my dating life back. So I decided I was going to face all of this with a sense of humor. Scammers? So, what! Terrible dates with rude people? Yes, but there are some hysterical stories.

As the months ticked by, I was not meeting anyone who interested me on Match.com, so I turned to eHarmony.

This proved to be another dead-end, so I tried other dating apps like Bumble and Hinge, only to be equally disappointed.

As a year was approaching with online dating and all I had were funny stories, my CFReSHC friends urged me to try Plenty of Fish (POF). I thought this was a hook-up site but, after their stories of how they met their significant others, I was intrigued.



JENNIFER KYLE

After a week on POF a cute message appeared. After looking at his profile I thought we had lots in common, so I messaged him back. We did the obligatory texts for a day and then he called me. OMG what a sexy voice, I thought. We talked for a while and he asked if he could call me tomorrow. Yes! And so it continued every day leading up to our first date. I felt like I knew him already and couldn't wait to meet him in person.

Being as this was the height of the pandemic, I knew I was taking a risk but I just couldn't pass this up and I'm so glad I didn't. On our very first

date I decided to reveal my CF to him. There was something very special about him, so I wanted him to know about my CF. I was nervous, wondering if he would end things but knew I had to tell him. I soon found out that he was a truly wonderful, caring person and had no intention of ending our relationship based on my CF. In all the previous dates I went on in the past year I did not reveal my CF. I knew these relationships were not going anywhere so I felt it was my choice not to tell. There were times I actually enjoyed being on a date and the person not knowing about my illness—it was a mini vacation from CF. But this time it was different. We are still together and our relationship continues to grow stronger. I am so glad I did not let age, abuse, or CF stop me from meeting a wonderful person. ▲

Jennifer Kyle is 57 years old, has CF and lives in Somerset, NJ. She was diagnosed at 37. Jennifer was a health, physical education, and dance educator for 16 years in the New Jersey public schools. For four years she served as an adjunct professor for Montclair State University, served as President for the New Jersey Association for Health, Physical Education, Recreation and Dance (now known as SHAPE NJ), and served as the VP of Dance for the Eastern District Association for the national SHAPE organization. Since retiring on disability, she runs a dog-sitting business and has helped organize BreathCons, ResearchCons, MiniCons for the CFF. She has also been a peer mentor for the CFF. Jennifer currently serves on the Governance Board for CFReSHC as the Meeting Coordinator and is working as a patient advisor for an outreach study with the CFF for improving the collection of spirometry results for telehealth appointments. She has also participated in five clinical trials and hopes to be a part of many more.



PHOTO BY DEREK POWAZEK

Needles

I am surrounded by physical pain,
Pain that pierces my body,
With every breath, with every touch, with every movement.
But I feel no pain,
For I have become stronger.
Strong enough to look pain in the face and smile,
To smirk at its witty desire to cause me discomfort,
to invade my space,
to destroy my inner being.
For only by tolerating pain, by befriending pain,
can I survive.
After all, the emotional pain of this disease,
Now that hurts.

-A. Stenzel, 1997

FROM OUR FAMILY PHOTO ALBUM...



MICHAEL, ERNIE, AND NICOLE KOWAL, WITH HER PARENTS, ERNIE AND CONNIE MATTHEWS AT THE 2022 GREAT STRIDES CF WALK IN ORCHARD PARK, N.Y.



PAT MULLEN AND JENNIFER KYLE.



JUSTIN AND AUGUST LORIO, FORMER POLE VAULTERS WHO TRAINED WITH JERRY AT IONA PREP, HIKE WITH JERRY CAHILL AT HIS "HIKE 2 BREATHE" TEAM AT THE ROCKEFELLER ESTATES IN NEW YORK STATE.



ROY BERKELEY-HADDOX



DOMINIC QUAGLIOZZI



JEANIE HANLEY AND HER HUSBAND, JOHN, IN ROME, ITALY, WITH THE COLOSSEUM IN BACKGROUND, IN JUNE 2022.

COLLEEN ADAMSON IN FRONT OF HER SERVICE PLAQUE FOR 26 YEARS OF SERVICE AT THE PENTAGON.





My Time During The Pandemic

By Roy Berkeley-Haddox

During the pandemic lockdown of 2020, I was free from the burden of hiding my cystic fibrosis in public for the first time. In isolation, I no longer needed to give drawn-out explanations for my CF symptoms or lie to cover them up. I no longer had to abruptly leave events or college classes to cough up cups of blood or take care of another symptom. I no

longer had to deal with the embarrassment of being taken out of class in an ambulance. Among people without CF, I'd always felt like an alien masquerading as a human. Ironically, right before the lockdown, my worst symptoms began disappearing because I'd started taking Trikafta. Now largely healthy and alone, I had all the time in the world to cope with the past and

wonder about who I really was and what it all meant. ▲

Roy is 31 years old and has CF. He lives in Los Angeles, California, and is passionate about filmmaking that explores issues of trauma and stigma. He is a recent graduate of the USC School of Cinematic Arts. You can email him at royberkeley@gmail.com.



ROY BERKELEY HADDOX

tion, metabolic control, and quality of life. Given that CFRD increases the risk of complications after a lung transplant, these findings suggest that a combined lung-pancreatic cell transplant may be an effective way of improving life for this group of end-stage CF patients. CFRD is associated with poorer lung function, poor nutritional status (slower growth and/or weight loss), and a greater risk of death. In addition, for CF patients undergoing a lung transplant, its presence decreases patient survival, while promoting infection and post-transplant pulmonary rejection. Restoring satisfactory glucose control through a transplant of pancreatic islets—groups of cells that produce insulin and other hormones—may help improve outcomes of a lung transplant in CF patients with CFRD. Combined lung-pancreas cell transplantation, however, is technically more complex, carrying a higher risk of complications due to the simultaneous thoracic and abdominal procedures in already very weak patients. Pancreatic islets can be successfully grown and maintained in the lab for up to 10 days, and transplanted into the liver through the portal vein. This allows implantation to be delayed until the patient's condition has improved. To date, only case reports of combined lung-pancreatic islets have been published, and questions remain as to the efficacy and safety of this procedure in this particular patient population. Researchers launched a Phase 1/2 clinical trial, called PIM (NCT01548729), to assess the feasibility and effectiveness of combined double lung-pancreatic islet transplant from a single donor in end-stage CF patients with CFRD and on insulin therapy. Using the same donor for a combined transplant is thought to reduce the risk of immune responses against the transplanted tissues. The trial's main goal was to assess the combined transplant's metabolic efficacy, measured by a composite score

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DIAMOND SUSTAINING PARTNER



ENDOWMENT PARTNERS





Pain Resolution

By Andrea Eisenman

Have you ever been in so much pain that you couldn't breathe? This was a new sensation for me and it had nothing to do with my lungs; rather, it was my gut. I had no idea what was happening when these attacks would come on suddenly, but they were debilitating and scary.

Initially, the pain was mild but grew in intensity over time, warranting a trip to the emergency room (ER) of the closest hospital. I was doubled over in agony, the main source of which seemed to be emanating from my diaphragm. The pain started slowly and escalated to a cramping type of pain where it felt like a tree trunk was pushing through my core, clamping it shut. To work through and ease this severe discomfort, I had to get on my hands and knees and have someone rub my back while I tried to sip the air. The person rubbing my back noted that my back felt hot to the touch.

During these situations, I couldn't eat or drink anything, including necessary medications, making a difficult situation if the episodes lasted longer than six hours. If I did try to drink once the pain eased slightly, within 10 minutes I started vomiting whatever small gulp of liquid I was able to get down. After about four hours, I called the transplant doctor on call at my clinic. They usually tell me to get to an ER to get some IV fluids, which is typical. Although I've been dealing with this situation for several years, I have seen it progress to the point of a hospital admit.

The random nature of these episodes was the scariest thing. I tried to note what was setting my system off each time. What can I avoid eating or doing so I can live preventatively? The answer, as it turns out, was more complicated.

All of this started in 2006, about a year after I had my gallbladder removed. I was told that eating fatty foods (ribs, fried foods, etc.) would be problematic without a gallbladder. I didn't eat many of those items and, when I did, I took more enzymes. I couldn't discern the pattern—after eating ribs I was fine, but there were a few times when I noticed that eating smoked fish on a bagel with cream cheese seemed to set off these painful episodes.



ANDREA EISENMAN

On one occasion, while visiting my mom in Hampton Bays, I was on IV antibiotics and the doctor on call in NYC told me to go get hydrated. When I arrived at the local ER to get fluids, my bloodwork indicated extremely elevated liver enzymes, so they kept me overnight and admitted me. They did an emergency abdominal CT plus a sonogram of my liver. They were concerned I had a blockage in my biliary tree (the gallbladder, liver, and bile ducts, collectively). I told them no, that whenever I vomit for several hours (and I had been vomiting for close to

five hours already), my liver numbers rise. It was so frustrating. I know my body and this was par for the course. By then, I was hydrated and no longer vomiting, thankfully. But if I left, I was signing out "against medical advice" (AMA), which meant insurance might not have covered my visit.

Because I was on IV antibiotics and the hospital didn't have that particular antibiotic in their formulary, I was put on a different one, which made me nauseated from the change midway through the course of antibiotics. Sadly, this happened on a Saturday and the attending clinician in the hospital claimed that I needed to stay until Monday so the surgeon could read the CT scan and see whether an operation was advised. So I stayed. There was no blockage, no stone or biliary duct problem, and my blood work returned to normal.

I was still no closer to understanding what was happening. There were several incidents after that admission, and I was becoming fearful of eating certain foods as I didn't want to end up in the hospital again. My transplant doctor ran every GI test under the sun—a colonoscopy, a stomach-emptying test, a nuclear test, and probably some I cannot recall. I had an episode happen while I was playing tennis, fortuitously with an opera singer. She saw I was distressed and coached me through breath work to get through it. It was amazing—I was able to relax my diaphragm and the pain slowly dissipated. I was convinced I had pancreatitis, but my transplant doctor said that doesn't go away with breathing or just spontaneously within a day.

After a few times, I was able to breathe through it, especially if no one was around to rub my back when I was prone. I tried breathing like an opera

“I back slid a few times into pain, even though I was given opiates but was afraid to take them.”

singer and sometimes it worked, but not consistently.

The last time it happened was a disaster. I went into full pain and vomiting mode. I couldn't keep any fluids down and therefore couldn't take my immunosuppressants. I was slumped over in agony. This time I was in NYC, so I went to my clinic's hospital where I lay in the ER for 10 hours with no medical intervention or attention other than IV Dilaudid for pain. My blood work didn't have the markers for pancreatitis, even though I was certain I had that at this point. I was given a urine cup and my pee was brown. Since no one was doing anything in the ER, my husband and I decided I could cope with this at home, so I left. I was still in pain and was in bed for two days. Despite being thoroughly drained and exhausted, I could drink little sips of water, so I started taking my meds. I called my transplant coordinator who thought I was still in the ER! I told her what happened and said there was no way I was going back to that horrendous place—either admit me to the transplant floor or I am “checking out.” I cannot live this way. She got me in but first I had to drink a bunch of liquid contrast to get a GI CT of my abdomen.

I was admitted to the transplant unit and I had never been so happy to be in the hospital. There they immediately realized I had a full-blown urinary tract infection so was put on IV antibiotics for that as well as a constant IV saline drip. I was seen by a lovely GI doctor who told me I didn't have pancreatitis but they didn't know what was wrong—yet. Nevertheless, the interns treated me like I did have pancreatitis. I was in the hospital for 10 days! I was

put on a low-fat diet for pancreatitis! It was crazy. I back slid a few times into pain, even though I was given opiates but was afraid to take them. I was reassured I was not going to get addicted.

I left the hospital feeling better but had lost about 10 pounds and was emotionally and physically decimated by the whole affair. I was supposed to remain on a low-fat diet at home and I did. I was to go back to see the GI doctor once out of “Club Med.” It was only at his office that the doctor and I had a heart to heart and he gave me a magnificent little blue pill which was an anti-spasmodic. I was told to only use it when I was trying to ward off an “attack.” We decided my stomach was spasming from an unknown trigger.

The minute I start feeling twinges of gut tightening, I take the blue pill. Sometimes I don't quite catch it in time. But amazingly, after popping it, the pill works in about five minutes. I haven't been hospitalized for this since 2018 and I make sure I always have those pills with me.

I am no longer living in fear of ending up in the nearest ER. It was a long road to discovery. I am grateful to have the tools, a.k.a. my anti-spasmodic, along for the ride for the rest of my life. ▲

Andrea Eisenman is 57 and has CF. She recently realized that her initials are AGED: Andrea Gail Eisenman Downey (her husband's surname)! She lives in New York, NY, with her husband Steve and dogs, Willie and Roscoe. Andrea is the Executive Editor for USACFA. She enjoys cooking new recipes, playing pickle ball, biking, tennis when possible, and staying active as her health allows. Her contact information is on page 2.

YOU CANNOT FAIL

The **You Cannot Fail** program is based on a saying that Jerry Cahill's parents shared with him at a very young age. This saying helped keep him determined to push through all bumps along his path.

You Cannot Fail is an inspirational launch pad that empowers people to discover and embrace their inner hero; to face the challenges of life with strength and courage; to meet each day with optimism; to live a life of creativity, purpose and passion. **You Cannot Fail** collects, organizes and shares individuals' stories about specific aspects of their lives in order to motivate and inspire others to be the heroes of their own stories.

Visit: www.youcannotfail.com to share your story, inspire others, and to become a part of this official program of the Boomer Esiason Foundation.





IN THE SPOTLIGHT

With Dominic Quagliozi

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

Readers, I'm thrilled to introduce one of my closest friends in the CF community, artist and performer Dominic Quagliozi! We met in 2017 because he messaged me after I posted in the big public CF Facebook group. We bonded over our love of visual and performance art, our philosophy about intimacy and relationships, and our fascination with the evolving human body. We also have very different CF experiences—almost seven years ago, Dominic received a double lung transplant. He also started Trikafta this January. But that's not even the biggest change Dominic has made this year. He and his wife Deb returned to Massachusetts after many years in California. Their son Thaelo is now experiencing his first snowfalls at their new home in Worcester. Please welcome our newest star, Dominic Quagliozi. Spotlight, please!

Age: 39

Home: Worcester, MA

What would you most like our readers to know about who you are and what makes you feel connected to others with CF?

I've always been a big proponent of building communities. One of the biggest things artists can do is build a community; it's no different with CF. We've done a great job with resources and providing opportunities for community building over the years. That goes back to my days as a young kid in the early 90s, well before the six-foot rule. We had camps and events where all the CF kids were invited, so I met others my age. This made a difference developmentally, especially in my pre-teens and teens. It let me stay connected to other people with similar challenges; I felt like nobody could really relate to me except my CF friends. So



DOMINIC QUAGLIOZZI IN HIS ART STUDIO.

I've always reached out to build those relationships.

You're an accomplished visual and performance artist; these days you're also a dad and a homeowner! How do you balance all of that with the daily tasks of managing CF?

It's all about living these experiences in my daily life—my health life, my chronically ill life—and then bringing them into the studio with materials from the hospital. I love having Thaelo be part of all that. He has a huge play space with a carpet; he loves when I'm in the studio. Whenever I'm down here, he comes and finds me. So my studio basically has been completely infiltrated by his toys. But that just makes it easier to spend time with him while I create.

I know you and Deb went through a pretty complicated journey to owning your home. What was it like doing all that as someone with transplanted lungs in the middle of a global pandemic?

I built quality relationships, both in general and with medical professionals, during my 15 years in LA. That reduced my anxiety and depression while I was there. Moving away from my literal paradise was so hard. I felt like the most like myself there; it was a huge part of my heart and soul. I moved back here for specific reasons but I keep questioning, "Why did we do this? Why did we come back?" But the artist in me is saying, "These things are going to strengthen the art you make."

Can you tell us more about your transplant experience and how living with your new lungs has shaped your art? Where have these experiences taken you creatively?

I would joke about feeling scared to get transplanted—not because I worried about dying, but because I feared once my CF lungs were removed I would lose all my artistic power. They brought me so much struggle, perspective, and content. In a weird way, my CF lungs were very giving toward my art career. Taking that away scared me and still does; I'm wrestling with that and the trauma underlying it. But my new lungs have given me seven more years of making art; now my work is getting purchased by museums and I can reach a wider audience. My current work has similar threads—common themes like temporality, patienthood, and privacy. So I'm still doing those things, just differently. Pre-transplant, I did a lot of performance-based things where I used my body. Now I'm not

doing as many performances, but I'm using a lot of ephemera from patient life like hospital gowns, clinical tissue, and rubber gloves.

You've also been hard at work setting up your new studio space at home. What will your basement studio enable you to do artistically in the year ahead?

A major factor in choosing this house was that Deb gets a beautiful studio upstairs that has natural light all day long for painting, illustration, and pattern design. Then I have the basement space that allows me to do all kinds of projects at once, which is really how I work best. I've got about 200 square feet, probably five times more than before. So I want to do larger-scale projects like soft sculptures using hospital gowns. I can work on multiple projects at once by setting up different tables and hanging canvases on the walls. I have one table for drawings and one for sewing, plus big folding tables I'll use for other projects. Right now I'm developing ideas around gowns or tissue and themes of clocks and parachutes. In this space I can stretch out, get messy, and return to the fundamentals of my art practice.

What are your big goals with your art right now? Where would you like to go with your work in the near future?

Starting Trikafta has really influenced my goals for my art. One of my new drawings is in a show at the Rhode Island School of Design Museum. The director asked if we could extend the show until October 9, which is amazing. So I have to network and meet other influential art people in New England—because of that show, I can connect with more opportunities in the area. I would also love to get a job teaching art somewhere or develop my own program with Deb. The freelance life has been stressful. It's burning me out and making time management

tougher. I'm missing deadlines and feeling scatterbrained about qualifications and materials for different job and grant applications. I would also like to do a residency. There are some really good art residencies in New England, so I want to take advantage of living here.

Besides lung issues, what are your biggest challenges with CF? What is your unique presentation of the disease like? How has that changed in your experiences with CFTR modulators?

It's this constant flow of problems that move around my body. I'll have sinus issues and then start a treatment for those. Then the sinuses clear up a little, but I'll have something else—a severely low white blood cell count, for example. Then I'll need a bunch of shots and medication changes to stimulate my bone marrow. That'll clear up and then I'll be dealing with arthritis in my back, causing pain and weakness. Next, I'll have similar issues with my knees. My new thing is entering Stage 4 kidney disease—it's getting pretty serious. Most days my legs swell up so much that I have trouble even wearing my shoes.

You've had a lot of changes with your health since starting on Trikafta. How have your experiences with Trikafta differed from what you noticed with previous modulators you were taking? How have your expectations evolved as you've been on the triple combo longer?

In 2015, Orkambi came out for folks with double delta F508, but that was a couple weeks after my transplant. So I didn't try that one or Symdeko. But when Trikafta came out in 2019, I was excited and wanted to know if I could get on it. I didn't actually get approved until I moved here. I started January 1, 2022, and, so far, it's been hard. I'm getting physical issues with

fatigue—my muscles feel like I've been doing squats. I know that's a possible side effect, but I haven't talked to anyone who's had it this badly. And I've been falling asleep like twice a day. I went to my endocrinologist and my labs look fine—slightly low on iron, but that's it. It's a lot emotionally and mentally; sometimes I question why I'm doing all this. I will say, though, it's nice to feel a breeze in my sinuses! Breathing through my nose again feels weird but awesome. Other things are the same—I haven't gained any weight or changed my diabetes regimen.

What helps you cope with your CF? Are there any community resources you've found helpful in living with CF—like websites, newsletters, apps, social media groups?

Being online, definitely. In the late 90s, when computers and AOL chat got bigger, I stayed in touch with more CF friends virtually. I used MySpace, but it was really Facebook groups that helped a ton. I knew maybe a dozen people growing up with CF just from hospital functions. But once I got on Facebook, I suddenly knew hundreds of other CFers worldwide. That's been really amazing. It also helped me through the transplant process; I went through it with four people who were going through it at the same time. We became friends and bonded very closely because of that. Of course, there's a dark side to that. I've had friends I'd chat with every day online and then I'd see their wall with messages saying, "RIP" and "I can't believe you're gone." I'd think, "We were just talking last night."

You and I really connected about the closeness we share with our spouses and how that intimacy supports us in managing our health. Can you tell our readers about how you and Deb met and how you first shared about your CF with her?

Continued on page 28

Until Deb, I basically withheld my CF. I'd only had more casual relationships; there was definitely something different about Deb. Even right when we met, I thought, "This might be a time where I need to say something." I needed to be completely honest—and I was hesitant because it was scary. On our second date, she was feeling down and told me she was having some health issues. I said, "That's awesome because I have cystic fibrosis! I'm used to being a patient, so whatever you're going through, I'll be there for you." We connected over this really dark subject matter over dinner and just kept talking. Thankfully, her issue resolved, but, shortly afterward, I had an exacerbation and ended up hospitalized. She started coming and hanging out; she brought her pads and we would sketch each other. I loved making art together, still do—it was a partnership

from the beginning. We've taken care of each other so much; now with Thaelo here, it's a new adventure.

As a first-time dad, how are you planning to talk with Thaelo about CF as he grows older? How does living with transplanted lungs—and now being on Trikafta—shape what you'd like to share with your son about your health?

I just want to share as I go. I wanted to see what kind of kid Thaelo would be and choose a pace he could handle. Now that I know him, I think that's a good approach. He's only two, but he's completely there for me. If he notices I'm tired or not feeling great, he'll bring me some water or pat me on the leg. It's wild to see how empathetic and emotionally intelligent he is. So I'll go step by step with him—more in kid terms now, then more specific language over

time. He already sees me taking pills every time I eat. I'll bring that up: "You know how you went to the doctor for your physical? Daddy has to go to the doctor, too." We'll read books or watch shows about healthcare and talk about them. I want him to know he might take care of me at some point—he already does, in a way. I want him to know the expectations as we get older. But he's a very easygoing kid; he can adapt to change.

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

I ran into a cliché because of COVID-19! I was making all this work before the pandemic even happened. Now people think I'm just remarking on COVID-19—not looking at the year it was made, or the context. There's a

including metabolic and nutritional parameters. A transplant was defined as successful if, at one year, three of the following four criteria were met: weight increase of at least 5%; fasting blood-sugar levels lower than 110 mg/dL; a 30% or greater reduction in insulin requirements; and at least a 0.5% drop in HbA1c. Secondary goals included changes in these individual measures: insulin production, lung function, and health-related quality of life, as well as safety assessments. 70% of the patients had a successful combined transplant. They showed an increase in body-mass index (BMI, a ratio between weight and height), better control of blood-sugar levels, and a 38% drop in daily insulin doses. One person was able to stop taking insulin. Successful lung-pancreatic islet transplant was also associated with better lung function and gains in health-related quality of life, particularly its physical aspects. No one died or rejected their transplanted lungs during the

follow-up year, and no complications related to the pancreatic islet injection procedure were reported. The safety profile of the combined transplant was similar to that of a lung transplant alone, with lung infection as the most common complication and no reports of unexpected adverse events. All serious adverse events, most commonly occurring within the first month post-transplant, were successfully treated. These findings indicate that this is an efficient and viable therapeutic option for patients with end-stage CF and CFRD.

<https://tinyurl.com/29dpv4gs>

AND

<https://tinyurl.com/ynj3xamc>

High Levels Of DMBT1 Protein In Lungs May Mark CF Progression

Elevated levels of DMBT1 in the lungs, a protein previously linked with inflammatory processes, may be a biomarker of progression in cystic fibrosis.

High DMBT1 levels impaired the movement of cilia, the finger-like projections that help to clear mucus from the airways. DMBT1 is a protein found in several lung cells, including alveolar type II cells that play key roles in lung function, including the regeneration of the epithelium—which lines the respiratory tract—following injury. In healthy lungs, DMBT1 is found at low to moderate levels that rise during inflammation and with bacterial and viral infections. Besides its role in inflammation, DMBT1 promotes blood vessel formation and epithelial cell maturation, highlighting its role in tissue repair. Researchers examined post-mortem lung tissue from a man with CF and tissues from CF patients who underwent a lung transplant. Compared to samples from people without lung disease, those from CF patients had markedly higher levels of the DMBT1 protein. Results showed that DMBT1 expression was upregulated in CF which is in line with

weird traumatic response to that. Now everyone's making work about illness; they probably have paint-by-number about COVID-19! It's a weird time for me as an artist and I'm trying to feel my way through it. I was using masks in my work or making portraits of my hospital team—like those photos where my providers didn't have the bottom half of their faces in 2010. I feel protective and resentful; I'm struggling with that. I want to let it go, but I can't—this is my life. Did people realize all these COVID-19 protocols were my transplant protocols? I didn't complain about them; I wore a mask literally everywhere except indoors in my own home for three months straight. Not just a mask that you could pull down under your nose, but a two-canister respirator.

Living with CF constantly requires us to change our plans and accept

difficult things. What would a perfect day look like for you? Where would you go and what would you do? Would your answer be different if the COVID-19 pandemic were under better control?

That's a hard question for me to answer right now, because obviously I would go away from here. I'd fly to LA and spend the day there. Despite the air pollution, I think LA is the perfect place to have cystic fibrosis because the climate is dry. The humidity in Massachusetts gave me pneumonia every year on my birthday in April. There's also a daily struggle getting from one place to another. The mobility issues that I have with CF are common, like arthritic knees. Even though I'm post-transplant, I do have chronic rejection plus shortness of breath with activity. Struggling with snow and ice getting to my car, needing to

shovel out my walkway and snowblower the driveway, are very challenging for me. Those big environmental changes make me notice my CF way more here. ▲

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

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

the known functions of DMBT1 during inflammation.

<https://tinyurl.com/nhzbtr92>

Association Between Elevated Peripheral Blood Eosinophil Count And Respiratory Outcomes In Adults With Cystic Fibrosis

Elevated blood eosinophil counts are linked to worse outcomes in asthma and COPD, but have yet to be well characterized in CF. Scientists hypothesized that higher stable visit blood eosinophil counts are associated with increased rates of lung function decline and pulmonary exacerbations (PEx). A retrospective analysis of adult CF patients enrolled in a prospective cohort study focused on blood biomarkers was performed. Those with high eosinophil counts experienced increased respiratory symptoms, but the rates of lung function decline and PEx were comparable between the group with high eosinophil counts and the one with low levels.

<https://tinyurl.com/2p993j43>

Home Monitoring In CF May Help Detect Pulmonary Exacerbations

Home monitoring with a mobile phone-linked spirometry device may provide an effective way of detecting pulmonary exacerbations in people with cystic fibrosis. The findings showed more pulmonary exacerbations were identified through home monitoring compared with standard CF care. The use of home monitoring was generally well-accepted by patients, for whom it gave a sense of control and independence. Several factors have been identified that can predict whether a patient is more likely to fail to return to baseline after an exacerbation. These include being female, infection with bacteria such as *Pseudomonas aeruginosa* and being malnourished. Another predictive factor is a large decrease, before the start of treatment, in FEV1. According to the researchers, since symptoms often

gradually worsen during an exacerbation, home monitoring would allow faster treatment initiation and possibly improve patient outcomes. Researchers hypothesized that home-monitored patients would need fewer days hospitalized and have a better health-related quality of life (HRQoL) compared with patients receiving routine CF care. In this pilot study (NCT02994706) adults with CF were randomly assigned to the home monitoring intervention or to standard care. Home-monitored patients were provided with a Bluetooth-enabled digital spirometer and a mobile phone, and instructed to examine their health status twice a week during the yearlong study period. Participants were required to record their FEV1 results using the digital spirometer. Their symptoms also were recorded with the use of the Cystic Fibrosis Respiratory Symptom Diary – Chronic Respiratory Infection Symptom Score (CFRSD-CRIS). A

Continued on page 34



CF: THE MIND GAME

When The Seeds Of Pain Sprout Anger

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

I have struggled with chronic pain for a big chunk of my life just like many of my fellow warriors, particularly those who've been fortunate enough (or unfortunate enough, depending how you look at it) to hang around for five-plus decades. At the risk of losing my audience at the outset for committing the crime of winning, I will briefly describe the sources and causes of my chronic pain from most to least favorite.

To begin with, I have several lingering sports injuries that I am particularly proud of: several hockey injuries—a banged-up shoulder, fractured kneecap, and back pain from slipping too many discs—and a broken wrist from a kickboxing tournament. I also have constant pain and tightness in my abdomen from being stabbed during a blackout and pain in my left elbow from being thrown through the rear glass door of the Apalachin Tavern. While all of these injuries are quite painful, I relish them the same way some folks lick their canker sores, because it makes me feel alive and brings to mind memories of the full life I once lived, bad choices and all.

On the other hand, my least favorite pain is CF-related because it was not by my choosing and it prevents me from living life more fully. Specifically, at present, my CF-related pain primarily consists of near-constant GI bloating and pain due to my enzymes no longer working as well and sharp, unpredictable right-lung pain, which began about ten

years ago and has only increased in frequency and acuity ever since.

Quite naturally, the seeds of pain have always inevitably sprouted anger within me if I do not take effective action to prevent it. Beginning at age nine, I started flying into fits of rage occasionally that seemed to come out of nowhere, not fit the circumstances, and last way too long. For example, one time I flew into a rage because we were out of strawberry Nesquik, which I discovered upon arriving home after a clinic visit in which I saw my best friend on oxygen for the first time. I

deeply regretted having that outburst and the many others that would follow, which never failed to cast a shadow of shame for days afterward. In their eagerness to console me, my nurse and my mom reassured me that those reactions were normal. Their reassurance did nothing to reduce the number of outbursts but did make me feel less guilty afterward.

Interestingly, when I was nine, my dad, who rarely helped with my chest physical therapy (PT), accidentally bruised three of my ribs while doing my chest PT. Not surprisingly, that

made it excruciatingly painful to breathe, let alone cough, for several weeks. Prior to that, on the eve of my first day of Christmas break, my parents got a surprise visit from my bus driver complaining about me. At this point, you're probably wondering what these two events have to do with

one another and, even more poignantly, the topic at hand.

For starters, a few days prior to my bus driver's ill-timed Christmas visit, some kids on the bus were playing with a radio-controlled plane and, as kids often do, were teasing me by keeping it from me. After begging and pleading with them to let me play with it, I yanked it out of one of the kid's hands while the propeller was spinning and it accidentally got badly knotted into the hair of a girl sitting across the aisle. When she burst into tears and began screaming, the bus driver flipped out, stopped the bus, and yelled at me—even going as far as threatening to kick me off the bus. After he finally extracted the propeller from the girl's hair by using another kid's scissors, which unfortu-

“The seeds of pain have always inevitably sprouted anger within me if I do not take effective action to prevent it.”



MARK TREMBLAY

nately left a gaping bald spot on the back of her head, he leveled another threat at me, which barely registered at the time: "I'm going to pay your parents a visit this Christmas to let them know what a monster you are." I did not try to talk back because the incident had been resolved and I never imagined he would follow through.

Later, his ill-timed visit did ruin my Christmas break and, worst of all, my parents returned all my Christmas presents as punishment. When that dreadful Christmas break was finally over, I was bitter due to what my bus driver did and my whole body ached from what my dad did, so I was completely miserable and my mind kept slipping into darkness, involuntarily contemplating ways I could exact revenge on my bus driver.

I've always known that whenever anyone hurts you, if you're patient and act at the exact right moment, you'll have the opportunity to exact vengeance, which is why I also fervently believe the key to not acting in anger is letting go of the hurt swiftly and forgiving the offending party quickly. However, in the throes of my rib pain, when that perfect moment arrived, I did not hesitate and let loose two simple words, "it's clear," that I had no doubt would exact the vengeance I sought. Several factors contributed to maximize the impact of those words at the exact moment I uttered them. The bus driver's regular short bus, which had minimal blind spots, was in the shop and a senior, Jimmy, who liked me and took me under his wing, cajoled the bus driver into letting me sit in the back with him while he spotted for the bus driver, who needed help with blind spots while backing up to turn the bus around at the top of the Jewitte Hill, where Apalachin's upper crust lived with their fancy extra cars occasionally parked on the streets. Unfortunately for my bus driver, I

quickly responded, "it's clear," when he asked Jimmy to look out the window—just as we began rolling over the hood of Dr. Rosen's 1965 Morris Garages (MG) Roadster. This sounded remarkably like a bus rolling over a frozen snowbank. Jimmy and I shot each other a knowing look that seemed to acknowledge instantly that neither of us ever saw the little sportscar now deeply wedged under the bus. Needless to say, our bus ride literally came to a screeching halt that day until a rescue bus came to take us the rest of the way to school, and our bus driver never drove that route again.

In retrospect, I do not believe that I would have been able to sustain my anger for as long as I did, plot my revenge so thoroughly, or act in as timely a manner if I had not been in such intense pain, which made it hard to focus on positive things and made it even more difficult to move past my anger. I am thoroughly convinced that this angry act, which was admittedly devastating to many undeserving people, was as much a reaction to my physical pain as it was to my anger at any one individual.

As I mentioned earlier, the frequency and intensity of my chronic pain has once again escalated, which has made me increasingly irritable and has made it difficult to stay focused on positive things. I am so concerned that the seeds of pain may once again sprout anger, that I enrolled in an online anger management class for adjudicated (convicted and sentenced) clients. I found through experience that when traditional counseling does not work to curb anger, anger management courses, particularly those that meet court-mandated criteria, can be extremely helpful. In fact, when I was a young psychologist in my mid-20s, I used to teach court-mandated anger management courses at the drug and alcohol treatment center where I

worked, so I have first-hand knowledge of how effective they can be when counseling doesn't seem to work.

During my six years teaching anger management and counseling adjudicated clients, I came across one dynamic in particular that I have always found deeply relatable. Aptly named the "victim perpetrator paradigm," it is a concept I believe some CF patients may find relatable as well. This occurs when a former victim, most often a severely traumatized one, becomes the perpetrator who traumatizes another person (but not necessarily the person who victimized them) with the intent of inducing in them the level of hurt they themselves felt when victimized and, in many cases, continues to feel.

If I want to prevent my pain from sprouting anger, I have to take proactive measures to prevent it from growing into chronic bitterness and rage, which triggers a cycle of isolation and even more bitterness. However, if I do take proactive steps to nip my anger in the bud by initiating counseling, enrolling in an anger management class, and/or sharing openly and transparently about my feelings early on, I can mitigate harm to my loved ones, myself, and at times the community at large. ▲

Mark Tremblay is 53 years old and has CF. He lives in Troy, NY, with his wife, MaryGrace. He has a Master of Arts in Psychology from Marywood University and a Master of Public Administration from Syracuse University. Mark worked in the New York Governor's Division of Budget for six years and 22 years as a Bureau Director for New York State Department of Health. He is the President of "CF Vests for Life," which collects donated therapy vests, nebulizers, and oxygen saturators for distribution to CF patients around the world. Additionally, he is the leader of the Attain Health group, "CF Warriors for Recovery and Freedom." Mark is also a director of USACFA. His contact information is on page 2.



TRANSPLANT TALKBACK

My Conversations with Jerry— The Triple Threat!

By *Andrea Eisenman*

This interview is intended for all individuals with CF, but especially for those who might need a new liver and kidney. Lifesaving transplants of both a liver and a kidney at the same time can be successful—never give up.

Jerry Cahill has been a friend for many years. He's been running marathons and biking centuries for as long as I've known him. He pushes hard to excel and, as he likes to put it, "I am relentless." He knows how to train his body and mind to accomplish his goals.

I was already transplanted with two lungs by the time I met Jerry. It became evident he would need a bilateral lung transplant, too. I tried to share as much about my transplant experience as I could to help him understand what to anticipate with the surgery and what recovery might be like. Everyone is different—even though we all have CF, there are so many variables that come into play.

Jerry proved to be a responsible and compliant owner of his newly transplanted lungs. So much so, I knew that when he needed to have a liver and a kidney transplant at the same time, he would be seen as a good candidate. But there are always other obstacles and I am not on the surgical committee. Because there is a severe shortage of organs, he was possibly facing a long wait while feeling sick.

I saw Jerry a few times prior to his recent surgeries and what I saw shocked me. My athletically active friend was wasting away in front of me and was the color of yellowing leather. Would he be able to survive the long wait for these organs? I was not sure he could withstand the rigors of two surgeries possibly taking 16 hours in total. I thought, I have to have faith and that if anyone

could do it, it would be Jerry.

How were you feeling during this time?

About two years ago, I started to get a lot of itching and started ripping at my skin. I had marks all over from scratching. I was constantly fatigued and didn't feel right. I was already seeing a liver transplant doctor, recommended by my lung transplant team, because I had abdominal swelling from my liver. The liver team did an ultrasound of my liver and they found I had a clot in the portal going into my liver. On top of starting daily Lovenox injections to break up the clot, I still had a lot of swelling in my body. The injections didn't work so I was given oral anti-coagulants. However, the bleeding got worse and oral medications were causing major nose bleeds. My fatigue worsened. The portal clot was not dissolving and I kept swelling. The hepatologist eventually drained my abdomen to combat the fluid buildup and swelling. All of my existing symptoms worsened when I was in a car accident. I was rushed to a local hospital

in Westchester. The doctors saw I was bleeding internally so they quickly transferred me to New York Presbyterian (formerly Columbia) Hospital. While in the hospital, I then met with the liver transplant surgeon who, after reviewing my CT scan, said I had to have surgery to try to stop the internal bleeding. After a week in the hospital recovering from the first surgery, I had more abdominal pain and was brought back into surgery again to stem the bleeding. Unfortunately, they could not fix the portal clot as it was too risky. They felt I could have bled out.

Before being discharged from the hospital, the oral blood thinner was adjusted. But, if I scratched my skin, I bled. Things were getting worse. I was still itchy and I was getting confused and not able to remember things. The liver team told me I would eventually need a liver transplant.

What was your quality of life at this point?

Thankfully my lung function was very high. And because I never think I am that sick, I was doing a planned bike tour to raise awareness for CF and transplant called, "Alive at 65." The irony was that I was barely alive. Just before the event, I was retaining a lot of fluid. And I could barely bike and barely get into the van after biking. The support team wanted me to stop but I would not stop—I had to finish it. I called my doctors at New York Presbyterian and they wanted me to come in and get diuretics.

It was at this point, even though I didn't realize it at the time, that my kidneys were failing, hence the extreme fluid retention and edema. Once I finished the bike tour or, rather, it almost finished me—my kidneys were not filtering out toxins, my creatinine was high, and my



JERRY CAHILL

energy was low—I was admitted to the hospital. I was immediately moved to a different floor to undergo dialysis and, at this stage, my memory is spotty. At one point I woke up in the ICU and was receiving dialysis 24/7. I only have a vague memory of this time. I was blacking out a lot. Without a functioning liver or kidneys, my body was becoming toxic. When I was awake and aware after a few days, I was told I would be needing both a liver and kidney transplant. I had three transplant teams visiting me in the hospital—my usual lung team plus the addition of liver and kidney teams. Both organs had to come from the same donor due to antibodies matching.

How did you take this news?

It was not a shock to hear I might need a new liver and then a kidney. I think I am strange. I never think I am that sick so I push through and want to get things done. I barrel through to get past the unpleasantness of waiting—I just want it over.

When they told me I would need both organs and the surgery could last 16 hours, I told them that it was ridiculous, and they were looking at me like I was crazy. They said, “Mr. Cahill, I don’t think you understand how sick you are.” I replied, “It can’t be that bad. What is my MELD score?” For those that don’t know, a MELD score (which stands for Model for End-Stage Liver Disease) ranges from six to 40, with the higher numbers indicating a more urgent need for a liver transplant. They told me my score and it was high. They said without these two transplants, I would not be here anymore. I was told that I would not be going home without a kidney and a liver. But because I was running a mild fever, the kidney doctors would not transplant me until I no longer had one. After 14 days, they wanted to wait six more days to be sure before activating me on the organ donor list. I was already listed for the liver. It then became a waiting game.

I was very out of it and it was all during COVID. There were only two visitors allowed each day. I was finally out of the ICU and on the transplant floor, receiving dialysis daily through a machine next to my bed. I had already been in the hospital for six weeks. And I was told it could be a two-to-three-month wait for both organs.

I was finally activated (by liver and kidney teams) and was put higher on the list due to needing both organs. I’m claustrophobic in small places like the MRI tube so I worried and wondered—how would I get through this, being in a small room for months? I figured I would have to make the best of it and try to build myself back up the way I did for lung transplant. But... three hours later...a doctor from the liver team came in—she said they had found a donor. I was baffled; unsure of what they meant. The attending explained that the surgeries were happening today! They came to get me at night and it was dark while I waited by myself with no visitors allowed.

Stuff got real. What were you feeling then?

At this point, I didn’t think I would survive as I was not in good shape. How would I make it through a 16-hour surgery? Even before I was listed, I was told lung transplant recipients are not good candidates for a liver transplant. They didn’t want to list me, but the liver surgeon who knew me said that they do not normally do it and I was at risk due to already being a transplant recipient. He also said, “But because of your mental state and your willpower, you are so unbelievable we are gonna give it a shot.”

They wheeled me in and put the liver in first, then they checked to make it sure it was functioning properly, before the kidney team came and put in a kidney (but did not remove my own and so I have three!). I was off intubation after only four hours.

Once you knew you were alive, what was going through your mind?

After surgery, after ICU, and on the transplant floor, a doctor asked me how I was and I said, “I’m ok.” And I added, “I didn’t think I would get through it.” The doctor confided, “Honestly, none of us thought you would!” When I could get out of bed, I started walking a lot in the hospital. I have learned in life to never obsess over things. I just try to get them done. I thought back and knew I had a good life and did a lot of things. I just try to keep reinventing myself and staying positive. My coping mechanisms are all centered around reinventing myself.

Each time I had to stop doing something, I tried to find other ways to move through life. When I could no longer work, I volunteered. When I could no longer run due to my knees, I started biking. Once that became too hard, I started hiking and walking. Any exercise is good for one’s mind and body. So I do what I can to stay healthy. I feel I am good at pivoting. I began walking in the hospital to build my strength back and now I am walking to raise money for BEF’s transplant fund.

What’s going on now?

I am now 10 months post-liver and kidney transplants. I am not where I want to be. I look fine and feel good, but my strength is not where I had envisioned it. I am now 66 and grateful to still be alive. But it’s a long haul. I am doing freelance work to survive. Living with CF never goes away. I have two to three appointments each week for different transplant teams. That’s a fulltime job in itself!

It’s not over, the journey with Jerry continues... ▲

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



MILESTONES

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

ANNIVERSARIES

Birthday

Jeanie Hanley
Los Angeles, CA
60 years on June 12, 2022

Mark Tremblay
Troy, NY
53 years on June 6, 2022

Transplant
Tanya Cunningham
Sandy, OR
5 years on May 14, 2022

TILLMAN continued from page 29

drop in FEV1 of 10% or more from baseline and/or a total score of the CFRSD-CRISS worsening by more than 10 points automatically triggered an alert to the participant’s care team. The team would then discuss the symptoms with the patient and decide on the best approach. Secondary outcome goals of the trial included assessments of antibiotic requirements, measured by the number of days on oral and intravenous antibiotics, and the identification of protocol-defined pulmonary exacerbations. This was described as the presence of four or more specific criteria, including a change in sputum, increased coughing, increased shortness of breath, fatigue, and fever, and changes in lung function. The mean number of hospital inpatient days was similar for patients in the home monitoring intervention group and for those receiving standard care. To determine HRQoL, the team used the Cystic Fibrosis Questionnaire-Revised (CFQ-R). No differences in HRQoL were found between the two groups. Protocol-defined pulmonary exacerbations were detected more often in home-monitored patients during the study period. Participants in the home-monitored group also received oral antibiotics for protocol-defined pulmonary exacerbations for longer compared with patients receiving routine care. However,

no differences in FEV1 over the 12-month study period were found. A qualitative analysis showed that most patients felt that home monitoring was beneficial. The findings of this trial confirm that home monitoring is effective in detecting pulmonary exacerbations in adults with CF.

<https://tinyurl.com/547rbm83>

Method To Detect Cystic Fibrosis Infection Within Minutes

The present methods for diagnosing acute and chronic infections are complex and time-A multi-disciplinary team set out to develop a diagnostic tool that would be rapid, accurate and simple-to-use for doctors. The multi-excitation Raman spectroscopy is the analysis technique produced; it’s a non-invasive method that emits a scattering of multiple colours of light into a patient’s sample. When light is applied to a sample’s molecules they can vibrate which helps researchers understand their characteristics. By using different colours of light, a different set of such vibrations can be triggered, meaning more information can be obtained about their composition than previously possible. This then allows ‘finger-printing’ that can be used to identify the properties of the pathogens. In many current techniques, a reagent needs to be added to a

sample or a tag needs to be attached to the molecules of interest to analyse their composition. This is not required under this new approach which uses natural properties of the molecules to analyse them. This new Raman spectroscopy based method offers many advantages over resource-intensive, culture-based methods, allowing rapid and label-free analysis. It avoids complex sample-preparation steps with sophisticated equipment.

<https://tinyurl.com/yzxtune9>

AND

<https://tinyurl.com/5n6jptmf>

AND

<https://tinyurl.com/yburjx49>

AND

<https://tinyurl.com/2h77fva5>

Effectiveness Of Antibiotics Significantly Reduced When Multiple Bugs Present

In the study researchers say that even a low level of one type of microbe in the airways can have a profound effect on the way other microbes respond to antibiotics. The results highlight the need to consider the interaction between different species of microbe when treating infections with antibiotics-and to adjust dosage accordingly. Chronic bacterial infections such as those in the human airways are very

Meet Our New Treasurer: Colleen Adamson

I am 53 years old and was diagnosed when I was 13 months old. My mom diagnosed me after looking up my symptoms in Dr. Spock's book! I had all the symptoms of CF: failure to thrive, sick all the time, salty skin, etc. Despite having all the classic symptoms, my doctors concluded that I did not have CF because we had no family history of it, but my mom insisted I be tested. As it turns out, I did have relatives on both sides of the family who died of lung issues, but these issues were not called CF at the time.

I had a bilateral lung transplant on July 3rd, 1998 and a living kidney transplant from my friend Kelly on March 7th, 2006. Both transplants were done at Fairfax Hospital in Falls Church, Virginia. I am very thankful for my two donors!

I worked full time for the U.S. Government as an Operations Research Analyst, developing cost estimates for major government systems such as ships



COLLEEN ADAMSON

and aircraft. I retired on disability a few years ago because I was having issues with my speech and memory. I was having a very hard time remembering numbers and talking about them, which is kind of important when developing and briefing cost estimates! Now I am glad

to not have the job stress anymore and grateful to have time to take better care of myself.

I spend my time volunteering for Fairfax Hospital and for my church, although I did take a break from in-person activities during the pandemic and I have not been back yet per my doctor's orders because the pandemic is still ongoing. I am a Fairfax Hospital Lung Transplant Center Mentor, speaking to pre- and post-transplant patients about my experiences with lung transplantation. I am also a Donate Life Ambassador for the Washington Regional Transplant Community (WRTC), promoting organ, eye, and tissue donation so that others may have the gift of life that I have been given. I am married to Scott Adamson and we will celebrate our 25th anniversary in June 2022. We have a furbaby named Penny; she is a Miniature Schnauzer and brings us joy every day. ▲

difficult to cure using antibiotics. Although these types of infection are often associated with a single pathogenic species, the infection site is frequently co-colonised by a number of other microbes, most of which are not usually pathogenic in their own right. Treatment options usually revolve around targeting the pathogen, and take little account of the co-habiting species. However, these treatments often fail to resolve the infection. The model allowed them to grow a mixture of different microbes, including pathogens, in a stable way for weeks at a time. This is novel, because usually one pathogen will outgrow the others very quickly and spoil the experiment. It enabled the researchers to replicate and study infections with multiple species of microbe,

called 'poly-microbial infections', in the laboratory. The three microbes used in the experiment were the bacteria *Pseudomonas aeruginosa* and *Staphylococcus aureus*, and the fungus *Candida albicans*. The researchers treated this microbial mix with colistin, which is very effective in killing *Pseudomonas aeruginosa*. But when the other pathogens were present alongside *Pseudomonas aeruginosa*, the antibiotic didn't work. The same effect happened when the microbial mix was treated with fusidic acid - an antibiotic that specifically targets *Staphylococcus aureus*, and with fluconazole - an antibiotic that specifically targets *Candida albicans*. The researchers found that significantly higher doses of each antibiotic were needed to kill bacteria when it was part of poly-microbial infection,

compared to when no other pathogens were present. At present antibiotics are usually only laboratory tested against the main pathogen they are designed to target, to determine the lowest effective dose. But when the same dose is used to treat infection in a person it often doesn't work, and this study helps to explain why.

<https://tinyurl.com/4c5v2nte>

AND

<https://tinyurl.com/ytaaytet>

AND

<https://tinyurl.com/2p8c2wph>

Bacteria Boosts Anti-fungal Medicine's Killing Power In CF Study

Co-infection with the bacterium *Pseudomonas aeruginosa* can increase the

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New Jersey State Organization Of Cystic Fibrosis Grant Opportunity

<https://www.njsocf.org/>

Pharmaceutical Services For Adults With Cystic Fibrosis

Since 1990, the New Jersey State Organization of Cystic Fibrosis has administered a special state-funded program, "Pharmaceutical Services for Adults with Cystic Fibrosis."

As more and more individuals with CF reach adulthood, they face increasing financial burdens and difficulty financing the high costs of living with cystic fibrosis. In response to the increasing needs of adults with CF, NJSOCF developed a plan for an adult program, now funded by the State of New Jersey.

Services are available to New Jersey residents with CF, age 18 and over, whose incomes meet state eligibility requirements. Direct service components of the program include assistance with paying for prescription medication; office visits and diagnostic copayments; home IV out-of-pocket expenses; medical equipment and supplies; nutritional supplements; and extra nutritious food. The program will also pay a health insurance deductible. Adults from every county in New Jersey are currently enrolled in the adult program and new applicants are always welcome to apply.

1. Diagnosis of Cystic Fibrosis verified by CF doctor
2. Must be a New Jersey resident
3. Must be 18 years or older
4. Individual annual income less than \$51,952/year

Fill out an application here: <https://www.njsocf.org/adult-program/>
Or you can also e-mail them at das@njsocf.org or dtimr@njsocf.org

Once you are accepted into the grant the following will be covered:

- Copayments for prescription medications related to the treatment of cystic fibrosis
- Copayments for office visits related to the treatment of cystic fibrosis
- Copayments for diagnostic testing, i.e., routine lab work, scans, x-rays and sputum cultures, related to the treatment of cystic fibrosis
- Out-of-pocket expenses for home IV antibiotics
- Copayments for respiratory equipment and supplies
- Insurance deductibles up to a maximum of \$1,000 per year

You will also receive \$125 each month for food and nutritional supplements. Here are the requirements:

- Shopping should be done every month for approved foods and/or nutritional supplements
- You must send back your itemized grocery receipt(s) each month in the postage paid envelope provided. You will not get another card until a receipt is received.

Please note that only nutritious and non-taxable food items should be purchased. If for some reason you cannot shop in any one month, please call or email us.

EXAMPLES OF UNACCEPTABLE PURCHASES

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Coffee and regular tea
Spices and condiments
All candy

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Laundry products
Cleaning products
Grooming and cosmetic aides

EXAMPLES OF ACCEPTABLE NUTRITIONAL SUPPLEMENTS

Vitamins
Boost
Ensure products
Carnation Instant Breakfast
Glucerna
Scandishake
Power bars and equivalents
Weight gain supplements



potency of therapies that kill *Candida albicans*, an infectious fungus, according to a new study. Little is known about how co-occurring infections affect the response to treatment. Scientists conducted tests using fluconazole (FLC), an anti-fungal medication that is commonly used to manage *Candida* infections. In lab dish experiments, the researchers used FLC to treat *Candida* fungi, either alone or in the presence of *Pseudomonas*. When *Candida* was grown with *Pseudomonas* in the absence of treatment, the bacteria had little to no effect on the fungus' growth. Treatment of *Candida* alone with FLC was fungistatic, but not fungicidal — that is, it stopped the fungus from growing, but did not kill it. However, when FLC treatment was given in the presence of *Pseudomonas*, the medication powerfully killed the fungus. Thus, fungal-bacterial interactions can drive an unexpected enhancement in anti-fungal susceptibility during treatment of infection. The fungicidal effect of FLC during co-infection suggests that *Pseudomonas* blocks *C. albicans* tolerance to FLC, leading to death rather than persistence or slow growth during treatment.

Research showed this effect may be driven in part by the two microbes competing for iron, a nutrient that both *Pseudomonas* and *Candida* require to survive. Supplementing the microorganisms with iron reduced, but did not eliminate, *Pseudomonas* and FLC's synergistic anti-fungal effect. This work demonstrates that polymicrobial interactions can indeed affect treatment efficacy and, most importantly, it highlights the importance of nutrient availability in the environment — such as iron — and how it modulates treatment efficacy.

<https://tinyurl.com/ymrz8cb8>

AND

<https://tinyurl.com/2p95vzjy>

AND

<https://tinyurl.com/mryfryyv>

Molecules Found In Mucus Can Thwart Fungal Infection

Researchers have now identified

components of mucus that can interact with *Candida albicans* and prevent it from causing infection. These molecules, known as glycans, are a major constituent of mucins, the gel-forming polymers that make up mucus. Mucins contain many different glycans, which are complex sugar molecules. A growing body of research suggests that glycans can be specialized to help tame specific pathogens — not only *Candida albicans* but also other pathogens such as *Pseudomonas aeruginosa* and *Staphylococcus aureus*. Within the mucus that lines much of the body, there are densely packed communities of different microbes, many beneficial but some harmful. This study, combined with previous work on *Pseudomonas aeruginosa* and ongoing studies of *Staphylococcus aureus* and *Vibrio cholerae*, suggests that different glycans are specialized to disable different kinds of microbes. Taking advantage of these mucins could help researchers design new antifungal medicines, or make disease-causing fungus more susceptible to existing drugs.

<https://tinyurl.com/297wnwrs>

A New Perspective On Opportunistic Pathogens Of The Genus *Bordetella* In Cystic Fibrosis

The *Bordetella* genus, closely related to the genus *Achromobacter* including the emerging pathogen *Achromobacter xylosoxidans*, has not been studied much in Cystic Fibrosis. The current literature suggests that *Bordetella* spp. are able to colonize and persist in CF patient's airways and a few studies have correlated pulmonary exacerbations with their presence. Others have shown *Bordetella* spp. to exhibit virulence-associated traits compatible with a potential contribution to CF clinical pathology using CF mice models. Regarding sources of infection, host-adapted species are thought to be acquired through zoonotic transmission but some host-associated species have the potential to survive and grow in the environment and were shown to be transmit-

ted from amoeba to mice, suggesting that the natural environments could be transient reservoirs for dissemination.

<https://tinyurl.com/2p8b4fe8>

Serological Biomarkers For The Diagnosis Of *Mycobacterium Abscessus* Infections In Cystic Fibrosis Patients

Culture conditions sometimes make it difficult to detect non-tuberculous mycobacteria (NTM), particularly *Mycobacterium abscessus*, an emerging cystic fibrosis pathogen. The diagnosis of NTM positive cases not detected by classical culture methods might benefit from the development of a serological assay. High antibody titers against two specific antigens were obtained in *M. abscessus*-culture positive CF-patients, allowing scientists to consider these serological markers as potential tools in the detection of CF-patients infected with *M. abscessus*.

<https://tinyurl.com/2dk5ybpp>

Changes In Airway Metabolites May Predict Likelihood Of NTM Infection

Cystic fibrosis (CF) patients with non-tuberculous mycobacteria (NTM) lung infection show significant changes in airway metabolites relative to those without such infections, a study shows. Metabolites are intermediate or end products of cellular metabolism, and some of these altered metabolites play roles in immune responses and bacterial growth. Combining metabolic and microbiota changes also allowed for an accurate distinction between patients with and without NTM infection, further suggesting that these may be risk factors of NTM infection in people with CF. Airway microbiota comprise the community of bacteria, fungi, and viruses that colonizes the lungs. NTM are increasingly being recognized as common and worrisome infectious agents in people with CF, as they cause infections that are challenging to treat, associated with a poorer prognosis, and often become chronic. A research team evalu-

Continued on page 38

ated whether airway metabolites and microbiota – reflecting the lung environment – could potentially represent risk factors for NTM infection in people with CF. The cause of nearly 60% of NTM cases was *Mycobacterium avium* complex, followed by *Mycobacterium abscessus* complex; both being the most common NTM types. The NTM cases and NTM-negative controls did not significantly differ in most clinical characteristics. Several of the 902 total metabolites detected in these patients significantly differed between NTM cases and controls, the researchers found. Notably, some of these alterations were present both before and after NTM infection, suggesting that they may represent risk factors of such infection. Their findings highlight significant differences in metabolic patterns between CF patients with and without NTM infection, including metabolites that play important roles in the host immune response and in bacterial growth. Metabolites showing differences even before the infection may represent risk factors and therapeutic targets for preventing and/or treating NTM infections in people with CF.

<https://tinyurl.com/eu6un7eh>

Breath Biomarkers Show Potential For Identifying NTM Lung Infections

Several potential biomarkers of nontuberculosis mycobacteria (NTM)-associated lung disease were identified in the breath of people with cystic fibrosis. This approach may offer a faster way of reaching an NTM diagnosis, which has historically relied on slow bacterial culture techniques. Breath holds great potential as a source of information on an individual's health status. In the breath collection procedure, patients were asked to breathe into a bag through a drinking straw mouthpiece. The air in the bag was pulled through a filter using vacuum into a tube that can absorb molecules from the breath sample. While the exact chemical identity of the biomarkers could not be definitively

confirmed in this study, several molecules had likely been previously identified in breath samples from cows, monkeys, and humans with mycobacterial infections. Overall, the findings suggest that the biomarkers may be breath signatures of mycobacteria infections generally and may not be specific to NTM, the researchers noted. They added that while the results suggest the breath test may be a “promising” way of identifying NTM-associated lung disease, a study involving more patients and the use of authentic chemical standards or high resolution analytical tools to absolutely confirm biomarker identity is necessary. <https://tinyurl.com/4cz54w7p>

Unprecedented Case Series Advances Promise Of Phage Therapy

An international team of researchers report promising results from the largest case series yet of patients treated with bacteriophage therapy for antibiotic-resistant infections. Non-tuberculosis *Mycobacterium* (NTM) infections are increasingly common among patients with cystic fibrosis. Treatment of NTM infections, particularly those caused by *Mycobacterium abscessus*, are difficult due to growing bacterial resistance to antibiotics. Bacteriophages are viruses that have evolved to target and destroy specific bacterial species or strains. Each phage species seeks and destroys only one bacterial species and the current armamentarium of known therapeutically useful phages is relatively small. As a result, phage therapy testing is currently constrained to experimental treatments where all other viable alternatives are failing or have failed. Phages were administered to the study participants intravenously, by aerosolization through a nebulizer or by using both methods twice daily over an average course of six months, though some patients had shorter or longer treatments based on clinical or microbiologic response. Patients were monitored for adverse effects, signs of symptomatic improve-

ment or reduced bacterial presence, emergence of phage resistance and/or neutralization of phages by the patients' immune systems. The authors reported no adverse reactions to phage therapy in any of the patients, regardless of type of bacterial infection, types of phages used or method of treatment. The team determined that phage treatment of mycobacterial infections shows promise, should be explored further, and provided several insights into how therapeutic phages might be effectively used.

<https://tinyurl.com/bdz3jhjt>

The Effect Of Antibiotic Changes During Treatment Of Cystic Fibrosis Pulmonary Exacerbations

Antibiotics are often changed during treatment of pulmonary exacerbations (PEx) in people with cystic fibrosis who have a poor clinical response. The authors aimed to describe the reasons CF providers change antibiotics and examined the effects of antibiotic changes on lung function recovery. The co-primary outcome measure was absolute and relative change in forced expiratory lung volume in 1 s (FEV1) at the end of treatment and follow-up. Secondary outcome assessed the proportion of patients returning to > 90% or > 100% previous baseline FEV1. Reasons for antibiotic changes included change in antibiotic route prior to discharge, drug reactions, poor FEV1 response, targeting additional microbes and lack of symptom improvement. In the researchers analysis, among non-responders, a change in antibiotics was not associated with any significant difference in absolute or relative FEV1 at the end of treatment or at follow-up. Antibiotic change in non-responders was not associated with improved return to 90% or 100% baseline FEV1 at end of treatment or follow-up. The researchers concluded that changing antibiotics during CF PEx treatment in those with poor clinical response was not associated with any improved FEV1 response or return to baseline lung function.

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Ivacaftor Withdrawal Syndrome: A Potentially Life-Threatening Consequence From A Life-Saving Medication

In 2018, a case series of three adult patients with cystic fibrosis suggested an ivacaftor withdrawal syndrome (IWS)

perpetuated by abrupt cessation of ivacaftor treatment. An additional case published that year proposed IWS in the setting of rifampin use, demonstrating this may also be induced by drug-drug interactions. While this syndrome is a rare complication of highly effective modulator therapy (HEMT), it is impor-

tant to recognize as increasing numbers of patients are prescribed HEMT. <https://tinyurl.com/yn5pc4rs> ▲

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

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- Share your ideas for **Focus Topics**, feature articles or any suggestions for improvements you may have to help make *CF Roundtable* more relevant and interesting to you.
- You can reach **USACFA** and *CF Roundtable* at any time by email at cfroundtable@usacfa.org
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IMPORTANT RESOURCES

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

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

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

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

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