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

Living And Aging Well With CF

By Edward Canda, Ph.D.

I am a 67-year-old living with CF. I retired two years ago after a 33-year career as a social work professor and I continue activity as a professor emeritus at the University of Kansas. I would like to share several stories about life phases in adulthood and lessons learned that support living and aging well with CF. I don't assume that my perspective will fit for others with CF. My perspective is shaped by a life and career honoring many spiritual traditions and philosophies and applying their wisdom to health and service.

EDWARD CANDA

Diagnosis in Early Adulthood

My childhood was marked by frequent colds, severe seasonal allergies, nasal polyps, and low weight. My older brother Tom had a similar medical picture, though more severe. Our family pediatrician, and later respiratory specialist, did not correctly diagnose our condition. This was a frustrating time of mistreatments, harmful side effects, and worsening lung infections.

When my brother moved out of our parents' home he went to see a free clinic doctor. Thankfully, this doctor referred him to a CF clinic for evaluation, which led to both of us receiving a correct diagnosis. Thus, at age 18, I embarked on treatments with our outstanding physician, Dr. Robert Stern of University Hospitals in Cleveland. He remained my physician for about 45 years until his retirement.

Initially, the diagnosis was shocking. I was relieved because our strange array of symptoms finally made sense and I knew there were treatments available. But this also threw my life expectations askew. I was trying to figure out who I wanted to be and how I wanted to live as I was entering adulthood just when I found out that I might not survive for long. Dr. Stern never claimed certainties about life expectancy or pushed limitations on life goals. He was honest and frank about the clinical outlook for CF, but he always individualized care. Rather than telling me what I could not do, he helped me figure out how medical care could help me accomplish what I wanted to do.

I generally had a wonderful time during my undergraduate years living on campus, full of adventures and intellectual growth. I decided to make the most of whatever time I had to live, while not being stuck in any life-limiting assumptions. Still, I had many quandaries about prospects for career, marriage,

Continued on page 6

INSIDE THIS ISSUE

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EDITOR'S NOTES

March, April, and May bring quite a few celebrations: March highlights Rare Disease day, April is National Donate Life month, and May is CF awareness month. I know I speak for quite a few of us adults with CF when I say that all three of these are close to our hearts every year. And, with the advent of modulators and watching more and more people with cystic fibrosis live longer and fuller lives, it seems fitting that our focus topic this issue tackles aging with CF.

Personally, I can't honestly say I contemplated life past 40. In my article this issue, I share about my myriad health complications—some related to CF and others are simply part of aging generally and my genetics. Most of these health challenges were never on my radar as something I need to handle or worry about in the future. This issue, former President and USACFA founder, **Kathy Russell**, writes about the many ways all of us with cystic fibrosis have endured, especially as we age. **Sonya Ostensen** tackles her most challenging issues with aging alongside CF—menopause and impaired vision. Long-time reader **Kim Nunnari** writes about her ongoing challenges as she ages with CF, despite being a late-diagnosis patient. **Jennifer Kyle** discusses the important role that both exercise and better nutrition have played in her own health, both before her CF diagnosis and as she has aged. Also in the focus topic this issue, **Andrea Eisenman** sheds light on the delicate balance required in managing all of her care, especially post-transplant.

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, **Alex Gantt** opens up about her CF diagnosis after years of keeping it a secret. **Dr. Nowakowski** writes about the seemingly endless hamster wheel of loss and adjustment when it comes to cystic fibrosis in addition to their oral health struggles, which are compounded by CF. As a follow-up to the winter 2022 issue, **Beth Sufian**, in our "Ask the Attorney" column, details the Trial Work Period for SSDI recipients in part II of her III-part series of articles regarding working while receiving social security disability benefits. **Isabel Stenzel Byrnes** writes about her long view of life, especially now that she has reached 50, a birthday she coins as "the new 70!" **Mark Tremblay**, in his column, talks about his lifelong battle to age all the while listening to peers around him lament aging. Spoiler alert! It repulses him.

In our "Voices from the Roundtable" section, **Faith Ettlich** and **Rowan O'Bryan** weigh in on how our Arts scholarship has helped them in their studies over the past year. And, in other noteworthy news, USACFA is very excited to welcome our newest director, **Ela Castillo**. Ela brings a passion for advocacy and strives for representation of all groups, including the underrepresented groups within the CF community.

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

A Year In Review: Real World Evidence, Functional Monitoring And Emerging Therapeutics In 2021

This is a comprehensive review of: 1) infection with SARS-CoV-2, 2) impact of COVID-19 on clinical care, 3) CFTR modulators, 4) extrapulmonary complications, 5) diagnosis, 6) early lung disease, 7) pulmonary exacerbations, 8) microbiology, and 9) novel therapies. While lengthy, it is well worth reading.
https://tinyurl.com/mu7r62es

CF Patients Satisfied With Care, But Not Life Quality, US Survey Finds

Most people with cystic fibrosis responding to a U.S.-based online survey were generally satisfied with their treatment plan and were not considering changing their medication.

LAURA TILLMAN

Shortness of breath was the most commonly reported symptom among CF patients, as well as the one that had the greatest impact on their daily life. Despite overall treatment satisfaction, nearly a third of survey respondents were dissatisfied with their quality of life, suggesting that more should be done in terms of symptom management and patient support/care. Shortness of breath also was most commonly reported as having the greatest impact on daily activities, followed by diabetes, persistent coughing, frequent lung/sinus infections, and damaged airways. Nearly all of the CF patients surveyed reported seeing a pulmonologist, and more than three in five were seeing a general or primary care doctor. Others reported being seen by a respiratory therapist, an endocrinologist, an ear/nose and throat doctor, and a cardiologist. All patients were taking medications to treat CF. Patients were on the same treatment plan for a median of 25.5 years. The

Continued on page 12

LOOKING AHEAD

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

Spring (May) 2022: Aging and CF. (Current issue)

Summer (August) 2022: Dealing With Pain. What types of pain do you deal with regularly? What has helped you with pain management? How does pain affect your quality of life? What alternative therapies have helped manage your pain? What, if any, problems do you have with tolerance to medications or addiction and how are you dealing with it? What advice do you give to someone dealing with addiction to pain medications?

Autumn (November) 2022: Transitioning from Pediatric to Adult Care. What resources do you wish were available to help with the transition to adult care? What resources helped the most? What were some of the obstacles you encountered along the way? What was the biggest change between pediatric care and adult care?

Winter (February) 2022: Accessibility and CF Care.

ASK THE ATTORNEY

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

Part II of III: Trial Work Period For SSDI Recipients.

By Beth Sufian, J.D.

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

In the previous issue (Winter 2022) of *CF Roundtable*, the first part 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 (SSI) Benefits.

This second part addresses continued monthly cash benefits and continued Medicare coverage during a trial work period for those receiving Social Security Disability Insurance (SSDI). Before going further, it is important to understand that the rules that apply to SSDI and Medicare coverage ***do not*** apply to SSI and Medicaid benefits. Please take care not to confuse the two because 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) that appeared in the winter edition of *CF Roundtable*.

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) extended period of Medicare coverage. This article focuses on the trial work period. Some people do not know if they receive SSI or SSDI benefits. A person should make sure he or she knows their benefit program so that they are using the correct work program rules when considering the effect of work on their ability to receive benefits.

For an application for SSDI benefits to be approved, the claimant must show that, because of an impairment, he or she is incapable of work or substantial gainful activity. If circumstances change after benefits are approved and the beneficiary returns to work, the Social Security Administration (SSA) may terminate benefits. However, a beneficiary may not know if he or she can return to work without trying, and he or she may not want to try if attempting a return to work will cause them to lose his or her benefits.

BETH SUFIAN

The SSA allows a person receiving SSDI benefits to attempt a return to work without losing the monthly cash benefit or Medicare eligibility. This is called a Trial Work Period (TWP). During a trial work period, a beneficiary receiving Social Security disability benefits (based on his or her own earnings history) may test his or her ability to work and still be considered disabled by SSA. SSA generally will not consider work performed during the trial work period as showing that the disability has ended until after nine months of work activity.

1. What Months Count As Trial Work Months?

Generally, if an SSDI beneficiary earns less than $970 per month in 2022 and works less than 20 hours per week, the work activity is typically regarded as less than substantial by the SSA. Earning less than the allowable amount generally will not affect SSDI eligibility or trigger a trial work month, provided the beneficiary continues to be disabled under SSA rules. If an SSDI beneficiary earns more than the allowable amount (such as $970 in 2022) during a month while on benefits, that month will be regarded as a trial work month.

2. How Long Is A Trial Work Period?

The maximum TWP is nine months. The nine-month period is cumulative and SSA will count each month that a beneficiary earns over the

Amount Of Monthly Earnings That Trigger A Trial Work Period*	
Year	Monthly Earnings
2022	$970
2021	$940
2020	$910
2019	$880
2018	$850

*20 C.F.R. §404.1592

monthly allowable amount ($970 in 2022) as a trial work month. Each month that a beneficiary earns more than the allowable amount is counted as a trial work month, even if those months are not consecutive.

It is important to note that the counting of trial work months is automatic. If you make over the allowable amount during a month while on SSDI, that month will be counted as part of a trial work period. Also, it is important to note that the dollar amounts are gross monthly earnings before taxes and other withholdings. The SSA will not necessarily give notice to a beneficiary that a particular month is counted as a trial work month. SSDI beneficiaries who are working must be aware of their gross monthly earnings to avoid an inadvertent trial work month or an unanticipated termination of benefits.

3. Trial Work Periods Do Not Preclude Medical Reviews.

SSA can consider medical evidence that might demonstrate a medical recovery at any time. Therefore, it is possible for SSA to conduct a medical review and stop benefits due to a medical recovery before the end of a TWP.

4. What Happens At The End Of A TWP?

SSA may determine the work attempt successful without additional inquiry. At the end of a TWP, the attempted return to work will either be considered successful or unsuccessful. SSA may conduct an inquiry at the end of a TWP to determine if the beneficiary's work attempt was successful. However, SSA may believe that it already has sufficient information to assess the beneficiary's disability status without a further inquiry.

SSA may make an additional inquiry—medical or earnings and expenses. In the case of a medical inquiry, SSA may need additional information to determine if the work attempt was successful. SSA may conduct an inquiry regarding the beneficiary's disability status and it may collect information about the disabling condition, limitations in daily activities, and any medical improvement. This inquiry is to determine if the beneficiary's condition is still disabling under SSA rules. When SSA determines a beneficiary is no longer disabled, the person is no longer eligible to receive benefits and the benefits are terminated.

In the case of an earnings and expense inquiry, SSA may also make an inquiry to determine if it correctly calculated earned income during the TWP and if it correctly counted the number of trial work months. To do this, the SSA may request pay stubs or other records to confirm that the beneficiary's gross earned income was over the allowable monthly amount for each of the nine months of the TWP. To determine if the months were properly counted as a trial work month, the SSA may also inquire about any impairment-related work expenses (IRWE).

5. Impairment-Related Work Expenses.

SSA deducts IRWE from gross earned income to determine if the beneficiary's income was over the allowable amount in a month. IRWE are described in detail in the SSA regulations (20 C.F.R. §404.1576), but generally include payments made by the beneficiary for items or services that are needed to engage in work and are incurred because of the disability.

SSA will consider an expense to be an IRWE if the item or service: (1) enables the beneficiary to work; (2) is needed because of the disabling impairment; (3) is paid for by the beneficiary and not reimbursed by another payer (such as Medicare, Medicaid or a private insurance carrier); and (4) is a reasonable cost for the item or service.

While the range of expenses that count as IRWE is broad, some common examples of expenses incurred by a person with CF are durable medical equipment (such as nebulizers) and payments for medications and medical services. Even if most of a medical expense is covered by insurance, the copays and deducible should count as an IRWE if they are costs paid by the beneficiary, not reimbursed by another payer, and otherwise meet the criteria for an IRWE.

There are limits to IRWE and the expenses must be verified. However, the important point is that IRWE can offset gross income that is over the allowable amount and possibly result in the countable income being less than the allowable. Another important point is that a small amount of IRWE

Continued on page 7

and other long-term goals. I felt awkward as friends provided postural drainage therapy and observed my illness close-up. Sometimes I felt despair and anger at a combination of my personal fragility and the larger context of the Vietnam War and rampant racial and social injustices. I started exploring what I could do to help our society and world.

A Second Life

My major was cultural anthropology. I explored not only what varied cultures had to offer about the meaning and purpose of human existence but also various techniques of meditation, healing, and spiritual transformation. Serendipitously, in my junior year at college, my advisor offered me the opportunity to study in South Korea as a graduate fellow and Fulbright Scholar. Immediately after graduation, I flew there with large bags of oral antibiotics and preparation for self-administered respiratory clearance therapies. When first offered this opportunity, I was dubious that I could undertake such travel and international living. But Dr. Stern and I agreed that the only thing we knew for sure about my future was that if I did not try, then I would forever regret the missed opportunity.

For 14 months, I studied East Asian philosophies, religions, and arts. I often visited cultural events with a Korean woman whom I later married (Hwi-Ja). I found ways to live and work around many bouts of fatigue and illness.

During my first summer there, in 1976, I traveled with a friend to beautiful places for a few weeks. Near the end of our journey, I stood on a beach looking out at the spectacular scene of the East Sea. Suddenly, a high wave crashed onto shore and knocked me down. It pulled me out and twirled me in deep water. I struggled to call for help and was crushed down by waves many times. I felt my life was being pulled down a bottomless drain. Yet an inner voice assured me this was not the right time to die and an unexplainable energy kept me going. My friend alerted a marine, who happened to be nearby, to rescue me with an inflated tube. The marine dragged me to shore, pushed the water out of my body, and then laid me on a cot. Someone asked, "How does it feel to come back from hell?" I said, "Great! Amazing! Wonderful!" In that moment, I realized the utter joy and mystery of just being alive. The sound of a nearby radio, the bright sunlight, the faces of rescuers looking at me—every sensation was completely wonderful.

The next morning, I watched a brilliant sunrise over the ocean, red-orange ripples flowing at me and filling me with new life. It was not CF that almost killed me, but rather a totally unexpected calamity. About two weeks later, I was hospitalized for acute pancreatitis, to which I am susceptible. It was probably triggered by the trauma of drowning and resuscitation. Once I came through the nausea, delirium, and acute pain, I passed fully into a new phase of life.

Hwi-Ja and I came to the U.S. in 1977 and soon married. Over the next nine years, we formed a new life together by connecting our diverse cultural and spiritual backgrounds. Hwi-Ja has been a constant supporter for me in all things. I completed graduate studies, including a Ph.D. in social work, and took my first faculty position in 1986. We both retired in 2019. During that career, much of my work focused on Southeast Asian refugee resettlement, health resilience, disability and independent living services, and mental health recovery. My main goal was to prepare social workers to support people's efforts to survive and thrive through chronic illness, crises, and trauma, especially by drawing on diverse religious and nonreligious spiritual perspectives, while always respecting clients' own goals, beliefs, and interests. Hwi-Ja was a medical social worker for 30 years.

This goal has been central to my personal life as well—my brother died when he was 41. I felt devastated. We had a final meeting that affirmed our brotherly love and care. That helped me work through survivor's guilt and to make a commitment to survivor's responsibility—the responsibility to live life well and meaningfully and to help others. In our family life, we missed having no children. However, unexpectedly, about 12 years ago, we became "adopted" grandparents for the new baby of close friends, and later also for her younger sister.

In adulthood, I established a lifestyle that combines the best of medical care, consistent use of medications, self-administered respiratory clearance (such as the Flutter and autogenic drainage breathing technique), meditation, yoga-inspired stretching, nature walks, Christian and Buddhist spiritual retreats, and many other holistic healing practices. About four years ago, after Dr. Stern retired, I shifted care to a wonderful team led by Dr. Deepika Polineni at the Adult CF Clinic at the University of Kansas Medical Center, who helped me transition to my current phase.

Retirement and Older Adulthood

About a week after my retirement at the end of 2019, I was hospitalized briefly and continued home IV antibiotics. Soon after my recovery, the COVID-19 pandemic hit and many aspects of activity shut down. I used the time to set new priorities. I'm serving on the CF Foundation's Adult Advisory Council and the Access Committee. Additionally, I served on the 2021 ResearchCon Organizing Committee. I also participated in a Trikafta clinical trial. The appearance of Trikafta, which I continue to receive post-trial, has been an amazing and unanticipated boon. I feel healthier than I did ten years ago.

Retirement presents new quandaries involving the intersection of older adulthood and CF-based challenges. My body is changing with age and more friends in my age cohort are dying. We shifted to a fixed income based on social security and retirement savings. Now there are complications of coordinating retirement-based Medicare, supplemental insurances, and patient assistance programs. I lost access to copay programs due to restrictions on Medicare recipients. These are issues

requiring advice, support, and geriatric medicine, all of which aren't readily available for older adults with CF. Fortunately, my wife and I had employer-supported retirement savings plans for decades, without which we would be in serious financial jeopardy at this point.

Conclusion: Mindfulness and Living/Aging Well

Underlying my life stories are some lessons learned that have been very important for my living and aging well:

- Seek the best of CF medical care provided by teams who are skilled, knowledgeable, creative, sincere, and empathetic, and who address the whole person, including CF, but not defined by or limited to CF.
- Complement medical care with a wide repertoire of holistic approaches to health and wellness.
- Establish and maintain a consistent lifestyle and daily behaviors of wellness.
- Do all this to maximize the quality and meaning of life for myself and others.
- Cherish loved ones, share feelings, and leave no cause for regret when they or I pass.
- Do not assume anything based on CF or any other situations, while realistically adjusting to each situation.
- Be ready to respond to possibilities and opportunities for learning and growth.
- Plan carefully for the future regarding life goals and retirement, but don't be attached to expectations.
- Access older adult services, such as through area agencies on aging, and coordinate with CF-based services.

All of this implies living and aging in a mindful way. Mindfulness practices can reduce symptoms related to anxiety, depression, and distress, and they can promote positive coping and daily life satisfaction. Mindfulness cultivates awareness that is clear, gentle, and nonjudgmentally present in each moment. It opens a space in my mind between an immediate experience and my reaction to it. This frees me for how to respond to pain and discomfort and how to enjoy the beauty and wonder of living. I can be aware of my feelings, thoughts, and sensations whether pleasant, unpleasant, or neutral, while not being trapped in them. Then I can respond with greater compassion and skill to myself and others.

When mindfulness becomes a way of life, every moment becomes an opportunity for growing insights through the ups, downs, steady times, and doldrums of life. For example, daily respiratory clearance and antibiotic inhalation treatments are a time-consuming inconvenience, but they are also opportunities for gratitude and self-care. I view my medicine and nebulizer with appreciation for the medical community and all people and beings and the earth itself that made this treatment possible. When I do my respiratory clearance and then inhale the nebulized antibiotic, I take this as a time to get centered, and to pay caring attention to myself, my breath, and my condition.

Mindful living does not mean always feeling happy or having a positive attitude. With mindfulness, even times of pain, worry, fatigue, anger, or discouragement can be acknowledged, loosened, let go, and recovered from more easily. We can regard ourselves with acceptance, compassion, and care at each moment. Mindful living and aging are about appreciation for the journey of life and experiencing growth and fulfillment along the way.

However, wellness is not just about individual and family effort. We need to have public health and social welfare policies and programs to promote a high quality of life for everyone, including people with disabilities and chronic illnesses. Medical services and insurance should be affordable, accessible, and equitable to support surviving and thriving. My personal and professional experiences have shown me that each person's wellness is interconnected with everyone's wellness. ▲

Dr. Ed Canda is 67 years old and has CF. He is a retired professor and lives in Lawrence, Kansas. He enjoys playing with grandchildren and performing meditative percussion. Many of his publications are open access at KU ScholarWorks.

SUFIAN continued from page 5

can make a big difference. Even a small amount of earnings over the monthly allowable will cause the month to be counted as a trail work month and, similarly, even a small amount of IRWE can offset income and prevent the month from being counted as a trial work month.

6. Grace Period.

If SSA determines a beneficiary has made a successful return to work at the end of a TWP and terminates benefits, then SSA will pay the monthly benefit for the month in which the disability ceased and for the following two months. This is known as the grace period.

In the next issue of *CF Roundtable*, Part III of this series will address what happens if after a TWP a beneficiary remains disabled under SSA rules and continues to have incomes from work. Part III will discuss SSA's extended period of eligibility and continued Medicare eligibility. ▲

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.

SPIRIT MEDICINE

The Long View

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

I'm going to say the F word...yep. FIFTY. Hallelujah, I made it to 50! This is something I never expected as a child with aggressive CF born in 1972. I have lost so, so, so many peers from my generation. And though I have many more elderly mentors with CF, I consider myself "old." Being 18 years post-transplant means to me that "50 is the new 70."

Growing up, all I wanted was a long life. I yearned to survive. Now, my youth is behind me. My body is changing. The transplant meds are taking their toll. Cancer has attacked my eye and I have lost my eye. But I'm still breathing easy. The future promises more challenges—the loss of my parents, the decline of my ability to function, and changes in appearance. I feel both dread and anxiety, and simultaneously laugh at my once-youthful innocence about a long life. What guides me as I reach this age?

One important source of insight is my perspective of the "long view." This means looking at life with a pair of binoculars, seeking the details of the expansive life that I've lived within the context of the wider human experience. I'm on the top of a mountain looking down to the trail I've climbed, and eyeing with trepidation the trail I must find to climb back down. I try to take the long view of my life, as if I were hovering above in a hot air balloon and enjoying the sights from a superior perspective.

The long view is the approach that no matter what I am dealing with in the present moment, I look backwards to see how good I've had it earlier in my life. Life with CF was, and is, hard as hell, but I did my very best and it paid off in the long run. I was sick growing up but surrounded with hope, love, compassion, and care. My community always looked at sick kids tenderly, so I was given attention and opportunities that never would have happened if I was healthy. I look back at all of us within the CF community and see so many kids in my old camp photos who are no longer here. I could've been one of those ten-year-olds who died. Instead, I'm here at fifty. I feel proud that I am still here to tell their stories.

The long view helps me realize I had my turn to be healthy during my transplanted life. I had a chance to breathe easy and to feel invulnerable. I had my chance to be a competitive athlete at the Transplant Games. I also had the chance to be healthy enough to have a career; to be a real professional.

The long view also helps me recognize how fortunate I am to be a woman in 2022, and how far we have come thanks to courageous feminists of the past. To be a woman of color brings more privilege today than ever; I get to marry whomever I want to, reach my fullest academic and vocational potential, and have worth despite being childless. And compared to human history, we live in relative peace, abundance, and security. No matter what I will go through, my basic needs—food, shelter, clothing, clean water, and air—are being met. How blessed I am!

> *"The long view helps me to normalize daily pain and stiffness as a part of aging."*

ISABEL STENZEL BYRNES

As I get older, the long view also means questioning my perceptions. If I think I need to be ashamed of my illness or my physical changes, I ask from where did these messages come? At this age, I question our cultural norms and ask whether they are fair. Is my value as a woman based on what I look like? What patriarchy! Was I "better" when I was younger and prettier? What ageism! I can call BS on the social values that try to diminish my sense of worth. That's why I love women who don't dye their hair.

As I mature, I am shifting from the desire to achieve and to accomplish—all with a sense of urgency to make the

most of my one precious life with CF—to simply enjoying this present moment. The long view means being on the other side of achievement and feeling content and satisfied with the life I've lived. True maturity means saying yes to saying no. Saying no so I can do more of what I enjoy and want to do.

The long view means looking at my past habits and patterns and shifting them to something healthier and more adaptive to my present and future needs. The overthinking, the need to please, the desperation to make the most of every moment…those things aren't helpful anymore. It means letting go of the resistance and fear of medical issues that constantly arise; leaning into difficulty gives us a growth mindset. My relationship with pain and discomfort is changing.

The long view helps me to normalize daily pain and stiffness as a part of aging. It helps to find compassion in myself when I can't remember things or can't do what I used to do. It means trusting that my body's got this—the natural process of aging, menopause, and loss. I have adapted and I will adapt to whatever change arises.

I look to ancient times to give me perspective. The long view helps me remember that for most of human history, fifty was a very old age. Elders who couldn't see well or move fast didn't survive in nature very long. In nature, when animals procreate, they have completed their purpose. Death gives rise to new life. That is the divine order. All living things age and die. We are no different.

As I age, I constantly strive to live a spiritually informed life. Linda Graham, a spiritual teacher and psychotherapist, says that "AGE" stands for Awe, Gratitude, and Equanimity. I feel awe at still being here; awe is really a deep love for beauty and life itself and all those in it. This love is how I commune with God. Nothing separates us from the love of God. Gratitude is intentionally looking at what is given to us, not what is taken. I have had a chance to be here—to grow and learn and know myself. How thankful I am for self-awareness and for all that life has to offer. Equanimity is reflected in "let me taste the whole of it" (Robert Browning). Equanimity is a goal and not easy to attain. I have to remind myself that all struggles and triumphs are part of this package of life; that each experience can still be blessed with teachings and bring me closer to God.

We know that adversity can enhance spirituality and spirituality can be a healing salve for adversity. English historian Arnold Toynbee says that generation after generation, there is "opportunity to open souls by way of the learning that comes through suffering, for getting into closer communication with God during their brief passage through this world." Through all the chapters of life, human beings are linked with God.

My spiritual goal is to age without life's hardships beating me down. I want to come out of my life difficulties fully spiritually alive, even if it kills me. I'd like to believe that hardships are polishing me; Confucius says, "A gem cannot be polished without friction, nor man without trials." As I age, my spirit becomes shinier; the edges are smoothed out.

Another spiritual goal is to leave this world a better place. Aging means being aware of my continuation. What does that mean? That by being kind to others, mentoring others, and making a difference in my community, I will continue no matter how long this body survives. Thich Nhat Hahn, a Vietnamese monk and global spiritual leader, says "my body will disintegrate but my actions will continue me." Life will continue.

I aim to embrace the sacredness in this bodily journey. I love that Susan Willson, in her book, *Making Sense of Menopause*, says that "menopause is a harvest, a time of life to pick the juiciest fruit, savor it and take the best seeds you have and plant them for the next generation." Ain't that the truth?

The older we get, the more we gain *and* the more we lose. As I cope with the loss of my eye, I tell myself that I had two eyes for fifty years. What a blessing! And the cancer was removed—how lucky I am! And, at the same time, I mourn what I've lost. I am being tested now in a different way. My eye is gone but replaced with an internalized long view, a new insight and vision. I'll do the best I can, like I always have.

With all the grief I carry, I find solace in different ways of looking at grief. The Lakota Sioux see a person who is grieving as most "wakan"—that means "most holy." Grief creates an openness to that which is beyond this everyday world. The pain of grief is seen to give access to a sense of the timeless, to the eternal. Grieving peoples' prayers are considered especially strong. Lakota Sioux tradition maintains that it's exactly these grieving people whom we should ask for help and prayer. So, as we age, there is potential for usefulness and purpose with the griefs we experience.

The long view is accessible to anyone, regardless of age. Even a 15-year-old can reflect on their full life or what they've overcome since age five. It is a spiritual act to remove oneself from being mired in present-moment emotions. My prayer is that everyone with cystic fibrosis can cultivate wise vision with the passage of time. In sum, I leave with theologian Harvey Potthoff's wise words: "Good aging is a quest for greater quality of life in the midst of the natural cycles of life and death, growth and diminishment, love and loss, joy and grief. It affirms the sanctity of existence and the continuing love of God in the midst of adversity, diminishment and even death." Amen to that. ▲

Isa Stenzel Byrnes is 50 years old and has CF. She lives in Redwood City, California. She is 18 years post-lung transplant.

PEARLS OF WISDOM

Rotten Roots: Oral Health Challenges In Aging With CF

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

Remember that new house I wrote about in my last column? About 12 hours before I was supposed to pick up the keys, my left front dental prosthetic broke apart in my mouth and then fell out completely. I wish I could say this was my first experience with having my teeth—whether the original ones or their porcelain replacements—fall apart. This story also did not sound unusual to any of my age peers in the CF community with whom I shared it. Welcome to aging with CF.

Getting older with a progressive chronic disease is alternately glorious, bizarre, rigorous, and horrific. Having outlived the initial life expectancy for people with CF in my birth cohort by a good margin, I find myself in uncharted territory. Several of my clinicians have humbly admitted that they are learning primarily from us patients about what to anticipate and how to respond as we grow older. This is a wonderful attitude and more than a little scary for everyone involved.

People with CF become pretty tough because our lives demand toughness. Honing our ability to laugh at all kinds of terrible experiences tends to help us live better with the physical burdens of this disease and also cope better mentally with the internal struggles we face. When my full crown came apart, I placed it in a little plastic jar that I continue carrying around in my shoulder bag. I come by my morbid fascinations honestly; growing up in a medical research lab often makes one embrace the body and its various odd products. But underneath the dark humor and the quirky fascination with decay, there is pain and heartbreak. My teeth are not the only broken things.

Growing older with a disease that barely lets you be young can feel like an endless spin cycle of loss and adjustment from childhood onward. The hardest thing for me right now is figuring out what is even "normal" for a body that did not receive coordinated CF care until its early 30s. I still struggle to balance my heightened tolerance for discomfort and pain with my acumen for self-advocacy. And I continue to process the cognitive dissonance of sounding the alarm about a problem for years before it became bad enough to merit more serious intervention in the eyes of clinicians.

> *"Looking back, I realize that I should have trusted my instincts more about something being wrong and pushed harder for an extraction."*

When that prosthetic broke after nearly eight years of continuous issues, I felt considerable relief both physically and mentally. I knew I would no longer have to chew each bite of food in fear of it breaking apart. I also felt less pressure around the socket after losing the crown than I had previously. But over time, the pressure became worse again and the area started to ache. I was just on the verge of calling the surgical practice back to say that I needed an urgent care appointment, when they called me to offer an earlier consult due to last-minute cancellations by other patients. I adjusted my work schedule accordingly and took the next-day appointment.

XAN NOWAKOWSKI

It took several rounds of CT scanning to give a clear image of the socket. All of the films also showed destruction in the upper jawbone. This surprised neither the surgeon nor me, given that my entire sinus network was essentially fossilized by congealed infected mucus until the summer of 2020 when my ENT care protocol changed. Beneath the socket with the diseased root, the bone was almost completely gone. Surgery was sched-

uled for the first available appointment, about three weeks later. We made a plan for how to address CF-specific infection concerns and for early follow-up to assess my healing.

This initial consult took exponentially longer than the surgery itself. Because I had worked arduously to keep my gum tissue—which is also the product of extensive reconstructive surgery—as healthy as possible, the socket was in good enough condition to support a combined extraction and bone graft procedure. My surgeon consented to operate with only local anesthetic so that I could remain as independent and functional as possible on the day of my procedure. So during the surgery, I was wide awake and able to follow what was happening. I even got to take a picture of the diseased root after it came out. My sense that this would help me cognitively proved very accurate when I saw what was attached to the remnant.

We expected to see a considerable amount of infected tissue, but not necessarily a cyst taking up the entirety of the empty space around the destroyed bone. Even with my face numbed, I could immediately tell that the pressure in the socket had vanished. I gave my surgeon a thumbs up on starting the bone graft, and only got through one more song in my headphones before he began stitching me up. The most challenging thing I did that day was pick up my antibiotic at a pharmacy in my neighborhood after multiple technical issues arose with the surgical team's effort to submit an electronic order for it. This was a welcome change after years of voicing the same concerns about that socket being infected and unstable.

To age with CF is to learn our own bodies painfully well, both literally and figuratively. Looking back, I realize that I should have trusted my instincts more about something being wrong and pushed harder for an extraction. Even though I am aging with a deadly progressive disease, I still face destructive norms about growing older that make many clinicians and patients prioritize cosmetic interventions over other types of care. If I had allowed my dentist to try to "save" this root by putting a deep post into it, I could have wound up hospitalized with a severe infection in the jawbone after having that cyst rupture.

As I told every provider involved with this process, I care about only two things: quality of life and functionality. This is what aging well with CF means to me. I absolutely notice how the continued progression of CF changes my physical appearance. But I would rather look different or be "missing" something than live in pain or go back to the soft-foods-only diet that was my only option after infections ate away enough gum tissue to expose bone below each root. Aging well with CF is about embracing every bit of freedom and comfort I can find. As I explained to my surgeon, there is absolutely nothing I will "miss" about this prosthetic or the tooth that was once there.

Of course, I say this from a perspective of economic privilege. I am getting treatment because I can afford treatment, both via the dental insurance coverage I get from my employer and via my own income and my family's resources to cover out-of-pocket costs. Many of my peers in the adult CF community do not have access to these resources. Because of broader patterns of systemic oppression of racial and ethnic minority communities, as well as queer and trans people, our adult peers who struggle most to access basic dental care experience these challenges within a larger context of injustice and hardship.

Basic dental care also does not suffice for people aging with CF. Getting the root remnants of my other teeth stabilized has required years of working with committed dental and periodontic professionals who embrace learning from me about the unique oral health concerns CF introduces. The need for provider education on CF dental issues cannot be overstated—and neither can the barriers to conducting such training. Because of the general classist stigma surrounding tooth loss, dental damage remains a largely silent issue even within the adult CF community.

In a world where so many of us are experiencing transformational change in our ability to keep breathing as we grow older, we find ourselves increasingly confronting the aggressive damage CF does to all the other parts that keep our bodies going. The joy of celebrating each new birthday comes with tremendous uncertainty about the new challenges we will face in the year ahead. I just celebrated 38 years myself, my surgery a slightly belated gift that left me smiling brightly around the hole in my bite. I remain grateful for the integrity of my remaining dental prosthetics and the little bit of natural enamel I still have. But this year, I feel more grateful for that hole and for the progress it represents toward my own goals for aging successfully. ▲

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

Arts Scholarship Update: Exploring Disability Through Art

By Rowan O'Bryan

I am so incredibly grateful and honored to have been one of the first two students awarded the newly established Scholarship for the Arts. I want to thank USACFA and the late Helen E. Eisenman for their work to support artists with cystic fibrosis through this scholarship. This aid has dramatically impacted my educational journey, giving me the financial support necessary to get my Bachelor of Fine Arts. I have had to become financially independent due to my parents not being able to find work during the COVID-19 pandemic. It has been a struggle to pay for college tuition; however, this scholarship has made it possible.

ROWAN O'BRYAN

As a student enrolled in UCLA's Department of Art, I am constantly investing in my art because not all of our supplies are given to us for free. Last quarter, in the fall of 2021, I made a wall sculpture related to my identity as someone with chronic illness. The sculpture is about three feet by four feet and made out of wood, nails, spray paint, and my used oxygen tubing, which I dyed in different colors. I used my old oxygen tubes as a way to repurpose the trash I create from my medical devices. I dyed the tubes in bright colors as a metaphor for my transition from pediatric care to adult hospitals and care teams. I wanted to make a statement about my relationship with the oxygen machines to which I have to be connected 24/7, in addition to highlighting the stark difference between the art in pediatric hospitals versus that in adult hospitals. After I switched to UCLA medical hospital for my adult care, I immediately noticed the white, sterile aesthetic of the interior of adult hospitals. I missed the colorful walls and playful art at Children's Hospital Los Angeles, which is sup-

TILLMAN continued from page 3

most common side effects were anxiety, joint and muscle pain, sinus congestion, nasal congestion, depression, and bloating. Joint and muscle pain, along with extreme tiredness, were the most bothersome. Among the patients who answered a question about their quality of life, more than half reported being satisfied. However, nearly a third of patients reported being "somewhat" to "very unsatisfied". More than half expressed optimism about their future. More than a quarter were "neutral," and only less than one fourth of patients reported being "somewhat pessimistic," with none saying they were "very pessimistic." Also, most patients were "somewhat" to "extremely satisfied" with their current treatment plan. The remaining were "somewhat dissatisfied" or "neither satisfied nor dissatisfied"; none were "extremely dissatisfied." More than three-quarters of patients also reported being satisfied with their health insurance provider. Nearly three fourths of the patients who answered a question about clinical trials expressed interest in participating. Overall, the survey results highlight generally high rates of satisfaction with care and health insurance among CF patients, but also indicate that a relevant proportion of patients are dissatisfied with their quality of life. These findings highlight areas of unmet needs and call for better care and support to improve the lives of these patients.

https://tinyurl.com/yb56beqd

Cardiovascular Complications In Cystic Fibrosis: A Review Of The Literature

Cystic fibrosis is a genetic disease caused by mutations in the cystic fibrosis transmembrane conductance regulator (CFTR) gene, leading to dysfunction of the CFTR protein. Disorders of the cardiovascular system in individuals with CF are usually attributed to secondary effects from progressive lung disease. However, CFTR has been localized to vascular endothelium and smooth muscle, suggesting that CFTR dysfunction may directly impact cardiovascular function. As treatments for CF improve and life-expectancy increases, the risk of vascular disease may increase in prevalence related to primary and secondary CFTR dysfunction, chronic

Continued on page 15

posed to make kids more comfortable. I think adults deserve that as well. When I finished the piece, I realized it helped me change my negative perception of my oxygen therapy into something positive that makes me feel unique. The funds from this scholarship allowed me access to the resources necessary to build this piece of art.

I plan on graduating in 2023 with a major in Fine Arts and a minor in Disability Studies. After I graduate, I want to find work and/or a graduate program that incorporates both art and disability as well as continue my studies and involvement with both the ASL and Deaf Communities. I want to continue to advocate and support the greater Disabled Community, including the CF and Deaf Communities, both in school and afterwards. I have always enjoyed fundraising as a form of supporting others in my various communities. I have considerable experience with fundraising through my involvement with my Great Strides team "Rowan's Roses" when I was a child and, more recently, events to help me pay for my future lung transplant. Another way I aim to show support is by getting involved in the larger disabled art community. Through my Disability Studies minor I plan to do an internship next quarter working with a collective of disabled and chronically ill artists and activists who have a goal of curating a library of knowledge on lived experience of disability as well as creating community-building workshops. I hope this internship will connect me to the network of disabled artists and kickstart my career goals.

I will continue to celebrate my identity through my art on public social media platforms to encourage others to embrace their differences. I have received so much love from others in the Disabled Community because of the importance and impact my art has in the community. I feel accomplished knowing others are benefiting from what I love to do. This scholarship has aided my artistic journey by giving me the freedom to explore and establish my passions without financial burdens. ▲

Rowan O'Bryan is 22 years old and has CF. She is a fourth-year undergraduate student majoring in Fine Arts and minoring in Disability Studies. Her artwork is largely centered around her identity as a person with disabilities. She repurposes her medical supplies and devices to offer an alternative and creative viewpoint on what it means to be chronically ill. She is a co-leader of a student group pushing for American Sign Language to be recognized as a major. She is also co-signatory of the Disabled Student Union where she works alongside other disabled students to fight for UCLA to become a more accessible university. She hopes to continue her advocacy and support for the Disabled Community in her career after receiving her Bachelor of Fine Arts degree in 2023. You can follow her on Instagram at @ row.1.

Meet A New Director: Ela (Gabriela) Castillo

Hi! I'm Ela Castillo. I'm an adult diagnosed with CF in early childhood. I have a passion for advocacy and fundraising within the CF community. Previously, I worked at Vertex Pharmaceuticals in Boston. I have rare CF mutations and I am currently ineligible for any modulator therapies.

I am American and I come from a mixed family—my father's side is Colombian and my mother is ethnically Ashkenazi Jewish (from eastern Europe). In addition to CF, I'm hard of hearing and neurodiverse. I was diagnosed with epilepsy and ADHD in adulthood. I strive to gain representation for all the groups I am a part of, as well as other underrepresented groups within the CF community.

I am passionate about accessibility and helping adults with CF find appropriate support, especially for the deaf and hard of hearing and for those navigating multiple disabilities. Another strong interest is researching and recommending practical recommendations for young professionals with CF. ▲

ELA CASTILLO

FOCUS TOPIC

AGING AND CF

Endure

By Kathy Russell

At the beginning of this year, one of my favorite radio hosts asked her listeners what their word for the new year was. I thought about it for a moment or two and decided that "endure" is my word. We all have had to endure so much over the past two years. Some of us have had to endure for all of our lives. Living with CF means learning to endure a lot.

I was diagnosed with CF in 1956, when I was 12 years old. At that time people who had CF were not expected to live long enough to enter school. Since I already was past the expected age of death, I decided to live. I am in my late 70s and have no immediate plans to stop living any time soon.

Because of new ways of treating CF, young people today can expect to live long lives. I read an article that quoted Kors van der Ent, professor in pediatric pulmonology at the University Medical Centre Utrecht in the Netherlands and coordinator of the multi-disciplinary HIT-CF project: "We're effectively shifting therapeutic trials from patients to the laboratory. Thanks to these drugs, in some patients there's a lung-function improvement of 30–40% and life expectancy can increase from the age of 30–40 to 60–80. In other words, there can be a normal life expectancy." This kind of thinking encourages me. I really hope that the young people of today will live into their 60s, 70s, and even their 80s.

April 17 is my 78th birthday-anniversary. At that time, I will have made 78 trips around the sun totaling 45,552,000,000 miles. That number does not even take into account the trip of 25,000 miles that we all make every day on our earth. That is a lot of travel and I have seen a lot on those trips. So much has changed over those years.

KATHY RUSSELL

When I was first diagnosed, it was fairly easy to recognize the cough of someone who had CF. Now, thanks to good medicines and treatments, it is much more difficult to identify the sound of a CF cough. Many of us cough only infrequently and our coughs sound almost like any "normal" person's cough. Many of us have fewer lung infections, and those that we do have are less severe and last a much shorter time than they used to.

There are some changes that I have noticed in my health that I don't know if they are a part of having CF or just related to general aging. The first one is my bone and joint pain. My back, neck, hands, and feet give me discomfort. I know that I have suffered compression fractures in my spine and neck and that those can be painful. So I guess I'd have to blame them on age.

The discomfort in my feet and hands may be CF-related arthritis. My thumbs are quite painful at times and make it feel as if I don't have opposable thumbs. That can make it difficult to pick up or hold onto a glass or cup. I know that I have to be cautious when I pick up or hold items. The balls of my feet feel as though I am walking on marbles. My soles feel as if they are completely coated in stiff plastic. The large joints of my big toes get quite stiff and make walking a trial. I believe that this discomfort is just another thing to endure.

A big change in my living as I've aged is that I get tired much more quickly than I used to. I find that I need to sit down or even take a nap after even a small amount of exertion. Some of this is just getting older and some is having less lung function than I once had. However, I am better now than I was for several years and I attribute that positive change to taking Trikafta. I have been taking it for two years and it has given me more energy than I've had since I was in my 50s. These new medicines that are developed for specific mutations are doing wonderful things for us. I hope that there soon will be such medicines for everyone who has CF.

My physicians and I have good working relationships. They trust me to be honest with them (and myself) and to let them know when there is a problem. They are quick to respond to my requests and needs. I have seen my PCP only twice in the last two years. In that time, I have seen my ENT doctor twice. I no longer need to see him as often as I once did, because my sinuses are almost normal. I have had virtual visits with my pulmonologist every four

months. I could never have managed with only virtual visits with him before Trikafta. Can you tell that I really like Trikafta?

So it seems that endure is the word for what I am living with, regardless of whether it relates to CF, aging, the pandemic, or what is happening in Ukraine. There is little that I can do other than hope for the best and endure whatever occurs.

Aging with CF is something that I hope will continue to get easier for everyone. Being an old person with CF who still has their original parts (no transplants) may become the norm if we can just endure.

Stay healthy and happy. ▲

> "Because of new ways of treating CF, young people today can expect to live long lives."

Kathy is 78 years old and has CF. She and her husband, Paul, have been married for 57 years. They have lived in the same house in Gresham, OR, for 45 years. She volunteered for USACFA and CF Roundtable from the beginning until a few years ago. She enjoys reading, doing word puzzles and jigsaw puzzles, and she loves cooking and baking, especially bread. Yum!

TILLMAN continued from page 12

systemic inflammation, nutritional health and hyperglycemia in individuals with CF related diabetes.
https://tinyurl.com/y8kdnuf9

Testicular Cancer In Men With Cystic Fibrosis

Researchers reported on the incidence and risk factors for cancer in individuals with cystic fibrosis in the United Kingdom. Using a case-control approach, they observed a high incidence of lower gastrointestinal cancers, confirming and extending results from a large cohort study in the United States. Besides, they reported testicular cancer in six men with CF during the 19 years of observation. Although the authors did not mention whether this number was greater than in the control group or than expected based on cancer rates in the general population, it is consistent with a two-fold excess risk based on numbers and rates observed in the US cohort.
https://tinyurl.com/yvs6dbnt

What Is Cystic Fibrosis–Related Arthritis (CFRA)?

Cystic fibrosis–related arthritis (CFRA) is a complication of cystic fibrosis. Pain attacks can be infrequent or happen regularly. There are two types of CFRA: cystic fibrosis–related arthropathy (CFA) and hypertrophic pulmonary osteoarthropathy (HPO). It is estimated that between 2% and 8.5% CF patients develop CFA. Signs of CFA include: 1) episodes of joint pain, swelling, or tenderness, 2) limitation of movement, and 3) fever. CFA is more common in people with more severe lung disease and can get worse during an active lung infection. CFA is more episodic, with joint pain lasting less than one week at a time. Symptoms arise during flares but may completely go away. HPO is less common in CF patients, at between 2% and 7% and generally causes severe symptoms which include: 1) episodes of joint pain, 2) dull bone pain, 3) digital clubbing, and 4) worsening in cold weather. It is unclear what causes CFA and HPO since little research has been done on these conditions. The following tests are used to diagnose CFRA: 1) full blood count (also known as complete blood count, or CBC), 2) measuring acute phase reactant levels (APR): These are inflammation markers in the blood that increase during an infection or injury, 3) X-rays, and 4) synovial fluid analysis: This test shows whether the fluid between your joints is normal. The color, thickness, and presence of bacteria are examined. CFRA may also increases the risk for bone diseases, such as osteoporosis and osteopenia. Treatment for CFRA includes NSAIDS, corticosteroids or DMARDs (disease-modifying antirheumatic drugs).
https://tinyurl.com/ycoztk69

Prevalence And Impact Of Rheumatologic Pain In Cystic Fibrosis Adult Patients

The aim of this prospective study was to evaluate the prevalence of spinal and joint pain, and their impact on disability, anxiety, depression, and quality of life in CF adult patients. The investigators found that rheumatologic pain is frequent in CF adult patients, and may affect daily living, anxiety and quality of life. Systematic assessment of rheumatologic pain should be included in the management of CF patients.
https://tinyurl.com/2p9esrmn

Urinary Tract Infections In Cystic Fibrosis Patients

To date, there has not been a published report of urinary tract infections (UTIs) in CF patients. Investigators performed a retrospective chart review

Continued on page 20

Exercise And Diet Effects On Aging With CF

By Jennifer Kyle

> "I am a true believer that even with CF we have control over our bodies and can still make positive changes to help combat what this disease and all our medications throw at us."

I am 57 years old. I have CF and I'm post-menopause. Many changes come with age, and we are paving a new path to figure out how CF affects those changes. Are the changes we experience normal? How does CF play a role in how our bodies will adapt to aging? What can we do to help combat the aging cycle and help manage our CF at the same time? It's something I have been passionate about for a long time.

I was diagnosed with CF at age 37, yes, 37! Since then, my life has been a series of connect the dots with past experiences, only to realize it was CF causing all of my medical problems. The most interesting revelation was that my doctors felt that exercise and diet saved my life prior to diagnosis. Those two factors motivate me daily.

When I turned 40, I was still dealing with getting used to having CF—the frequent infections, medications, and hospitalizations. I knew I was well past the survival age, and, on top of this, my body decided it was time to enter perimenopause. Along with all the changes in my menstrual cycle, I started to experience my first issues with arthritis in my knees and hips. I thought this was unusual but decided I would just keep exercising and eating healthy and not give it much thought. I was soon diagnosed with osteopenia, which surprised me, but my doctor explained this was "normal" with CF. I knew exercise helped with combating osteoporosis so I decided I would increase my exercise to help stop the progression. In addition to taking prescription Fosomax, I decided to increase my running from three to five miles. I slowed my pace so I could get through the five miles without too much exhaustion and, after six months, my body was used to the change. Instead of increasing my weights, I increased repetitions to promote lean muscle mass and bone density, again hoping I was doing what was right for the osteopenia in conjunction with the arthritis. I ate fruits and vegetables daily, drank at least eight and sometimes up to 12 glasses of water, and kept chicken and fish as part of my diet.

JENNIFER KYLE

By 48 I was in full menopause. Realizing the lack of estrogen was going to play another role in bodily changes, I decided it was time to get back to dancing and started studying ballet and modern dance with the Princeton Ballet School. Having a degree in dance made this a relatively easy transition, but it had been a long time since I was in a studio. The dance was just the thing my body needed. I realized I was needing more flexibility and posture exercises with my workouts and dance provided the missing piece. So along with running and weightlifting, I felt I was at my peak performance for my fitness. Still, I wondered about my diet. It was then I decided to start following a more vegetarian-focused diet. I stopped eating chicken—eating just fish—and incorporated completely vegetarian days into my week. I noticed how much easier my digestion was on those days with less gas and bloating. So, I kept that routine and, as the years ticked by, I ate mostly vegetarian.

It wasn't until a few months ago that I did some research into plant-based eating. I had been on Trikafta for nine months and the side effects were starting to show—increased liver counts, blood sugars, and cholesterol, along with neuromuscular tears, were plaguing me and I was worried. My doctor and I decided to decrease my dose and I decided to go full plant-based eating (vegan).

Since making these changes I am the healthiest I have ever been. All my counts are down in the low to normal range. My joint pain is gone and only

mildly flares up if I do an extra hard workout. The swelling in my hands has decreased and the pain no longer wakes me up at night. My workouts have even improved, and, for the first time in years, I am able to start increasing weights, which is increasing my muscle mass and helping me improve my cardiovascular exercise. My skin looks better and my gastro problems have all gone away.

I am a true believer that even with CF we have control over our bodies and can still make positive changes to help combat what this disease and all our medications throw at us. I see exercise always being a part of my life. I know as I age my exercise routine may change and I will gracefully accept those changes. I will continue to work hard at keeping myself fit and feeling the best I can feel. I know it's because of diet and exercise that I am the healthiest I have ever been and I am so happy for it. ▲

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.

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CARROLL GROEGER

IN MEMORY OF
LAUREN
MELISSA KELLY

Aging Is A Privilege, Mostly

By Sydna Marshall

I was diagnosed in 1991 after my surgeon discovered polyps during a routine adenoidectomy. Polyps aren't common unless you have cystic fibrosis, in which case they're likely the bane of your existence. At that time, the life expectancy for someone born in the 80s varied between 10 and 14 years, depending on the source.

At the time of my diagnosis, no one told me that I likely wouldn't survive into my 20s. I remember finding out at camp that first summer after my diagnosis when a cabinmate announced that she had read in her science textbook that I would die before I'm 20. Needless to say, I came home from camp with all kinds of anger at not knowing that and even worse, finding out that way.

Most of my early life with cystic fibrosis was relatively easy, comparatively. I had high lung functions. My weight was stable, if not slightly over much of the time. I got by with oral antibiotics when I had an exacerbation. And treatments didn't take nearly as long. Granted, most of the advancements in nebulizer treatments came in my 20s. My last semester of college, fall of 2001, I developed my first drug allergy—Cipro. I was miserable. I had hives everywhere, and I mean *everywhere*. I was on steroids and Benadryl for a solid month. That last semester I didn't take care of my health much and I paid the price in the end. By December, I was in my doctor's office (at that time, we didn't have a CF clinic yet so I saw my pediatrician who also specialized in CF) every other day. Nothing was working and I had constant pleuritic pain—I felt like someone was stabbing me in the sternum. By April, I was admitted for two weeks for my first ever tuneup in the hospital.

Let's fast forward to my mid 20s. My sinuses are textbook CF sinuses, or worse. Honestly, it's a miracle I lasted as long as I did without too many sinus issues. I was plagued by daily, debilitating headaches. Like many with CF, my dark sense of humor saved me. I joked often that I'm a human barometer and can feel the pressure changes in my head. I started seeing my ENT monthly for scopes of my sinuses. I racked up quite a few sinus surgeries and more drug allergies along the way. Finally, I agreed to have a frontal sinus obliteration when I was 31. There aren't too many of us in the CF community who have had this "pleasure." It's a brutal surgery with a very long recovery. I'll skip the details here but essentially my forehead sinuses are now filled with fat from my tummy. This was lifesaving for me, on several levels—I got quality of life back for a time and, during surgery, my ENT found a pocket of infection that was sealed off but close to rupturing, causing either blindness or spinal meningitis. In the many, many CT scans we did pre-surgery, that pocket always looked like bone growth. Surprise! During my 20s, I developed consistent pain that felt like 24/7 heartburn. After gobbling down a box of Gas-X, with zero relief, I went to the ER in tears. Turns out, I had a golf-ball-size gallstone—the doctor immediately scheduled an emergency cholecystectomy to remove my gallbladder. I was so happy to say goodbye to that pain!

SYDNA MARSHALL

Let's fast forward again—this time to my 30s. This is where the aging bit really started cropping up in my health. In addition to my CF, asthma, and chronic sinus disease, I was also diagnosed with osteopenia, which isn't terribly surprising since my vitamin D levels, until recently, have always been low. Sometimes, as low as five. After having a dozen or so PICC lines and rounds of IV antibiotics, the team in IR (interventional radiology) commented that placing a PICC was challenging from existing scar tissue from prior PICC lines. The last PICC they placed hit a lymph node, causing lymphatic fluid to leak consistently. I went back to the ER within the week, after having a dressing change daily because everything under the adhesive was wet. The ER physician refused to place a new line saying I needed to save my veins. Eventually, they took me to IR where they applied Dermabond to the site and assured me that it wouldn't leak anymore. The next day I had my home health nurse pull the line because the Dermabond wasn't sealing the leak as promised. That was the impetus for my decision to have a portacath placed—preserving veinous access and, as an added bonus, no more PICC line snafus. It's one of the best decisions I've made regarding my healthcare. It's so much easier and my husband is a pro at

accessing and deaccessing me for IVs and monthly flushes. In my 30s, I was also diagnosed with CF Liver Disease (CFLD) and had two endoscopic retrograde cholangiopancreatography (ERCP) surgeries to clear out the debris in my biliary ducts.

On top of all of that, I was also diagnosed with Graves disease in my 30s. This ranks second to CF for me as it was, and still is, a miserably long road back to feeling normal. For about 18 months prior to my diagnosis, I kept complaining about being hot and cold and I was lethargic beyond what was normal for me. I was dismissed as being "menopausal" from the outset. It took a year before my pain specialist at the time finally heard me and ordered blood work for my thyroid. I've never been so grateful for bad bloodwork! Near the end of that time period, I remember having to sit on the floor to blow dry my hair before work because I only had about three spoons (according to the spoon theory) for the entire day and drying my hair used one of those up! Did I mention work? I also still worked full time. It was exhausting, and I needed an exorbitant amount of support from my husband to make it through a single day. Eventually, I left my full-time job so I could focus on my health and getting better. I had my thyroid nuked in 2017. When my TSH levels skyrocketed from hyper (I was in the negative) to hypo (my levels jumped to 91 in one week's time), I again felt absolutely horrible. The effects of a thyroid hormone imbalance are far-reaching: hair loss, weight gain, constipation, and fatigue, to name a few. Over time, I've learned how to pinpoint what feels off and whether we should recheck my TSH levels. A lot happened in that decade of my life!

Meanwhile, my lady bits are also a hot mess. In the last 15 years, I've had a lot of procedures down there: cervical cryotherapy to remove dysplastic cells; a LEEP procedure to remove cancerous cervical cells; an ablation to cauterize my uterine lining; an Essure placement (coils placed in the Fallopian tubes to prevent pregnancy); removal of ovarian cysts; and, finally, a hysterectomy, in which my ObGyn discovered I had a bicornuated uterus. If this entire paragraph reads to you like the screenplay for an obscure sci-fi film, you're not alone.

Let's fast forward to now. I was recently diagnosed with hypertension, bordering on malignant hypertension. It's been increasing over the last few years and I'm now on a blood pressure medication and, thanks to genetics, a cholesterol medication as well. I'm in the process of finding a cardiologist to add to my cadre of specialists. I've also scheduled a skin test for allergies as the blood test didn't give the doctor a lot of information. I can't take my blood pressure medication the day before or day of as it interferes with the allergy testing! This will be my second round with allergy shots but I'm hoping that my quality of life in the allergy capital of the world (Austin, TX) will improve, especially with the spring pollen.

> "I'm finally in my 40s and I wouldn't trade it for anything—I've made it!"

All this to say, I'm finally in my 40s and I wouldn't trade it for anything—I've made it! Despite the myriad health problems I've endured, on top of cystic fibrosis, I have outlived several life expectancies. Do I feel like sometimes I got the raw end of the deal with my genetics? Absolutely. I also know that I'm fortunate—while my sinuses are some of the worst CF sinuses, my lungs are rather resilient. As a kid, I didn't anticipate the additional health complications that come with aging as I didn't think I'd be around long enough. I'm overjoyed that I was wrong. My medical history and complications can be frustrating, to put it mildly. I also watch other adults with CF around me struggle with far worse on a daily basis. I've reached the point where it takes me a few hours to fill out the medical history questionnaire for new doctors and hospital admissions. Thank goodness for online patient portals—I can't even fathom having to spend that much time filling out a paper form at the doctor's office! But, as I've told every single person who complains about grey hairs and wrinkles, aging is a privilege, mostly. It's not, however, a guarantee that every aspect of it will be smooth sailing. As I age and see the changes in both my health and appearance, I'm continually reminded of how lucky I am to be alive in this time with the medical advances, technology, and treatments I have at my disposal. I know for a fact that my life-saving frontal obliteration wouldn't have been a possibility if I had been born just ten years earlier.

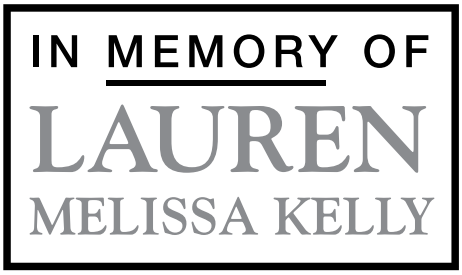
Mostly, I'm extremely grateful that my mom lived just long enough to celebrate my 40th birthday with me. And I am grateful for my dad, my husband, and a milestone birthday that was never a guarantee and likely a pipe dream when I was first diagnosed. Here's to you, Mom. ▲

Sydna Marshall is 41 years old and has CF. She is the President and Managing Editor of USACFA. She lives in Austin, TX, with her husband, Adam, and her two furbabies, Husker and Cutty. She's an avid reader, loves jigsaw puzzles, firmly believes that the heart of the home is in the kitchen, enjoys walks with her pups, and treasures her time on her yoga mat. Her contact information can be found on page 2.

The Higher Education (Formerly The Lauren Melissa Kelly) Scholarship

The application deadline for the Higher Education (formerly the Lauren Melissa Kelly) Scholarship is June 30, 2022. Any student seeking a degree in higher education, from Associate to Ph.D., is welcome to apply. We look for students who demonstrate tremendous academic achievement, community involvement, and a powerful understanding of how their CF—matched with these achievements—places them in a unique situation to gain leadership roles within the community. We believe that any higher education is a strong foundation for advocacy and involvement in the CF community.

Nancy Wech established this scholarship in honor of her daughter, Lauren Melissa Kelly. This semester's winners



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

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

TILLMAN continued from page 15

at a major academic medical center during 2010-2020 to determine the features of UTIs in 826 CF patients. They identified 108 UTI episodes during this period. Diabetes, distal intestinal obstruction syndrome (DIOS), and kidney stones were correlated with increased risk of UTIs. UTIs in CF patients were less likely to be caused by Gram-negative rods compared to non-CF patients and more likely to be caused by *Enterococcus faecalis*. The unique features of UTIs in CF patients highlight the importance of investigating non-respiratory infections to ensure appropriate treatment.
https://tinyurl.com/mr2xtye8

Two Anaerobic Bacteria Species Tied To Declining Lung Health In Adults

Two types of bacteria—*Porphyromonas pasteri* and *Prevotella nanceiensis* — were found to be highly prevalent in the mucus of adults with cystic fibrosis and associated with a progressive decline in lung health. These two bacteria differ from previously identified, disease-exacerbating bacteria in being anaerobic species, meaning they do not require oxygen to survive. Researchers evaluated anaerobic bacteria in CF sputum and its potential association with lung function decline over eight years. Sputum samples from 70 adults were analyzed and compared with patients' clinical data. Among the 10 most abundant anaerobic bacteria in the samples, *P. pasteri* and *P. nanceiensis* alone were associated with lung function declines, as measured by yearly changes in each patient's forced expiratory volume (FEV). Data showed the presence of *P. pasteri* or *P. nanceiensis* correlated with an average FEV loss of 52.3 and 67.9 milliliters each year, respectively, indicating a decline in lung health. The greater the abundance of either bacteria in an individual's sample, the faster the rate of decline.
https://tinyurl.com/yc8m4kg4

Higher BMI Associated With Favorable Clinical Outcomes In Patients With Cystic Fibrosis

In a systematic review and meta-analysis, researchers found higher BMI was associated with favorable clinical outcomes in patients with cystic fibrosis, urging reconsideration of the currently recommended target BMI, which in children older than 2 years, is at least the 50th percentile; in adults, the target BMI is greater than or equal to 22 for women and greater than or equal to 23 for men. However, BMI does not distin-

Continued on page 25

PHOTO BY ALEJANDRA CHAVERRI

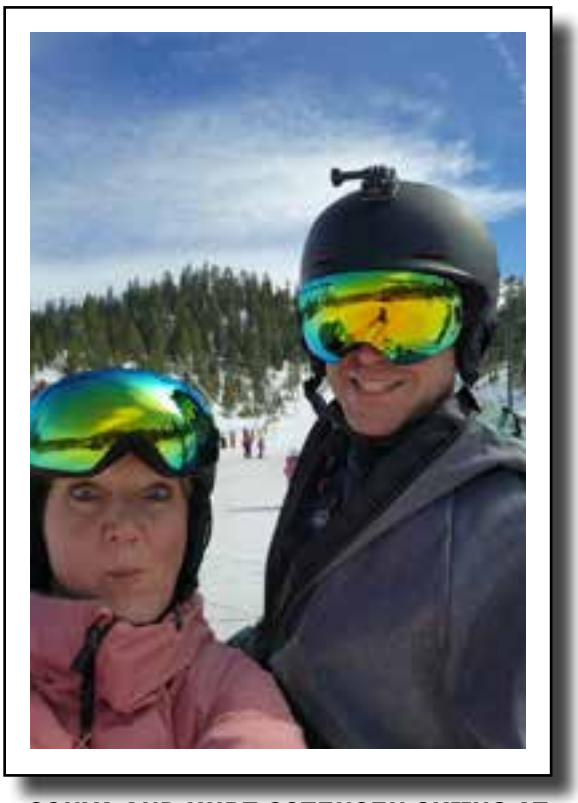
For My Granddaughter

Your world has never
Been on hold
You dive into the deepest
Water like the
Swimmer you were born
to be
Your grace under pressure
And all you must endure
Keeps me in awe
Your sparkling self dances
Into my heart with
Love,
Grannie

-E. Dougherty, 2002

FROM OUR FAMILY PHOTO ALBUM...

ROWAN O'BRYAN NEAR HER SCULPTURE "UNTITLED," AT THE UCLA 2022 UNDERGRADUATE JURIED EXHIBITION.



SONYA AND KURT OSTENSEN SKIING AT LAKE TAHOE, NEVADA, IN MARCH 2022.

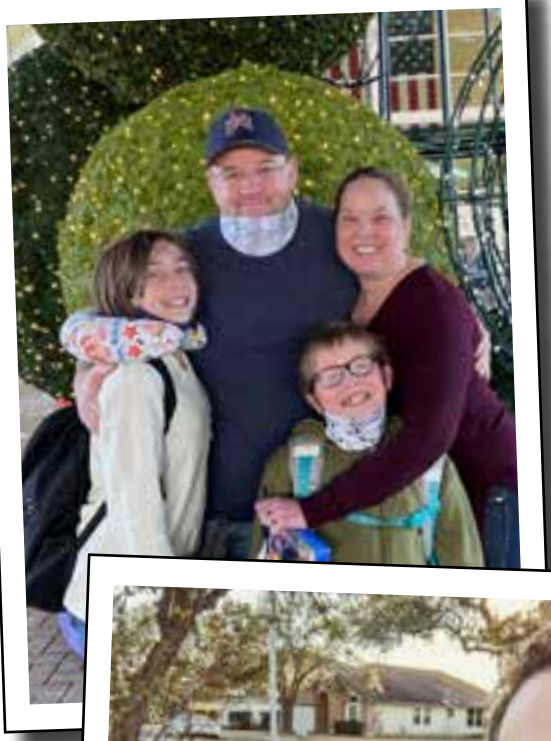
FAITH ETTLICH

ED AND HWI-JA CANDA VISIT THE OHIO STATE UNIVERSITY (ED'S ALMA MATER FOR M.S.W. AND PH.D. DEGREES) WHERE HE RECEIVED THE DISTINGUISHED CAREER AWARD FROM THE COLLEGE OF SOCIAL WORK IN 2021.

PAUL AND KATHY RUSSELL.

ELA CASTILLO IN NEW YORK CITY.

JENNIFER KYLE AND HER DOG, SCARLET.



ALEX GANTT WITH HER HUSBAND, DANIEL GANTT, NIECE KYLIE AND NEPHEW HUDSON RINGING IN THE NEW YEAR.

KIM AND ED NUNNARI.

SYDNA MARSHALL AND ADAM KEYS.

Aging With CF

By Kim Nunnari

Each quarter, when *CF Roundtable* lands in my mailbox, I sit down as soon as time allows and read it cover to cover. No matter the topic, each issue has articles that resonate with me and they're written by people like me. While I realize everyone has different experiences, there is comfort in knowing that each person who is generous enough to share their story *understands* what it's like to live with CF.

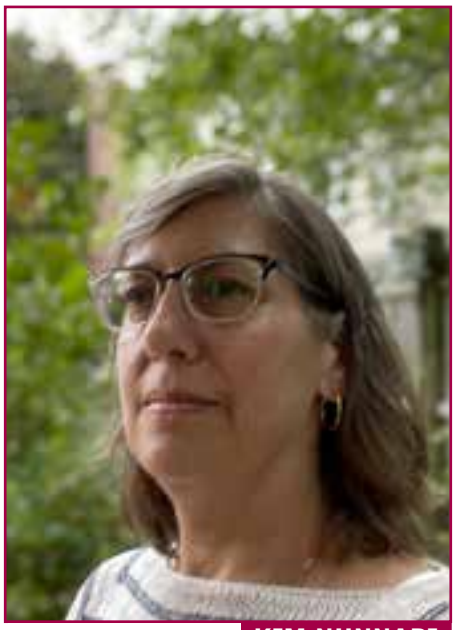
I appreciate the honesty and insight of everyone who has generously contributed over the years and have felt a vague sense of guilt for not doing so myself. When I saw the topic for Spring 2022—Aging with CF—I knew that I needed to sit down, write something, and finally contribute back.

I'm 57 years old. Although I wasn't diagnosed until the age of 19, I was sick throughout my childhood and adolescence and was misdiagnosed for many years. I grew up in a small town without a big medical center where CF might have been recognized earlier. I was diagnosed in 1984 and, despite the relief of finally knowing what was wrong, it was a terrifying diagnosis. There weren't any treatments other than IV antibiotics and chest PT at that time. In 1984, I had already outlived the life expectancy of 18.

I was able to connect with a few other patients through an international publication for CF adults, and it became apparent that one of our common coping mechanisms was a dark sense of humor. A favorite joke back then was that at least we wouldn't have to worry about wrinkles, dementia, or any of the other age-related problems that people without CF endure. Every cloud has a silver lining, right?

Even with my occasional dark sense of humor, I have always tried to do what I could to stay as healthy as possible, while living as normal of a life as CF would allow. In the 90s, Pulmozyme seemed like a miracle and helped space out hospitalizations. Hypertonic saline was another arrow in the quiver; before I stopped working, I would get up 30 minutes earlier to add them to my morning treatment regimen. The Flutter provided a bit of independence with airway clearance, but, unfortunately, I never really mastered the technique. Once the Vest was available, I found that was much more user friendly. Although I've never been one who enjoys exercise, I recognized early on the benefits in terms of airway clearance, fitness, and mental health. I have practiced yoga for 25 years, walk regularly, and have started doing high-intensity interval training in the last eight years to increase strength, endurance, and functional capacity.

> "It is a privilege to be able to experience getting older, despite having to put up with age-related degeneration (and wrinkles!) in addition to CF and its complications."



KIM NUNNARI

Despite my best efforts, living with CF for this long has taken a toll on my body. My intestines are cranky and don't work as well as they used to. My lungs are damaged from repeated infections. My veins are scarred. I have both CFRD and osteopenia. I also have a portacath. In the past few years, Trikafta has made a significant difference: fewer hospitalizations, no cough, improved quality of life, and more energy. This is almost as good as the cure we dreamed about back in the 1980s. What a gift!

My CF symptoms may have improved, but my youthful joke of being spared age-related issues hasn't come to pass. I had a complete hysterectomy after being diagnosed with uterine cancer in my late 40s. In my 50s, I have developed cataracts and arthritis in my hip. In the past year alone, I had a hip replacement and two cataract surgeries, both of which have improved my quality of life immeasurably. It's amazing to have a surgery that can actually repair or fix an issue, not just alleviate symptoms. I never anticipated any of this, but I'm not complaining; it is a privilege to be

able to experience getting older, despite having to put up with age-related degeneration (and wrinkles!) in addition to CF and its complications.

One of the richest experiences of having CF has been the connection with other patients. No matter the difference in age, geographical location, or health, there is an understanding and bond that is like no other. I am very fortunate to have a loving family, friends, and healthcare professionals who are incredibly supportive, but, try as they might, they aren't able to understand what it is like to go through life with CF. My gratitude extends to the CF friends I have had for years, with whom I can reminisce about the "old days" of sharing a hospital room during admissions. I am also grateful to those who are younger and bring a fresh perspective to living with the disease.

I'm incredibly grateful to be as healthy as I am at this age. Every life is full of ups and downs and mine is no exception, but I believe CF has given me a perspective and resilience that has served me well and will help me face whatever lies ahead.

Wishing everyone resilience, health, and happiness in 2022! ▲

Kim is 57 years old and has cystic fibrosis. She lives with her husband and two cats on the coast of Massachusetts, just north of Boston. They have a wonderful adult son who lives in New Hampshire. In the warmer months, you can find her outdoors in her garden or at the beach. When she's indoors, she likes to bake, knit, quilt, and read. She's looking forward to spending more time with friends once the pandemic is under control.

TILLMAN continued from page 20

guish between the major components of the body, namely, fat mass, fat-free mass, total body water, bone mineral density and bone mineral content. Studies of patients aged 2 years and older with cystic fibrosis with altered BMI or body composition were compared with patients with measured parameters in the reference ranges. The primary outcomes were pulmonary function, exocrine pancreatic insufficiency and cystic fibrosis-related diabetes. Compared with those with normal weight, researchers observed higher FEV1 among those who were overweight and obese. In addition, likelihood for cystic fibrosis-related diabetes and pancreatic insufficiency was higher among patients with normal weight compared with those with overweight. These findings suggest that nutritional status plays an important role in maintaining organ function in patients with cystic fibrosis. Because a higher BMI is associated with better clinical parameters, clinicians are advised to reconsider increasing the currently recommended target BMI. The use of a nutritional strategy that increases BMI, at least until the upper limit of normal BMI is reached, should be included in the daily protocol. The results suggest that careful evaluation of body composition should be incorporated into everyday clinical practice. However, obesity is a major global health concern and is associated with increased morbidity and mortality due to higher rates of heart disease, hypertension and diabetes. Thus, emerging concern over patients with cystic fibrosis who are overweight or obese experiencing these detrimental health conditions is justified, particularly in the era of modulator therapies. … Further longitudinal studies to address the consequences associated with overweight status in patients with CF to optimize nutritional approaches and treatment plans will be required to help guide further clinical recommendations.

https://tinyurl.com/5hd6372m

Women With CF Need Counseling To Preserve Fertility, US Study Finds

With continual advances in treatment, people with CF are living longer and those of childbearing age are increasingly choosing to become pregnant. CF itself can cause problems with fertility. In addition, many patients eventually require a lung transplant to maintain their health, and transplant recipients typically need to take immune-suppressing medications for life to prevent organ rejection. A common side effect of these medications is infertility; as such, CF patients may wish to explore options to preserve fertility — processes like freezing eggs or sperm, which can be later used to have children. A team of researchers conducted a two-part study to better understand what woman with CF know about fertility preservation. In its first part, the team surveyed 50 women with CF between the ages of 18 and 35 and recruited nationwide. Most of the women reported being in a relationship. The majority were not mothers, but most said that they wanted to have their own children in the future. Despite their desire for a biological child, nearly three out of four survey respondents reported never discussing fertility preservation with their healthcare providers. About one in three said that they had never discussed options for parenthood with anyone. The women generally had little knowledge about options for preserving fertility. The researchers noted that women who had conversations about fertility tended to be more knowledgeable about its preservation. Married women also tended to be more

Continued on page 27

Growing Old Is More Than I Ever Expected

By Sonya Ostensen

SONYA OSTENSEN AND HER DOG, TALA.

At 47 years young, with respect to CF, I am the healthiest I have ever been. Obviously, there were many times in this journey I would have said there is no way I would make it to this point. And yet, here I am attacking life with a vigorous energy: working out, raising our now eight-year-old daughter, belly laughing, working, traveling, taking care of the dog and three cats, volunteering, cleaning the entire house without wheezing and hacking up green goblins, sleeping without coughing, running around with my daughter and her friends, and breathing deeply. And yet these happy events have, paradoxically, taught me to expect the unexpected—because there is one thing we can count on, and that is change. Therefore, I am soaking this up and going full speed ahead while it lasts, regardless of age.

Nonetheless, growing old is proving to be interesting. Obviously there are the annoying joint aches and the beginnings of hearing loss, possibly resulting from years of antibiotic and steroid treatments. Lack of circulation is also a bit concerning—my feet and legs tend to go numb and tingly throughout the day, both during activity and while being stationary.

For now, the two biggest challenges of age and CF are vision damage and menopause. I currently wear bifocals to help with astigmatism and nearsightedness, in addition to having regular screenings for glaucoma. However, in the past two years, I have also developed an abnormal neurologic pupil dilation. The right pupil dilates to an unusually large size while my left pupil is sluggish and remains small. Unfortunately, this impairs my ability to read as the words begin to multiply and blur together almost daily. Fortunately, all scans for brain tumors and pinched optical nerves are negative. The neuro-ophthalmologist I see recommended I take beta-carotene, a precursor for vitamin A, which is already in my CF ADEK vitamins. The cause of this optical phenomenon remains enigmatic; however, I refuse to give up and continue to expect the unexpected.

I hate to bring up the elephant in the room but, whew, perimenopause is a *big* one that tends to sneak up with impeccable timing: I'm dealing with our second grader when she's testing the boundaries; I'm having a technology breakdown during an important project; or I'm just hearing the annoying sounds my husband makes when he chews (poor guy, he never knows what is in store for him). At such moments, I suddenly feel there is a volcano erupting in my very being and I am unable to control its explosion. Any small annoyance and *boom*! It can trigger this loss of patience. It is not pretty, just as night sweats are not sexy. When I realize I am unable to control my inner demons, I am repelled by my own actions and feelings. At that point, I become my own worst enemy and double down on angry self-criticism. That's when the psyche is its own worst enemy and causes me to recoil. Of course, this withdrawal is not opportune as life demands that we are always on point. This is where I struggle to control my rage of imbalanced hormones and try to appear normal and functioning. The aftereffect almost always results in depression. Just when I started feeling like a confident woman who knows what she wants, menopause slaps me in the face as if to say, "take that!"

Of course, I realize that as I write this, I am not the only woman who is going or has gone through this change. I am not convinced menopause is

much different in an individual with CF as opposed to anyone else, except that a lot of women with CF are living long enough to experience this life transition that, is ultimately a gift. After my CF care team determined that avlimil (a natural supplement with low-potency ingredients like black cohosh, soy isoflavones, and valerian root) is not compatible with my modulator medication, I chose not to take any hormones or supplements and just try to deal with this stage of aging as naturally as possible. Thus, for now, I am just working on my patience (or lack thereof) with diet, exercise, camaraderie, sleep, meditation, crying, letting go of what others think, and sometimes laughing at myself during these hormonal episodes.

Aging is nothing to fight against—it will happen regardless of whether we are on board with it or not. I believe as women—especially as women with CF—we can grow with this transition; it may not be elegant and it's certainly not always fun, but we can do our best to appreciate this new challenge. At the very least, we can expect the unexpected because you never know what miracle might be around the corner that may open more opportunities to continue our individual missions and enjoy this life a little longer. ▲

> "Just when I started feeling like a confident woman who knows what she wants, menopause slaps me in the face as if to say, 'take that!'"

Sonya Ostensen is 47 years old and has CF. She is the interim Secretary for USACFA. She lives in Melbourne Beach, FL, with her husband, daughter, dog, and three cats. She holds a Bachelor of Science in Environmental Sustainable Resource Management from Ohio State University. After working in environmental health, she retired due to CF complications. Sonya has a passion for wildlife rehabilitation and loves to travel with her family and experience new cultures. Her favorite activities include gardening, baking, walking the beach, and climbing trees with her beautiful daughter. Her contact information is on page 2.

TILLMAN continued from page 25

knowledgeable. In the study's second part, the researchers interviewed 20 women with CF, and summarized some of the broad themes that emerged from the discussions. One major theme was that many women with CF are not adequately knowledgeable about fertility and its relationship to CF. The patients commonly reported that medical resources were scarce, and they often turned to other patients or social media for guidance. The women also noted that fertility in general was not viewed as a priority by their healthcare team. The women recommended that CF care providers need to be educated about fertility in CF, and take care to bring up these issues with patients. The women stressed the importance of clinicians viewing patients as complete human beings with unique wants and needs, and not as merely a collection of symptoms to be managed. The interviewees also highlighted the importance of incorporating women's healthcare into CF healthcare, and vice versa. Findings from the study support that additional education is needed for women with cystic fibrosis who are considering parenthood. Clinical care models should include early, regular, and thoughtful discussions about reproductive health issues, including fertility preservation.
https://tinyurl.com/yaokkx7l
AND
https://tinyurl.com/2p8ph3dm

National Jewish Health Pulmonologist Guides Development Of Innovative Tool To Help People Of Color Screen Their Own Symptoms For Cystic Fibrosis

Cystic fibrosis is most common within the white population, but it affects people of all races and ethnicities. The recognition of CF in Black individuals and people of other racial and ethnic backgrounds has been markedly inadequate. Following the virtual North American CF conference in the fall of 2020, Dr. Taylor-Cousar met Terry Wright, an Arkansas resident in his 50's whose symptoms were misdiagnosed as asthma in combination with common gastrointestinal problems for decades. After Dr. Taylor-Cousar was introduced to the couple, they discussed the possibility of developing a free online tool that would be accessible for anyone to self-screen for symptoms of CF. She offered her medical expertise to create and refine the tool that could help reach the broader population. Then, Dr. Taylor-Cousar and the Wrights presented the tool to the Cystic Fibrosis Foundation's (CFF) education committee, which found that it meets their guidelines and standards of an effective screening tool for public education about CF. Now known as the Wright Cystic Fibrosis Screening Tool©,

Continued on page 30

Aging Gracefully

By Andrea Eisenman

Every time I turn around, my husband reminds me that I am old. What he means is that I can no longer expect to easily do what I did in my youth. I understand that. I do not expect to play tennis or swim like I did in my 30s. I know that the last few years brought many new health challenges. I did not get this far in life to say, oh, well, I guess I can no longer do this or that. I should just give up? No way!

When I complain I am tired, he says, "you are old." When I comment that my back hurts, he says, "it's because you are getting older." Yes, I am old-ish and certainly old for someone with CF, but he doesn't take into account that I am being treated for a recurrence of Post-Transplant Lymphoproliferative Disease (PTLD), which causes fatigue because my Epstein-Barr virus (EBV) load becomes high. This is common when someome is immune suppressed and their own body cannot fight off the EBV. The oncology team felt that I may have gotten EBV from my lung donor 22 years ago.

I had a spinal fusion at age 16 and now have resultant back pain. Most of my back is fused, and playing tennis with the necessary torque can cause back pain. It doesn't help that my fusion is separating from my lower spinal column. Plus, I also have degenerating discs and arthritis, both of which can cause extreme nerve pain that radiates into my hips. However, the way I feel during and after I play tennis brings me such joy that I want to keep playing for as long as possible. I play tennis with many women, most of whom are 20-25 years older than me, and they always call me the young'un! Obviously, age is relative. Until recently, they said they were

ANDREA EISENMAN

amazed watching me dash around the court after tennis balls while wearing two leg braces (I had a fractured right knee and a meniscus fix on my left knee). Thankfully, these braces assist me in not injuring myself further. People see the braces and immediately assume I do not move much. I love proving them wrong. Although, I hate to admit it, I am slowing down and do not run for everything any longer. Tennis has been on hold for me since July 2021 when radiology placed a PICC line in my right arm to treat the PTLD. And I am right-handed. I am just starting to wade back into playing. I'm not back to playing a full game, but I'm active in a tennis clinic where an instructor runs several women through drills and eventually we play mock games or "queens of the court." After I played last week, I felt great mentally but knew the blow back on my body might be intense. I am happy to write that I was only a little sore—nothing that I don't sometimes experience, anyway.

Still, when my husband says I am old, I do what I need to do to function daily. It is a big effort to constantly monitor how I am feeling—do I need to call my doctors or what I can do to help myself breathe better or how I can build up stamina to be able to play tennis again? These are things I focus on in the moment. And I remain grateful I am still alive.

At my age and with my physical limitations, I know in order to enjoy these kinds of physical exertions without the ensuing pain, I have to put more work into my aging body prior to stressing it. This is a change from 15 years ago. I have been going to physical therapy and learning exercises to support my core and build up my glutes, quads, and hamstrings to keep me strong while I was unable to play tennis or swim due to my PICC line. Eventually, I began to focus on what I could do to get into better shape for when I could play again. This was tedious as I was fatigued for at least four to six months when I was in treatment for PTLD and my EBV was high. Thankfully, I was in a clinical trial and I received infusions of cytotoxic T-cells that were taken from donors whose own bodies were besieged with EBV. These cells were administered via PICC line through a 50-ml syringe push with the hope that the T-cells would target and kill the EBV in my body. Even though my EBV was dropping, receiving these cells each week was extremely tiring and I barely had the energy to do much else aside from eating, sleeping, napping, and, at times, cooking. Exercise was an afterthought and something that was just too much to consider.

> With my physical limitations, I know in order to enjoy these kinds of physical exertions without the ensuing pain, I have to put more work into my aging body prior to stressing it.

Eventually, I could see that my infusions were going to be ending soon and I forced myself to get back on my stationary bike. I was winded and discouraged. But I made myself find the time, after a nap, as many days a week as possible to just ride and not judge how I felt. I also forced myself to spend time on my mat on the floor with focused muscle-group-strengthening techniques. I was amazed at how fast my body had lost its strength during the prolonged treatment.

I went into menopause after my birth control pills were stopped in 2017 due to a deep vein thrombosis—this likely contributed to my muscle weakness. The birth control pills were used for maintaining bone health and keeping osteoporosis at bay. I was put on a bone-building medication called Alondronate (generic for Fosamax) to help improve my bone density. However, since stopping the birth control, my muscles no longer react the same and I feel generally weaker. I do not feel I can build the same muscle I had before menopause. And I am more blobby in the middle—dare I say, paunchy?

Each year I am alive, I see that most physical things do get harder. But do I give in and just not do them? Or do I try to find a workaround to keep doing what I love? No and yes. Staying active is important for me physically but also mentally and socially. I have a lot of great friends whom I have met through playing tennis. They keep me going.

Prior to my lung transplant, I was lucky if I could walk on a treadmill for 20 minutes while wearing supplemental oxygen. Now that I am old(er) and hopefully wiser, I try to manage my expectations and be more realistic with physical goals. And I have to be careful with my osteopenic bones when I am active. I don't want to fall and fracture anything. Regardless of what my husband tells me about being old, I am so grateful I am able to be active and enjoy the life I have— limited as it may be compared to my younger self and to my healthy peers. I am just trying to make the most of it and constantly learning to adapt. ▲

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.

CF Roundtable Speakers Bureau

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

Please contact us at cfroundtable@usacfa.org if you or your organization would like to request a speaker. You can find more information on our wonderful speakers and topics on our website: https://www.cfroundtable.com/speakers-bureau. If you have CF and are interested in joining our Bureau, contact us at cfroundtable@usacfa.org!

Voices from the Roundtable

Attending College With CF: How Scholarships Made Music School A Reality

By Faith Ettlich

College? I'm not sure I want to attend a four-year college...that doesn't sound appealing in the least. These were some of my first thoughts in high school when the topic of "college" was brought up. After an entire education of homeschooling, I knew I wanted a higher education; however, with my family's military career, the idea of staying in one town for four years was an extremely new concept. After all, I had never lived in one place for more than two or three years. It took quite some time before I realized that all I had ever wanted to study was music, and it was the education pathway I needed to pursue. Once I came to that realization, I ambitiously applied to schools and scholarships. And now, thanks to scholarship programs, like those offered by USACFA, I am able to attend college.

FAITH ETTLICH

I attend Gordon College, located in Wenham on the North Shore of Massachusetts, and am getting my Bachelor of Music in Piano Performance. I love the school as it is a small, Christian, liberal arts college and focuses on students and their individual achievements. It fosters a very personal atmosphere and brags a small population, making it an intimate experience. This was important to me as I knew my family would probably not always live close to me. That prediction came true—the day my first semester ended, I received news that my family would be moving from Maine to Hawai'i in three months. My community of friends and support of professors at Gordon, however, is a blessing, and I look forward to my next three years here.

The college and career path I have chosen is rigorous and competitive. My life currently consists of dozens of hours of practice a week, a 20-credit workload, and two music ensembles (not to mention my honors requirements). Every school day is filled with three to five hours of practice, an hour of rehearsal, four hours of classes, and homework! Because of this, I rarely

TILLMAN continued from page 27

the tool is being shared with CF care teams through the CFF and with the general public on social media. The new tool will help people self-identify symptoms that could be related to CF and aid medical providers in identification of potential individuals with CF, especially those who are Black, Indigenous, and People of Color (BIPOC). Once the screening is completed and the symptoms are consistent with CF, the individuals can take the information to a doctor, who can refer them to get a test to confirm if they have CF.
https://tinyurl.com/y6vzgv7p

How Do Patients Decide On Treatment For Cystic Fibrosis Exacerbations?

In a study of patient decision making involving people with cystic fibrosis, researchers found that concerns over difficulty or pain with breathing had the greatest impact on patients' decision making with respect to the treatment of pulmonary exacerbations, and that gastrointestinal problems also figured prominently in influencing treatment decisions. Given the varying toxicities and burdens of therapeutics for cystic fibrosis, researchers sought to determine how these patients valued different aspects of their health status and the trade-offs they made when choosing between alternative therapies. Toward that end, researchers conducted a discrete choice experiment survey (DCE) in order to quantify how those with CF and their caregivers hypothetically viewed the relative importance of outcomes resulting from treatment of CF pulmonary exacerbations. Participants were asked to make a series of hypothetical decisions about treatment for pulmonary exacerbations to assess how they make trade-offs between different attributes of health. The outcomes studied were: breathing difficulty/pain, fatigue, pain unrelated to breathing, lack of motivation/demoralization, reduced appetite, presence of fevers/night sweats, high treatment burden,

have free time and certainly have no spare time to work to earn money to pay for college. This is the main reason I am so thankful for the Arts Scholarship offered by USACFA; without this scholarship and others, I would feel much pressure to earn extensive money for school in order to graduate free of debt. Instead, I can dedicate my time and energy to an extremely demanding major without the major burden of potential college debt.

After college, I hope to move on to a graduate program in piano either here in the States, or potentially in Europe. I would love to accompany for ballet and opera company rehearsals, teach private lessons, perhaps fill in when needed in an orchestra, and, of course, perform. With this said, it is imperative that I continue pursuing quality education in order to be competitive in the extremely intense industry—the world of music.

For now, I stay healthy by consistently doing my treatments, attending routine doctor visits, and taking my medications daily. Even with CF, I can participate in "college life" and be as productive and accomplished as ever. Additionally, I can focus on my intense workload and enjoy the area in which I live. When I'm not studying or practicing, you can find me having a good laugh with my friends, teaching piano lessons in town, attending church, trying every café near me, planning another trip (it looks like Hawai'i and London this summer!), or spending the evening in Boston to see a show at Symphony Hall.

In high school, it was clear that I would be mostly responsible for taking care of my college bill, and, thankfully, here I am, attending a liberal arts college in New England, studying what I love. Thank you USACFA, for the amazing scholarship, and thank you for supporting cystic fibrosis students all across the country! ▲

Faith is 19 years old and has CF. She currently attends Gordon College near Boston, MA. Faith was one of first two people to receive the Scholarship for the Arts in 2021. When she's not studying or practicing piano, she enjoys traveling and exploring the local area.

inability to meet goals, sputum production and clearance, and gastrointestinal symptoms. The study found that for people with CF, breathing difficulty/pain had the greatest influence on their preferred health outcome states followed by gastrointestinal symptoms, then presence of fevers/night sweats. For caregivers, the greatest influence on their preferred health outcome for their child was breathing difficulty/pain, followed by pain unrelated to breathing, then gastrointestinal symptoms. The researchers concluded that the health outcome preferences reported here should guide decision-making when seeking agreement on personal goals of therapy between patients and clinicians for treatment of pulmonary exacerbations.
https://tinyurl.com/yckxrerp

CBD And Cystic Fibrosis - Can Cannabidiol Help?

Clinical interest in cannabis and CBD is rising, and the data in favor of its healing power continues to trickle in. CBD even holds the potential to alleviate some of the symptoms of cystic fibrosis. CBD, short for cannabidiol, is a natural cannabinoid that can be found in the Cannabis sativa plant. It is non-psychoactive, meaning that CBD is

Continued on page 36

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

CF: THE MIND GAME

Perdition To Salvation: The Road Less Traveled

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

Aging. I'd rather be tortured by aliens with a penchant for sadism than think about it. In fact, to be fully transparent, I can barely stand when others mention it, let alone complain about it. Sometimes I imagine that hell is actually spending an eternity locked in an uncomfortably cramped space listening to healthy people complain about aging, like a bad remake of Jean Paul Sartre's play, *No Exit*.

Aging repulses me; but before I explain why, I have to warn you there is no sexy or compelling way to write about aging. However, to quote my favorite professor's advice on teaching, I will attempt to "do well, be funny, and, failing that, at least give a good book recommendation." Given the topic, I will most likely fail on the first two counts, but hopefully you'll at least get a good book recommendation if you hang in there until the end.

I have spent hours mulling on this topic, including questioning whether my visceral repulsion to it is some sort of antisocial, inhuman, sociopathy; however, I ultimately concluded that my reaction is quite simple. It comes down to this: I cannot fathom complaining about aging as someone whose entire existence has been spent striving to age. I have spent a lifetime trying to extend my time as the sands slip beneath my feet in the hourglass of my existence even as my hold on this life fades and my physical faculties falter.

However, aging with CF has many paradoxical, easy-to-overlook rewards as well. In five decades, I've survived long enough to earn many distinguished service medals, including diabetes; hearing loss due to aminoglycosides; sight loss due to diabetes; early-onset chronic arthritis; and my two most recent prized trophies—chronic pain in my right lung and acute digestive problems, which are untreatable with enzymes.

As some of you may know from prior columns, I lived in a halfway house in the Alternatives to Incarceration Program (ATIP) program in Onondaga County, New York, while I attended college and played hockey and rugby. During rugby practice one cold fall afternoon, I got into a fight and broke one of my teammate's wrist. Immediately, the halfway house got a call to have me report to my probation officer (PO) downtown where I expected him to cuff me and escort me across the street to jail to await transfer to an upstate corrections facility. My best friend at the time, who is currently doing a 20-year stint in a California penal institution, offered to drive me, which, inexplicably, the halfway house permitted. On the way, we had what you might call a serious existential discussion. He started the conversation abruptly with an offer to buy me an 8 ball (a heroin/cocaine cocktail that's taken intravenously) so I could overdose in order to avoid the eight-year stint I was facing. I was shocked and flattered he would consider spending nearly all his money to give me the option, but I couldn't take him up on it. At the time, I do not believe my mind had the tiniest cluster of neurons in the frontal lobes of my cerebral cortex to begin to fathom such a notion. Considering all the fighting I had done just to age into my 20s, the thought of giving up was cognitively walled off from consideration. Exasperated, he exhaled loudly and said, "what the hell are you going to do upstate with CF?" I thought about it for

> *"One very dark, desperate night I withdrew the gun from my mouth and veered back onto the road less traveled, not because I feared or couldn't fathom giving up but because I had grown to love the fight."*

MARK TREMBLAY

a second and responded much more calmly than I imagine he expected, "I don't know…I guess I'll be the only CF patient in prison…what other choice do I have?" Visibly upset, he half shouted, "how long do you think you'll make it up there, man?" I quickly shut down the conversation by saying, "I really don't know but one thing's for sure, I'd rather take my own road (the road less traveled) than the easy way out you're offering." As you can probably imagine, the rest of the 15-minute car ride felt like two hours and was spent painfully, yet perfectly, silent until he let me out, at which time we neither shook hands nor hugged, but rather parted ways like strangers who had never so much as seen each other before. In the end, the whole incident was blamed on a rough scrimmage and charges were never filed—my PO had no grounds for writing up a probation violation.

More than a decade later, after years of therapy, in the middle of a messy divorce, bankruptcy, and a lengthy health crisis, my mind had finally softened enough to weigh more thoughtfully the option of taking the easy way out, but once again I did not. Due to God's grace and mercy, one very dark, desperate night I withdrew the gun from my mouth and veered back onto the road less traveled, not because I feared or couldn't fathom giving up but because I had grown to love the fight and, even more than that, the person the fight made me. I simply longed to age more even though it most certainly would entail more time in the trenches battling CF and other challenges as I aged into my 40s.

A decade later, I made it to yet another penultimate milestone in my life—a day I never thought I would see. On the day I finally received my first dose of Trikafta, I felt an unparalleled sense of personal relief, but my sense of relief extended far beyond that. To contextualize this, I, like thousands of other patients, had prayed my whole life for a miracle that would lift the cross of CF from my shoulders. However, even through many intense prayer sessions, I harbored a deeply held reservation that I dared not admit to myself nor share with others. The truth was, in my heart of hearts, I did not merely want a personal miracle; I longed to see all CF captives set free so the remaining 10% of CF patients who can't take Trikafta aren't left behind. On October 21, 2019, the day the FDA approved Trikafta, I pulled over on my way home from work and uncontrollably wept for what seemed like hours until I noticed it was pitch dark outside. I had never experienced such overwhelming joy. I knew that God had heard the secret desires of my heart by following through on the impossible promise that I never actually voiced to Him. Trikafta would not only provide me relief, but it would also provide relief to tens of thousands of fellow warriors and prevent still tens of thousands more young, would-be warriors from ever setting foot on the battlefield. When I finally pulled back onto the highway again, I took such a deep breath I coughed vigorously when I exhaled and then I softly whispered, "okay, God, you have done more than I ever could have asked or imagined so you can take me home now." At that moment, I recalled Simeon who saw his life's desire fulfilled the moment he held the infant Jesus in his arms: "*Sovereign Lord, as you may have promised, you may now dismiss your servant in peace. For my eyes have seen your salvation…" Luke: 2:29-30.*

In the past two years I have aged greatly. I can no longer read a hardcover book due to diabetic-related progressive sight loss, my right lung hurts so much it makes me wince if I move suddenly, and the gastrointestinal side effects of Trikafta cause nearly constant abdominal bloating and pain. Another unexpected side effect of Trikafta for me is that the nominal bump in PFTs meant I was no longer eligible for disability or early disability pension so I will be stuck indefinitely trying to hold down a fairly demanding job while living at a functional level as bad as, if not worse than, before Trikafta. Nevertheless, I am forever grateful to have aged long enough to see Trikafta and how it has brought hope to thousands of fellow warriors.

Just a few closing thoughts on this terminally unsexy topic—if you want to avoid aging, at least in spirit, don't ever let yourself believe your best is behind you. Wear your scars and badges with humility and gratitude and live in a state of perpetual expectancy that something wonderful is unfolding even if you are blind to it. Finally, as promised, if you, like me, have lived life traveling down "The Road Less Traveled," read the book by the same name by M. Scott Peck. His novel spent 10 years on the *New York Times* Best Seller list, sold over ten million copies and was translated into 20 languages. I promise you will not regret it. ▲

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

IN THE SPOTLIGHT

With Alex (Alexandra) Gantt

By Jeanie Hanley, M.D.

We were thrilled when Alex contacted us to be interviewed. Having been secretive about having CF for most of her 36 years of life, Alex's most recent mission has been to spread the joys and benefits of talking about CF to whomever will listen. And we obliged! She was diagnosed with cystic fibrosis at nine months old and was raised in Austin, TX. She and her husband of 16 years, Daniel, and their black lab, Bella, now live in Colorado. Alex has an extraordinarily open and charming take on sharing all aspects of life and CF. Please welcome our newest star, Alexandrea Gantt. Spotlight, please!

Are you eligible for CFTR modulators like Trikafta?

Yes. I started taking it at the end of December 2019. Prior to Trikafta, I was never eligible for any modulators, so this was a game changer for me.

What benefits have you noticed while on Trikafta?

I've definitely noticed less coughing! I feel so incredibly quiet now and my friends and family certainly notice. If we get separated in public, my husband can no longer find me by following my cough! In Texas, I was on IV antibiotics five to six times a year. After moving to Colorado, it was three to four times a year. Now that I'm on Trikafta I still consider it a win with only once or twice a year!

What side effects have you had?

I've experienced major weight gain plus a debilitating headache a few times a month. I did have the rash in the beginning and had to take Benadryl for over a week to combat it. I also had dizzy spells for over 70 days, but they finally faded.

ALEX GANTT HIKING IN THE SNOW WITH HER DOGS IZZY AND BELLA.

Why did you move to Colorado?

For the last few years that I lived in Texas, I was on IVs pretty much every other month. We went to visit my friend in Denver and I could breathe better with the dry air. It was significant enough that my husband and I decided to move here. Now when I fly back to Austin (well, pre-COVID-19), the humidity immediately makes my lungs tight and I almost always have a CF exacerbation once I'm back home.

Why were you closed off about sharing that you had CF?

Growing up, I never really had to tell anyone about having CF. In school, everyone always just knew (mostly from another student who had CF). Anyone I met outside of school was usually a friend of my brother so they already knew about me. After high school, it felt nice to keep a part of me "hidden" when I met new people and it almost became an obsession to see how long I could go without revealing that part of me. It usually wasn't long though because I would inevitably get sick and need to go to the hospital.

In high school, I met a few people with CF through the message boards on CFF.com and became very close to one of them in particular. When she passed away suddenly, I was devastated. I decided I would never befriend anyone with CF ever again to protect myself from that pain. I truly thought I was helping myself, but, in reality, I was actually hurting myself by denying those friendships.

What led to you opening up about your CF?

I did a speech about not having kids for my CF clinic, which led to someone reaching out about speaking at a mini-con, which led to me facilitating a session and meeting other people with CF who just "got it." From there, I threw myself into the community and have never looked back. I now have many close friendships and I can't imagine not having them in my life.

What would you say to others who are not open to talking about their CF?

A good place to start is by attending virtual events, such as CFF MiniCons, offered by CF organizations. I know how scary it can be to take that first step, but I have never regretted it for a moment. I know that there is a chance I could lose a CF friend again, but I have finally realized that I would rather have known them and lost them, than to never have known them in the first place.

How else are you involved with the CF community?

I frequently participate in the work groups for CFF's BreatheCon and FamilyCon. I am also a mentor for CF Peer Connect. In the beginning of 2020, I was featured on a podcast and shared my CF story, which was completely out of my comfort zone but very rewarding! I will also be one of the cochairs for 2022's FamilyCon.

What profession were you in and how was it affected by CF?

I was in HR for almost ten years. I loved it, but I was always putting work before my health. I finally had to stop working. It was a really hard adjustment at first and I especially hated meeting new people because I knew they would ask the inevitable: "So, what do you do?" I don't want to share with people I've just met why I don't work, so I found myself just stammering and changing the subject. I kind of resent that because, if I had kids and was a stay-at-home mom, no one would bat an eye—you can't just stay home as a wife in this day and age. So, I started saying I'm a housewife because I find it amusing to see the confusion and/or judgment in their faces. It's the little things that give me pleasure in life, I guess.

What effect/impact has the COVID-19 pandemic had on you?

The pandemic has been really hard on me, but I know I'm not alone in that, which helps. I didn't leave my house for 15 months and then I had the summer of 2021, where I actually lived life again and thought everything was on its way to being over. Then the Delta variant hit, followed by the Omicron variant, and I have again been homebound ever since. Luckily, I keep very busy! I have a virtual CF group that meets every Wednesday, a monthly Bible study that I run, a writing group that I help run, and a book club, all virtual. I also FaceTime with my niece and nephew and watch 80s kids movies over Zoom with some of my high school friends.

We recently had to put down our beloved rat terrier, Izzy, and it was heartbreaking. After losing her, however, I realized how lucky I really was to have had almost two full years of quality time with her because of the pandemic. I will never regret having that time with her, and, because of that, I don't have the same harsh feelings over being stuck at home anymore.

Who is your greatest support?

My husband. I know it's cliché, but he is my best friend and I couldn't do life without him.

Any hobbies?

I am an avid reader! With all my treatments (and the fact that I read very fast) I always log over 200 books a year on Goodreads. I also love jigsaw puzzles, hiking, board games, crossword puzzles, and playing video games, especially with my niece and nephew over FaceTime! We bought an RV this last summer, and now my new favorite thing is taking the RV out to camping sites and just getting away from it all.

Do you have a funny CF story?

When I was 16, I had to be hospitalized for a tuneup. When I returned back to school, a boy from one of my classes came running over to me and said, "I'm so sorry, I had no idea you had hemophilia!" (!?!) I just kind of stared at him and said, "Neither did I." Turns out, one of my teachers told the whole class that I was in the hospital for hemophilia, and, let me tell you, that was a fun rumor to handle.

How do you cope physically and emotionally?

I am a big proponent of therapy and I also started an antidepressant about two years ago. For so many years I was opposed to taking something for my clinical depression, mainly because all I'd ever heard were horror stories about SSRIs. Now that I've been taking it and it's been so amazing for me, I want to scream it from the rooftops so other people know it's a real option and they don't have to be scared of it! I wish more people had been open about it working for them so I wouldn't have been resistant to trying it for so long.

Who or what inspires you?

My niece and nephew. I love watching them grow up and figure out who they want to be in this world, and they continue to surprise me every day. They are the funniest, smartest, and sweetest kids I know. I am blessed to have such an incredibly close relationship with them. We get them for a few weeks every summer and spending time with them is one of my favorite things in the world.

Anything else you want to add?

A few years ago, doing an interview like this would have been unheard of for me. It's kind of funny to look back and remember how closed off and shy I was, because now I'll tell complete strangers that I have CF! I wish I could go back and tell myself that I didn't need to hide. I hope that anyone who has the fears that I did will hear my story and change their mind. ▲

Dr. Jeanie Hanley is 58 years old and has CF. She is a director and former president of USACFA. She lives in Los Angeles and welcomes your comments. Her contact information is listed on page 2.

If you would like to be interviewed for our "In The Spotlight" column, you can fill out the form on our website. Alternatively, you can contact Andrea Eisenman, Jeanie Hanley, or Xan Nowakowski directly. Their contact information is on page 2.

Mailbox

"Your quarterly publication is a godsend to people with cystic fibrosis and their families by providing them with valuable information regarding the disease, latest treatments, research and, most importantly, a community that understands. So grateful to you!"

Marina Forstmann Day
Los Angeles, CA

"Thanks to all the improvements, more CF patients become adults, and dealing with the demands of adult life along with CF is a huge challenge. Thanks for helping those patients. The Wise Foundation is happy to invest in the future of CF patients through your cause. Keep up the great work!"

Scott Knight, parent of an adult with CF
Tyler, TX

TILLMAN continued from page 31

devoid of the 'high' that cannabis' mind-altering molecule, tetrahydrocannabinol (THC), is known to cause. Both cannabinoids – CBD and THC – have been shown to have powerful therapeutic action, either when administered alone or in combination. Cannabinoids interact with our endocannabinoid system (ECS), a complex cell-signaling network in our body that helps to regulate our mood, immune function, appetite, sleep, memory, pain, and much more. CBD can indirectly modulate the function of our cannabinoid receptors, which can be found all over the brain and body – including within the airways. Back in 2002, a paper published in the Journal of Cannabis Therapeutics detailed a theory that cystic fibrosis causes a deficiency in endocannabinoids and "by elevating these levels, symptoms may improve". CBD indirectly increases levels of anandamide, an endocannabinoid that is produced naturally in the body. By this token, CBD could supplement the ECS and subsequently improve CF symptoms. Since this paper was published, the effects of THC administration have harbored positive results in animal models of cystic fibrosis. There is existing evidence to suggest that CBD can reduce pain and inflammation, support the immune system, and relieve gastrointestinal issues in countless other clinical conditions. CBD will not be a cure for CF, but it could, in theory, help to relieve some of the symptoms, without the unpleasant side effects of existing drugs. CBD has potent anti-inflammatory action. It has been shown to reduce inflammatory pain and reduce the severity of the inflammatory response in numerous experimental and clinical studies. Inflammation is a major driver of cystic fibrosis symptoms, so CBD could help to relieve inflammation and inflammatory pain in the lungs and the digestive system in patients living with CF. Cannabis is a well-known appetite stimulant. This is largely due to THC, but there is some evidence to suggest that CBD can also stimulate appetite. CBD has also been shown to suppress nausea in animal models, which suggests that CBD could support those with CF who struggle to eat. Cystic fibrosis is a hugely debilitating and isolating disease that can cause anxiety or depression in many patients and their families. Where CBD surpasses other pain relief medications is its ability to support our mental health. By interacting with serotonin receptors in the brain, CBD can help to support the signalling of serotonin. This is thought to be why CBD can relieve anxiety and depression, as demonstrated by various human and animal model studies. Currently, the only evidence in favor of using either CBD or cannabis to manage symptoms of cystic fibrosis is anecdotal. For now, there is no clinical trial evidence. While many people choose to self-medicate with cannabis-based products, there is insufficient data to support its safety or efficacy in managing CF symptoms in the clinic. As a supplement, CBD is considered to be safe and well-tolerated. The side effects of taking CBD are rare and mild. These can include nausea, fatigue, and appetite changes. This is, obviously, a potential contraindication of using CBD to manage CF, since problems with appetite and digestion are common symptoms. There is no definitive answer for how much CBD to take for cystic fibrosis, or how to take it and researchers can't say for certain whether CBD could improve symptoms of cystic fibrosis.

https://tinyurl.com/bdzxtp4j

Ivacaftor, Tezacaftor, Elexacaftor (Kaftrio) In Combination With Ivacaftor (Kalydeco): Risk Of Serious Liver Injury; Updated Advice On Liver Function Testing

Cases of serious liver injury with elevated transaminases and bilirubin have been reported during treatment with Kaftrio–Kalydeco combination therapy. In all patients, measures of alanine aminotransferase (ALT), aspartate aminotransferase (AST), and total bili-

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rubin levels should be done before starting treatment, every 3 months during the first year of treatment, and annually thereafter.

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Trikafta May Be Safe And Effective For Liver Transplant Patients

People with cystic fibrosis (CF) who had a liver transplant can be safely started on Trikafta (elexacaftor/tezacaftor/ivacaftor). For most patients in the study, Trikafta's use resulted in an easing of symptoms, better quality of life, and healthier body weight and lung function. Doctors with the CF and transplant teams should work together in closely monitoring a patient's progress. Regulatory approval was based on positive results from clinical trials showing its efficacy and safety. However, liver damage can occur as a side effect of Trikafta's use, and patients who have or had liver problems are advised to talk with their doctor before starting with it. For those getting a solid organ transplant, possible problems include a worsening of liver function and drug–drug interactions with immunosuppressants, which must be taken to prevent the immune system from attacking the transplant. Despite the risks, Trikafta may offer benefits to some CF patients who have had a lung transplant. However, there are no recommendations provided for CFTR modulator use with patients undergoing a liver transplant.

https://tinyurl.com/234yk8rs

Cystic Fibrosis Transmembrane Conductance Regulator Modulators Decrease Hospitalization Rates

For patients with cystic fibrosis (CF), using a CF transmembrane conductance regulator (CFTR) modulator was associated with reduced hospitalizations for acute pancreatitis (AP). Patients were stratified by pancreas-insufficient CF (PI-CF) and pancreas-sufficient CF (PS-CF) status. More patients with CFTR modulator use had PI-CF. Stratified by pancreas sufficiency, 1.4% of AP admissions occurred among the PS-CF and 5.3% of admissions among the PI-CF groups during CFTR modulator use. The highest AP hospitalization rate occurred among patients with PS-CF who had discontinued CFTR modulator use. The lowest rates of AP hospitalization were observed among the patients with PI-CF who were currently using CFTR modulators. Thus, this study found that patients with CF benefited from CFTR modulator use, regardless of established pancreas sufficiency. These data justify a cost-effectiveness analysis to better assess the feasibility of expanded CFTR modulator use for preventing AP hospitalization among patients with CF.

https://tinyurl.com/56w47raz

Trikafta Leads To Lesser Need For Antibiotics, Fewer Hospitalizations

Treatment with Trikafta is linked with fewer infection-related hospitalizations and a reduced need for antibiotics among people with cystic fibrosis. Trikafta is an oral combination of three CFTR modulators (elexacaftor, tezacaftor, and ivacaftor) that works to improve how the CFTR protein functions in patients with at least one copy of the F508del mutation in the CFTR gene, a CF-causing defect found in roughly 90% of all patients. Clinical trials and post-approval data have shown that Trikafta significantly improves lung function and reduces pulmonary exacerbations. However, fewer studies have assessed its impact on lung infections

Continued on page 38

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

Sonya Ostensen
Melbourne Beach, FL
47 years old on March 12, 2022

Ed Fleischman
Long Island, NY
80 years old on December 24, 2021

TILLMAN continued from page 37

and the use of antimicrobials in CF patients. Researchers analyzed real-world data and identified 389 CF patients who started treatment with Trikafta between July 1 and Dec. 1, 2019, and continued using it through March 14, 2020. That timeframe allowed researchers to analyze data collected 15 weeks before and after the start of the therapy. The 15-week observation window was selected to avoid potential overlap with the COVID-19 pandemic, which led to a significant decline in healthcare utilization in March 2020. A total of 389 CF patients not taking Trikafta during the same period were included as sex- and age-matched controls. Specifically, the team compared the number of total healthcare visits, inpatient visits, infection-related visits, and antimicrobial prescriptions before and after Trikafta was initiated. This analysis showed that a 15-week treatment of Trikafta was associated with fewer healthcare visits, inpatient admissions, infection-related visits, and antibiotic prescriptions. By contrast, these parameters remained generally unchanged or worsened in control patients, although initial data of overall infections and healthcare utilization suggested that patients on Trikafta had more severe disease. Increased age was not significantly associated with any of the outcomes, but being a female was linked with a more infection-related visits. Also, co-existing conditions were associated with worsening across all outcomes.
https://tinyurl.com/2p95yaa3

Targeted Exhaled Breath Analysis For Detection Of *Pseudomonas Aeruginosa* In Cystic Fibrosis Patients

Pseudomonas aeruginosa (PA) is an important respiratory pathogen for cystic fibrosis (CF) patients. Routine microbiology surveillance is time-consuming, and is best performed on expectorated sputum. As an alternative, volatile organic compounds (VOCs) may be indicative of PA colonization. This study consisted of 1) a literature review to select VOCs of interest, and 2) a cross-sectional CF study. The investigators concluded that biomarkers from exhaled breath can yield high sensitivity for bacterial infection in cystic fibrosis patients.
https://tinyurl.com/yxe6k2xc

Cystic Fibrosis Genetic Modifiers Identified In Novel High-Throughput Screening Platform

Using an updated version of a recently developed technology called MaMTH-HTS (Mammalian Membrane Two-Hybrid High-Throughput Screening) that allows the detection of membrane-associating factors directly in living human cells, scientists have identified hundreds of new proteins that could lead to a better understanding of the cellular effects of cystic fibrosis and help identify new drug targets for precision medicine treatments. More than 400 proteins were identified that were associated with either healthy or mutant CFTR, and have shown that some of them could predict the variability seen in patient symptoms and treatment responses. One of the CFTR associated proteins that the team has identified is Fibrinogen-like 2 (FGL2), a protein believed to play a role in hepatitis, liver disease and immunity. Despite tailored CF treatments for the nearly 2000 known CFTR mutations, patient responses vary widely even among patients who share the same mutation. Researchers have long suspected variations in treatment response hinge on genetic modifiers and environmental factors. The identification of CFTR interacting proteins in the current study will help pinpoint these modifiers. In addition to FGL2, the researchers are working on other CFTR targets.
https://tinyurl.com/3zjkacnv

AND
https://tinyurl.com/y2w9ka9p
AND
https://tinyurl.com/46c63x9x

Specific Biologics Inc. Announces Therapeutic Development Award From The Cystic Fibrosis Foundation

Specific Biologics Inc., an early-stage biotechnology company developing novel gene editing technologies, announced that it will receive funds to support the preclinical development of its Dualase™ gene editing platform to target a set of cystic fibrosis-causing nonsense mutations where patients currently have fewer therapeutic options available. Dualase™ cuts DNA differently than existing gene editors. This feature enables the high frequency insertion of new sequences into precise locations in the human genome. Specific Biologics Inc. two-site Dualase™ platform gene editors cut DNA in a way that optimally exploits the cell's naturally occurring DNA repair pathways. This enables two gene editing outcomes, precise DNA deletions to disrupt genes or increased repair to correct genes. Specific also develops lipid nanoparticles to deliver the gene editor to target cells. Specific will deliver its gene editing technology using an inhalable lipid nanoparticle carrier, which will aid in delivering the gene editing machinery inside the cells of the lung. It is believed that if the CFTR nonsense mutations are corrected in enough cells, a gene editing therapeutic could provide a long-lasting benefit for CF patients.

https://tinyurl.com/262fw3r6

Pulmocide Receives Investment From The Cystic Fibrosis Foundation

Pulmocide Ltd. announced a $3.5 million investment from the Cystic Fibrosis Foundation for the development of opelconazole for lung transplant recipients. People with cystic fibrosis, particularly those who have undergone lung transplants, are highly susceptible to invasive *Aspergillus* fungal infections in their airways. Opelconazole is a novel azole specifically designed for inhaled use to prevent and treat pulmonary aspergillosis with potentially fewer side effects than standard-of-care azole treatments. This profile is expected to enhance efficacy and reduce the toxicities and drug-drug interactions seen with systemic antifungal therapies. The CF Foundation's funding will support a multicenter Phase 2 study of Pulmocide's opelconazole. The study will assess the treatment's safety and tolerability when used to prevent invasive pulmonary fungal infections in lung transplant recipients.

https://tinyurl.com/3ttn4mwz
AND
https://tinyurl.com/4sn3yhj4
AND
https://tinyurl.com/2pf6uns5
AND
https://tinyurl.com/2p8j5svp

Cystic Fibrosis Foundation Invests In Felix Biotechnology As They Advance New Therapies Targeting Chronic Infections In Cystic Fibrosis

Felix Biotechnology announced it received a strategic investment from the Cystic Fibrosis Foundation as part of a Therapeutics Development Award. The award will bolster Felix's work creating a novel therapy to manage chronic *Pseudomonas aeruginosa* lung infections. Felix takes a unique approach by engineering the natural predators of bacteria, tiny viruses called bacteriophage (or phage) into durable, broadly effective treatments. Felix's phage therapies are specially designed to consider and drive advantageous evolutionary outcomes of phage treatment. This results in a more durable therapy, which is key given the need to repeatedly administer phage to address these chronic infections. This therapy is currently being evaluated in a clinical trial at Yale.

https://tinyurl.com/ydfmynrh
AND
https://tinyurl.com/2nus77pn

CSU Researchers Discover That Anti-Malaria Drugs Can Fight Pulmonary Disease

A research team at Colorado State University has discovered that drugs used to treat malaria are also effective at treating a pulmonary disease similar to tuberculosis. The study is a significant development in the fight against infections caused by non-tuberculous mycobacteria (NTM). The research targeted an NTM known as *Mycobacterium abscessus*. Few drugs are effective against this mycobacterium, and the ones that are tend to be toxic and cause bad side effects. The researchers believe that the bacterium is capable of sensing and responding to threats in its environment, such as lowered oxygen levels, oxidative stress and acidic pH, which are our body's natural ways of fighting disease. It does so by activating, among other things, a regulator known as DosRS which controls many essential functions in the bacterium such as its respiration, ability to form biofilms and ability to enter a dormant state when the conditions are not favorable to bacterial multiplication. They found that in mice, two existing antimalarial drugs were able to prevent DosRS from responding to stresses, meaning that the bacterium struggled to fight off antibiotics and the immune system's natural disease response. One of the things the treatment did was to lower the bacterium's ability to form biofilms, thereby reducing its ability to resist killing by antibiotics. The treatment alone was just as effective at dropping bacterial loads in the lungs as the combination of antibiotics currently used to treat the disease.

https://tinyurl.com/2p9d77ny

Rapid Implementation Of Telemedicine Intervention For Cystic Fibrosis Shows Success

Continued on page 40

TILLMAN continued from page 39

Rapid implementation of a cystic fibrosis care model including telemedicine was successful for monitoring lung function and identifying exacerbations during the COVID-19 pandemic. The care model also enabled monitoring of BMI and decreased use of antibiotics. The implementation of telemedicine during the COVID-19 pandemic reduced patient and staff interactions and by doing so preserved personal protective equipment. Triple therapy (elexacaftor/tezacaftor/ivacaftor) likely contributed to the overall clinical stability in patients with cystic fibrosis during the COVID-19 pandemic. Telemedicine also increased access to care for adults with cystic fibrosis living in regions remote to a cystic fibrosis specialty center.

https://tinyurl.com/4jeaa7bp

AND

https://tinyurl.com/mr32sf75

A Cross-Sectional Investigation In Cystic Fibrosis: The CFTR Genotype, And Maximum Exercise Capacity Are Linked.

It was determined that human skeletal muscle cells carry the cystic fibrosis transmembrane conductance regulator (CFTR). Variations in CFTR functioning among cystic fibrosis patients may be a key factor of cystic fibrosis patients' maximal exercise capacity. There have been few conflicting investigations on the association between CFTR genotype and maximal exercise capacity. This study looked at the factors influencing maximal exercise capacity, as measured by peak oxygen uptake (V.o2peak), in children and adults with cystic fibrosis. In an international, multicenter, cross-sectional study, researchers collected data on CFTR genotype and cardiopulmonary exercise tests in cystic fibrosis patients aged 8 and up. Functional classifications I–V were assigned to CFTR mutations. They found that in patients with cystic fibrosis, the functional genotype class of the CFTR gene was not

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linked to maximum exercise capacity; however, for those with at least one copy of an F508del-CFTR mutation and a single class V mutation had a reduced maximal exercise capacity.
https://tinyurl.com/v5kkxk6t

Boost In Vigorous Exercise Fails To Improve Lung Function In Cystic Fibrosis

Increased vigorous physical activity did not improve lung function for sedentary patients with cystic fibrosis. ACTIVATE-CF, an international, parallel-arm, multicenter, randomized controlled trial, enrolled 117 relatively inactive patients aged 12 years and older with cystic fibrosis. All patients were randomly assigned to an intervention group in which patients added 3 hours of vigorous physical activity per week, or the control group in which patients did not change their physical activity behavior. The primary outcome was change in percent predicted FEV1 at 6 months. Secondary outcomes included physical activity, exercise capacity and motives, time to first exacerbation, exacerbation rates, quality of life, anxiety, depression, stress and blood glucose control. Compared with the intervention group, patients in the control group demonstrated higher FEV1 at 6 months. However, patients in the intervention group reported increased vigorous physical activity compared with the control group. In addition, patients in the intervention group had higher exercise capacity at 6 and 12 months and had higher physical activity in total steps at 12 months. Researchers observed no effects on all secondary outcomes during the study period. These findings indicate that a steep increase in vigorous physical activity represents the wrong approach to improve lung health for the majority of people with cystic fibrosis who are relatively sedentary. In contrast, the significant but counterintuitive improvement in FEV1 in the control group possibly induced by a moderate increase in physical activity suggests a less stringent 'motivational approach' may effectively modulate an increase in physical activity.
https://tinyurl.com/4tzh2mc8

Continued on page 42

ARTICLES

Acute ST-elevation myocardial infarction in two young women with cystic fibrosis and cystic fibrosis-related diabetes. Sirisha Reddy Thambuluru, Sayyad Kyazimzade, Katherine A. Despotes, Deepa Kirk, Jennifer L. Goralski. Journal of Cystic Fibrosis. VOLUME 21, ISSUE 1, E44-E47, JANUARY 01, 2022

Ischemic heart disease is rarely reported in people with cystic fibrosis (PwCF) despite multiple potential risk factors. Researchers report two cases of ST elevation myocardial infarction (STEMI), both in young women with cystic fibrosis (CF) and cystic fibrosis related diabetes (CFRD). These cases illustrate the importance of considering myocardial injury/infarction in the differential diagnosis of patients with CF and chest pain or shortness of breath, and addressing the growing risk of cardiovascular disease (CVD).
https://tinyurl.com/hfe9v67a

Comparison of microbial composition of cough swabs and sputum for pathogen detection in patients with cystic fibrosis. Dominic Fenn, Mahmoud I. Abdel-Aziz, Paul Brinkman, Renate Kos, Anne H. Neerincx, Josje Altenburg, E. Weersink, Eric G. Haarman, Suzanne W.J. Terheggen-Lagro, Anke H. Maitland-van der Zee, Lieuwe D.J. Bos. Journal of Cystic Fibrosis. VOLUME 21, ISSUE 1, P52-60, JANUARY 01, 2022

With the continued advancement of CFTR modulator therapies there is likely to be a burgeoning population of adult cystic fibrosis (CF) patients unable to expectorate sputum. Consequently, the detection and surveillance of pulmonary colonisation, previously reliant on sputum culture, needs re-examining. The authors hypothesised that cough swabs analysed with culture-independent analysis of the 16S gene could serve as a surrogate for colonisation of the lower airways. Cough swabs and sputum samples were collected from consecutive adults and children with CF across two sites at regular outpatient appointments. The researchers discovered that culture independent analysis of cough swabs provides an inaccurate diagnosis of lower respiratory tract colonisation and should not be used as a diagnostic test in patients with CF.
https://tinyurl.com/mveu4fc8

A comparison of clinic and home spirometry as longtudinal outcomes in cystic fibrosis. Alex Paynter, Umer Khan,Sonya L. Heltshe, Christopher H. Goss, Noah Lechtzin, Nicole Mayer Hamblett. Journal of Cystic Fibrosis. VOLUME 21, ISSUE 1, P78-83, JANUARY 01, 2022

The COVID-19 pandemic has accelerated the transition to telehealth, including the use of home spirometry in cystic fibrosis. Evaluating the accuracy and precision of longitudinal home spirometry is a requisite for telehealth-based research. This secondary analysis of a CF study (eICE) evaluates whether there are cross-sectional or longitudinal differences between home and clinic spirometry. Participants were issued a home spirometer, and asked to complete spirometry efforts twice per week for one year. Clinic spirometry was collected at baseline and every three months. Cross-sectional differences between clinic spirometry and the closest home spirometry measurement were analyzed. Home spirometry is estimated to be 2.0 percentage points lower than clinic spirometry cross-sectionally. Longitudinally, the estimates of 12-month change in home spirometry varied by analysis method from -2.6 to -1.0 ppFEV1/ year. In order to leverage the potential cost, feasibility and convenience of home spirometry, the differences with clinic spirometry must be acknowledged. Significantly lower ppFEV1 in home devices shows that direct comparison to clinic spirometers may induce a spurious change from baseline, and additional variability in home devices impacts statistical power. The effect of coaching, setting, and equipment must be understood to use and improve home spirometry in CF.
https://tinyurl.com/yc6u6jtj

Quality of home spirometry performance amongst adults with cystic fibrosis. Jody M Bell, Sheila Sivam, Ruth L Dentice, Tiffany J Dwyer, Helen E Jo, Edmund M Lau, Phillip A Munoz, Samantha A Nolan, Nicole A Taylor, Simone K Visser, Veronica A Yozghatlian, Keith KH Wong. Journal of Cystic Fibrosis. VOLUME 21, ISSUE 1, P84-87, JANUARY 01, 2022

Spirometry is usually performed under the supervision of a trained respiratory scientist to ensure acceptability and repeatability of results. An observational study was conducted to evaluate the quality of spirometry performance by adult cystic fibrosis patients with and without observation by a trained respiratory scientist. Patients were instructed to perform spirometry without supervision within 24 h of their remote CF clinic consultation. Spirometry was repeated at their consultation, supervised by a respiratory scientist using video conferencing. The majority of patients achieved grade A (excellent) or B (very good) spirometry quality with and without supervision independent of lung function severity. Similarly, forced expiratory volume in 1 second demonstrated no significant differences with paired spirometry performed within a 24 hour period. For a large proportion of adult CF patients, unsupervised portable spirometry produces acceptable and repeatable results.
https://tinyurl.com/5d226ej2

Clinical characteristics of *Pseudomonas* and *Aspergillus* co-infected cystic fibrosis patients: A UK registry study. Dominic A. Hughes, Olga Archangelidi, Matthew Coates, Darius Armstrong-James, Stuart J. Elborn,

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Siobhán B. Carr, Jane C. Davies. Journal of Cystic Fibrosis. VOLUME 21, ISSUE 1, P129-135, JANUARY 01, 2022 *Pseudomonas aeruginosa* (Pa) and *Aspergillus* species (Asp) are the most common bacterial and fungal organisms respectively in CF airways. The researchers aim was to examine impacts of Asp infection and Pa/Asp co-infection. They determined that co-infection with Pa and Asp was not associated with reduced lung function compared with Pa alone, but was associated with additional use of IV antibiotics. Asp infection itself is associated with several important indicators of disease severity.
https://tinyurl.com/tvwb9du5 ▲

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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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/
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