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

How Self-Insured Group Health Benefit Plans Limit Access To CF Care

By Beth Sufian

The Affordable Care Act ("ACA") reformed the individual and group health insurance industry in an effort to make health insurance coverage more available and affordable. However, some of the ACA's reforms do not apply to self-funded health benefit plans.

A. Self-funded health benefit plans do not have the same degree of consumer protection as group health insurance plans.

Self-funded or self-insured plans are a type of employer-sponsored health benefit plan where the employer bears the risk of employee health costs and the employer pays the benefit claims itself. Self-funded plans are an alternative to fully insured health benefit plans, where the employer purchases a group health insurance policy from an insurance company, such as Ætna, United Healthcare, or Blue Cross.


BETH SUFIAN

Employers may choose to self-fund plans because they are able to avoid costs and some regulations imposed on insurance companies by the ACA, among other reasons. A company that self-funds its health benefit plan is not regarded as an insurance company, and therefore is not subject to some of the ACA regulations that apply to insurance companies. The result is that self-funded plans do not have the same degree of consumer protection as fully insured plans. However, self-funded plans cannot exclude a person due to pre-existing conditions and must follow many other ACA marketplace reforms.

B. Self-insured plans are not required to provide essential health benefits.

One of the most important reforms under the ACA is that health insurance companies include a comprehensive minimum set of covered services in all of their health insurance benefit plans. This minimum set of services is known as "essential health benefits." Essential health

Continued on page 12

INSIDE THIS ISSUE

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EDITOR'S NOTES

Happy New Year from all of us at *CF Roundtable*! Our focus topic this issue tackles the complexities of insurance battles we all face in managing CF. On our cover, our President, **Beth Sufian**, highlights what self-insured group health benefit plans are and how they could potentially hinder access to care for individuals with CF. **Jeanie Hanley** shares a horrifying story involving a drone spy. **Lora Moser** writes about her battle with coverage through her Healthwell grant. In her column, "Speeding Past 50," **Kathy Russell** shares her insights on security relative to health and insurance. **Leah Sands** reminisces about the huge learning curve and responsibility in taking care of her own insurance as a teen.

In her wellness column, **Julie Desch** breaks down anxiety and the importance of using various tools, such as mindfulness, to work through it. In this issue, **Isabel Stenzel Byrnes** talks about setting goals in her column "Spirit Medicine." **Molly Pam** discusses potential insurance pitfalls and things to consider when starting down the fertility path in the Family Matters column. **Kat Porco** writes about the effect of Trikafta in blood sugar management in her CFRD Education column. In his column, **Mark Tremblay** writes about emotional exacerbations and what they look like in CF. As always, **Laura Tillman** does a wonderful job compiling all the latest research and information about CF from the internet.

In our "Voices From The Roundtable" section, **Linda Stratton** shares her perspective as an "elderly" CF patient starting Trikafta along with one of her original poems. **Colin Maydahl** poses the question "what will you do now that you're on Trikafta"? In our column, "In The Spotlight," read about how **Lorena Wood**'s life was shaped by her having CF and her challenges and joys of being a missionary.

CF Roundtable couldn't happen without the board of directors and the help, support, and commitment of everyone in the community, especially our donors. As we start off 2020, we'd like to extend our sincerest gratitude for all of the financial contributions throughout the past year. If you're interested in writing for *CF Roundtable*, check out page 3 for a list of our future focus topics. Don't see anything on the list that interests you? Our "Voices From The Roundtable" section features the voices of everyone in the CF community. With that in mind, send us your thoughts, whether they're in the form of an article, poetry, song lyrics, or anything else. The stories in our newsletter are the heart and soul of our organization. From all of us at *CF Roundtable*, we wish you all health and happiness in the new year.

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

Sydna

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

Information From The Internet...

Compiled by Laura Tillman

Average Life Expectancies For Cystic Fibrosis

Over the past few decades, life expectancy for people with CF has improved dramatically. Recent research suggests that by 2025, the number of adults living with CF will increase by approximately 75%. Several factors — including sex, lifestyle choices, any infections, and the type of CF gene mutation that a person has — can influence life expectancy. The median predicted survival age is an internationally accepted way to estimate life expectancy. Unlike a mean average, the median uses the midpoint in a set of numbers. It more accurately reflects the age that a person with CF can expect to reach. The data indicate that half of all babies born with CF in 2017 will live to be 46 or older. Other statistics suggest that more than 50% of babies with CF born in 2018 and 50% of people with CF aged 30 or older in 2018 will likely reach at least their fifth decade of life. These predictions do not take into account the potential for improvements in care and treatment that may occur as people age. It is also important to note that these figures are just averages. Some people will live longer. In fact, some people with CF are living into their 70s. When discussing life expectancy, it is also important to consider a person's quality of life. How an individual views their quality of life depends on a number of factors, including their age and general health status. Many people with CF develop health complications as they age. Some of these can contribute to reduced quality of life and early death. Potential complications include:

LAURA TILLMAN

- bile duct or intestinal obstructions
- bronchiectasis, a condition that causes airway damage

Continued on page 7

LOOKING AHEAD

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

Winter (February) 2020: Insurance Issues. (Current issue.)

Spring (May) 2020: Weight Issues. (Submissions due March 15, 2020.) Do you have trouble gaining weight? Trouble losing weight due to CFTR modulators? How do you gain/lose in a healthy way? Can you share some tips? Recipes? Exercise routines?

Summer (August) 2020: Diversity in the CF Community. (Submissions due June 15, 2020.)
We are looking for articles by individuals who live with cystic fibrosis and who either are a person of color, are LGBTQ+, have a national origin that is other than the United States, or have another disability in addition to CF.

Autumn (November) 2020: People with CF Who Have Started a Business. (Submissions due September 15, 2020.)

SPIRIT MEDICINE

The Spirit Of Striving: Setting Goals

By Isabel Stenzel Byrnes

This autumn, I had the privilege of pursuing a goal that I've had for a very long time. My father has been an avid mountaineer all his life and has climbed all 247 peaks of the Sierra Nevada over the last 40 years. But high elevation has made my lungs bleed since I was 12, and I never could accompany him on any hikes. One of my dad's greatest feats was hiking Mt. Whitney, the tallest peak in the continental United States, which he did six times. Joining him on Mt. Whitney had always been a dream of mine and, as luck would have it, a friend of mine won permission to climb the mountain — this after several years of applying via lottery for a permit. Unfortunately, instead of an overnight permit, it was day-use only. The hike was 22 miles long and could take 18 hours...but sure, we could do it in a day!

This hike was one of the greatest physical challenges of my life. While a hike is just a hike, it is also a grand metaphor for life itself. With or without CF, life can be kind of dull sometimes. To me, it's important to always have something to look forward to. Setting goals and pursuing them gives life its juiciness and spark. We set our minds on something, we plan, we prepare, we train, and we go for it. Sometimes we feel good and sometimes it hurts like hell, but we keep going.

In this article, I'd like to explore the spirit of striving. A goal is just that...something far off to strive for. Greg Reid, the motivational speaker, once said, "[a] dream written down with a date becomes a goal. A goal broken down into steps becomes a plan. A plan backed by action becomes a reality." We all have the ability to create our reality by setting realistic goals and pursuing them.

The first step of my goal was planting a seed of desire. Why did I want to hike Mt. Whitney? Because my father did it. Because I can breathe now. Because the mountain is there. Because I'm hungry for adventure. Somewhere, the motivation was born.

The next step was planning. I gathered several close friends who were capable of this physical endeavor and thought it would be fun! Together, we set out a strategy to train together. Many emails were exchanged to rally our enthusiasm and divert our fears. This stage of life is the dreaming stage: bonding together by wondering what it would be like, imagining our victory, feeling the emotions of pursuing this goal. There was a great deal of laughter involved, as we questioned our craziness to want to do this.

> *"These desires to set physical goals are deeply engrained in my DNA. If I don't push, I won't survive."*

ISABEL STENZEL BYRNES

As the hike became closer, our planning became more detailed. What would we bring? What would we share? We booked high-altitude campsites before the hike in order to acclimate. We opened ourselves up to surprises. We all agreed that it would be okay not to reach the summit, despite the inherent disappointment stemming from unmet expectations. We would just do our best and go for it. We knew safety and health were our greatest priorities — for my 79-year-old father, for my friend who is a heart-lung recipient, and for me as a 15-year-post-transplant person with CF.

There is the CF. I had to think deeply if I could do this. Could I handle 14,500 feet? Once I could hardly handle 3,000 feet. Was this an attempt to prove something? Did I want to do this as just anybody, or as a person with CF? CF itself has been the source of my hard-headed determination.

I recently learned about a term called "trauma mastery." This means that many of us may have been exposed to some hardship or trauma early in life and spend much of our lives trying to

overcome that trauma. A child who is abused may, as an adult, repeatedly get involved in abusive relationships. Each time, she thinks, "this time, I'll be strong. I'll fight back." But instead, the cycle continues. I wonder if my desire to push my body is a form of trauma mastery. With CF, it took effort and determination just to live. I was always fighting my body, pushing it to cooperate, endure, and stay alive. It worked, for 47 years. And these desires to set physical goals are deeply engrained in my DNA. If I don't push, I won't survive. So, I strive to survive...the next life-threatening effort.

Another step toward a goal is the training, which started months before this hike. I'd do the Stairmaster at the gym. I'd walk up a long staircase near my home. I'd hike locally every weekend, with the mileage increasing over time. I went on a backpacking trip and hiked 17 agonizing miles in one day. A few weeks before Whitney, my friends, father, my basset hound, and I hiked two of Lake Tahoe's highest peaks, each over 10,700 feet, over two days. Our rewards were the beautiful views on the summits of these peaks, surrounded by migrating butterflies. The training helped to build confidence and trust in our bodies and abilities. And like life itself, there was pain. An old foot injury spoke to me every step and reminded me of my fragility.

Finally, the weekend of Whitney arrived. On the long drive to the Eastern Sierras, we slept, ate, and bonded in anticipation of what would come. We compared our meal plans and how we'd fuel our bodies. I ate steak several days in advance, something I never do. I packed electrolytes and stopped taking insulin. But mostly, we explored the psychology of a goal — focusing on our confidence, squelching our self-doubt, pumping up our determination, imagining victory. *We got this.*

And at 2 a.m. on Labor Day, we woke up in the dark to eat a hearty breakfast and were at the trailhead by 3 a.m. The slowest (me) lead the way. Headlamps lit the blackened trail. The only things we could see were the flickers of dust in the air and our feet stepping forward on a path. For three hours we hiked slowly uphill until a stretch of orange appeared on the horizon below. Little by little, more light appeared. By 7 a.m., we turned off our headlamps. We stopped every hour for a snack. The higher we went, the more I focused on my breath. I could simply *breathe*.

Up and up we went. Life is literally one step forward at a time. Sometimes I stared down at my feet and forgot to pause and look behind me at the glorious view. Fatigue set in and the time between breaks grew shorter. I panted and gasped, reliving my old CF days. I couldn't keep up a conversation — just how I had felt during end-stage CF. We filtered water and hydrated often, which meant stopping for the bushes. Soon, we were so high, there were no bushes!

Our minds had to stay focused on the goal. *Keep going. Don't stop. How much longer?* Each of us hiked at our own pace. People passed us. I was slow. *Really* slow. My SpO2 was 83 at 12,000 feet. I felt lightheaded, almost drugged, and hypnotized. But I could keep going. This drive wrestled with the constant desire to stop. *Ok, I'm done. This is too hard.* Everything hurt and I was too nauseated to eat. There were 99 switchbacks until the saddle (the top of the mountain before the final trail to the summit), and we'd walk a little, turn a corner, walk a little and repeat.

My mind also had the power to distract from the pain. I thought about my father and grandmother who were refugees in the war, and about the many people like them who've had to walk for miles and miles, fleeing for their survival. Here, such difficult perseverance is optional. My mind focused on those who could not walk, who didn't have the equipment and nutrition we had that day to hike for 17 hours. I thought of all my CF friends who gasped at sea level for air. Focusing on perspective helped me connect to the suffering of all of humanity and keep going.

There was something magical going on during this journey. I was deeply mindful of everything going on in my body, and everything around me. My heart rate, my breathing, my steps forward, my stomach. This was being in the moment — allowing the pain, the discomfort, and being with it.

The sky started to darken eight hours into our hike. Some people were turning around. One woman walking down was in tears from exhaustion. One of my friends pleaded to retreat. Thunderstorms and lightning are life threatening at this altitude. I just wanted to make it to the saddle. The wind started to pick up. Like life itself, obstacles started to present themselves. We were on decision road. What do we do? Keep going or stop? Our goal had to change. Reaching the summit seemed impossible. Reaching the saddle seemed doable. My body felt so depleted, I prayed that God would make a decision for me. And I started second-guessing myself. *I should've trained harder. We should've left earlier. Darn, CF and diabetes weaken my muscles. I wish I could be faster. I should've done this five or ten years ago.*

Ultimately, after nine hours of uphill hiking, we made it to the saddle at 13,600 feet. I praised my donor for this feat. We celebrated as the rain started to come down, snapped photos and, just as we turned around, the hail balls started to fall. We heard thunder. Our seven-hour race down the wet, cold, and slippery mountain added to the challenge. My dad slipped twice. My foot screamed in pain. My rain jacket soaked through. The creeks we crossed in the dark on the way up had

Continued on page 15

SPEEDING PAST 50

Feeling Secure Helps One's Health

By Kathy Russell

An aura of security can help one to feel healthy and may actually assist with maintaining better health. I have found that a sense of security can help me to feel much healthier and happier. Many things contribute to one's feelings of security. Certainly one important component is an income that is steady. Another would be having a good, safe place to live. Having a steady diet that is nourishing is quite important, too. Having good health insurance is right in there among the things that are most important.

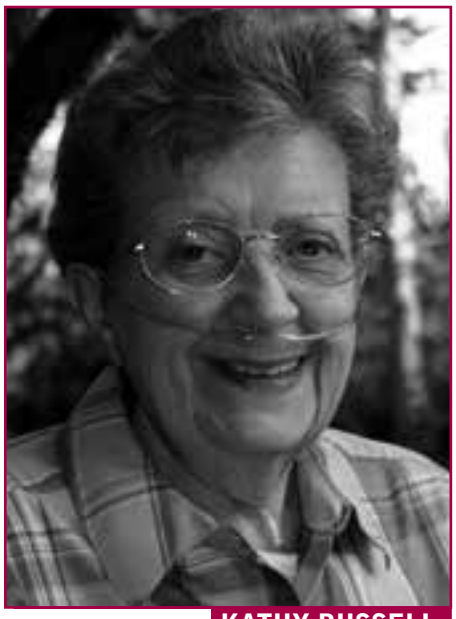
At my age, feeling secure always is important, but sometimes it is difficult to achieve. Being sure of one's healthcare is one way to add to the feeling of security. I am grateful I have Medicare and good supplemental health insurance. With the rising costs of hospitalizations, medicines, and doctor visits, it is possible to feel very insecure, and sticking to a budget can be almost impossible.

It is important to budget our energy as well as our income. One thing that helps me save energy is having an advocate who handles almost all of my insurance questions and problems. My husband, Paul, is my advocate, and he is excellent at it. He has taken on so much of the load of worries that insurance can cause. He is the one who spends hours on "hold" waiting to speak with someone about whatever problem is vexing me at the time. He explains what the trouble is and sticks with it until he gets a satisfactory conclusion. I really appreciate what he does for me.

We all know that planning ahead can help to save energy. Being prepared for unexpected charges can help with protecting our limited energy. I offer a couple of suggestions for getting the most out of your health coverage. First, it is important that you know what your policy covers and how to access all of the benefits you need. When you have to dispute anything, and the chances are you will, you will be able to cite the specific information from your policy that covers the particular problem. It certainly has helped me in the past.

Also, here is a piece of advice Beth Sufian offered at a CF conference several years ago — "pick your battles." She cited a charge for a lab test that was only $15 or $20 (obviously this occurred several years ago, when lab tests could be as little as $15 or $20), and the company wouldn't cover it. She pointed out the immediate cost was not that much and one might be tempted to overlook it. If it is a one-time-only test, you could decide that it wasn't worth fighting for and just let it go. However, if you knew you were going to get that test several times a year, it could add up. So this would be a case of a charge that would be worth fighting for. Expending the energy needed to get the company to cover the charges will save you time, money, and energy in the long run.

> *"Being sure of one's healthcare is one way to add to the feeling of security."*



KATHY RUSSELL

The past three months have flown by for me. I am amazed at how much time I have for various things since I no longer am so involved with the newsletter. After 29-plus years of concentrating so much of my time and energy on USACFA and *CF Roundtable*, suddenly I am getting more of my projects done. I love it! That isn't saying that I regret any of the time that I have spent on the newsletter and other USACFA chores. It's all good. It is nice to be able to spend time on other chores, though.

We were blessed with a very pleasant summer. Autumn has been pretty nice, too. I am hoping that winter will be mostly mild. We did get a light dusting of snow the night before Thanksgiving. It was gone by midmorning and we've seen none since. Of course, other parts of our state got walloped with a "bomb cyclone" that came in shore down on the southern coast and made a fine mess of road travel. Since we had no plans to be away from home, it really didn't affect us. We do

hope that we get some precipitation during winter, however, because we are in a mini-drought in most of Oregon. We're about nine inches below our normal rainfall for the water year. It is important to have enough snow in the mountains and enough rain in the lakes and reservoirs to last through the drier seasons. When we do, I feel more relaxed and secure.

My health has been so much better in this year since I started taking Orkambi. I notice I really have more energy and don't have to take as many naps as I used to. When I saw my pulmonologist, recently, we discussed whether I should try Trikafta. He and I both wanted to read more about it before we make any major changes. That's okay with me. I hate to take a chance on "upsetting the applecart" by stopping Orkambi and starting Trikafta. It all is rather scary in a way.

When I read all the information I could find, I discovered that two of the meds I take may have negative interactions with Trikafta. The doctor and I discussed that and decided we will leave things as they are for now. I am doing well and I don't want to do anything that might change that. Maybe when there is more empirical data available, I may make a change. I feel there is no hurry to change and am sure it is best to get as many facts as I can before I make any changes. My doctor concurs.

Having a physician who talks things over with me before ordering any changes helps me to feel more secure. My doctor sits down and talks with me. He asks questions and listens to my answers. He listens to any questions I may have and answers me with his best knowledge of whatever subject we are discussing. He also asks Paul if he has any questions. It really helps me to have Paul understand what the doctor is saying. It helps me to be sure that I understand and remember what the doctor said. Having that kind of relationship with my doctor definitely adds to my sense of security.

I am switching gears now. I would like to hear from anyone who has CF and is at least 50 years old. I would like to know what issues interest you, what would you like to read more about, and what are your CF concerns? Also, are you interested in writing a "Speeding Past 50" column? I am happy to share the space with anyone who is interested. You may contact me at krussell@usacfa.org. I hope to hear from you.

Until next time, stay healthy and happy,

Kathy ▲

Kathy is 75 and has CF. She served as a director and as president and treasurer of USACFA. She and her husband, Paul, live in Gresham, OR. You may contact her at krussell@usacfa.org.

TILLMAN continued from page 3

- chronic infections, including bronchitis and pneumonia
- diabetes
- infertility, particularly in males
- nutritional deficiencies
- osteoporosis, a type of bone condition
- pneumothorax, which involves air collecting in the space between the lungs and the chest wall
- respiratory failure

Despite the possibility of these complications, some research suggests that the perception of quality of life improves as people with CF get older. In the later stages of CF, complications often cause serious problems for people. These complications typically affect the lungs, but they may also affect the:

- endocrine (hormone) system
- intestines
- liver
- pancreas

The leading causes of death among people with CF are respiratory failure and chronic progressive pulmonary disease.

Several factors can influence a person's quality of life and life expectancy. These include:

Age: As people get older, there is an increased risk of complications, some of which can be fatal.

Sex: Women with CF have a poorer outlook than men with the condition. Some research suggests that this is due to the increased risk of death in women with CF-related diabetes.

Health status: Conditions associated with a lower survival rate include:

- certain infections, including Staphylococcus aureus
- diabetes, in women
- being female
- malnutrition
- low body mass index, or BMI
- pancreatic insufficiency
- low lung function
- frequent pulmonary exacerbations, or episodes during which lung symptoms get worse

Although there is no cure for CF, people with the condition are now living longer than ever. Their self-reported quality of life is also much better than in previous decades.

https://tinyurl.com/y3zox6bn

New Clinical Tool Predicts 1- and 2-Year Mortality In Cystic Fibrosis

Researchers have developed a novel clinical tool that uses patients' overall health status and the risk for intermittent shock events to predict

Continued on page 19

CFRD EDUCATION

The Impact Of Trikafta On Blood Sugar Management

By Kat Porco

On December 19, 2018, Maylie, my daughter, who has CF and diabetes, took her first dose of Trikafta. Our lives changed in that moment. It became about living, not simply surviving. But, with the overnight health that suddenly manifested itself, so did some inconsistencies in our previously accepted norms for disease management.

Following the first month, I began to notice that the control of diabetes, of which I had become so proud in the past, seemed impossible to achieve. I was seeing highs that did not correlate with our vigilance around bolus dosage timing, our well-honed carb-to-insulin ratio, or the long-standing basal dose that had given us ideal fasting numbers for years. I witnessed Maylie's first hypoglycemic event that included disorientation when her sugars dropped below 40. We sat together waiting for the low to resolve and, despite being three-year veterans in her diabetes management, we felt the same frustrations we had experienced when she was newly diagnosed and we were not in control. We spent the next few weeks trying to make sense of this new pattern by vigilantly following her CGM data and tracking both her food intake and her dosage timing, all to no avail. After a couple of long and frustrating months, Maylie's blood sugars and insulin dosages stabilized back to their pre-Trikafta state. I would love to have the answer everyone is seeking, because, as of the 12-month mark, her need for insulin has not decreased. However, time will tell. Everyone will have a unique experience based on the severity of their CFRD when integrating modulators. The majority of my clients who are on Trikafta are currently facing these confusing trends. While it is super frustrating, I hope that everyone can give themselves a little grace as their bodies adapt to this new drug.

Admittedly, no one is really sure how Trikafta is impacting the body on all these levels; but Vertex has hired a full-time endocrinologist to start studying the impact of modulators on CFRD. For now, I will try to make a little sense out of this confusing process and hypothesize as to the inconsistencies we are all seeing with the addition of the new modulators. The hope is that this drug has the capability to potentially restore some pancreatic function and/or stimulate beta cell production for any cells that are still functioning. Additionally, as the sodium chloride channels become more efficient, there may be a decrease in infections, thus minimizing some of the inflammation.

As we have discussed in the previous newsletter, CFRD is essentially a mix of Type 1 (deficiency of insulin production, due to beta cell death) and Type 2 (resistance to insulin, due to inflammation and steroid therapy). So, if the modulators lower the infection and inflammation, resistance may minimize, which in turn would increase sensitivity to endogenous insulin, as well as the supplemental exogenous insulin. Feeling better may also lead to an increase in exercise, thus activating the muscle cells that store insulin, ultimately decreasing your resistance. All of this could lead to unexpected hypoglycemia. Many individuals on Trikafta are seeing an increase in appetite, leading to an increase in food consumption and weight gain. Insulin dosages are generally calculated based on weight and titrated due to resistance. So, if you gain weight from the addition of modulators, you may see a need to adjust your carb-to-insulin ratio (i.e., go from 1:30 to 1:25).

I think the most important point during this time of flux is to minimize hypoglycemia at all costs. While my message is generally focused on mitigating the majority of hyperglycemic excursions above 140, safety dictates

> *"Everyone will have a unique experience based on the severity of their CFRD when integrating modulators."*

KAT PORCO

that it is critical to monitor for hypoglycemia as well and, in this scenario, you may need to ride a little higher for a while, as your body transitions. If you are ever having more than two hypoglycemic events per week that need treatment and are not induced by bolus insulin/incorrect carb counting, your basal dosage needs to be reviewed with your physician. Having multiple hypoglycemic incidents in an effort to avoid hyperglycemic incidents is not a safe way to manage blood sugars. If you see multiple hypoglycemic incidents after three hours of dosing bolus insulin or in morning fasting numbers, please reach out to your endocrinologist to discuss therapeutic adjustments.

All of this is a starting point for further discussion on how the new modulators affect CFRD in the long run. I look forward to the research that Vertex shares with us in the future. In the meantime, if you are feeling the rollercoaster of blood sugars that most are experiencing, know that you are not alone and be patient with the process. ▲

Kat Porco is a diabetes educator and the co-founder of Attain Health Foundation. She received a Bachelor's in Social Work followed by a Master's of Science in Health Communications. Her work over the past ten years has been solely focused on supporting and advocating for the cystic fibrosis community. Throughout these years, she has seen the disconnect between the recommendations of the medical community and applicability for the patient community. Because she does understand this complex relationship, Kat felt that she could assist in bridging the gap to reach ultimate health goals through health coaching and diabetes education. She is a Duke Certified Integrative Health Coach, as well as being Nationally Board Certified in Health & Wellness Coaching through the National Board of Medical Examiners. Kat is a Certified Diabetes Care and Education Specialist (CDCES) through NCBDE.

FAMILY MATTERS

What To Know About Obtaining Insurance Coverage For Fertility Services

By Molly Pam

Many men and women with CF struggle with infertility. It is an emotionally challenging experience that is often not discussed. For many, the most wrenching aspect of it is not the condition itself but paying for treatment. One round of in vitro fertilization ("IVF") can cost $15,000 or more, depending on your insurance and coverage, and multiple rounds might be required to yield a healthy pregnancy, thus totaling upwards of $100,000. It is important both to plan financially and to consider the demands of parenting as you embark on this life journey. Additionally, fertility support is not necessarily a covered benefit and/or coverage is only minimal. It's essential to calculate what you may have to pay before you start making fertility appointments. In this article, I will outline many of the ways in which fertility services may be covered by insurance. There is such a wide variety of coverage scenarios that this article is in no way a definitive guide as to whether your services will be covered.

Does my health insurance cover fertility services?

This might seem like an obvious question, but it is an important one to consider, as many people assume their insurance won't cover it. While newborn and maternity care are considered "essential services" under the Affordable Care Act, and thus their coverage is required, fertility services are not. However, 16 states have laws requiring health plans to cover some aspects of fertility services. Even if you do not live in one of those states, some plans may cover some or all of the costs (lucky you!). Some will cover only the fertility-inducing hormone shots but not the IVF process itself, and several insurance plans have a cap on either total costs or on the number of IVF rounds covered.

It is important to call your insurance company and thoroughly read your plan details to figure out what is and is not covered. Placing a phone call to check which specific services are covered *before* starting the process and also *before* subsequent rounds of tests, helps reduce the possibility of ending up with a surprise medical bill. Make sure to get a reference number for the call so that if you call back, the customer care agent can pull up the record of your conversation.

Asking anyone other than your insurance provider about coverage can result in confusion. A woman with CF gave me the best piece of advice that I could possibly share: "If you're not sure about coverage, don't take anyone's word for it until you speak directly with the insurance company. The clinic we worked with for IVF was not very helpful, and actually incorrectly told us our insurance would not cover any of it. Ultimately, we had to figure it out with the insurance company directly, by calling and reading our policy."

> *"Asking anyone other than your insurance provider about coverage can result in confusion."*

MOLLY PAM

What "catches" should I be aware of?

Make sure your fertility clinic is in network; they will typically complete the authorization before making your appointment. Commonly, employer plans that cover fertility services have stipulations that employees be at the company for a defined period of time before those services are covered — and this goes for paid parental leave, too. For example, my employer-based insurance covers fertility services through Progyny, a fertility benefits company, but only after one year of employment.

Prior authorization is also commonly required for fertility treatments. Additionally, many fertility clinics will only see couples after they can document an inability to successfully sustain a pregnancy: 12 months without having conceived for couples under 35 and 6 months for those who are older.

Some insurance companies will

cover ultrasounds, blood tests, and specialist visits until there is an official diagnosis of infertility, at which point they may stop covering those services. It can be particularly distressing for that to happen, so you can discuss medically appropriate diagnostic codes with your doctor to optimize insurance coverage. I know one woman who had three miscarriages and wanted testing to figure out the cause. Her insurance would not cover anything to do with infertility; however, it did cover diagnostic tests to find out why an ectopic pregnancy had occurred. Her doctor resubmitted the insurance paperwork with ectopic pregnancy as the diagnosis and the procedure was later covered.

While the above information is important for all people exploring coverage for fertility treatment to consider, there can be many specific tests and services required to get pregnant, with variable coverage. Here are some examples with common limits seen in insurance policies:

Intrauterine insemination ("IUI")

During IUI, semen from a woman's partner or a donor is placed through the cervix into the uterus to facilitate fertilization of an egg in the fallopian tubes. Colloquially, people call it the "turkey baster" method. It is often the first step taken by couples trying to become pregnant and, in some cases, can be required by insurance prior to coverage of IVF. It is usually less expensive than IVF, typically costing around $500-$1,000 per insertion, but is also somewhat less successful. However, IUI can be successful when the primary factor preventing natural pregnancy is the thick cervical mucus reported by many women with CF, and is therefore a worthwhile first attempt.

In vitro fertilization ("IVF")

IVF is when eggs and sperm are combined in a lab setting, and the fertilized eggs are implanted into a uterus. Many insurance companies that cover fertility services will set a limit on the number of rounds of IVF they will cover or cap the total coverage. One woman I know said her insurance covered *"50% of up to two rounds of IVF, and after that we were on our own."*

There are many types of egg retrieval that can be done but, generally, women take hormone injections to stimulate the ovaries and produce several eggs at once. This can be a physically and mentally exhausting time, a factor to take into account with your health and other CF-related issues.

MESA and TESA

97% of men with CF are infertile due to a blocked or missing vas deferens. If they want a biological child, they will need a procedure to extract sperm from the testicles (MESA, TESA and others). The good news is that it is a viable option for men with CF. It requires a minor surgical procedure that results in a sperm sample that can be used for IUI or IVF. The costs for this are usually bundled with IVF, IUI, or sperm storage, so the costs vary.

Surrogacy and adoption

In surrogacy a third party carries the pregnancy. Couples may pay for not only egg and sperm retrieval from themselves or donors, but the IVF itself, in addition to the cost of medical care during pregnancy, labor, and birth for their surrogate. This is not always covered by insurance. One person with CF who became a parent through surrogacy told me, *"I assumed my insurance would not cover the costs of surrogacy, so I bought a secondary insurance plan to obtain coverage. Near the end of the process, I realized my insurance did cover it and I had needlessly paid for the second plan."* Another said, *"We had to buy an independent plan for our surrogate because her insurance did not cover surrogacy even though pregnancy is a federally mandated condition all insurance companies must cover."*

Surrogacy also comes with unexpected non-covered items. One woman warned, *"Keep in mind that if you go the surrogacy route, a lot can happen that could cost more money, such as, if your surrogate is put on bed rest and cannot work. Or if she has post-op complications"* (such as from a C-section).

If you are considering adoption, you may be able to use money you have in your Healthcare Savings Account, also known as an HSA. Some larger employers may provide coverage for adoption fees.

Genetic testing

Many people with CF opt to get their partners tested for CF mutations, either with the standard panels of 32 or 82 mutations, or a full sequencing to see any abnormalities on the CFTR gene. If you have CF and your partner carries a mutation, the odds of having a child with CF are higher. Spouses, children (yes, there are adult children of people with CF!), siblings, and even extended family such as cousins can opt to get tested since they may have inherited the same mutation as you. A referral from their primary care physician ("PCP") will likely be required by the insurance carrier in order for the genetic counseling and tests to be covered. When my partner and I were looking into getting him tested as a potential carrier, he went to his PCP, and they informed him his insurance would not cover the testing if they ordered it, so they referred him to a genetic counselor so testing could both be ordered and covered by insurance.

Genetic testing for fertilized eggs is rarely covered, though many with CF opt for this route when they know their partner is a carrier so the embryo(s) selected do not have CF.

Egg and sperm storage

Egg storage can cost approximately $600 per month out of pocket, while sperm storage is much cheaper, around $300-400 per year.

Continued on page 17

benefits include the following ten benefit categories designed to ensure that all consumers have a minimum standard of coverage regardless of the health insurance plan in which they may enroll:

1. Ambulatory out-patient services;
2. Emergency services;
3. Hospitalization;
4. Pregnancy, maternity, and newborn care;
5. Mental health and substance use services;
6. Prescription drugs;
7. Rehabilitative services and devices;
8. Laboratory services;
9. Preventive and wellness services and chronic disease management;
10. Pediatric services, including oral and vision care for children.

However, self-funded benefit plans are not required to cover these essential health benefits. Even if a self-funded plan elects to offer services in any of these categories, it may substantially limit and restrict the amount of coverage.

For example, a self-funded plan is not required to offer any prescription drug coverage. If a self-funded plan did offer prescription drug coverage, the amount of coverage could be limited to a specific dollar amount per year. The prescription drug coverage could be limited to only specified drugs, may exclude individual types of drugs, or may exclude whole classes of prescription drugs such as CFTR modifiers and correctors. Consequently, a self-funded plan may have much less coverage and fewer benefits than fully insured plans offered by health insurance companies.

C. Self-funded plans may not cover the whole cost of a claim.

With a self-funded plan, the employer is responsible for all of the cost of benefits and is exposed to the risk of unexpectedly high treatment costs. Examples of such unexpectedly high treatment costs are: high-cost/low-frequency treatments, such as organ transplant (often referred to as "shock claims"); and low-cost/high-frequency treatments (often called "high utilization claims"). The new CFTR modulator drugs that modify and correct the faulty CFTR gene functions are both high-cost *and* high-frequency items, which makes them particularly disfavored by self-funded plans.

To protect against being liable for these costs, self-funded plans will typically purchase stop-loss insurance. Stop-loss insurance reimburses self-funded employers for benefit claims (such as shock claims or high utilization claims) that exceed pre-determined expenditure levels either for each person, for the whole group, or both. However, stop-loss insurance does not provide a benefit to a covered employee. Stop-loss insurance insures and benefits the employer only. It does not provide any insurance benefit to the covered employees. If the stop loss does not pay, the responsibility for payment of the claim is with the employee.

When a group of employees includes an individual, who has high-cost claims, the employer and its stop-loss insurer may establish a specific stop-loss level on that individual. This practice of establishing an individual stop loss is known as "lasering" because it focuses on the specific individual like a laser beam.

A laser stop loss assigns a higher specific deductible for an individual with a known condition that is likely to exceed the usual deductible. It is used when an individual on a plan possesses a higher pre-disposition for illness or higher health care costs than other employees.

While the ADA prohibits employers from firing employees or refusing coverage due to illness, employers who have self-funded health insurance plans have a substantial financial incentive to find a way to terminate employment for employees who have high health care costs. The employer may find legitimate, non-discriminatory reasons to terminate employment (such as an employee's failure to regularly and predictably appear for work as scheduled) and be relieved of the higher costs associated with that individual's continued employment.

> "A self-funded plan may have much less coverage and fewer benefits than fully insured plans offered by health insurance companies."

D. Self-funded plans have no consumer protection from unreasonable increases in premiums.

The ACA requires the U. S. Department of Health and Human Services ("HHS") to annually review all increases in premiums by health insurance companies. Any rate increases of more than 10% must be justified with evidence presented to the HHS. However, this review process for rate increases applies only to small group and individual insurance plans. It does not apply to self-insured plans. Consequently, employers with self-funded plans can raise the amount of employee contributions for health coverage by any amount without submitting to an

annual HHS review, in addition to increasing employee contributions by more than 10% annually without restriction or justification.

E. Covered employees may not know if they are enrolled in a self-funded plan.

Many people do not even realize their employer-sponsored plan is self-funded. Many employers who self-fund their employee health benefit plans do not administer or manage the benefit plan themselves. Self-funded plans are typically administered by a third party.

Third-party administrators are often insurance companies — such as Ætna, United Healthcare, or Blue Cross — that have the expertise in administering benefit plans. So the summary plan description, explanations of benefits, claim denials, and other communications come from an insurance company that is acting as the plan administrator for a self-funded plan. These communications usually have the insurance company's name and trademark on them.

To the ordinary employee, it may appear that the benefit plan is a fully insured plan provided by the health insurance company, when in actuality the plan is self-funded by the employer and only administered by an insurance company.

F. What should a person with CF do if enrolled in a self-funded plan?

1. Do not raise unnecessary concerns.

If a person with CF is enrolled in a self-funded plan, there may be no immediate cause for concern. If the plan is paying for CF care claims and CF prescription drugs according to the terms and conditions of the plan of benefits, then it is unlikely there is anything the person with CF would want to do. If a self-funded plan is paying CF-related claims, a person with CF would not want to raise any unnecessary concerns about potential future high-cost medical treatments that have yet to be prescribed by the CF care team.

2. How to learn whether a plan is self-funded.

Even if your employer's plan is paying claims as it is supposed to do, the covered employee may still want to know whether their plan is a self-funded plan. The employee can look at the Summary Plan Description (the booklet that explains coverage and claims procedure) for important information about the benefits, including whether the plan is self-funded or fully insured. The summary plan description should contain a statement that the plan is self-insured or self-funded by the employer — usually in the very beginning or very end of the booklet where it states who is insuring or funding the benefits. This information may also be found in the section of the summary plan description that describes who makes the final decisions of whether a particular claim will be paid. There the summary plan description may say something like: "…because this is a self-funded plan, the employer makes the final decision on whether a claim is to be paid."

The insured person may also examine communications from the third-party administrator for information about self-funded status of the plan. The insurance company that is acting as a third-party administrator will often want to clearly state that it is only administering — and not insuring — the employee health benefit plan. The covered employee can look closely at explanations of benefits and enrollment cards to see if there is a statement something along these lines: "Insurance Company, as third-party administrator of the Employer Company, Inc., health benefit plan."

3. Avoid futile efforts.

Self-funded plans have fewer consumer protections than fully insured plans. If a particular consumer protection is not present in a self-funded plan because it is not required by law, a person should understand it may be very difficult to obtain the protection. For example, the employer is the final decision maker on claims in a self-funded plan — even if the employer lacks the medical expertise to make the decisions. Contesting the employer's role as the final decision maker in a self-funded plan is often unsuccessful as the employer is the final decision maker. If a claim is denied, an effective use of time and effort may be determining an effective appeal of the denial, or exploring new coverage options — such as switching to a spouse's dependent coverage or enrolling in individual coverage through an ACA insurance exchange.

G. Conclusion.

While self-funded plans lack important consumer protections found in fully insured plans, they still offer important levels of coverage and benefits to people with CF enrolled in employer-sponsored health benefit plans. Regardless of whether the employer's plan is fully insured or self-funded, individuals with CF enrolled in an employer-sponsored plan should learn about the type of coverage the employer offers and what limits and restrictions are placed on benefits. This is an ongoing effort, because plans and benefits are renewed annually and often change. The goal is always to assure that the plan of benefits, however it is funded, allows a person with CF to visit their CF Care Center, be admitted to a hospital that treats CF patients, and covers prescription medication. ▲

Beth is 54 and has CF. She is an attorney who specializes in disability law and is a Director and current 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.

FOCUS TOPIC

INSURANCE ISSUES

The Fight Of My Life

By Lora Moser

Twenty-eighteen was the worst year of my life, mentally and physically. In 2017, my husband accepted a position with a new company that required us to relocate to Austin, from Fort Worth, TX. His new salary put us outside the income guidelines (by a mere $650 annually) to qualify for the Healthwell Foundation grant that I and so many other CFers depend on for life-saving medications.

I received a letter from Healthwell requesting our tax documents pursuant to their audit of my file. After reviewing the files and discovering my husband's income, they voided my grant in December 2017. At this point, I had already been on Orkambi for 17 months. I wasn't quite in full panic mode because I didn't have a complete understanding of how Medicare works in combination with a private insurance plan. I knew that my husband's private plan went into effect on January 1, 2018. I thought I was going to be okay because I was listed as a dependent on my husband's plan, as well as having my own Medicare coverage. I convinced myself that *surely* having both policies meant my copays for the more expensive drugs (Orkambi, Pulmozyme, Cayston, and Creon) would be manageable due to the dual coverage. I was wrong and misinformed. Over several phone calls, I had numerous employees at our private plan's company, as well as Medicare customer service, confirm that I could be a dependent and could have the dual coverage — that the private plan would be my primary coverage and Medicare would be my secondary coverage. Proper training could have saved me so many tears, grief, and roughly $3,000 in insurance premiums.

In reality, dual coverage is not allowed when you have Medicare concurrently with a *private* medical insurance, private being the key word. In other words, you cannot be listed as a dependent on a private insurance plan if you also have Medicare as it is considered "double dipping." We paid about $550 in monthly premiums for that private plan from January-June of 2018 with zero benefits in exchange.

LORA MOSER

After attempting numerous times to fill those four expensive drugs, we quickly realized that the copays were not financially possible. I had to do without those four drugs from January-August 2018. We also discovered that Medicare recipients are automatically excluded from receiving any sort of copay card from drug manufacturers, and my husband's income prevented me from qualifying for any assistance plan. It seemed the only way around this predicament was divorce. Neither my husband nor I wanted a divorce and were wholeheartedly opposed to that idea. Why should I have to be divorced in order to get the drugs I so desperately need?

I started 2018 with approximately 56% FEV1. Because we exceeded Healthwell's income guidelines by $650 annually and therefore struggled to obtain my medications throughout the course of the year, my lung functions took a huge hit. Immediately prior to my hospital admission in August of 2018, my FEV1 had dropped to an all-time low of 31%. Healthwell says they take extenuating circumstances into account and will reconsider applicants on a case-by-case basis. I provided numerous documents, including copay quotes from my Medicare plan for the costly drugs, which totaled thousands of dollars each month. I made zero progress.

Thankfully my lungs have rebounded, but I know I won't always be that lucky. Miraculously, my Healthwell grant was reinstated a few weeks after

> After attempting numerous times to fill those four expensive drugs, we quickly realized that the copays were not financially possible.

being discharged from the hospital. After the grant was reinstated, I attempted to refill one of those four drugs. Healthwell then informed me that since I hadn't had any activity with my grant in 90 days, they dropped my grant balance to zero dollars. I was *livid*. I told them that they were the reason I haven't had any activity, and that I should have still had a balance of $13,100 remaining for 2018. I felt so defeated. So much for fighting for my life, only to have the one lifeline I thought I was getting back be slammed in my face. They told me that they could make no promises, but to reattempt to fill the script. I couldn't believe it: the script processed, and the remaining balance was reinstated for the remainder of the enrollment year.

Fortunately, Patients for Affordable Drugs got wind of my scenario. Without their assistance, I don't know that my grant would have been reinstated. My unfortunate predicament boils down to pharmaceutical greed. And my ordeal inspired articles about me in the *New York Times* and the *MIT Technology Review*. The NBC affiliate (KXAN) in Austin, Texas, interviewed my husband and me within a couple days of being discharged from the hospital. I was the lead story in that night's 10 p.m. news.

I feel like this happened to me for a reason. I've always been a fighter. I was raised to talk to people about my CF, not to shy away from it. Maybe, somehow, my willingness to scream this situation from the rooftops to those who are willing to listen will help bring light to Big Pharma greed. As CFers, we fight for our lives every single day. Pharmaceutical greed is killing us faster than CF. Drugs do not do us any good if we cannot afford them.

As for 2019, my Healthwell grant was depleted in early August, so I am back in the exact same scenario as I was in 2018: no access to costly drugs until I am eligible to reapply for the Healthwell grant in November.

I am a 41-year-old CFer living in the Austin, TX, area. I was diagnosed in 1979, at age two with "failure to thrive." I will continue to fight this fight for all CFers past, present, and future. ▲

Lora is 43 years old and has CF. She lives with her husband, Casey, and dog, Bella. When she's not visiting her stepsons, Conner and Jace, she's gardening, kayaking, snuggling with Bella, or working on kitchen and bath design with her husband.

BYRNES continued from page 5

become roaring rivers. The final mile felt like ten miles. But we made it to the car before dark, by 7 p.m.

This was life's essence. With goals, there is always potential for disappointment. We didn't make it. But we came so close. The summit was two more miles from the saddle and 900 feet of elevation gain. Every goal requires judgment and careful realistic assessment of what's possible. Younger and faster people passed us going down the mountain and told stories of how their hair stood up and cell phones stopped working due to the electricity in the air. We made the right decision. Our fitness apps said we burned 6,500 calories that day. We were all overflowing with gratitude to be able to attempt this mountain and be healthy enough to do it. Though everything hurt, thankfully, we all returned home safely.

Climbing a mountain is just a story, but it was also a spiritual experience. It was an act of faith, hope, trust, and personal belief in our ability to set a goal and pursue an aspiration. During the agony on the trail, I had conversations with God. Something bigger than I was going to keep me going.

We all have our own mountains to climb. Maybe they are health related, but they could be related to work, relationships, parenting, fitness, or other goals. We know there are obstacles in the way. We know there's a destination, but it's the journey toward the destination that matters. Just a few months ago, Trikafta was approved and thousands of CF patients are hoping for a new chapter in their CF. Many of you have worked so, so, so hard for this turning point. One step at a time, one treatment at a time, you have arrived at this summit. Enjoy the view! Prayers are hung, and we wait for new chances for goals, hopes, and dreams to come true. Whatever those are, may your own striving and determination be part of the adventure.

This article is dedicated to Mary Convento and Jessica Martens, devoted CFRI superstars and fellow Mt. Whitney mountaineers. Isabel is 47 and has CF. She lives in San Mateo, California, with her husband of 20-plus years, Andrew. She can be reached at isabear27@hotmail.com.

Droning On

By Jeanie Hanley

It was that kind of late winter day in Southern California — sunny with clear, blue skies after a recent rain — perfect for "exercising" outside. Exercising is a term used loosely, since any walking for me at the time was considered exercise whether it be at a snail's pace or brisk. That was four years ago. My health could change on a dime, from one day to the next, even from morning to nighttime. On this particular morning — a morning that would haunt my memories for months and cause indescribable anxiety — my adult kids all happened to be home. I felt good, so off we went to the beach-side cliffs and started down a steep walk down to the shore.

In hindsight, red warning flags followed me throughout that walk. But I'll come to that in a second. First, here's a little background on what was foremost on my mind, and what I was determined not to dwell on that day: insurance issues.

My long-term disability ("LTD") insurance company recently changed hands and their annual request for information (to explain why one is on disability) did not satisfy them. They felt that I could work full time despite my daily symptoms, hours of treatments, therapies, and regular hospitalizations. Yeah, sure. They requested more information and details and still didn't seem satisfied. I couldn't fathom what else they wanted.

And so I welcomed the distraction of my family visiting. Off we went on the 20-minute drive to the cove. After parking, we headed to the steep dirt path (fortunately, wide enough to fit the five of us) cut into the cliffside. We descended slowly but steadily. I was clearly the one slowing down my fit family. Still, it was easy to be in the moment with the seagulls hovering, the waves crashing, and seals barking.

About half of the way down, the buzzing of an overhead drone briefly interrupted my reverie. Although an unwelcome noise, the louder sounds of the ocean shore drowned out the incessant buzzing the further we descended.

JEANIE HANLEY

The drone later appeared above the ocean swells, again disrupting our jaunt to the shore. We all hoped a big wave would appear and engulf this intrusion.

Ascending the steep cliff on the return walk proved more challenging than I anticipated. My kids decided to help me out by counting each step, pushing me to make it to 10 steps before we rested, then another 10, followed by rest, and so on. During one of our rests, my son spotted the blows (aka spouts) of two grey whales in the distance. It was beautiful. We also took a selfie picture at the edge of the path, the angle of which looked as if we had rock climbed the cliffside!

Days later, during my CF clinic appointment, the social worker told me that the LTD folks had both video and pictures of me hiking that day. The culprit — that annoying drone. Talk about a violation of privacy, something you'd expect in a sci-fi movie! Yes, that was the drone! One of the pictures was similar to our selfie. And yet the videos showed neither the rest periods nor my labored breathing. Based on these, LTD spies had posed the question that if I could hike, then why can't I work? It enraged me and felt like an invasion of privacy — that they would use the drone to follow me and my family and snap pictures and video of us. As a result, for many months afterwards, I felt that I had another set of eyes watching me while on my walks or when I tried to push myself exercising or, really, anytime I was outside. I cringed every time I heard a drone.

Fortunately, my CF clinic responded swiftly and strongly to the company, stating that indeed part of the CF treatment program is to exercise as much as possible. The clinic added that they were very pleased to see I was following the prescribed regimen and that, no, they didn't give the go-ahead to my returning to work full time.

Still, it wasn't enough. The LTD insurance company then set up an appointment with one of their pulmonologists for what they called an "outside review." I learned I would have to subject myself to a doctor I don't know, didn't want to see, who was at least an hour's drive away (under the best L.A. freeway conditions), and let this stranger decide whether I should continue on disability.

This is when I called Beth Sufian

from the CF Legal Hotline for help. She guided me through the process — what to expect, what to bring, and basically every aspect of the upcoming visit. I had to wait over a month for the mandatory pulmonologist appointment, which meant that I drove myself crazy with worry as evidenced by my hair falling out more and more every day.

Normally I am quite an optimist and summon my energy every day to feel as well as possible by completing my self-care, treatments, and exercise. I don't think about how difficult my life is, nor do I think of myself as sick and certainly, I don't dwell on symptoms. But the purpose of this forced LTD company visit meant that I had to prove my illness and put on display how sick I am to a stranger, which went against every bone in my body.

> "The purpose of this forced LTD company visit meant that I had to prove my illness and put on display how sick I am to a stranger, which went against every bone in my body."

By the day of the visit, I was an utter mess. My husband drove the 1-½ hour scenic drive through L.A. traffic. At the doc's office, the front desk had the nerve to ask me for my health insurance information, which I refused to give. They wanted to get paid by my health insurance and by my LTD insurance company? Fortunately for them, they didn't insist.

It turned out the pulmonologist was not knowledgeable about CF. He was a community physician who had only come in contact with CF when he was in training over 40 years ago. What a different disease it was then. With such cursory knowledge, he should've signed me off as a miracle, still alive at 53 years old, and let me be on my way immediately.

Instead, I had to power through the stressful two-hour visit. He thought the main cause of death was due to intestinal and not respiratory issues. I wish I had brought charts and teaching slides. They would've been useful during my explanation of what CF was: the genetics, the mutations, the clinical symptoms, and the treatments. Not knowing whether he was understanding the complexity and severity of CF throughout our visit was nerve-racking to say the least.

By the end of the visit, he informed me that he approved the continuation of my disability. I walked out of there feeling as if I could breathe again and that a huge weight had been lifted off my shoulders. My hair started growing back. The spike of adrenaline whenever I saw a drone eventually stopped.

All the preparation with CF Legal resulted in the LTD insurance company continuing my status. What an outstanding resource for our CF community. I believe their guidance ensured that it all worked out. ▲

Jeanie, 57, lives in Los Angeles, in a city that has recently placed heavy restrictions on the use of drones. She also is a physician and Treasurer of USACFA. Feel free to contact her anytime. Her information is on page 2 of this issue.

PAM continued from page 11

Are there other options if my insurance does not cover fertility services?

Ask your employer! If you have an employer-based plan, ask your HR team if they would consider adding it to the benefits plan. This does not require you to be open about your CF, because infertility is common. One person told me, *"We talked to our employer about adding IVF to the insurance plan. While they were unable to add fertility coverage to the insurance due to it being a religious organization, they quietly slipped us a check from a discretionary fund to help us afford it."*

If talking to an employer is not an option, secondary insurance plans can help defray costs. There are also grant-giving organizations that help fund fertility services.

Finally, be kind to yourself during this process. It takes an emotional and often physical toll. I want to end with some advice from others with CF who have gone through this: *"Make sure you are taking care of yourself because this process can wear you down. Even if you do not carry the baby, you will need all the support you can get." "Everything you do in your fertility journey is personal." "Things do not always go as planned and your emotions will be all over the place." "Prioritize your relationship and up your self-care by 10x."*

If you are struggling with infertility and need resources, Resolve: the National Infertility Association, is a good place to start. https://resolve.org/

CFF covers the basics of fertility services, with special consideration to CF: https://www.cff.org/Life-With-CF/Transitions/Family-Planning-and-Parenting-With-CF/Alternative-Ways-to-Build-a-Family/Assisted-Reproduction-and-CF/ ▲

Molly Pam is 31 and lives in San Francisco with her husband Adam and betta fish Sally. You can reach her at mpam@usacfa.org or find her on Instagram at chefmollypam.

The Insurance Binder

By Leah Sands

One morning, years ago when I was still a teenager, I came downstairs into the kitchen to see my mother with her hands over her face. I tried to tiptoe into the room, because I didn't know the context of her issue and wasn't quite in the mood to sort it out with her. Despite my best efforts to be quiet, she heard me, as I have been cursed with heavy walking feet. She peered up at me with tears in her eyes and began to unload. She told me how stressful the morning had been: she had fought an ongoing battle with our pharmacy in an attempt to get my medications covered by insurance. I knew just as well as she did that these medications were necessary to keep me alive and well. She told me how the pharmacy couldn't help and that she wasn't sure when I'd be able to get my medications. Frustration was written all over her face. As we talked through the stresses of my medical needs and the insurance companies, she decided to show me her spreadsheet. It was an attempt to make sense of, and gain control of, her child's terminal disease — a mighty task indeed. The sheet had dates, addresses, doctors, treatments, and medications. I was a young woman nearing my 18th birthday, and my mother thought it was time to pass on the torch and let me take on my medical care. She spent an hour or so explaining each detail of the sheet and then printed fresh copies, slid them into sheet protectors, and then into a binder. She handed over the bible of stressful, complicated, and pertinent information. I was excited and scared all at the same time, but ready to take on every aspect of becoming an independent woman with cystic fibrosis.

Over the next several years, I learned the ins and outs of my insurance plan, which was still my father's because I was a college student. I seemed to have it figured out and was able to get my prescriptions, doctor's visits, and procedures covered without much hassle. When I graduated college with my nursing degree, I landed my first real job at a hospital. When signing my papers with HR, the decisions were overwhelming, and the most stressful was picking the insurance coverage. Which one should I choose? From PPO to HMO, with or without deductibles, and premiums — what's a premium? I picked one, hoping I had made the right choice. Luckily, I had, and everything was covered for that period in my life.

LEAH SANDS

I continued with life — got married, moved to another state, and got a new job. The only difference with my coverage was that I was now under my wife's plan. She had picked it when single, and we hoped that all my medical needs would be insured. We came across hurdles with getting specialty medications that could only be obtained from pharmacies that specifically dealt with CF. My wife would yell on the phone things like, "if you don't cover this medication, then you'll be paying a lot more when she's hospitalized!" After weeks of pushing, the insurance company finally gave in. We spent many more years and phone calls fighting for coverage, pleading with the woman who had the ability to hit the "approve" button.

I have spent years working in the healthcare industry, including medical billing and medical insurance underwriting. I have seen the front- and back-end inner workings of insurance. I have fought for my patients, just as my wife has fought for me. It is an exhausting task, especially when you're sick. I feel fortunate that I have the experience and knowledge to now handle insurance challenges easily, but it doesn't take away the number of denials or the fact that insurance companies are businesses too.

Businesses — all about the Benjamins. But, in my case, it's so much more than that. In addition to my full-time job, I started my own business in IT support. I would love to push my business further, focusing my time solely on its growth. However, because I need good health insurance, it's in my best interest to keep my full-time job so I can keep my healthcare benefits. I spend my time bouncing between jobs, sometimes burning the candle at both ends, so I can keep my coverage.

Fortunately, with the recent CFTR modulators, I have been able to pursue my dreams. I never would have had the energy or health to have my own business and do the things I'm doing without these medications. I'm so thankful for these advancements, and grateful

that my insurance challenges have been minimal. However, I know that is not the case for everyone with CF. Some have been fighting diligently for their life-changing pills. From government insurance programs to private payers, most people are having to push through challenges to get their medications. Social media conversations are peppered with people asking for assistance with insurance appeals and for answers from those who have managed to beat the system. And then there's a fear, a nightmare really; the fear that the pills will be available, then suddenly stripped away because they're no longer covered. The price of the medications is very high, and without insurance covering them, no one would be able to afford the out-of-pocket cost.

I know that not everyone has that person in their life who will hand over a well-thought-out, sheet-protected binder of pertinent information. In fact, most things in life don't come in a nicely packaged guide. But if I were to hand over a binder of insurance information to my fellow CF warriors, this is what it would contain:

- Keep a couple months stock of meds in case there are issues with coverage, like needing prior authorizations
- Talk to people on the phone at the insurance companies — sometimes human-to-human contact is most helpful
- Get help from your medical team and social worker
- Find programs that will help with copays, like patient assistance programs
- Be as educated as you can about insurance processes
- Get advice from others with CF
- Lean on family and friends for help fighting when you need help
- Stay updated on information and treatments for CF so you have evidence to fight
- Keep pushing — anything can be overridden ▲

Leah Sands is 38 and from Detroit, Michigan. She loves photography, team sports, and traveling. She can be contacted by email at leah.sands@gmail.com.

TILLMAN continued from page 7

1- and 2-year mortality in cystic fibrosis. There is a great need for developing an accurate prediction model for mortality to identify patients who would benefit from expedited referral to a lung trans-plant program. The researchers sought to develop a clinical tool for predicting 1- and 2year risk for death. They found that the combined effect of CF chronic health status and CF intermittent shock risk provided a simple clinical scoring tool for assessing 1-year and 2-year risk for death in an individual person with CF. The prediction model has some limitations. For one, although the majority of the predictor variables were obtained in the previous 12 months for the 1-year model or in the 12-month interval 1 year before for the 2year model, the annual measurements do not reflect acute deterioration in health status. The researchers noted that it would be helpful to use encounter-based records in the future. In addition, the study population includes observations over the course of 30 years, during which CF prognosis and treatment have changed considerably.
https://tinyurl.com/rovg3tk

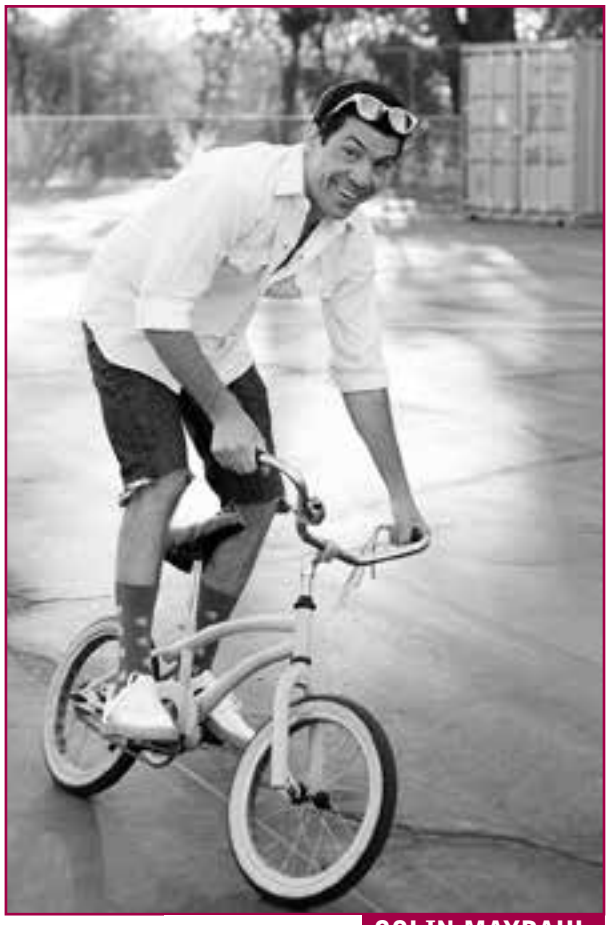
Modifier Gene May Explain Why Some With Cystic Fibrosis Are Less Prone To Infection

Cystic fibrosis is caused by an inherited mutation in the cystic fibrosis transmembrane conductance regulator (CFTR) gene. Due to this mutation, the CFTR protein doesn't embed in cell membranes to form a channel for chlorine ions the way it should. As a result, mucus-producing cells secrete a thicker-than-normal mucus that can create blockages in the lungs and digestive system. In the lungs, this thicker mucus can help bacteria thrive, making lung infections a serious and chronic problem for many people living with cystic fibrosis. Yet some people with cystic fibrosis don't develop lung infections as early or as frequently as others. Researchers discovered that genetic variations dampening expression of another gene, called RNF5, offer a likely explanation. In the study the team found that individuals with cystic fibrosis who carry specific genetic variants lowering expression of RNF5 have more mutant CFTR protein on their cell surfaces. Even if the CFTR protein isn't totally functional, it's likely better than having none at all. The RNF5 gene is located in the Major Histocompatibility Complex (MHC). Genes in this region encode molecules that are displayed on the surface of most cells in the body. They play an important role in how the body responds to infections. Scientists have long known that everyone has their own set of MHC gene variations, and that they make people more or less susceptible to infections or autoimmune diseases. But be-cause the genes present in the MHC region are so dense with variations, they have not been well-studied for their direct link

Continued on page 24

Dance Yourself Clean

By Colin Maydahl



COLIN MAYDAHL

The last four years have been a bit of a roller coaster. In December it will be four years since my ex-wife and I split. I didn't want the marriage to end at the time, but we were not meeting each other's needs and are both now in better places. I went to counseling, leaned on friends, and started buying more tickets and saying "yes" to invites from friends (even if I later had to cancel because I got sick). Orkambi (which I was in the study for) had given me a lot more freedom from CF life, but I knew I had to keep putting one foot in front of another. It started with an Iggy Pop concert in San Francisco, which I attended with a very good friend; then a plane ticket to San Diego to see my dad; and two tickets to see LCD Soundsystem (my absolute-favorite band) with my best friend of 20+ years. As the summer progressed, I started realizing that having all these new experiences fed my soul and made me a happier person. I had things to look forward to and was making new memories all the time. I wasn't just wondering when would be the next time I would end up on IV antibiotics with a hospital stay (which was my life before Orkambi).

Now, as 90% of us have either begun or, it is hoped, will soon begin, taking Trikafta, our lives are going to start changing, and we won't be the same group of sick people living day-to-day, survival-type lives. Our identity and relationship with CF will change, and we will hopefully be able to take the lessons that CF has taught us and apply them to more of a normal life. We will be able to buy tickets, plan things, go on trips with a smaller CF monkey on our backs, not the gorilla we have been carrying our entire life.

Think about what you want to do. The limitations on our bodies will be changing and you will be able to have some more fun in life. Get tickets to a concert, a destination, or even just say yes to a party at a friend's house. Put more energy into having fun and celebrating life. You will enjoy looking forward to the next show and remembering the last. Our lives have already been cut short but now lots of us are getting a second shot at life. Where are your friends tonight? ▲

Colin is 37 years old and lives in Auburn, California. He works part time as a bicycle mechanic, loves to cook, and loves to dance. He can be reached at colinmaydahl@gmail.com.

Cystic Fibrosis Mothers

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

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

If you would like to join our Facebook support group, please e-mail Karen Vega at: kvega@usacfa.org.

Going Through Hoops

Sometimes it seems that we always
are going through hoops to just be.
There are meds to be taken and treat
ments to do, you see.

Then phone rings. I answer and some
one says, "Let's get together."
I say, "I'd love to, but what do you
think about the weather?
Will it be too warm or too cool?"
(Having to ask that makes me feel
like a fool.)
"At the place we are going, do they
allow smoke?"
(You know that most 'No Smoking'
areas are such a joke!)

"Do they have good air circulation
there?"
(Or will we all be re-breathing each
other's air?)
"Will everyone follow guidelines for
cross-infection protection?"
(Or do we have to worry about com-
ing home with some new infection?)
Before you can invite someone new you must
say,
"Do you have pan-resistant bugs, B. cepacia
or MRSA?"

Going to a conference requires even more
work.
Sometimes I feel like I am a jerk.
First it's a trip to a doc to collect sputum for
culture,

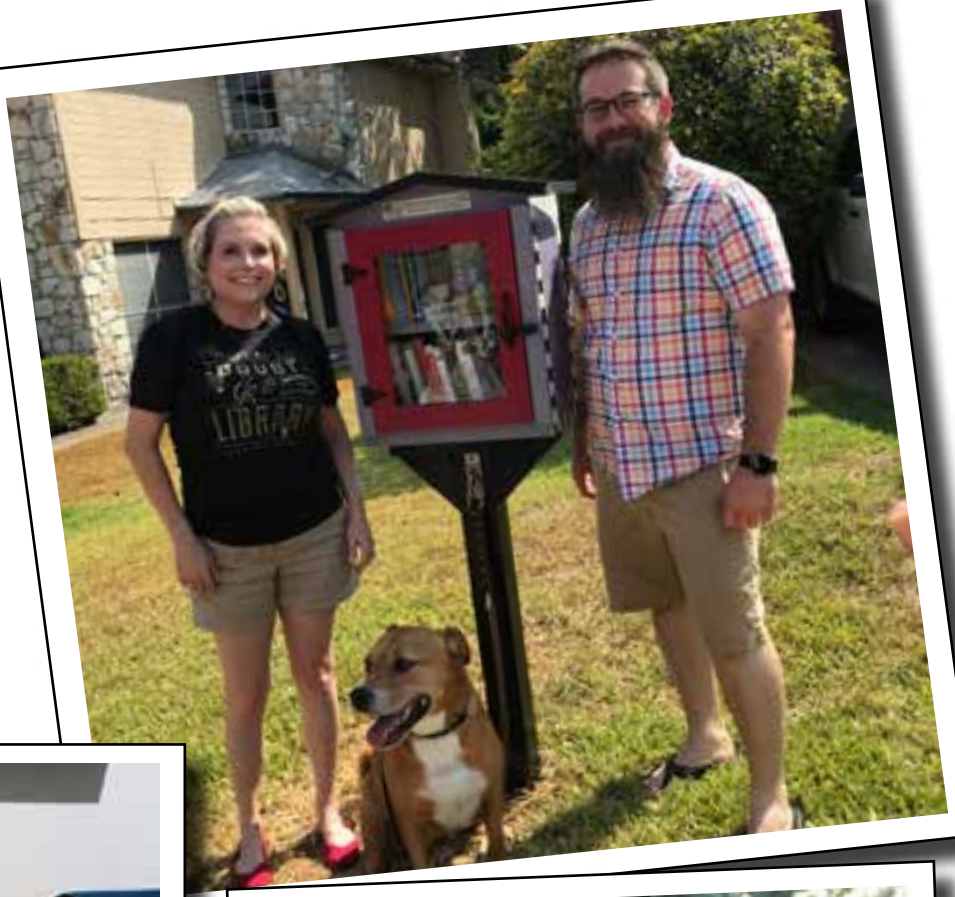
PHOTO BY STEPHEN BOYER

Then it may mean a visit to a blood-drawing
vulture.
Forms to be filled out, releases to sign.
We may risk our lives on the dotted line.

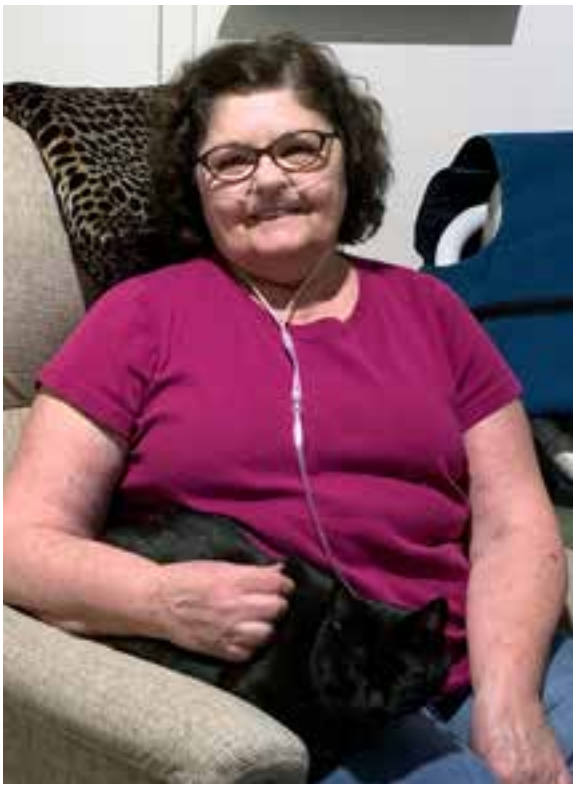
Seeing friends may be less work for you than
for me,
Since I must go through so many hoops, just
to be.

-K. Russell, 2000

FROM OUR FAMILY PHOTO ALBUM...



SYDNA MARSHALL, HER FUR-BABY HUSKER, AND HUSBAND ADAM KEYS AT THE OPENING OF HER LITTLE FREE LIBRARY ON HER 39TH BIRTHDAY.



LINDA STRATTON WITH KITTY, GRACIE, RESTING AT HOME DURING THE HOLIDAYS.

KRISTINE, RICK, AND LORENA WOOD.

MOLLY PAM AND HER HUSBAND, ADAM COHEN, VISITING THE PYRAMIDS AT GIZA IN EGYPT.

LEAH SANDS WITH HER WIFE, HEATHER, AND SONS, LANDON AND GAVIN, AT CHRISTMAS 2019.

MARK AND MARYGRACE TREMBLAY CELEBRATING AFTER CONQUERING THE 2019 CF CYCLE FOR LIFE IN SARATOGA SPRINGS, NY, ON SEPTEMBER 22, 2019.

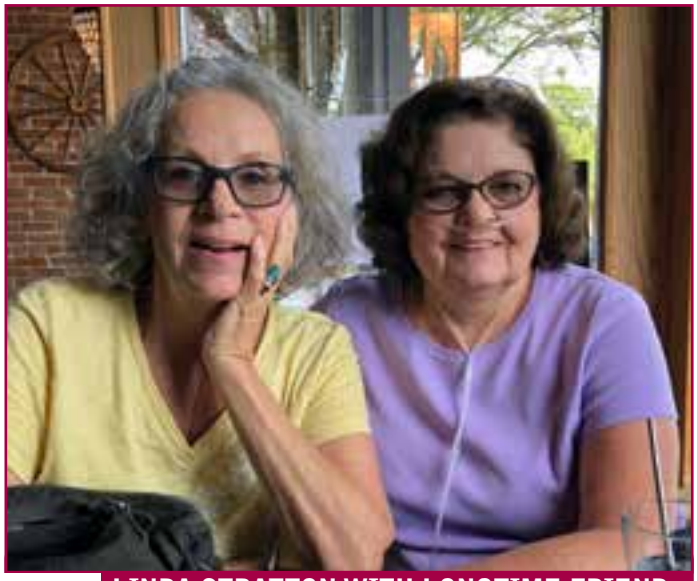
My Trikafta Story

By Linda Stratton

As an elderly" CF patient and a recent recipient of the new medicine, Trikafta, I feel the need to share my story. I wrote the following poem last summer when my FEV1 was hovering between 20 to 24%. Unable to function with any activity, I was truly making plans for end of life. Having finished my Last Will and Testament, I made sure my sister was aware of all of my wishes. The day I turned 65 years old, I had a port placed, and I felt I knew exactly what my future held.

Façade

Abundant makeup, curls in my hair, clothes that cover a belly bloated,
all a façade of how I truly feel.
Cystic fibrosis – a vicious circle: hospital stay, feel good, lung congestion, sick, then hospital again.
Through the years circle gets smaller, latest pulmonary test the gauge.
One day out equals several at rest, lungs rebelling, bloody results.
Over and over people ask, "how are you, how are you, how are you?"
I reply, "I'm good, I'm good, I'm good."
They comment on my healthy look, unaware of the energy it takes to get there.
Only those closest see the day-to-day struggle – nebulizers, pills, a tight schedule to keep, just to breathe.
Through steroid-laced eyes I look in the mirror, this puffy face unrecognized.
Yet, people still say "you look so good," why do I feel they lie?
I wait for the day God calls me home, my heavenly body running with glee, leaping with joy, façade no more, finally free!



LINDA STRATTON WITH LONGTIME FRIEND, DEBORAH FORSYTHE, SUMMER 2019.

TILLMAN continued from page 19

to diseases.

The research team took a new approach to analyzing MHC gene variants by grouping them. That allowed them to more easily identify associations between genetic variation, gene expression levels and their effects on complex diseases.

https://tinyurl.com/yx2cr5x7

Youngest, Oldest Adults Experience Lowest Survival Rate After Lung Transplantation

Of all factors analyzed, age emerged as the most significant risk factor for death at all timepoints after transplant, with the youngest and oldest having worse survival than middle-aged patients. In addition to age, risk factors for death for all patients included higher creatinine, single lung transplant, hospitalization before transplant and in-creased bilirubin. Notably, the researchers also found that risk factors associated with death varied across age categories, but social determinants, including lower education and government insurance, disproportionately affected patients younger than 30 years.

https://tinyurl.com/t3dsjrv

Inhaled Cyclosporine May Increase Survival, Pulmonary Function After Lung Transplant

Inhaled liposomal cyclosporine may prevent bronchiolitis obliterans syndrome (BOS) progression following lung transplantation. Cyclosporine is traditionally given orally in pill form as part of the post-lung transplant standard-of-care regimen as 1 of several anti-rejection medications transplant recipients must take for the rest of their lives to prevent chronic organ rejection. Despite these measures, the immune system often succeeds in attacking the transplanted organ, causing nearly half of lung transplant recipients to develop BOS within 5 years of getting their transplant. For the current study, 21 patients who underwent lung transplant and were in the early stages of BOS for 48 weeks. All of the

At the end of last year and into the beginning of this year, I learned of a medication in clinical trials that had a lot of promise for me. It was made for one of my specific mutations, DF508, as opposed to the Kalydeco that I had been taking for at least three years. In September, I was informed I had been approved for early dispensation of Trikafta. Because I am allergic to all inhaled antibiotics, my doctor and I anxiously awaited this new medication, viewing it as my last hope. Kalydeco helped in some areas, but it didn't perform as needed for my lung function. As stated previously, I had high hopes for this next step in care.

To my disappointment, the day I went to pick up the Trikafta, I woke up sick. I could feel a cold coming on fast — my sinuses were raw and my airways inflamed. Worried they wouldn't allow me to start the new medication with these symptoms, I hesitated to say anything to anyone. During some required testing, such as PFT, a sinus scan, blood work, etc., I finally blurted out that I was getting sick and was told not to worry: it wouldn't be a problem. What a relief!

I started the Trikafta, and — no surprise — the anticipated cold virus came on strong and brought on an abundance of mucus! And yet, I noticed my sputum wasn't getting dark and ugly as it had in the past. In fact, each day it was getting lighter and lighter. I had been preparing for a hospital stay, but I was quite amazed when sputum production started to decrease as well, and I felt better with each passing day. I showed no signs of infection, no fever, and no bloody sputum, which meant no hospitalization! After a week on the new medication, I was feeling normal, like how I imagine people who don't have CF feel when recovering from a cold. After a month of treatment, with the addition of this incredible medication, I had no wheezing or coughing, day or night. My last FEV1 was an amazing 36%. WOW! I can't wait to see how well I do on my next PFT.

There are a few minor side effects with Trikafta, though nothing to really complain about. I have, however, experienced some weight gain, which for most CF patients would be a good thing. I'm one of a small population of patients who are overweight for some reason. The extra weight is acceptable considering the improvement in my lung function. I sit in amazement each morning, and several times during the day, when doing my nebulizer treatments. For the very first time, my sputum production is minimal and almost white. I don't ever remember it being so devoid of color...not in my entire life! Being several weeks in, at this point I believe Trikafta will be an amazing blessing to a major population of CF sufferers. I'm very thankful for my CF doctor, who's willing to pursue new avenues and keep trying.

So, I find myself daydreaming about a new life —a bout what I want to do in the near future. I'm feeling good, strong, and courageous most days and truly think there are a lot of possibilities for me now that I wouldn't have considered two months ago. Then, I was just waiting to die; but now, I have hope! ▲

Linda Stratton is 65 years old and has CF. She lives in Louisville, Colorado, with her three-year-old cat named Gracie. They enjoy early morning exercise and afternoon naps.

patients were given conventional oral immunosuppressants, including tacrolimus, myco-phenolate mofetil, and prednisone; 11 were randomised to also receive the inhaled cyclosporine twice daily for 24 weeks. The researchers found that BOS progression-free survival was 82% for patients who took inhaled cyclosporine versus 50% for those who received standard care alone. Lung function measures of forced expiratory volume and forced vital capacity stabilized in the treatment group, but worsened with the controls. Most importantly, the median survival for those who received the inhaled cyclosporine was 4.1 years compared with 2.9 years for those who did not receive the added therapy.
https://tinyurl.com/tcwdwo8

New Approach To Treat Cystic Fibrosis Lowers Risk Of Lung Transplants

In people with cystic fibrosis (CF), a new approach was found to reduce inflammation, which has the potential to reduce the need for lung transplants and lower the risk of death. The main cause of death in people with CF is lung disease, which is driven by severe inflammation and chronic infection in the airways. The researchers found that one of the most aggressive bacteria found in the lungs of those with CF caused certain immune cells to change their metabolism. This change caused the immune cells to produce a protein that causes more inflammation. They identified that high levels of the protein were associated with worse lung function and a higher risk of death or need for a lung transplant. The team then used a small molecule called MCC950 to reduce levels of the protein in a laboratory model of CF. In addition to reducing inflammation, this also helped clear the lungs of bacteria. This marks the first time that researchers were able to stop this protein in CF in vivo by targeting cell metabolism, which could potentially lead to a new approach to treating inflammatory diseases like CF.

Continued on page 27

WELLNESS

Anxiety Sucks And What To Do About It

By Julie Desch

I have thought about writing an article about anxiety for *CF Roundtable* for years but, for one reason or another, it was never the right time—either I was caught up in my own anxiety and didn't have the focus required, or I was at a safe distance from anxiety and didn't want to risk visiting it.

Anxiety and depression are two major mental health downsides of living with CF for both a large percentage of CF adults and an even bigger percentage of mothers of individuals with CF. Anxiety is no joke. Having lived over 59 years with CF, I have personally experienced plenty of both anxiety and depression. I've also witnessed how anxiety wreaks havoc in the lives of many of my friends with CF. The way I see it, anxiety is a form of fear. It exists on an emotional continuum ranging from mild anticipation to outright terror, but its position is closer to the latter.

Living with CF sometimes reminds me of that horrible (and downright cruel) "learned helplessness" experiment. In brief, there were three groups of dogs in this experiment: a control group that wore only a harness; a second group that wore a harness, which occasionally shocked the dogs, but they learned to stop the shock by pressing a lever with their noses; and a third group of dogs, each of which was harnessed to a dog in group two, thereby receiving random electrical shocks that also stopped at apparently random times. The dogs in group three, therefore, had absolutely no control over either their suffering or its relief.

The shocking (pun intended) outcomes were discovered after this initial "conditioning." Dogs from each group were placed in a box with a shallow divider in the center. One half of the box was electrified and would provide a small shock as each dog was placed in it. The other half of the box was shock free, so all the dog had to do was step over the divider and the shock ended. Dogs from the control group and group two easily learned this maneuver and quickly got out of harm's way. But the dogs from group three, the ones who had learned that suffering and its relief were equally random, didn't even try to escape. They simply lay down and proceeded to get shocked. This was termed "learned helplessness." You could perhaps suggest that these dogs had actually "learned" that depression and anxiety were part of life and, as a result, would not even try to make their life situation better. Observing these dogs, you would be justified in calling them depressed and would, of course, imagine their minds being in a state of continuous anxiety.

Now I'm not calling us helpless by any means. But it seems to me that CF exacerbations are somewhat like those shocks given to the dogs in group three. Despite doing everything we can to stay healthy, exacerbations happen, often "out of the blue." And if I think about my hospitalizations, it seems to me that my baseline, after I get out of the hospital, is completely up to fate and not really due to anything I have done. This is why it makes sense to me that anxiety and depression are so prevalent in our community: it might seem as if it were just hanging out and waiting for the next "shock," and this provokes anxiety. It doesn't even have to be, and usually isn't, a conscious thing. It's not as if we're really just sitting around wondering when we are going down — it's more like a subtle tension in which our subconscious minds marinate. Anxiety then pops up in strange ways, like social anxiety, body image anxiety, or just random "free-floating" anxiety. For me, it rears its head randomly in occasional panic attacks.

JULIE DESCH, M.D.

So what's a body to do in these scenarios? Lie down in a box, accept our fate, and not try to help ourselves? Of course not. Instead, we attempt to work with the anxiety (and any emotion, for that matter) in a way to make it less unpleasant by utilizing available techniques, such as the mindfulness technique of "deconstructing" emotion. And really that is what we are after…not to be bereft of emotions, but to experience our emotions with less suffering.

Mindfulness of an emotion such as fear involves being aware of its components in real time (the "now") with an attitude of "allowance," without judgment. Imagine you have two sets of earbuds in your gym bag, one white

and the other bright red. They are in a completely tangled ball of wire that, from a distance, takes on the color pink. In this metaphor, the pink color is the emotion of fear. But when you get close to the tangled mess, you see that, really, the pink color does not exist — rather, there is a combination of both white and red. The white wire signifies thoughts, and the red body sensations. The emotion of fear, in this case, is then seen to consist of a very tightly congealed collection of thoughts and bodily sensations. When you allow yourself to turn toward the fear with full awareness and move in close, you can discern and better understand the intertwined sensations comprising fear. As you carefully and methodically untangle the bodily sensations from the thoughts, you become very clear about what is present. This is when "allowing" comes into play. The bodily sensations may certainly be unpleasant. Maybe you feel tightness or queasiness in the stomach area, or maybe there is a pulsation in the throat region that feels uncomfortably constraining. The "allowing without judgment" attitude toward these sensations lets them be as they are, as big as they need to be; and when there is no internal resistance to them, they then simply move through the body. With continued practice you can then become able to "see" the individual thoughts that arise with these sensations, and you begin to discern those that contain useful information vs. those that are unhelpful and very likely untrue. After all, thoughts are simply words or images that arise in the mind. Just because they are there does not mean they are true.

It takes quite a bit of courage to turn toward fear in this manner. After all, the emotion of fear has evolved to convey a very specific message: *run*. We naturally avoid fear and any situations that give rise to it. If you don't take the risk and settle into the discomfort surrounding fear, however, you can't see the interlocking elements and therefore you have no standing to challenge the thoughts that arise.

When kayaking, a keeper hole can be dangerous territory. A keeper hole is a very powerful hole or hydraulic in which the foam pile or backwash surrounding the hole is so strong that it doesn't easily release kayaks, debris, or bodies as it recirculates them in the hole for a long time. In other words, if you get thrown from your kayak and end up in a keeper hole, you can't swim out of it normally. Albeit counterintuitive, to escape the hole you have to fight your normal instinct and instead swim directly to the bottom of the hole — you literally swim toward your apparent demise. Doing so pops you out of the circulating current away from the keeper hole. Getting close to fear feels a lot like this. Clearly seeing exactly what comprises your fear, helps release you from its viselike grip.

I am no pro at this, but I have experienced the benefits of this technique directly, and I continue to be impressed at how effective this process is when working with difficult emotions such as anxiety, depression, and all of their components. ▲

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

TILLMAN continued from page 25

httpv s://tinyurl.com/yx7mcbxy
AND
https://tinyurl.com/tgk9wgs

CF-Related Diabetes Affects Girls More Often, At Younger Age Than Boys

Girls with cystic fibrosis-related diabetes (CFRD) are exposed to risk factors of early death — including chronic infections, poor lung function, and poor nutrition — at a significantly higher proportion and younger age than boys. While CFRD shares some features with both type 1 (impaired insulin production) and type 2 (insulin resistance) diabetes, it is a clinically distinct condition. Several studies have shown that CFRD, which affects nearly half of all CF patients, becomes more prevalent as patients age, and is associated with poor lung function, lower weight (poor nutritional status), and higher mortality. Researchers evaluated the occurrence of CFRD, as well as its association with previously suggested risk factors. Results showed that a total of 21.6% of CF patients had CFRD, and that this frequency increased by nearly 10% every decade — affecting 9.7% of those aged 10 to 19 years, 24.1% of those age 20 to 29, and 32.7% of those 30 or older. Moreover, CFRD developed significantly earlier in females than males. The data also showed that carrying severe CF-associated mutations (both mutations of class 1–3), having pancreatic insufficiency, and being a woman increased the likelihood of developing CFRD by up to threefold. In addition, CF patients with CFRD had a higher risk of developing chronic infections, poorer lung function, and a poorer nutritional status, than those without CF diabetes.

Overall, the findings point out that female CFRD patients are exposed to these known risk factors for early death at a younger age and in a higher

Continued on page 32

IN THE SPOTLIGHT
With Lorena Wood

By Jeanie Hanley and Andrea Eisenman

Lorena has been a *CF Roundtable* subscriber for over 20 years and wrote an article in 1998 on how CF is a "chisel." During this interview, we enjoyed hearing her insightful theories about how her struggles have shaped her life, and the reasons why she was asked to face them in the first place. Lorena is a missionary and puts her "trust in a relationship with Jesus," which has helped her overcome many difficulties. Her sense of humor is exemplified in a CF experience she shared — something only those in our CF community could understand. It's worth the read. Meet our newest star. Spotlight, please!

Stats: Age: 53 miracles; mutations are double delta F508; latest FEV1 was 49%; CFRD since age 19; and married for 26 years. We have an adopted daughter who is 17 years old. We adopted her as an infant.

When were you diagnosed with CF?

My mom always wanted a petite girl. When I "failed to thrive" as a newborn and always tasted salty to my grandma, they took me to the doctor and did a sweat test. I was diagnosed at three months old. My mom got her petite girl.

Where do you live?

My husband and I have lived in Colorado Springs since 2005. We moved here from Pasadena, California, because the clinic doctors were talking "lung transplant." The smog was killing me. My husband had a twin brother in the area. The American Lung Association declared Colorado Springs to be one of the cleanest cities in the U.S., and the National Jewish Hospital had one of the shortest transplant waits. Our altitude is 6,800 ft. My oxygen level is the same as others here: 97%. My lungs cleared and stabilized, and the transplant team said no transplant needed.

You mentioned that CF was a "chisel" in an article you wrote for *CF Roundtable*. What did you mean by that?

CF, the chisel, has been used in my life to shape and carve me into the beautiful person I am. If a tree doesn't face any hard rains, storms, or drought, it doesn't develop a good root system. It's the same with us humans. CF has caused me to go deep in every area of my life. I can pass this strength on to others in their struggles.

What difficulties did you experience growing up that chiseled your life?

I grew up with a single mother who was an alcoholic and smoker. At diagnosis, my mom was told I had just a couple years to live. Once, when my mom was driving drunk, I prayed and asked God why I had CF. I felt like He said to me, "Lorena, it's so that others watch you and gain strength for their struggles." I accepted that as one of my life callings. CF has just been one of the tough things I've had to endure. I know I have a purpose and won't die until they all are accomplished according to God.

LORENA WOOD

Who raised you and got you through these tough times?

I lived mostly with my mom for 16

years of my life. My grandmother was instrumental in caring for us at critical times when my mom was drunk, hungover, or MIA. She took us to church. My mom was great when she was sober but, because of her painful teen years, she turned to alcohol to cover the pain.

What was your health like during this time?

My mom never really was consistent with my treatments. I kept active being on the swim and gymnastics team until my sophomore year in high school. My health took a turn for the worse as I neared 16. I had to enter into two foster homes because of my home life. It was stressful on my body. I ended up living with my older brother, his wife, and my younger brother in California for my remaining two years of high school. Glendora High didn't have a gymnastics team so I joined the badminton team. I was hospitalized more after high school. In Bible school (college), I had my first episode of hemoptysis. The blood splattering on the white snow in Minnesota was dramatic and scary.

When did you start receiving proper CF care?

I really got into a good CF care center in Orange County, California, when I was in high school.

What kind of work do you do?

I have homeschooled our daughter since first grade. My degree is in intercultural studies. My love is working with Muslims, especially refugees. I help in practical ways, assisting them in adapting to American culture.

What dreams did you have for your career? How did CF affect your ability to work and your career?

When I was younger, I wanted to be an actress or an Olympic gymnast. After high school, I became a CNA and started nursing. I then submitted to Jesus at age 19 and felt called into missions. I was going to be a midwife in Afghanistan or Pakistan. Because of CF, I had to adjust in some big ways. I have the heart for the work but not the lungs. Again, some of those dreams had to die, but others were birthed.

How did your husband handle your having CF? What did this tell you about him?

I've been open about my CF. It's a platform and springboard I use to share with others. We met in the dorm while I was doing an internship in Pasadena, California, for a Muslim ministry. I told him about my CF and it didn't stop him from pursuing me. He told me that "when we get married, I take on CF, too, and I'm going to fight for you." He really has been my top advocate, especially when I could not fight or advocate for myself. Based on previous failed relationships because of my CF, I knew he was my million-dollar man.

How did CF affect your ability to have children?

Positively. CF has taught me to fight through loss and keep getting up. We checked to see if my husband was a carrier. We then tried intrauterine insemination (IUI) twice and trusted it to God. We turned to adoption and nine years later, after six emotional miscarriages, our little preemie girl was born nine weeks early. The adoption went perfectly. We fostered after that, too, and wanted to adopt a little child named Marcel. My husband was having eye issues (he is legally blind), however, and it did not work out for us.

Did you stay in contact with your biological parents as an adult? Did the relationship with them help your health?

I lived with my mom until I was 16, when it was no longer possible. I started reaching out to my dad when I was around 20. I reconciled with them both and stayed in close contact until they both passed. My dad came to my college graduation and walked me down the aisle. My mom came to our wedding sober. I have always tried to honor my parents according to Ephesians 6:1-3.

What are your current CF health issues?

My lungs have always been my primary struggle. I've had two lung collapses, one which needed surgery. My average hospital stay in the last ten years is once every two or three years. Sinus disease plays a big part. And I have had major back issues because of the coughing. This past year has been super hard with two hospital stays and another close call. I praise God for my CF team. Dr. Saavedra is awesome, as well as others at National Jewish. Having a great CF team really matters. Right now, I'm trying to get my A1C down to 7. My biggest weakness is not exercising regularly.

Are you on a CFTR modulator?

I began Orkambi in 2015. This year I started to feel it just wasn't working. So we switched to Symdeko about two months ago. I'm doing much better.

Did going on disability help your health or have other benefits?

I was disabled long before I went on disability. Because we are missionaries, my work credits were unique. We were spending nearly $30,000 per year mostly on my care. It was crazy hard. I am so thankful for disability because it has helped save us tons of money. I can't thank Beth Sufian and her team at CF Legal enough. They helped tremendously in my getting approved for disability.

Continued on page 32

CF: THE MIND GAME

What Is An Emotional Exacerbation?

By Mark Tremblay

As CF patients, we are all familiar with pulmonary exacerbations. I know when I'm having a pulmonary exacerbation because, initially, I feel run down, and then I usually get a runny nose, as well as a scratchy or sore throat. If those symptoms persist, and my taste and voice change, followed by painful coughing, then I know the exacerbation has arrived and is about to stomp all over the village of my life like a giant in a fairy tale. At that point, I have to abandon the village and prepare for battle using the weapons with which we are all too familiar: hospitalization; IV, oral, and inhaled antibiotics; extra hours of airway clearance; more 7% saline sessions; protein supplementation; and, for me, Zofran to treat the nausea caused by the various antibiotics. If all goes well, I wound the giant sufficiently so that he retreats for a time and I can begin rebuilding my life. If need be, I adjust to a new normal — depending on how much pulmonary damage I've sustained.

Even though we're intimately familiar with the symptoms, treatment, and effects of our physical battles with CF, we tend not to be as cognizant of the mental and emotional toll those battles exact, nor what we can do if we're struggling with the psychological fallout. For example, on a recent crisp November morning, I was walking the trail on Hawk's Ridge in upstate New York where I live, admiring the pristine beauty of the first fresh snow that had arrived too early to blanket our vibrant autumn leaves, which only yesterday had set our mountain ablaze with a cornucopia of color. In the middle of my warm sentimental reverie, the horrifying face of the giant appeared and, along with it, memories of past battles and fallen soldiers. For I know all too well that the snow is a harbinger of winter and the giant's imminent return. Winter is when he has always waged the fiercest battles, and my memory is filled with the faces of friends he has claimed in winters past.

Regardless of whether you've only had a few encounters with the giant or you're a hardened soldier who's had so many encounters you've lost count, those battles have shaped you in many ways, both consciously and subconsciously. They might have made you a great fighter but, as many a decorated war hero knows, the strongest, best fighters struggle the most when they come home after the din of battle has died.

I'm reminded of something I used to say to a close friend who struggled mightily with depression, which often led her to hole up in her room for days, eating nothing but Ben and Jerry's and drinking heavily. Whenever she morosely talked on and on about why she was depressed, I'd say, "stop talking because you're depressing me." Interestingly, several years later, I was diagnosed with depression myself, which I now treat with antidepressants and counseling. In retrospect, I realize those talks likely depressed me so much because I was already depressed myself.

There is ample evidence to suggest that those battles traumatized you more than you know, particularly if they began at a young age. As a CF patient, you have spent your life under near-constant threat of physical harm and even death. Think of when were first diagnosed, had your first exacerbation, experienced restricted breathing, coughed up blood, had to go to the hospital for days or weeks, got a peripherally inserted central catheter (PICC) line or Port-a-cath placed, or had other unwelcome, painful, and invasive procedures.

> *"We need to be equally aware of the emotional symptoms of CF when the giant is in our heads and we're losing the fight."*

MARK TREMBLAY

Fortunately, many in our community have begun to see the tide of battle turn with the arrival of new powerful weapons, including the new modulator drugs such as Trikafta. However, some won't be able to deploy these weapons because of varying genetic mutations or unpleasant side effects. Also, despite adding the new modulator to their routine, many will find the mental and

emotional weight of the disease will not lift when/if their symptoms ease.

It is well known that when soldiers or people in general, including children in war-torn areas, are traumatized, they carry the trauma with them well after the immediate threat is removed. This trauma manifests in the form of heightened anxiety, depression, and sometimes substance use disorders, which are all considered symptoms of Post-Traumatic Stress Disorder (PTSD). Research shows that children in neighborhoods where traumatic experiences are more prevalent and varied in nature, (e.g., violence, food insecurity, environmental toxicity, housing insufficiency, and poverty) are at a heightened risk for emotional, cognitive, and developmental delays, as well as depression, anxiety, substance use disorders, and suicide. Similarly, CF children would be particularly susceptible to these effects as they are often exposed to trauma at a young age and over many years. Studies indicate the younger the child when first exposed to trauma and the longer the exposure lasts, the more severe and treatment-resistant the effects. However, the sooner a person gets help after exposure to trauma, including counseling and/or medication, the more quickly the person will feel emotional relief and stability, return to pre-crisis levels of emotional and mental functioning, and eventually achieve full recovery.

Just as we've all become well aware of the physical symptoms of CF when the giant is lurking in the woods or has crashed the gate, we need to be equally aware of the emotional symptoms of CF when the giant is in our heads and we're losing the fight. Some of the signs that you might be experiencing battle fatigue or an *emotional exacerbation* as I call it are:

- having intrusive thoughts of past battles or having undue fear of future battles during times of relative health;
- being unable to enjoy activities that you used to love or look forward to;
- finding it harder to fall asleep or stay asleep;
- having difficulty getting out of your head even when among your close family or friends;
- feeling a strong pull to withdraw or hide in socially isolating activities (e.g., video binging, game playing, social media obsessing, phone fidgeting, etc.); or
- turning to alcohol or substances for escape and emotional relief.

I, like many of you, considered myself battle hardened and mentally tough beginning in early childhood. Before I was even a teenager in karate tournaments and other sports, I proved I could withstand levels of pain that left my buddies and coaches aghast. Excessive drinking and drug use, to the point of dependency, were several of the many ways I dealt with the emotional weight of it all. I got sober after receiving inpatient treatment at the age of 18 and wrongfully assumed I had licked the mental giant. It wasn't until several decades later, when I sought counseling to deal with my divorce, bankruptcy, and business failure, that I realized I had been depressed and experiencing PTSD symptoms most of my life. In hindsight, had I been more aware of the emotional toll of my battles with CF and sought help sooner, I may have been able to avoid many failures in my life and saved my loved ones and friends a lot of heartache and pain.

As we all strive to bring our best fight to the giant, live our fullest lives, and thrive despite this disease, we have to be keenly aware of where our heads are at all times. If any of what I've described sounds like you or something you might be going through, you might be experiencing an *emotional exacerbation.* If so, I strongly encourage you to talk to your CF team and ask them for a referral to a licensed counselor, social worker, or psychologist. Based on my thirty-plus years' experience helping and counseling people in self-help groups, treatment centers, and church ministry, most folks I've known, particularly those who've sought counseling for trauma, have benefitted greatly from the emotional catharsis, personal insight, and behavioral change after only a handful of sessions.

Some may view asking for help as giving up, tantamount to defeat, but I assure you that the best fighters know that in order for you to survive the battle, you need to keep your wits about you and your head in the game and you can't do that if your mind is bogged down and struggling with an *emotional exacerbation.* If you're experiencing an *emotional exacerbation,* a friend's ear or family member's love can provide comfort and support. However, the only way you're going to push that giant out of your head and keep him at bay emotionally is to work with a licensed professional so that you can get back out on the battle field with a clear head. CF doesn't take a day off, so we can't either. ▲

Mark is 50 and has CF. He lives in Albany, NY, with his wife, MaryGrace, and stepson, Sean. He holds a Master's in Psychology from Marywood University and a Master's in Public Administration from Maxwell School of Citizenship and Public Affairs from Syracuse University. He was the Director of Rehabilitation, Vocation, and Family programs at Syracuse Behavioral Health. He is one of the first researchers in the country to study psychological functioning in adults with CF, including a presentation at the 1999 American Thoracic Society International Conference1. His contact information is on page 2.

He has a YouTube channel "Breathing Grace": https://www.youtube.com/channel/UCqxYndMVzFs7eu6kbWAHDDA/featured?view_as=subscriber

WOOD continued from page 29

Do you have a humorous CF story?

I was taking a hike one day with my hubby in a wooded area soon after being discharged from the hospital. IV antibiotics always wipe out my intestines, but I needed to get out. I thought I had prepared well enough but the urge came...so I went up this embankment to get a little privacy and out it flowed. As I was trying to clean myself up with leaves, I heard this barking and growling coming down the hill. I looked up and there he was: a guard dog coming right toward me. I mean what was I to do? "A girl has got to do what she has to do," to quote from the movie *Ever After*. As I was trying to stand up on a hill, pull my pants up and seek to protect myself from this dog coming right for me, I started yelling at him and picked up some of my ~ and hurled it at him. NO kidding, it was my best weapon. The dog took one whiff and stopped in his tracks. He had probably never encountered such a beast before. I scurried down the hill, told my husband and we proceeded back to the car.

What are your favorite hobbies?

I love romantic comedies and like to watch my favorites over and over again. The other hobby, which annoys some in my family, is moving the furniture around. It's like I get a whole new set of furniture and room layout each time. I have done it since my youngest age.

What motto do you live by?

My life passage can be found in Psalms 40:1-3.

How has *CF Roundtable* affected you?

I enjoy hearing others' perspectives and the help they have found regarding their particular issue that I happen to be dealing with at the time. Reading the section "In Memory" always reminds me that one day I will be there. Maybe not because of CF, but for some other reason. We will all die, so don't put your life, your dreams, or your ambitions on hold simply **because** of CF. You just may achieve them **with** CF! ▲

Jeanie Hanley is 57 and is a physician who has CF. She is a Director and the Treasurer of USACFA. Andrea Eisenman is 55 and has CF. She is a Director of USACFA and is both the Webmaster and Executive Editor of CF Roundtable. Their contact information is on page 2. If you would like to be interviewed for "In The Spotlight," please contact either Andrea or Jeanie.

TILLMAN continued from page 27

proportion.
https://tinyurl.com/qlfu7wj

Aspergillosis, ABPA-Related Hospitalizations In Cystic Fibrosis Confer Worse Outcomes

Patients with aspergillosis-pulmonary cystic fibrosis (CF) have worse outcomes. Significantly longer hospitalizations compared with patients with CF were also noted in those with aspergillosis-CF and allergic bronchopulmonary aspergillosis (ABPA)-CF. The trend of aspergillosis- and ABPA-related hospitalizations in patients with CF has significantly increased from 2005 to 2014. The adjusted risk for aspergillosis-CF was significantly higher in patients with chronic kidney disease. Patients with aspergillosis-CF had significantly higher rates of acute respiratory failure, mechanical ventilation, and inpatient mortality than those patients with CF but without aspergillosis or ABPA. Significantly longer hospitalizations were also seen in patients with aspergillosis-CF compared with patients with CF but without aspergillosis or ABPA. The adjusted risk for ABPA-CF was significantly higher in patients with asthma and in patients experiencing weight loss, but significantly lower in women. Patients with ABPA-CF had significantly longer hospitalization; however, the rates of acute respiratory failure, mechanical ventilation, and inpatient mortality were not significantly different compared with those of patients without aspergillosis or ABPA.
https://tinyurl.com/y4388nmh

Safety, Early Effectiveness Of POL6014 Confirmed In Phase 1 Trial

Single dosing of orally inhaled POL6014, an experimental therapy to treat chronic lung inflammation, can effectively block the activity of a pro-inflammatory enzyme in the lungs of people with cystic fibrosis (CF), results from a Phase 1 clinical trial show. Neutrophils, a type of immune cells, form part of the primary line of defense of the immune system in case of infection. These cells release an enzyme, called neutrophil elastase (NE), that normally digests damaged cells, and prevents the infection from spreading. However, neutrophils also are found in the inflamed lung tissue and sputum of CF patients. In CF lung, excessive release of NE causes additional tissue damage, leading to a progressive decline in lung function. POL6014 is a highly potent and selective inhibitor of human neutrophil elastase (hNE) and was shown to reach high concentrations in the lung when administered by inhalation via an optimized eFlow® nebulizer. At the highest dose the treatment triggered inhalation-related side effects including short transient decline in FEV1, cough, and respiratory tract irritation. At all lower doses, POL6014 was well-tolerated with no

MILESTONES

Please share the milestones in your life with our readers. Your successes and achievements may serve as a source of motivation for others in need of an infusion of "positive mental attitude" in the pursuit of their goals. Send us a note specifying your "milestone." Include your name, age, address and phone number. Mail to: ***CF Roundtable*, PO Box 1618, Gresham, OR 97030-0519. Or e-mail to: cfroundtable@usacfa.org**

ANNIVERSARIES

Birthday

Andrea Eisenman
New York, NY
55 on November 28, 2019

Lawrence Lafary
Riverton, IL
80 on December 5, 2019

Sydna Marshall
Austin, TX
39 on September 19, 2019

Transplant

Mike Schnitzer, 62
Vernon Hills, IL
Second bilateral lung transplant
1 year on November 29, 2019

clinically significant adverse findings as measured by clinical laboratory tests, electrocardiogram, and blood pressure measurements. Additional analysis showed that human NE activity was reduced in participant's sputum shortly after inhalation at all dose levels. Researchers also found that three and 24 hours after inhalation the concentration of POL6014 was 1,000 times higher in the lungs compared to blood levels. This showed that systematic exposure to POL6014 throughout the body was small and was mainly restrict-ed to the lungs.
https://tinyurl.com/yhtnebda
AND
https://tinyurl.com/vlu92dc
AND
https://tinyurl.com/u6zafhm

Copeptin A Potential Biomarker Of CF Symptom Severity During Pulmonary Exacerbation, Study Shows

High levels of the peptide copeptin are associated with increased severity of cystic fibrosis. These results suggest that copeptin can be used as a biomarker to identify patients who could benefit from more aggressive treatment. Copeptin, a type of peptide — a short chain of amino acids that is a fundamental component of cells — is a known marker of prognosis and disease severity across many inflammatory, respiratory, and metabolic diseases. Researchers measured copeptin levels in the serum — a component of blood — of 28 pediatric CF patients, 13 of whom were in stable condition; the remaining 15 were undergoing pulmonary exacerbation. Copeptin also was measured in the sputum of 10 CF patients. They also looked at the patients' complete blood count (CBC), body mass index (BMI), bacterial population in the sputum, lung function (using spirometry), and chest imaging (using the Brasfield score). Symptom severity was assessed using the Shwachman-Kulczycki score, and the children's quality of life was assessed through the Cystic Fibrosis Quality of Life Questionnaire-Revised (CFQ-R). The results showed that copeptin levels in both the serum and sputum were higher in CF patients during a pulmonary exacerbation, compared with a stable period. However, these results were not statistically significant. Furthermore, serum copeptin levels did not correlate significantly with any laboratory markers of inflammation, pulmonary function tests results, or BMI. Interestingly, however, the researchers found an inverse correlation be-tween serum copeptin levels and symptom severity during pulmonary exacerbation. An inverse, or negative correlation also was found with the Brasfield score, which measures the severity of structural lung changes, during ex-acerbation. Together, these results suggest that serum copeptin may correlate with the disease progression and patients' deterioration. The researchers also found that copeptin levels were inversely correlated with the quality of life in people with CF, including in the domains of vitality and eating habits — specifically, loss of appetite. The correlation of copeptin to quality of life supports the peptide's value as a measure of disease severity.
https://tinyurl.com/w56yz3l

Continued on page 34

TILLMAN continued from page 33

New CF Treatment Candidate AP-PA02 Targets *P. Aeruginosa* Infections

Armata Pharmaceuticals is going to target multidrug-resistant *Pseudomonas aeruginosa* infections in cystic fibrosis (CF) patients with its new enhanced bacteriophage candidate AP-PA02.

AP-PA02 is being developed to replace the company's candidate AP-PA01, which was shown to successfully treat a CF patient with resistant *P. aeruginosa* infection. Growing evidence suggests that using bacteriophages (al-so known as phages) — viruses that specifically infect and kill bacteria — may be a viable option to fight bacterial infections that are difficult to treat, particularly those resistant to multiple antibiotics. Armata used its proprietary phage library to screen hundreds of P. aeruginosa bacteria samples isolated from CF patients in the U.S. and Europe. With this approach, the company was able to identify AP-PA02, which has shown to effectively target ap-proximately 90% of the bacteria samples. This positive result prompted Armata to move forward with AP-PA02 development. The company expects to submit an Investigational New Drug application for AP-PA02 with the U.S. Food and Drug Administration in the fourth quarter of 2019. If the application is approved, the company will be cleared to start assessing the clinical potential of its new phage candidate in CF patients.
https://tinyurl.com/rwto6s4

Translate Bio Announces Pipeline Program Update

Translate Bio, a clinical-stage messenger RNA (mRNA) therapeutics company developing a new class of potentially transformative medicines to treat diseases caused by protein or gene dysfunction, announced that it is prioritizing the development of pulmonary disease programs including the ongoing development of MRT5005, its clinical candidate for the treatment of CF. MRT5005 is a first-in-class mRNA therapeutic designed to address the underlying cause of CF regardless of genetic mutation by delivering mRNA encoding fully functional cystic fibrosis transmembrane conductance regulator (CFTR) protein to cells in the lung through nebulization.
https://tinyurl.com/veh3q6l

AzurRx BioPharma Announces Positive CFF DSMB Review Of Final Phase 2 OPTION Trial Data

AzurRx BioPharma, Inc. announced that the Cystic Fibrosis Foundation (CFF) Data Safety Monitoring Board (DSMB) has completed its review of the Company's final results of the Phase 2 OPTION trial and has found no safety concerns for MS1819-SD for the treatment of exocrine pancreatic insufficiency in cystic fibrosis (CF). Additionally, the group supports the Company's plan to proceed to a higher 4-gram dose of MS1819-SD in its next planned Phase 2 clinical trial. MS1819-SD, supplied as an oral non-systemic biologic capsule, is a recombinant enzyme that is derived from the yarrowia lipolytica lipase, and unlike the standard of care, does not contain any animal products.
https://tinyurl.com/t4lwcos

AND

https://tinyurl.com/yys9vo8x

Corbus Pharmaceuticals Completes Enrollment Of Phase 2b Study Of Lenabasum For Treatment Of Cystic Fibrosis

Corbus Pharmaceuticals Holdings, Inc. announced the completion of patient enrollment in the Phase 2b study evaluating the efficacy and safety of lenabasum for the treatment of cystic fibrosis (CF). Lenabasum has Orphan Drug Designation and Fast Track status for treatment of CF. Lenabasum represents a potential new anti-inflammatory treatment option for people with CF and recurring pulmonary exacerbations. Its potential benefit is without regard to CFTR mutation or the current treatment the patient is receiving. Lenabasum is a rationally de-signed, oral, small molecule that selectively binds as an agonist to the cannabinoid receptor type 2 (CB2) and has been designed to resolve inflammation, limit fibrosis and support tissue repair. CB2 is preferentially expressed on activated immune cells and on fibroblasts, muscle cells, and endothelial cells. Lenabasum has induced the production of pro-resolving lipid mediators that activate endogenous pathways which resolve inflammation and speed bacterial clearance without immunosuppression. It can reduce expression of genes and proteins involved in inflammation and fibrosis. Lenabasum has demonstrated acceptable safety and tolerability profiles in clinical studies to date. Lenabasum treatment also was associated with a lower rate of and longer time to pulmonary exacerbations in a Phase 2 cystic fibrosis study. Additional clinical studies are being conducted to confirm these results and sup-port applications for regulatory approval. Patients in the study are randomized 1:2:2 to either receive lenabasum 5 mg twice per day, lenabasum 20 mg twice per day or placebo twice per day for 28 weeks, with 4 weeks follow-up off active treatment. The primary efficacy endpoint of the Phase 2b CF study is the event rate of pulmonary exacerbation. Secondary efficacy outcomes include other measures of pulmonary exacerbations, change in forced expiratory volume in 1 second (FEV1), % predicted, and change in Cystic Fibrosis Questionnaire-Revised respiratory domain score.
https://tinyurl.com/s6wlksf

Proteostasis Therapeutics Completes

Continued on page 36

Benefactors

BRONZE

Anonymous (in memory of Thaddeus Novak)
Anonymous (in memory of James B. Trecek)
Francis Birkner
Andrea Paes Borns
Darleen Boynton
Jeanne-Marie Bruno
Corr Family Trust
Kevin Corr
Mary E. Cummings
Shawna Ritchie Deck
Julie Desch
Sue Dietz
Tina Dimick (in honor of Eric Dimick)
John Dolan
Suzy Domenick
Nina Ferrell
Doreen Gagnon (in memory of Joe Kowalski)
Ann Grennan
Jeanie Hanley, M.D.
Richard Harris (in memory of Kathleen Harris)
Barbara Harison
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Elizabeth Hissing
Douglas Holsclaw, Jr., M.D.
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Douglas Johnson
Glen and Rhonda Keysor
Bonnie Larner-Langer
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Jessica Martens
Hedy McLaughlin
Bill Michelle
Xan Nowakowski
Anne O'Connor
Virginia & Thomas Otter
Judy Packer
Terry Reilly (in memory of Cris Dopher)
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Lisa Bassett Stackhouse
Stephanie Hamm Sutton
Amy Sylvis
Sharon Thompson (in memory of Melissa Thompson)
Laura Tillman
Carolyn Truesdell
David Versteeg (in memory of Sandy Turenhout-Roorda)
Devin Wakefield
Leigh Wynkoop (in memory of Peter McMurry)
Norman Young, Jr.
Jason Yu

SILVER

Edward Kinney
Karen Scott
James Yankaskas, M.D.

GOLD

Colleen and Scott Adamson
Bob & Jill Eknoian Lopez

PLATINUM

Genentech, Inc.
Hill-Rom Services, Inc.
International Biophysics, Corp.

SUSTAINING PARTNERS

Abbvie Inc.
Gilead
Mylan, Inc.
Two Hawks Foundation (in memory of Dr. Lisa Marino)

Encourage Family and Friends to Donate!

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

TILLMAN continued from page 34

Enrollment In Phase 2 CF Study

Proteostasis Therapeutics (PTI) has completed patient enrollment in a Phase 2 clinical trial evaluating its cystic fibrosis transmembrane conductance regulator (CFTR) modulator combination therapies in cystic fibrosis (CF) patients who have at least one copy of the F508del mutation in the CFTR gene. This Phase 2 clinical trial (NCT03500263) is a global, randomized, placebo-controlled study designed to assess the effectiveness, safety, and tolerability of PTI's combination therapy over a period of four weeks in CF patients homozygous or heterozygous for F508del. The goals of the study include safety, changes in sweat chloride concentration and changes in percent predicted FEV1. The combination treatment includes 600 mg of PTI-801 (third-generation CFTR corrector), 300 mg of PTI-808 (a CFTR potentiator), with or without 10 mg of PTI-428 (a CFTR amplifier). CFTR amplifiers are a new type of medicine that help increase CFTR protein levels in cells and tissues. CFTR potentiators are drugs that can help overcome gating defects that are caused by certain mutations of the CFTR gene. CFTR correctors help the CFTR protein form the right 3D shape so it can properly move to the cell surface. Of note, the triple combo (PTI-801, PTI-808, and PTI-428) received fast track designation by the U.S. Food and Drug Administration (FDA) in 2018.
https://tinyurl.com/t8urbkp

UI Researchers Develop New Delivery Tool For Gene Therapies For CF, Other Respiratory Diseases

A new, more efficient method for delivering gene therapies to lung tissues — based on tiny engineered proteins, called peptides —is creating therapeutic opportunities for people with diseases like cystic fibrosis. The tissue lining the airways, called respiratory epithelium, has natural barriers that can prevent viral vectors — the common vehicle used to transport gene therapies — from effectively delivering the wanted therapeutic gene to the targeted airway specialized cells. To overcome this limitation, researchers developed a new delivery tool that uses simple peptides to reach notoriously hard-to-access lung and airway cells. The team engineered these cell-penetrating peptides (CPPs) so they could retain their natural internalization proprieties, while also being able to escape a cell's mechanisms for removing foreign products. Using experimental cell models and mice, they showed that their engineered CPPs could safely and effectively transport CRISPR elements to different specialized airway epithelial cells. The team also confirmed that by using these "shuttle" peptides it was possible to achieve gene editing at levels high enough to be useful clinically without signs of short-term toxicity. The team is now screening approximately 100 new peptides to identify those that could represent the most efficient delivery systems for biological cargoes.
https://tinyurl.com/s63jd6b

Researchers Develop New Device To Eliminate Mucus Secretion In CF

Researchers have developed a new technology that can be used to unclog and eliminate mucus secretions from the airways of patients with various respiratory diseases, including cystic fibrosis (CF). There is currently no effective therapy to either directly or indirectly treat the small airways of the lungs. The idea behind the new technology was that concurrent application of low-frequency flow oscillations and high-frequency acoustic waves could help mucus detach from the airway wall. The mucus could then be removed, either by breaking it down or by agglomerating, or gathering together mucus lumps. The technology works by introducing air pressure and acoustic pulses into the airway and lungs over a low-pressure airstream. It is targeted to treat small obstructed airways by reducing the buildup of mucus. The use of this technology was found to be effective in a series of laboratory experiments in airway and lungs.
https://tinyurl.com/vkgca4p

Patients' Avatars Being Used To Test Cystic Fibrosis Drugs

Researchers are leading a revolutionary approach to managing treatment of cystic fibrosis using patients' derived lung and gut mini-organs or 'avatars' to test how they will respond to the latest drugs. They tested patients' stem cell derived mini-organs against various CFTR modulators in a centralized laboratory. Using recent breakthroughs in stem-cell biology, the researchers isolated cells directly from respiratory or gut tissue and encouraged large scale expansion of them to create mini-organs (organoids). Since these organoids were created from the cells of patients with CF, they are effectively an avatar for that person. In other words, if the drug works on their avatar, then it will likely work on the patient. These organoids are cryopreserved in the biobank and can be tested against new drugs in the future. The research lab uses the organoids to identify drug responsive from non-responsive individuals. Of the patients' avatars identified as responsive to the CFTR modulators, three have been ultra-rare CFTR mutations. The research team is also testing ways to correct the defective CFTR by adding a correct copy of the gene to the cells. These cells serve as an invaluable tool to enhance the current understanding of CF and the translational research efforts that aim to develop new therapeutic agents to fight the disease and shape the future for CF precision medicine.
https://tinyurl.com/w6mock2

Hexoskin Shirt Can Accurately Assess Lung Function, May Be Tool In CF,

Study Shows

A new study has shown that the Hexoskin smart shirt can accurately assess lung function in healthy people. Sup-ported by these positive results, the team plans to further explore the potential of this smart shirt to monitor lung function in people with lung diseases. Hexoskin represents a user-friendly alternative that allows the remote monitoring of lung function. The smart shirt has incorporated sensors that assess lung volume as the fabric stretches with each inspiration and expiration chest movement — basically with each breath. It also has incorporated sensors that can record heart rate and movement.

All data collected by the Hexoskin can be transferred to its own analytics software, which can be used with a mobile phone or computer for health monitoring. Using the smart shirts could help people with respiratory disorders to predict disease worsening so they can step up their treatment earlier. Smart shirt technology offers a promising, though relatively expensive, tool for monitoring patients' respiratory health status during normal activities in a way that does not interfere too much with their daily lives.
https://tinyurl.com/rgrmpna

BP Device May Be Useful For Managing Blood Sugar Levels In CFRD, Study Says

The bionic pancreas (BP), an automated system that controls blood sugar levels, may be useful in managing cystic fibrosis-related diabetes. Cystic fibrosis-related diabetes, or CFRD, is the most common non-pulmonary manifestation of cystic fibrosis (CF). Statistics indicate that up to 50% of adults and 20% of adolescents diagnosed with CF will develop CFRD at some point in the course of the disease. The current standard of care for CFRD consists of administering insulin, a

Continued on page 38

BOOMER ESIASON FOUNDATION'S PROGRAMS

CYSTIC FIBROSIS SCHOLARSHIPS

The Boomer Esiason Foundation Scholarship Program provides assistance to students with cystic fibrosis based on a number of criteria including: academics, athletics, arts, and more...
www.befscholarhships.com

LUNG TRANSPLANT GRANT PROGRAM

The Boomer Esiason Foundation Lung Transplant Grant Program provides grants to cover expenses including, but not limited to: temporary housing, food, and transportation costs.
www.befgrants.com

CYSTIC FIBROSIS PATIENT DISASTER RELIEF PROGRAM

In light of recent natural disasters, The Boomer Esiason Foundation created a fund that directly assists those affected families in the cystic fibrosis community.
www.esiason.org

TEAM BOOMER FIGHTING CYSTIC FIBROSIS

Team Boomer is a program that encourages people with cystic fibrosis to incorporate exercise into their everyday lives; provides an avenue for individual athletes in a variety of sports to raise money for cystic fibrosis; and offers assistance to grassroots athletic events looking for a cause to support.
www.teamboomer.org

BREATHE IN
GUNNAR ESIASON & SALTY CYSTERS
A CYSTIC FIBROSIS PODCAST

A series of audio and video podcasts, featuring Gunnar Esiason and the Salty Cysters, that highlight people with CF and the challenges they face.
www.gunnaresiason.com

JERRY CAHILL'S CYSTIC FIBROSIS PODCAST

A series of audio and video podcasts in which Jerry Cahill interviews people with CF who are living, breathing, and succeeding through the power of exercise, nutrition, and compliancy.
www.jerrycahill.com

TILLMAN continued from page 37

hormone produced by the pancreas that helps control blood sugar (glucose) levels. The BP employs continuous glucose monitoring (CGM) coupled with mathematical algorithms to determine when and how much insulin needs to be administered through its automatic pump to normalize blood sugar levels, when used in its insulin-only configuration (IOBP). However, the BP device also can be used in a bihormonal configuration (BHBP). That allows the automatic pump to administer not only insulin, but also glucagon, a hormone that also helps control blood sugar levels and counterbalances the actions of insulin. Researchers compared the effectiveness of both configurations of the BP device to standard of care at normalizing blood sugar levels in adults with CFRD. Results showed that both BP configurations enabled participants to control their blood sugar levels. All interventions kept the percentage of time participants spent with hypoglycemia to a minimum. The participants all reported treatment satisfaction in daily surveys, and said the BP device helped them manage their diabetes and have greater peace of mind.
https://tinyurl.com/t4gra3t
AND
https://tinyurl.com/wfsfg2a

Early Data Supports Effectiveness Of ARO-ENaC, Potential CF Therapy

Preclinical data shows that Arrowhead Pharmaceuticals' investigational inhaled treatment for cystic fibrosis (CF), called ARO-ENaC, accelerates mucus clearance and preserves lung function while being safe for the kidneys in animal models. ARO-ENaC is an investigational RNA therapy designed to reduce the production of the epithelial sodium channel alpha subunit (αENaC) in the airways of the lung. ENaC is a sodium transport channel that is hyperactivated in the lungs of people with CF. This increased activity contributes to dehydration and mucus buildup in the lungs. ARO-ENaC is a small molecule that works by preventing the production of ENaC channels, and by triggering a pathway called RNA interference (RNAi). It targets for destruction, or silences, αENaC mRNA molecules, which are genetic messengers that carry the information necessary for making αENaC proteins. The preclinical data shows that inhaled ARO-ENaC doubled mucociliary clearance (MCC) in healthy sheep, paralleling the magnitude of effect of Kalydeco (ivacaftor, an approved CF therapy) in humans. Moreover, the agent proved effective for preserving lung clearance in a sheep model of mucus obstruction. Data also showed that the TRiM platform increases the potency of RNAi-triggered silencing of mRNA, and durably reduces the production of ENaC in the lungs of rodent models. Importantly, data on animal models indicated that ARO-ENaC can target ENaC in the lung while avoiding negative effects on the kidney.
https://tinyurl.com/yx5u4jwr

CF Antibiotic Response May Be Predicted By IGF2R Levels, Study Says

Measuring the levels of insulin-like growth factor 2 receptor (IGF2R) during the first days of treatment with intra-venous antibiotics could help predict treatment response in cystic fibrosis patients experiencing pulmonary exacerbations. Identifying novel biomarkers that help assess if a patient is responding to a combination of antibiotics in the first days of treatment could help improve the patient's overall response and outcome. The investigators evaluated the changes in 346 proteins, and found that the levels of 47 of them changed considerably during the first five days of treatment. These included proteins involved in immune response and inflammation, such as interleukin-6, C-reactive protein, and calprotectin, which had previously been linked to CF activity. IGF2R was the only protein whose levels changed considerably during early treatment, and correlated with response to therapy and patient outcomes. Therefore, this protein could serve as an early marker of response to therapy during treatment with intravenous antibiotics. IGF2R regulates several processes such as wound healing, blood vessel growth, and response to viral infections. The team also noted that future studies, including more patients, should validate the findings, and assess other potential biomarkers.
https://tinyurl.com/v8kkftr
AND
https://preview.tinyurl.com/sjnjtjd

***P. Aeruginosa's* Stress Response Helps It Evade Antibiotics, Study Says**

Researchers have discovered a stress response mechanism that allows *Pseudomonas aeruginosa* to evade the action of antibiotics and to survive. Like other species of bacteria, *P. aeruginosa* spreads and grows by swarming, a process that describes the rapid, collective, and coordinated movement of a group of bacteria. While swarming, bacteria produce several substances that control how different bacteria sub-groups within the aggregate move individually.

In the study, researchers discovered that if attacked while swarming — by an antibiotic or a virus that infects bacteria (bacteriophage) — *P. aeruginosa* bacteria start producing a molecule called Pseudomonas quinolone signaling (PQS) that tells other bacteria in the group to steer away from danger. The researchers believe that this stress response mechanism is part of the reason why P. *aeruginosa* is so hard to eliminate with antibiotics and has become resistant to many of them over time. In the laboratory the bacteria simply swim around the 'dangerous area' with antibiotics or bacte-

riophages. When they receive the warning signal from their conspecifics they move in a neat circle around. In their experiments, researchers studied the movement patterns of *P. aeruginosa* aggregates in petri dishes that mimicked the environment these bacteria would come across in the lungs of people with CF. After studying these movement patterns, they found that healthy bacteria consistently steered away from bacteriophage-infected bacteria, as well as from those that had been killed by antibiotics. In addition, the team discovered that in both cases, bacteria that had been attacked produced large amounts of PQS that repelled the movement of healthy swarms of bacteria into the affected area. This type of stress response is a key survival mechanism of *P. aeruginosa* that may be explored further to help scientists find more effective treatments to eliminate these bacteria. The next step is to investigate how to target this communication between bacteria in large aggregates.

https://tinyurl.com/syb9ld3

AND

https://tinyurl.com/vx83mrr

Collaboration Between Crystecpharma And Iconovo Aims To Develop Better Inhaler Solutions For Patients

CrystecPharma signed a letter of understanding with Iconovo to develop innovative dry powder inhaler solutions for the treatment of patients with lung diseases. Direct delivery of medications into the lungs permits their rapid uptake via a non-invasive route. The two key factors that determine successful lung delivery of inhaled medications are the inhaler device itself and the formulation. Formulations require precise control of particle size, ease of aerosolization, and stability. CrystecPharma specializes in a technique called modified supercritical anti-solvent (mSAS) technology to engineer particles with improved characteristics. In mSAS, the medication is dissolved in a suitable solvent, such as ethanol or acetone, and added to carbon dioxide subjected to critical temperature and pressure conditions (known as supercritical fluid, or SCF, where distinct liquid and gas phases do not exist). The solvent is quickly extracted from the medication by the supercritical fluid (hence the name "anti-solvent"), leaving dry microscopic particles with well-defined crystalline morphology. In this way, mSAS allows precise control of particle size and characteristics.

https://tinyurl.com/wzuskqs

Can CF-tailored Talk Therapy Help With Depression And Anxiety? Drexel Study Seeks To Find Out

How well Acceptance and Commitment Therapy (ACT), or tailored "talk" therapy that can be delivered via tele-medicine, helps people with cystic fibrosis (CF) cope with mental health issues will be the focus of an upcoming study. Mental well-being is an area of concern with CF patients, who are known to be two to three times more likely to have bouts of anxiety or depression as people without CF. ACT, a type of talk therapy, may be helpful for people with chronic diseases like CF. And if used as a form of telemedicine, patients do not need to meet in a group, lowering their risk of exposure to infectious agents. ACT is a novel, interactive, and experiential treatment, different from other talk therapies.

A small, three-year pilot study found that ACT helped in easing anxiety and depression in patients, and seemed to also have a positive impact on lung function. The new trial is planned to take place at centers across the United States. Participants will complete six 50-minute talk therapy sessions delivered by a webcam, and will be randomly assigned to receive either ACT modified for people with CF or supportive psychotherapy. Researchers will analyze patients' levels of anxiety and depression, as well as the frequency with which they take their medicines, among other health measures.

https://tinyurl.com/vvyaptn

After Big Gains, Cystic Fibrosis Foundation Bankrolls Research Toward Cures — And Drugs For Those Left Out

The Cystic Fibrosis Foundation unveiled a $500 million initiative aimed at developing treatments for patients who aren't helped by the Vertex drugs and, ultimately, at finding cures for all CF patients. The new Path to a Cure plan will fund some basic research. But the majority of the money will go to support clinical programs. The $500 million will be doled out through 2025. The bulk of the patients who do not respond to the modulators have CF caused by nonsense mutations (also called stop or X mutations), which fail to generate a version of CFTR close enough to the healthy form for modulators to coax activity from. The modulators also do not work on some rare CF mutations; in others they have not been tested. The CF Foundation's plan outlines a number of clinical approaches that it intends to support. Some, including an approach called a readthrough therapy, would be specific to nonsense mutations. But others, including those involving RNA and DNA, would be "mutation agnostic," meaning they would work for all CF patients. While modulators have been transformative for some patients, they are not cures. Gene therapies (delivering a healthy CFTR gene with the help of a harmless virus) or gene editing with tools like CRISPR have potential as one-time therapies that can permanently overcome the underlying mutation, so patients would no longer need to take modulators or other drugs. Some of the foundation's funding will also go toward

Continued on page 40

TILLMAN continued from page 39

researching how to deliver therapies.
https://tinyurl.com/yx65fw5m

CF Foundation Awards Eloxx $1.61M To Support Trial In People With Nonsense Mutations

The Cystic Fibrosis Foundation (CFF) is giving Eloxx Pharmaceuticals up to $1.61 million to support its planned Phase 2 clinical trial program assessing the safety, tolerability, and chemical properties of ELX-02, Eloxx's lead investigational compound to treat cystic fibrosis (CF) caused by nonsense, or stop, mutations. The program is set to include two open-label, dose escalation, Phase 2 trials. Both studies will focus on the effects of multiple doses of ELX-02 in patients with at least one G542X allele. The G542X allele is an abnormal variant of the CFTR gene that is included in Class I, which comprises a group of nonsense mutations that insert a stop signal in the coding sequence of CFTR. Because of this premature stop signal, the production of the CFTR protein halts, leading to the production of a shorter, non-functional protein. ELX-012 is an experimental treatment that targets ribosomes — the small structures that are responsible for the production of proteins in cells — to tell them to bypass the stop signal in the mutated CFTR coding sequence. In this way, ELX-012 increases the amount of full-length CFTR protein that is being produced, potentially minimizing the effects of CF. If Eloxx's clinical trial program succeeds, ELX-02 may become the first treatment option for CF patients carrying at least one nonsense mutation.
https://tinyurl.com/tf3jj3u
AND
https://tinyurl.com/ta2c2o7
AND
https://tinyurl.com/rofduds

For Potential CF Therapy ARV-1801, Arrevus Secures Loan

Arrevus received a Small Business Research Loan to support the development of the company's Phase 3 therapeutic candidate ARV-1801 for the treatment of pulmonary exacerbations in patients with cystic fibrosis (CF). In February, Arrevus finalized the purchase of the ARV-1801 (sodium fusidate) program from Melinta Therapeutics. ARV-1801 has unique antibacterial, anti-inflammatory, and mucolytic (mucus-thinning) activities. As such, Arrevus considers this therapy an ideal candidate for the treatment of pulmonary exacerbations in CF patients. Additionally, ARV-1081 is considered safe, and has been used for more than half a century for treating infections in CF patients in markets outside the United States. Sodium fusidate has been incorporated into multiple treatment guidelines for the decolonization of Staphylococcus in patients with cystic fibrosis. Arrevus wants to demonstrate that this agent can also improve the lives of patients with cystic fibrosis who are experiencing pulmonary exacerbations.
https://tinyurl.com/s8mt5wq

CF: NIH Grants $2.7M To Research Anaerobic Bacteria's Role In Flare-ups

The National Institutes of Health (NIH) has granted $2.7 million to Michigan State University (MSU) scientist Robert Quinn to investigate the role of anaerobic bacteria in the development of flare-ups among patients with cystic fibrosis (CF). Anaerobic bacteria are a type of bacteria that live without oxygen, and are frequently present in the lungs of patients with CF. However, little is known about their role as a pathogen or contributor to the development of the disease. Despite the fact that anaerobic bacteria are found in all CF patients, their presence is of-ten dismissed by physicians and scientists as a non-factor.

During flare-ups there is a turnover in bacterial populations. Interestingly, while some bacterial species die, anaerobic bacteria start to thrive and dominate during flare-ups. Due to the high burden of bacteria and chronic lung infections in patients with CF, physicians treat patients with increasingly aggressive antibiotics, which is the traditional approach to treat infections in CF. However, many doctors treating patients aren't aware of these anaerobes. It's believed that physicians have not been treating the principal cause of the flare-ups — the anaerobes. The researchers will use a new CF model that was recently developed at MSU, which mimics a CF-infected lung and allows them to conduct studies in a more realistic model. The team will also use innovative bioinformatics data analysis platforms, sequencing of the microbiome (the population of bacteria that reside in the lung), and metabolomics (the study of metabolism in organisms) to gain more insight into the bacterial dynamics within a CF lung. The scientific rationale is that a better understanding of what causes microbial changes during these flare-ups will lead to more efficacious and targeted therapy against pathogens. It's also felt that it is important to treat not only the underlying cause of CF — the mutations that cause the disease — but also the lung infections in order to achieve long-lasting results.
https://tinyurl.com/ujvz2pp
AND
https://tinyurl.com/wrue5mk

Laurent Pharmaceuticals Receives A CAD 2.7M Financing From Biomed Propulsion Fund

Laurent Pharmaceuticals Inc. announced that it has received the final approval for a CAD 2.7M loan from the Quebec Government. This financing will help support the Company's Phase 2 clinical study evaluating LAU-7b in adult patients with cystic fibrosis ("CF"). LAU-7b is an oral drug acting on the resolution phase of the inflammation, with the potential to treat chronic pulmonary inflammation that leads to irreversible lung damage in patients with CF. LAU-7b was also shown to enhance functional CFTR expression in CF airway cells, an effect that was further improved in the presence of a CFTR modulator. The goal of the APPLAUD trial is to evaluate LAU-7b's effect on the preservation of lung function in adult patients with CF, by reducing persistent unresolved inflammation in the lung and stimulating the return to homeostasis. LAU-7b is administered on top of the standard of care, including all commercially available CFTR modulators.
https://tinyurl.com/wsarks5
AND
https://tinyurl.com/wnn6ooz

TREATMENTS

Omadacycline as a promising new agent for the treatment of infections with Mycobacterium abscessus. Hannelore I Bax, Corné P de Vogel, Johan W Mouton, Jurriaan E M de Steenwinkel. Journal of Antimicrobial Chemothera-py, Volume 74, Issue 10, October 2019, Pages 2930–2933

Despite intensive treatment regimens, the outcome of *Mycobacterium abscessus* infections is extremely poor and thus novel treatment regimens are needed. Although tigecycline seems to be one of the best options currently available, its long-term use is hampered by severe toxic side effects as well as the need for intravenous administration and the relatively high concentrations required for efficacy. The results of this in vitro study on omadacycline activity, together with its favourable (pharmacokinetic) properties, suggest that omadacycline is a potential new agent for the treatment of *M. abscessus* infections.
https://tinyurl.com/vevxslp

Ivacaftor Is Associated with Reduced

Continued on page 42

Calling All Writers

Have you written an article or story for *CF Roundtable?* If not, why haven't you written? Are you concerned that you may not be a great writer? Don't let that stop you. We have people who will work with you, on your article, to make it the best it can be.

Are you concerned because you can't think of a topic? How about if we give you a few ideas to start with? Here are some titles that go from head to toe and might pique your interest to write. Remember, these are only suggestions. You may come up with entirely different ideas and that is fine with us. All we ask is that you write about your experience with CF.

Here are a few possible topics for your use: headaches; understanding what you hear; pain(s) in the neck; arm twisting; the case at hand; a breath of fresh air; gut reaction(s); pain in the butt; oh, my aching back; getting hip to a subject; standing on one's own two legs; at the foot of the problem; toeing the line; my sole responsibility. As you can see, these are humorous suggestions that are meant to give you some ideas. You need not use any of these, but you may, if you wish. For other ideas, check out the Looking Ahead section on page 3. All submission dates for the coming year are posted there.

We ask that all submissions be in Microsoft Word or a similar program. Please also send a recent high-resolution headshot in JPEG format. You may send your submissions to: cfroundtable@usacfa.com

TILLMAN continued from page 41

Lung Infection by Key Cystic Fibrosis Pathogens. A Cohort Study Using National Registry Data. Freddy J. Frost , Dilip S. Nazareth , Susan C. Charman , Craig Winstanley , and Martin J. Walshaw . Annals of the American Thoracic Society. Vol. 16, No. 11 | Nov 01, 2019

Ivacaftor can greatly improve clinical outcomes in people with cystic fibrosis (CF) and has been shown to have in vitro antibacterial properties, yet the long-term microbiological outcomes of treatment are unknown. Ivacaftor use was associated with early and sustained reduction in *P. aeruginosa* rates via a combination of increased clearance in those with infection and reduced acquisition in those without infection. The improved prevalence of *P. aeruginosa* infection was independent of reduced sampling in the ivacaftor cohort. Ivacaftor was also associated with reduced prevalence of Staphylococcus aureus and Aspergillus spp. but not *Burkholderia cepacia* complex. https://tinyurl.com/u8ddflz

Efficacy and safety of the elexacaftor plus tezacaftor plus ivacaftor combination regimen in people with cystic fibrosis homozygous for the F508del mutation: A double-blind, randomized, phase 3 trial. Heijerman HGM, McKone EF, Downey DG, et al. The Lancet. November 26, 2019

In this phase 3, multicentre, randomized, double-blind, active-controlled trial of elexacaftor in combination with tezacaftor plus ivacaftor conducted at 44 sites in four countries of 113 individuals with cystic fibrosis homozygous for the F508del mutation, aged 12 years or older with stable disease, and with a percentage prognosticated forced expiratory volume in 1 of 40–90%, inclusive, experts ascertained the efficiency and safety of the elexacaftor plus tezacaftor plus ivacaftor combination regimen in individuals with cystic fibrosis homozygous for the F508del mutation. In comparison with tezacaftor plus ivacaftor alone, in the lives of persons with cystic fibrosis who are homozygous for the F508del mutation, a clinically strong advantage and a favorable safety profile by elexacaftor plus tezacaftor plus ivacaftor along with the potential to result in transformative developments was given. https://tinyurl.com/tcs7dys

PATHOGENS

Mycobacterium abscessus, an Emerging and Worrisome Pathogen among Cystic Fibrosis Patients. Degiacomi G, Sammartino JC, Chiarelli LR, Riabova O, Makarov V, Pasca MR. Int J Mol Sci. 2019 Nov 22;20(23)

Nontuberculous mycobacteria (NTM) have recently emerged as important pathogens among cystic fibrosis (CF) patients worldwide. *Mycobacterium abscessus* is becoming the most worrisome NTM in this cohort of patients and recent findings clarified why this pathogen is so prone to this disease. *M. abscessus* drug therapy takes up to 2 years and its failure causes an accelerated lung function decline. The *M. abscessus* colonization of lung alveoli begins with smooth strains producing glycopeptidolipids and biofilm,

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while in the invasive infection, "rough" mutants are responsible for the production of trehalose dimycolate, and consequently, cording formation. Using a *M. abscessus* infected CF zebrafish model, it was demonstrated that CFTR dysfunction seems to have a specific role in the immune control of *M. abscessus* infections only. This pathogen is also intrinsically resistant to many drugs, thanks to its physiology and to the acquisition of new mechanisms of drug resistance. Few new compounds or drug formulations active against *M. abscessus* are present in preclinical and clinical development, but recently alternative strategies have been investigated, such as phage therapy and the use of β-lactamase inhibitors.
https://tinyurl.com/sgmneon ▲

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

REMINDERS

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

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IMPORTANT RESOURCES

Partnership for Prescription Assistance: Phone: 1-866-218-0694 http://www.pparx.org/prescription_assistance_programs
The Partnership for Prescription Assistance brings together America's pharmaceutical companies, doctors, other health-care providers, patient advocacy organizations, and community groups to help qualifying patients without prescription drug coverage get free or low-cost medicines through the public or private program that's right for them.

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

Transplant Recipients International Organization, Inc. (TRIO): Phone: 1-800-TRIO-386 http://www.trioweb.org/index.shtml
An independent, nonprofit, international organization committed to improving the quality of life of transplant recipients and their families and the families of organ and tissue donors. For information, write to: TRIO, 7055 Heritage Hunt Dr, #307, Gainesville, VA 20155 or e-mail them at: info@trioweb.org

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

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