CF Roundtable®
A NEWSLETTER FOR ADULTS WHO HAVE CYSTIC FIBROSIS
AUTUMN 2018

"While I Was Fearing It, It Came"

By Roberto J. De La Noval

So begins one of Emily Dickinson's more famous poems, on the subject of fear. Most people with CF can sympathize, I think, with the thrust of these lines:

There is a fitting a dismay
A fitting a despair
'Tis harder knowing it is due,
Than knowing it is here.

To live with cystic fibrosis is, to a large degree, a waiting for tragedy. And often the shape this waiting takes is that of fear: fear of premature death, fear of lung failure, fear of transplantation, fear of losing functionality, fear of diabetes or arthritis, fear of incontinence, fear of intimate rejection, fear of causing suffering to and leaving behind those we love (perhaps the most severe and demanding of these fears, though I possess no survey data on the question). My intention in what follows is not to dispel these; I happen to think

ROBERTO J. DE LA NOVAL

that most human fears are eminently justifiable, and we rarely accord them the dignity they are due. My point is rather to reflect on the experience of fear itself within a life lived with CF—how it fills the days, coloring the imagination throughout the years in its darker tones. Only when the experience is known in its depth can the question of how to respond to fear be most profitably posed.

I can, of course, speak only autobiographically. Fortunately for me, my early childhood with CF was relatively painless: I experienced my first hospitalization only as a teenager, and before then I lived somewhat naive to the full implications of CF. Well, that's not exactly true. Around the age of eight I stumbled upon a book on CF for families with CF kids. The memory itself is spotty, but what is clear is a very tiny Rob lighting upon passages describing an achingly short life expectancy—32—and an obscenely high chance of infertility—98%. These numbers sank deep into my fallow child's mind, though the roots deposited there would not bear fruit for years to come. Like I said, in childhood I lived mostly oblivious to the darker realities of CF, and it was only my teenage years that propelled me into a more conscious state of simmering anxiety.

For me, fear is the color red. This

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INSIDE THIS ISSUE

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EDITOR'S NOTES

If you are receiving this newsletter later than you expected, I apologize. I was suffering from what I call "brain freeze." I would work on things for a little while and then I would just be unable to continue. I guess I was feeling the effects of age and season change. Again, I apologize.

Once again we have some changes to the Board of Directors. **Mark Levine** has left the board. We thank him for his service. We welcome a new director, **Jessika Biele**. You may read her bio on page 18.

I hope you have read the article on the cover. It is an interesting article by **Roberto De La Noval** that deals with fear and health.

In this issue we introduce a new column called "Savoring Serendipity." **David Tarnow** tells of a life-changing incident in his life and how he has used it to improve his living.

Our Focus topic in this issue is: Changes We Experience As We Age. **Jessika Biele** writes of turning 30 and how it affected her. **Jeanie Hanley** speaks of understanding her body better and learning how to use that knowledge. **Linda Stratton** writes of moving to a new environment and adjusting to it. I discuss changes both good and not so good in "Speeding Past 50."

As usual, **Laura Tillman** has compiled a wonderful column of "Information From The Internet" for us. You'll find much interesting news there. In "Spirit Medicine" **Isabel Stenzel Byrnes** writes of reaching one's full potential. **Julie Desch** writes of survivors' guilt in "Wellness."

Laura Mentch and **Jeanie Hanley** report on the 32nd Annual CFRI National CF Family Education Conference. Each of them gives us a peek into what the conference offered and how others can benefit from attendance at it.

The "Poetry Corner" has a poem by **Grace Knight**, who tells of her battles of life with CF.

On page 35, there is an invitation to participate in The Cystic Fibrosis Reproductive And Sexual Health Collaborative (CFReSHC). They are looking for some specific kinds of people. Please read about what they are looking for.

We are pleased to announce the recipients of the Lauren Melissa Kelly Scholarships for autumn. You may read about **Julia Ruggirello** and **Roberto De La Noval** on page 23.

Be sure to check out the information about Giving Tuesday on page xx. Your continued support keeps us going. Thank you.

I end on a personal note. By the time you read this, I will have celebrated my fifth anniversary of bilateral mastectomies due to cancer. Surviving five years, cancer free, is a wonderful accomplishment and I am grateful.

Be sure to check with your doctor about flu shots.

Until next time, stay healthy and happy,
Kathy

Publication of *CF Roundtable* is made possible by donations from our readers and grants from Sustaining Partners - AbbVie, Gilead Sciences, Kroger Specialty Pharmacy 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

Orkambi Taken For One Year Returned Glucose Levels To Normal In Group Of CF Patients In Study

One year of treatment with Orkambi (lumacaftor/ivacaftor) returned blood glucose (sugar) levels to normal in almost half of a small group of diabetic or glucose-intolerant cystic fibrosis (CF) patients with the F508del mutation in the CFTR gene. CF patients may show abnormal blood glucose levels due to poor secretion of insulin, the pancreas-secreted hormone that controls the levels of glucose in the blood to prevent it from getting too high (hyperglycemia) or too low (hypoglycemia). Treatment with Orkambi also resulted in a significant decrease in the need for intravenous antibiotic treatment.

http://tinyurl.com/yc5w573j

LAURA TILLMAN

GLPG2737 Add-on Increases Effectiveness Of Orkambi For Cystic Fibrosis, Phase 2 Results Show

Adding GLPG2737, Galapagos's investigational CFTR corrector, to Vertex's Orkambi (lumacaftor/ivacaftor) enhances the effectiveness of the treatment in cystic fibrosis patients with two copies of the F508del mutation. The Phase 2 PELICAN clinical trial (NCT03474042) evaluated the effectiveness, safety and tolerability of the combo therapy in 22 adults with two copies of the F508del mutation in the CFTR gene who were receiving stable treatment with approved therapy Orkambi. Participants were randomized to receive oral capsules of GLPG2737 or a placebo with Orkambi twice daily for 28 days. Analysis of the participants' lung function, evaluated by absolute change from baseline in percent predicted forced expiratory volume in one second ($ppFEV_1$), revealed a positive effect of the combo therapy. GLPG2737 also significantly reduced

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LOOKING AHEAD

Please consider contributing to *CF Roundtable* by sharing some of the experiences of your life in writing. Read the **Focus** topics listed below and see if there are any about which you might like to write. In addition, humorous stories, articles on basic life experiences, short stories, artwork, cartoons and poetry are welcome. We require that all submissions be original and unpublished. With your submission, please include a recent photo of yourself as well as your name, address and telephone number. Photos will be returned. Send all submissions to: **CF Roundtable, PO Box 1618, Gresham, OR 97030-0519,** or e-mail to: **cfroundtable@usacfa.org**

Autumn (current) 2018: Changes We Experience As We Age.

Winter (February) 2019: Organ Transplants. (Submissions due December 15, 2018.) Have you had an organ transplant or are you hoping to get one? Do you have advice or warnings about having an organ transplant? Have you decided to not have a transplant? Please share your experience with our readers.

Spring (May) 2019: Planning For Education And Careers. (Submissions due March 15, 2019.) Do you have any good information about being able to pay for college or career training? Are there career fields that you would recommend for people who have CF?

Summer (August) 2019: ENT Problems And Sinus Disease. (Submissions due June 15, 2019.)

SPIRIT MEDICINE

Reaching One's Potential

By Isabel Stenzel Byrnes

In this issue of Spirit Medicine, I'd like to ponder the theme of reaching one's potential. In the dictionary, *potential* is defined as "latent qualities or abilities that may be developed and lead to future success or usefulness." The most powerful example of potential is holding a newborn baby in your arms, wondering what this human being will grow up to be like and what difference he or she will make in the world.

We all once were that newborn baby, full of potential — except our parents were told that because of a diagnosis of cystic fibrosis, our lives likely would be shortened. Yet the potential of a child with CF is still full of possibilities. Most of us reading this have made it to adulthood. Some of us are athletes, college graduates, working professionals or parents; but others with CF struggle to maintain any normal life due to multiple hospitalizations and our complex medical routines.

I am very blessed to be 14 years post-transplant. Although I have minor medical issues that arise regularly, I am fortunate to live a very normal life right now. My normalcy is defined by working in a 32-hour-a-week job and being able to consistently work for the last six years. Up until the age of 40, I had a very difficult time maintaining the health (and confidence) to work regularly. I feared pursuing some jobs because of my health. Now, for the first time ever, my colleagues don't know I have CF; they know me for me and my skills and talents. For the first time, I'm being judged for my professional skills and personality, and that's it. I have had many opportunities at my job, including becoming a supervisor and gaining in-depth training with a professor on complicated grief treatment for grief counseling. I'm finally growing and learning and contributing. For the first time in my life, I feel like I'm reaching my potential.

> *"While I celebrate this positive place in my life, I also ponder where I would be in my life without my disease. I wonder if anyone else does that?"*

ISABEL STENZEL BYRNES

While I celebrate this positive place in my life, I also ponder where I would be in my life without my disease. I wonder if anyone else does that? This fantasy offers an escape from CF and allows me to dream about "what if." It can be totally depressing, because I'll never know what life is like without CF since it's a permanent part of my story. Yet, thanks to transplant, I have been given a break.

I think about the limitations CF has imposed on many of our lives. We have to plan around health insurance, proximity to medical centers or a career that is safe or part-time. At CFRI retreat, some of us even joked that we sought out a life partner because they had good health insurance! I dream I could have been really successful, breaking the glass ceiling in a high-powered career. Or maybe I could've had children and they'd be applying for college around now. Maybe I would've lived in West Africa or really made a difference in the world. I know I'd still have problems, because all people do, but I wouldn't have my health to set limits on my potential. Perhaps there would be other reasons for limitations, but I often remind myself that it's okay to grieve what I couldn't have or didn't have because of my CF. I'm not trying to feel sorry for myself at all; I'm just acknowledging that CF has shaped my life profoundly. I didn't have a carefree youth. I didn't have athletic prowess growing up. I couldn't do Peace Corps or Teach for America like I wanted to. I became a grief counselor instead of a park ranger.

Yet, I return to the positive and reflect on how we all can reach our fullest potential despite CF. Because of CF, I have become a damn good grief counselor. I may have severe Prograf brain, but at least I can listen and support my clients because of the wisdom granted to me by CF. Life threw multiple curveballs, but it led me down a path rich with life lessons, existential growth and self-awareness. I have finally reached a place where I understand why I'm still alive. There is meaning and purpose to living this long.

I remember spending my precious last days with my 92-year-old grandmother. For many years, she said that she was ready for Heaven. Yet she was still waking up each morning. So she told me, "Always ask yourself, why is God keeping me alive? What am I alive for?" I now can take all the teachings I've gained coping with life-threatening illness, loss and grief, and navigating the medical system, to offer this to my clients. Discovering our potential - at any stage of life and with any circumstance - is one of our greatest callings.

So how do we discover our fullest potential? We need to get creative with CF and explore what we can do despite our health limitations. First, we need to move out of our comfort zone. Joel Osteen says, "If you want to walk on water you have to get out of the boat." Reaching your potential means growing beyond what's easy or comfortable. We have to actively pursue whatever it is that will help us reach our potential. We have to remove distractions, focus on what's most important to us and find mentors who will teach us what we need to know. We all have a profound gift inside of us that is waiting to be released. We have to envision what is possible instead of focusing on what doors are closed. For those who believe this, God wants us to go further and fulfill His destiny for us. We are being asked to keep learning and growing. It takes faith, patience and hope to discover our potential.

The definition above uses the word "usefulness." Perhaps finding our potential means finding ways to be useful. With CF, it can be hard to feel useful when we constantly need help and medical care. Yet, there are many unimaginable ways to be tremendously useful to the world despite CF.

As I write this, the online CF community is mourning the loss of Claire Wineland, a CF superstar made famous by her blogging and Claire's Place Foundation. Claire was an extraordinary young woman who was eloquent, outgoing, authentic, intelligent and creative in sharing her message. Rather than succumbing to loneliness, isolation and estrangement in her hospital room, her life's mission was to educate the world about thriving despite a serious illness. Her death was a devastating blow to the world who hoped to see her reach her full potential.

In her final video, Claire talked about pursuing transplant. She stated, "I'm getting these lungs so I can do something, so I can give something to others...so I can help people." After overcoming her fear of transplant, Claire was genuinely excited to live. She said, "There's so much more I want to do...I want to live and I want to try because I think that I could do something with my life and it could be meaningful." The world wanted to see what her potential would be if she was healthy. Tragically, Claire experienced the worst potential of transplantation - a severe side effect actually shortened her life. This is such a sad end to a woman full of potential. And yet, one source of comfort for me is that, in some ways, CF gave Claire an avenue to reach her fullest potential as an inspirational public speaker, advocate, spokesperson and educator. In her message, Claire was "useful" to both the CF community and the larger community. Though we all wish she had lived longer, I find comfort in knowing that despite her aggressive CF, she did the best she could to live a meaningful, rich and happy life. Now in death, Claire's message will help continue her potential.

> *"Our parents were told that because of a diagnosis of cystic fibrosis, our lives likely would be shortened. Yet the potential of a child with CF is still full of possibilities."*

Like Claire, we all have unique gifts to offer this world. While we are alive, we are full of potential to do good things, to leave our mark and make a difference in this world. And we don't have to "do" anything extraordinary to reach our potential; we can simply "be" the best person we can be to reach our fullest potential. Our potential includes living well with CF.

Now, it is almost 2019 and this will be a new year. I encourage you to set out onto unchartered territory, believe that you possess a unique treasure to give the world and pursue your dreams. May you aim for your fullest potential. ▲

Isabel is 46 and has CF. She lives in San Mateo, California, with her husband of 20 years, Andrew. She can be reached at isabear27@hotmail.com.

SPEEDING PAST 50
...Oh, My Aching Back

By Kathy Russell

Aging is a subject that all of us will have to face or already are facing. In this context aging means getting "old." When I was young, getting old when you had CF meant becoming a teenager. Now, thanks to all of the research and improvements in our care, aging means the same for us as it does for other people. I have to say that it makes me very happy that people who have CF can expect to get "old."

CF-related arthritis is something that many of us have to deal with. It can start at a very early age, but it can get much worse as we age. Many of us have scoliosis and other spine problems. As we age we are more likely to have osteopenia or osteoporosis, which cause a great deal of back pain. I don't enjoy having back pain, but I am happy to be alive to feel it. I don't take any pain pills, so this level of pain is my new normal. I am accustomed to it...most of the time. The rest of the time, I sit with a heating pad on my most uncomfortable back muscles. It may not make it better, but it does make me notice it less. Also, I sit on a special pad so that my seat muscles do not get so sore. (I used to have more padding there, but I have lost weight and can feel the bones, once again.)

My hands are painful, too. I notice more pain in my thumbs than my other digits. There are times that my thumbs are so unreliable that it almost is as if I have no opposable thumb. I cannot rely on my thumb to keep a strong hold on cups, glasses or bowls. There just is no strength in that thumb joint, much of the time. I have not found a brace or supporter that helps with these painful thumb joints. I just have to stay aware of how they are doing on any given day.

My ankles are weakened, too. I have injured my ankles a few times in the past. I never have broken a bone or had a serious sprain, but I have twisted my ankles enough that they can get unreliable at times. I find that sitting with my feet up when I can and wearing prescription orthotic inserts in my shoes helps. I try to be aware of where I put my feet when I'm walking. I don't want to make a misstep and do some real damage to my old ankles.

I mentioned my orthotic inserts – they help me avoid some of the pain of arthritis in my feet. At times, my feet feel as if there is a marble underneath the ball of each foot when I step down. I used to look to see if there was something there. Now I know that it is just how my feet feel. I also have had to deal with occasional bouts of plantar fasciitis. That is when the heel of a foot feels as if you have jumped down on a rock. Yikes! It hurts.

Losing lung capacity is another problem that most of us will face. It is normal for one's lung capacity to decrease with age. It just happens that ours tends to decrease more and at a faster rate than the general population. Many of us do or will use supplemental oxygen. It is normal that we resent having to wear a cannula in our nose. However, it is much better than the alternative if we choose not to wear it. I have some friends who are on supplemental oxygen for other lung problems. They hate to wear it in public, so they choose to stay home or to go out without it. They then are unable to do much physical activity because they run out of air. I'd rather wear it and be able to partake of life. It may not be attractive, but it is what it is and I am grateful for it.

> *"I don't enjoy having back pain, but I am happy to be alive to feel it."*

KATHY RUSSELL

So far, I have mentioned many things that are not fun or pleasant, so I will now try to be more positive. I am happy that I have survived so long because so many positive steps have been made by researchers, physicians and other caregivers in the care and treatment of CF. When I was young there were only a few very basic antibiotics. I am sure that my docs tried all of them on me at one time or another. They were so sure that they would be able to "cure" me. Of course they really didn't know much about CF. Most of them had finished medical school before CF was "discovered," so they really didn't understand.

There was no Pulmozyme, no handheld inhalers, no therapy vests, no small compressors to run nebulizers and the list goes on. It seems as if there are new breakthroughs happening almost every month, now. If people are compliant and really stay active, they have every reason to expect to live a long, fairly normal life with CF. That may the best thing (for me) about living long with CF. When I was diagnosed at age 12, the median life expectancy for people with CF was about four years of age. (Since I already was 12, I figured that they didn't know what they were talking about.)

Now more than half of people with CF in the U.S.A. are at least 18! Life expectancy numbers continue to go up. Children born with CF now have a life expectancy of 43-plus years. Wow! Those are some great numbers. I never thought that I would live long enough to see those kinds of statistics.

Another great change in CF during my lifetime is that when one is around a group of kids who have CF, it is not evident that they have CF because there isn't the sound of constant coughing. I am so happy about this. These children may never know the stigma of having an almost constant, noisy, congested-sounding cough. They may never have people shy away from them because of their coughing. Also, they may never have the embarrassment of uncontrollable gas and odiferous bowel movements. Modern enzymes are such a blessing. This also makes me happy. I hope that everyone gets to live long enough to be like me and be able to talk about "back when I was young" about their CF.

Thinking of these positive steps in CF life makes me forget about my aching back and keeps me happy to be alive. I wish each of you a long and happy life that is filled with good health, love and laughter.

Stay healthy and happy,
Kathy ▲

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sweat chloride levels. In general, the combo therapy was well-tolerated, with adverse events mild to moderate in severity.
http://tinyurl.com/yd5ku6lc

Kalydeco's Benefits Include Lower Risk Of Death And Pulmonary Flares, Real-world Data Shows

Real-world results from the largest analyses of Vertex Pharmaceutical's cystic fibrosis treatment Kalydeco (ivacaftor) done in CF patients to date confirm the therapy's ability to significantly lower the risks of mortality, transplants, hospitalization and pulmonary exacerbations compared to untreated patients. Researchers analyzed data from a subset of patients, none with a lung transplant, undergoing treatment with Kalydeco for five (U.S. group) and four years (U.K.). Data from these patients were compared to that of age-, sex-, and disease severity-matched CF patients not using Kalydeco. They evaluated lung function (measured by the percent predicted forced expiratory volume in 1 second, $ppFEV_1$), the rate of pulmonary exacerbations and hospitalizations and infection with the bacteria Pseudomonas aeruginosa. Additional measures included body mass index (a measure of body fat) and the number of CF-related complications (such as CF-related diabetes). Analysis of data showed better preserved lung function, improved nutritional measures, decreased risk of PEx [pulmonary exacerbations] and hospitalizations and favorable trends in prevalence of CF complications and P. aeruginosa prevalence in patients treated with Kalydeco.
http://tinyurl.com/y8943cjs

Phase 3 Trials To Assess Potential Breakthrough Therapy For Patients With Cystic Fibrosis

Two Phase 3 trials are being conducted to explore a new potential breakthrough therapy of two compounds—VX-659 and VX-445—which will be used as part of a triple combination modulator therapy. Researchers postulate that the new therapy could potentially address the underlying cause of the disease as Cystic Fibrosis Transmembrane Conductance Regulator (CFTR) modulator therapies are designed to fix the malfunctioning protein created by a mutated CFTR gene. The hope for the trials is to develop a combination therapy capable of effectively treating CF patients with a single F508del mutation.
http://tinyurl.com/y8yy4hqz

VX-445

VX-445 is designed to treat the most common form of cystic fibrosis as part of a triple therapy with tezacaftor (VX-661) and Kalydeco (ivacaftor). VX-445 and tezacaftor increase the amount of mature protein that reaches the cell surface by targeting the processing defect that causes the faulty protein to be degraded. Ivacaftor is designed to increase the function of the protein once it reaches the cell surface. The combination of the three treatments should improve the flow of salt and water into and out of the cells. The triple combination therapy of VX-445, tezacaftor and Kalydeco has been tested in Phase 1 and 2 clinical tri-

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is because my primary CF symptom is massive hemoptysis. I cannot count the times I have suffered massive bleedings that have put me in serious risk. Those who have undergone the experience know it well enough that I will forego description. It is not grossly inaccurate to describe my situation as grave: given the right circumstances, I could bleed out, or asphyxiate on my own blood. (Perhaps it goes without saying, but for years I have studiously avoided drinking fruit punch). Yet it would also not be inaccurate to say that I lead an extraordinarily healthy life for someone with CF: my lung function is high, I am a serious salsa dancer, and I can perform basically any activity I please. This two-fold reality, of health and sickness, is tied together by the red bands of fear—when healthy, fear of sickness, and when sick, fear of death. If I am carrying a time-bomb in my chest, then this low-grade anxiety is the oppressive "tick-tock" of the hands making their way through potentially bloody revolutions. It's a jolt, a rude awakening, every time I am reminded that I am not an entirely welcome visitor in the land of the healthy; for, after all, I have dual-citizenship, and though I may feel rusty at first, upon returning to the land of the sick my quick recall of hospital colloquialisms and CF lingo reminds me that sickness remains my mother tongue.

Yet the violence of the transition from normalcy to crisis, from balancing my studies to bleeding at length, constitutes such a shock to the system that the psyche inevitably seeks some mediating term to negotiate the difference. Fear fulfills that role. I am constantly alert, perpetually scanning for signs of slight decrease in lung function, of joint pain, of tightness in the chest, of whatever, simply—I have concluded—as a way to pass the time, of alleviating the duller ache of waiting. These are, to employ Dickinson's words, a "fitting," a trying on of disappointment before it arrives. It has taken me years to realize how ingrained this habit of anticipation has become. It is now a reflex, second nature. And it's not all bad: some degree of anticipation makes for prudence in a life shot through with greater risks than normal. But like wearing a winter jacket at the beach, the bulk of this anxiety ill suits me in moments of health, making it difficult to play in the sand and shore.

More curious, and more pernicious still, is the way this coat hangs onto my skin. After a while of wearing it, it becomes too difficult to take off. In fact—if I'm honest—I've kind of grown to like it. For to see myself without it would be eminently strange.

SHAY NOTHSTINE AND ROBERTO DE LA NOVAL AT SOUTH BEND LATIN DANCE WEDNESDAYS.

While I was fearing it, it came,
But came with less of fear,
Because that fearing it so long
Had almost made it dear.

It is bizarre truth that fear can transform into comfort, can become so familiar that to live without it is itself fear-inducing. For the human psyche craves equilibrium, and it will take whatever it can get to achieve it. To describe my fear as dear is just to say that my fear has become a constitutive element of myself, the shades through which life's light is refracted for me. This is perhaps seen most clearly in the fact that today, though I am now fairly sure (always a dangerous thing to say with CF!) that I will live beyond my 32nd year, this year continues to bear a heavy symbolic weight for me. To find relief in the fact that things are going better than expected with my CF is, frankly speaking, hard to do: That fearing it so long had almost made it dear.

But this is just one apparition of subliminal fear. It can manifest in as elemental a fashion as feeling a pang of terror at the sight of red soon after a major bleed. Or in the anxiety of recurrent chest tightness and arthritic deterioration of my feet, a potential indicator that my dancing days are numbered. Or, in a more subterranean fashion, in avoiding taking trips due to an unconscious anxiety of suffering an exacerbation while abroad. The fear can co-exist with survivor's guilt, for living while others die and thriving while others barely survive (an experience described so well in Isabel Byrnes's "Spirit Medicine" column in the Summer 2018 issue of *CF Roundtable).* Coupled with this guilt, the anxiety can unconsciously cause me at times to distance myself from the CF community, to block them out of view so as not to catch glimpses of the road I too must walk in time. Worse still, fear can cloak itself in disinterestedness and evasions in the things of the heart—love and commitment, the haunts of intimacy. The fear flits in and out of consciousness, at times visible in its starkness and at other times masked by its permutations and its attendant emotions and attitudes. But it is pervasive, and it is a part of me.

All this is not meant to be a paean to fear. I describe this experience in detail, rather, to lay it bare and perhaps

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to invite recognition in others who read it, whether they have CF or not. For we all are afraid. But when we do finally become conscious of the fear that is always rattling its tail in the darker caverns of the self, a new agency emerges alongside this insight: fear can lose its dearness. A new self can slowly, painfully come to light. But that is possible only when a new fear is faced, the fear of living without the cold comforts of the normal, the expected, the dreaded. The fears that crippled and bound me served, in their time, a purpose; they helped me survive what was foreign and seemed unbearable. They were a way of making sense, even as they paradoxically caused me more suffering, only of a different kind. But now, when I can see they have outlived their usefulness, I must begin the difficult work of bidding them adieu or negotiating stricter terms for our cohabitation.

The trying on the utmost,
The morning it is new,
Is terribler than wearing it
A whole existence through

Fashions come and go, and at times the coats of anxiety just don't fit. To be present in health, to enjoy it, to pursue it, to take the risks accompanying it—these are better than wearing anxiety a whole existence through. Better the shock of surprise than a living death; better mindfulness than submerged panic; better reality than fantasy. While I was fearing it, it came—I hope instead that whenever the next tragedy comes, it will occur not while I am fearing, but while I am caught up in the business of life. ▲

Rob De La Noval is 29 and has CF. He is a doctoral candidate in theology at the University of Notre Dame. The son of Cuban immigrants, he grew up in Miami, FL, and now lives in charming South Bend, Indiana. He has yet to adjust to the snow. You may contact him by e-mail at RobetDLN@gmail.com

als. Improvements were observed in lung function measured by forced expiratory volume in one second (FEV_1), which measures the total amount of air exhaled in one second and sweat chloride content, a measure of disease severity.
http://tinyurl.com/yasak6q7

CF Study Compares Outcomes Of Lung Transplants

One-year survival after a lung transplant of cystic fibrosis (CF) patients chronically infected with antibiotic-resistant bacteria is similar to those without infections. Cystic fibrosis patients often develop pulmonary infections with bacteria that are capable of resisting the action of conventional antibiotics. These multidrug-resistant organisms may affect the long-term success of the procedure in these patients. Researchers have now compared the one-year mortality rate between CF patients who are negative and those who are positive for chronic infections with multidrug-resistant organisms. They analyzed data from 3,256 cystic fibrosis patients who had a lung transplant and found that, even before the surgery, there were significant differences between the two sets of patients. Namely, those who were chronically infected with multidrug-resistant organisms were more likely to be on extra-corporeal life-support and/or ventilation assistance. These patients were also more likely to have an infection requiring intravenous antibiotic treatment in the two-week period before the transplant, and had a higher rate of pneumonia episodes requiring an infusion of antibiotics in the 12 months preceding the surgery. Despite this, the one-year mortality post-transplant was similar between CF patients without infections and those chronically infected with multidrug-resistant organisms. Based on the results, the researchers concluded that the presence of multidrug-resistant organisms was not a predictor of one-year mortality, and suggested that patients with CF chronically infected with MDRO [multidrug-resistant organisms] should not per se be excluded from LTX [lung transplant] but undergo thorough multi-disciplinary evaluation including infectious diseases specialist input.
http://tinyurl.com/y7k5d7al

HOPE-1 Trial Shows Potential Of SPX-101 To Improve Lung Function In CF Patients

Results of a Phase 2 trial showed that treatment with Spyryx's investigative drug SPX-101 can improve lung function in patients with cystic fibrosis, regardless of their background genetic mutation causing the disease. SPX-101 is a small protein fragment that was developed to target epithelial sodium channels (ENaC) in the lungs and prevent them from taking up sodium. The drug reduces sodium absorption, allowing fluids to be retained on the airway surface, making mucus clearance an easier task. During the trial, SPX-101 was found to be safe and well-tolerated. The most common adverse events reported were increased sputum production and cough. No major impact on blood potassium was reported. The effectiveness and safety of SPX-101 is currently being evaluated in the HOPE-1 trial (NCT03229252).
http://tinyurl.com/y9773x4l

Inhaled Mannitol Can Help To Improve Lung Function In Adult CF Patients, Data Show

Treatment with inhaled mannitol is safe and can help to improve lung function in adult patients with cystic fibrosis (CF). Inhaled mannitol is being developed by Pharmaxis and is currently available in some countries (namely in Europe, Russia and Australia) under the brand name Bronchitol. It is still unclear exactly how the treatment works, but it is believed to modulate the underlying defect of CF by improving hydration of the airways, changing

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FOCUS TOPIC

CHANGES WE EXPERIENCE AS WE AGE

Turning 30

By Jessika Biele

I was born into a society that hates birthdays past the age of 30 and glorifies everything about being "healthy." I also was born with cystic fibrosis, so no matter how much I tried I could never be the poster child for what it means to be the epitome of health. I have also enjoyed and reveled in every birthday I have ever had. The honest truth about aging? For most people, CF or not, aging is this paradigm shift between being happy to be alive and being scared about getting closer to death.

For my parents, and even my boyfriend, aging is about this number that they want to keep secret, it is about all these things that they can't do anymore or they must do more slowly. For me, aging has been this beautiful, magnificent process of gaining more knowledge and more strength to live in harmony with this disease that I have had since birth. Doing things slower? Yes, please. Things I can't do anymore? I'll find new things. Keeping my age a secret? Absolutely not. Just like the wrinkles on our faces and the scars on our bodies tell a story, my age is a solid representation that I am still here, and I am still going. I celebrate every birthday with pizazz and excitement, and I encourage others to do the same. Sometimes I even celebrate half-birthdays because, well, life.

The day I turned 30, I coughed - rather lightly actually - and pulled a muscle in my shoulder. I had to ice and heat that thing for at least two weeks. Having shared this experience with a group of people who did not have CF, I was bombarded with similar stories – "I pulled my back out pulling clothes out of the dryer," "I drove for two hours yesterday and could barely walk." Sure, I am getting older, but so is everyone else around me. When I was younger I used to loathe when non-CFers would say things to me that implied that my journey – and what I thought back then was a struggle – was the same, if not just as bad as theirs. "I have asthma," "I'm diabetic." Yeah guys, but I have cystic fibrosis on top of those things. I did not like the comparison. Aging, however, taught me that it is not a competition, and in the moment where I expressed my pulled shoulder from coughing to that group of people, I realized that it was absolutely and serendipitously okay that I was almost like everyone else. My differences are amazing but so are my similarities. Sometimes I forget that, as I age, it is just as important to connect with people as it is to inspire them with my stories. Honestly, everyone has a story that is inspiring. Everyone has overcome something. Living and aging taught me that.

The month that I turned 30, I decided to start running. I know, right? Even though I thought I had prepared myself for the running journey, I was sadly mistaken when a leg pain that woke me up in the middle of the night turned out to be a broken hip. That's right, I had fractured my femoral neck at the ripe age of 30. I opted out of surgery – doctors agreed I did not need it – and instead was on crutches for upwards of six months. I even crutched my way through a European vacation. The injury is strange for someone my age, but with CF, maybe it wasn't. I feel like I have been living this paradox my whole life. At 16, I had already been in the hospital and in doctors' offices more than I had been home. So, at 16, my body age could have been anywhere from 40 – 50 years old. At 30, with FEV_1, weight, medications, manifestations of CF et cetera considered, I could estimate that my body age is upwards of 80. Who knows, right? What matters to me, though, is quality of life. My body may diagnostically show that I am not 30, but my heart and my

> "My body may diagnostically show that I am not 30, but my heart and my spirit show no age."

JESSIKA BIELE GETTING AROUND IN THE LOUVRE.

spirit show no age. Living and aging taught me that.

The year that I turned 30, I had to stop working so much. I thank my boyfriend, Paul, for this endlessly, probably too much he would tell you. Though I am not on supplemental oxygen and I do not go on IVs as much as I used to when I was younger, I still find that everyday activities, plus treatments, plus exercising, plus maintaining my weight, plus work and plus school, became overwhelming. It would be only natural that some of those things would have to take precedence over the others. When I was younger, it was my treatments, and exercise, and a balanced diet. At the age of 30, while work and school are important to my mental well-being, I could not very well continue doing those activities if I was not taking care of myself physically. Balance is key. Even though I am still in school, contribution to society was always important to me. To meet that emotional need, I started volunteering – when I was 30. Volunteering is flexible especially since it is not paid work. I can say "no" more and be helpful when I am able. Living and aging taught me that.

As I have gotten older with CF, I have noticed that I am a little slower, I hurt just a little bit easier, and I need just a little bit more recovery time than I used to. But as I have gotten older with CF, I have also become more compliant with therapies, I have become more involved in the cystic fibrosis community and I have appreciated the crazy days just as much as I have appreciated the days where I do absolutely nothing except watch Harry Potter movies. As I have gotten older with CF, I have learned quite a bit more compassion and empathy for those with and without CF, I have learned to listen to my body more and I have learned the importance of self-care.

Aging is amazing. ▲

Jessika is 32 and has CF and CFRD. She is a Director of USACFA. She is a ghostwriter, copywriter and freelance website designer. She lives in Florida with her boyfriend, Paul, and their puppy, Bhodi. You may contact her at jbiele@usacfa.org.

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mucus properties and easing its clearance. The treatment safety and efficacy were tested in a Phase 3 trial (NCT02134353). Researchers found that relative change in ppFEV$_1$ after six weeks of treatment was significantly associated with a sustained positive response over the trial's 26 weeks. Based on the findings, the researchers concluded that inhaled mannitol was shown to be a safe, effective treatment for adult CF patients.
http://tinyurl.com/ycgsfgbh

P. aeruginosa Strains In Cystic Fibrosis Patients Killed By Combining MS Treatment With Antibiotic

Combining the approved multiple sclerosis treatment glatiramer acetate with the antibiotic tobramycin improved its ability to kill antibiotic-resistant strains of Pseudomonas aeruginosa isolated from cystic fibrosis (CF) patients.

In a previous study, researchers showed that glatiramer acetate has antimicrobial activity against P. aeruginosa. In this study, the research team tested its potential in helping to overcome various P. aeruginosa strains that are resistant to antibiotics.

The team tested antibiotic sensitive and resistant strains of P. aeruginosa isolated from airway secretions of CF patients. The different strains were incubated with varying concentrations of tobramycin, both in the presence or absence of glatiramer acetate. Researchers incubated the cultures overnight and assessed the bacterial growth every hour. Adding glatiramer acetate to tobramycin increased the killing of sensitive P. aeruginosa strains by 56.6%, results showed. The tobramycin-resistant strains were also more effectively killed in the presence of glatiramer acetate, especially with higher doses of the antibiotic. The researchers are also testing glatiramer acetate's ability to work with other antibiotics, namely ceftazidime, ciprofloxacin and colistin.
http://tinyurl.com/y7nbd49g

Antioxidant Supplementation Associated With Reduced Respiratory Illnesses In Patients With Cystic Fibrosis

Treatment with antioxidant-enriched multivitamins may reduce respiratory illnesses in individuals with cystic fibrosis (CF). In the study, the researchers examined how a cocktail of multiple antioxidants affected inflammation and health outcomes in patients with CF over a 16-week period. Patients in the study do not ordinarily absorb dietary antioxidants, including carotenoids such as beta-carotene, vitamin E, CoQ10 and selenium, which can help neutralize inflammation in the body. The capsules used in the study were specifically designed for individuals with difficulties absorbing important dietary antioxidants, including carotenoids such as beta-carotene, vitamin E, coenzyme and selenium. According to the researchers, supplementation increased antioxidant concentrations in

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Wheelchairs, Walkers And Canes

By Linda Stratton

Recently, I moved into affordable housing, a senior (55 plus) building in Louisville, Colorado. As some of you already know, I had been caring for my parents—Mom passing in 2012 and Dad, more recently, in 2017. The thought of what I perceived this move would be and what it actually became brings a smile to my face today. In observation, while moving into my new apartment, I noticed several residents in wheelchairs, using walkers or canes. I thought, "This is perfect—my new purpose!"

When I applied to be on the waiting list at Kestrel Property, two years ago, I had several ideas about how to stay active and be helpful. Moving in, as one of the younger residents arriving, visions of getting to know my elderly neighbors and implementing activities and beneficial care danced in my head. In reality, those "elderly" neighbors have shown the ability to not only get around, but most of them are able to work circles around me and my broken down body.

One neighbor, in his 90s and caring for his wife as well as himself, makes daily treks to the grocery store, crossing an unusually busy street—he walks with a cane. Another couple spend their mornings going to an infusion lab for treatment, then most afternoons off to take part in numerous activities with their grandchildren. Around the outside of our building, there are elevated gardens filled with vegetables and flowers flourishing from nurturing care. Also, many of us meet every afternoon in one of the common areas for coffee and friendly fellowship. In addition, activities such as bingo, movie nights, potlucks and sock-hops are regularly planned solely by our residents. I become tired just thinking about the many ways my neighbors, and now new friends, stay active.

> "Having been a fairly active CF adult, I struggle with my new reality, I struggle with asking for help."

Turning 64 years old in July 2018 hasn't been one my best years. I made it through the move okay and had such high hopes for a new life. Unfortunately, a virus came my way this past spring and my lungs have been murky ever since. With another hospitalization looming in the very near future, I'm afraid this year is going to be over before I ever get started. During my last debilitating exacerbation, neighbors brought food, ice cream and many get-well wishes on a daily basis, checking to make sure I was okay. While hospitalized, one new friend collected mail and cared for my cat, Gracie. I was extremely thankful for their care and concern.

THE FIRST SOCK-HOP AT KESTREL. LINDA STRATTON ON THE LEFT, FRIENDS SUSI AND JODI ON THE RIGHT.

It's difficult to accept the changes in my health and body, wondering if it's the beginning of the end, if you know what I mean—pulmonary function is at an all-time low. Having been a fairly active CF adult, I struggle with my new reality, I struggle to admit I'm not able; I struggle with asking for help.

Attending a recent potluck/sock-hop, a workable solution suddenly dawned. It's not productive to focus on what I'm not able to do—I must focus on what I CAN DO! I didn't feel able to get out on the floor and do the twist, the pony or the stroll amongst many poodle-skirted dancers; lugging oxygen behind me somewhat hinders my agility. I could, however, get up, increase the oxygen one liter and dance in place, singing along with the relished songs of the '60s. A great time was had by all.

What a blessing it's been to have this experience of a new, though different from planned, beginning. What has transpired turned out to be the polar opposite of what I had in mind for myself and others around me. As I age, cystic fibrosis and the treatment thereof mercilessly ravage my body. Nonetheless, the good Lord has placed me in a world of care—a world where in a short period of time I've made many special friends. ▲

Linda is 64 and has CF.

YOU CANNOT FAIL

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

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

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

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IN MEMORY OF
LAUREN
MELISSA KELLY

Changes As We Age

By Jeanie Hanley

Although there have been many changes over the years for better or worse, the best changes have been understanding my body better and becoming tuned to what makes me feel better physically and mentally. I've learned what makes this body work best - exercising daily, sleeping at least eight to ten hours a day, being outdoors for at least two hours a day and, it goes without saying but I'm going to anyway – taking all my medications. And I've learned what circumstances make it worse. To mention a few, I know to avoid red wine (hemoptysis), carbonated drinks (GI issues) and certain foods and medications because they affect one or more areas – lungs, sinuses, abdomen and mental health.

JEANIE HANLEY

It takes constant fine-tuning to identify patterns of how foods, caffeine, activity etc. interact with each other and affect different organ systems. I have a good idea of what my limits and restrictions are. When I exercise over an hour at a time, it saps my energy. When it's between 30 minutes to under an hour, I feel energized throughout the day. When I don't drink enough water, dehydration occurs with symptoms of wheezing and chest tightness, abdominal pain and salt craving. When I have too much

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the blood of the participants and temporarily reduced inflammation in the blood at four weeks, but not at 16 weeks. Additionally, half as many of the patients taking supplemental antioxidants experienced a pulmonary exacerbation requiring antibiotics compared with the group taking the control multivitamin. Patients in the group treated with antioxidants also experienced a lower frequency of respiratory illnesses compared with the control group.

http://tinyurl.com/y9ym9dp4

AND

http://tinyurl.com/ybk7gwe7

Lipid-based Nanoparticles Enable Improved Therapy For Cystic Fibrosis Patients

Scientists and clinicians at Oregon State University (OSU) and Oregon Health & Science University (OHSU) have demonstrated the possibility of a far more accessible form of CF treatment, which does not attempt to rescue the mutant protein. Instead, it uses chemically modified RNA messengers for cystic fibrosis transmembrane conductance regulator (CFTR, the faulty gene that causes the disease) into nanoparticles. This results in a medicine that can be inhaled at home. Rather than trying to mend the patients' existing mutant protein behind the mucous and lung dryness, the mRNA-loaded nanoparticle approach causes the cells to actually make the correct protein—that allows cells to properly regulate chloride and water transport. Other types of treatments had to be patient-specific because of all of the different mutations, but this therapy works independent of patient genotype. These systems can be repeatedly administered to a patient and the effects are reversible if someone needs to stop the therapy for any reason.

http://tinyurl.com/y9kbdlr6

AND

http://tinyurl.com/ybf85ld2

AND

http://tinyurl.com/y7ozy6pq

Positive Top Line Data From The CARE CF 1 Clinical Study Of Oral Lynovex In CF Exacerbations

NovaBiotics announced that its oral therapy for cystic fibrosis (CF), Lynovex®, has met the study objectives of the CARE CF 1 clinical trial. CARE CF 1 assessed the effects of two weeks of Lynovex treatment as an adjunct to standard of care therapy (SOCT) in CF, compared to placebo plus SOCT. This trial was designed to determine whether the inclusion of Lynovex capsules alongside SOCT lessened the clinical impact of exacerbations in adults with CF, as measured by symptom severity and levels of bacteria and inflammatory mediators in sputum and blood. No drug-related severe adverse events (SAE) were

> “Another change for the better has been a willingness to try new things, learn what leads to feelings of happiness and passionate interests, and then do them as often as possible.”

caffeine or more than one glass of Prosecco, the same symptoms of dehydration occur. If I have insomnia and don't make up for the lost sleep during the day, it affects my concentration, my activities and my diet in that I start eating poorly. Bring on the sweets!

Another change for the better has been a willingness to try new things, learn what leads to feelings of happiness and passionate interests, and then do them as often as possible: reading at the beach, swimming, investigating ancestry, socializing, planning trips etc. This willingness has not always led to new interests. Sometimes it's made me realize what I don't like – cycling (vertigo and imbalance don't mix), calamari (too chewy), fish served with the eyes staring at me (too sad). On the other hand, fried green tomatoes and pickles worked for me; kayaking and reiki are new interests I enjoy.

Reaching out more to friends and socializing has been another beneficial change. I have come to realize that good friends don't care about my chronic coughing. They are considerate enough to give me a heads-up when they're sick. And, since I'm fairly outgoing, as long as I'm feeling well and know I'm not contagious, then I'll join social events at the drop of a hat. The end result of this willingness has been improved physical and mental health. ▲

Jeanie is 56 and is a physician who has CF. She is a Director of USACFA and is the President. She lives in Los Angeles with her family. Her contact information can be found on page 2.

reported. The initial efficacy outcomes measured in CARE CF 1 were the reduction in the number of bacteria in sputum, improvement in patient-reported outcome scores of CF respiratory symptom severity and improvement in lung function (FEV_1) following two weeks of treatment. A statistically and clinically significant dose and regimen-specific reduction in CF respiratory symptom severity was identified in patients taking Lynovex plus SOCT versus those on placebo plus SOCT after two weeks of treatment. The same dosing regimen also resulted in a clinical and statistically significant reduction in blood white cell count after two weeks when compared with placebo. These Lynovex specific improvements were mirrored by changes in health-related questionnaire scores, sputum levels of inflammatory mediators and an increase in lung function after two weeks of treatment. The sputum of patients taking Lynovex also contained less bacteria than sputum from patients who received only placebo plus their SOCT antibiotic treatment. These clinical effects are supported by the finding that Lynovex significantly reduces the blood white cell count indicating reduced inflammation in the lungs.

http://tinyurl.com/y9dgp9u5

AIT Therapeutics To Present New Data On Nitric Oxide's Ability To Clear Mycobacterium Lung Infections

Nitric oxide (NO) is a naturally occurring gas that plays a significant role in triggering the immune system. In addition to its involvement in several biological processes, when applied directly to the airway, it can attract immune cells and has the potential to clear bacterial, fungal and viral infections. Mycobacterium abscessus complex (MABSC) is a group of multi-drug resistant nontuberculous mycobacteria that cause very aggressive and hard-to-treat lung infections. There are currently no approved treatments against MABSC. AIT Therapeutics' new data supports the ability of nitric oxide to fight MABSC lung infections. The nitric oxide generator and delivery system developed by AIT Therapeutics is a patented drug-device combination that generates nitric oxide from normal air.

http://tinyurl.com/y7bddk9a

AND

Data From CF Patients Show Nitric Oxide's Potential Against Bacteria

Nitric oxide is a small molecule that is an important mediator of immune defense mechanisms against infections. The compound has been shown to have broad-spectrum antibacterial activity against several strains of bacteria. Researchers evaluated NO's potential

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POETRY CORNER

Only Morning

By Grace Knight

Only morning, only morning, only morning, only morning

Only morning is the place where I get to be by myself, for myself, and only myself
I look outside and see the trees, the sun, the birds chirping, and I hear the drone
The vibrations of my machine remind me that I'm alive,
It relaxes me
Its rhythm matches my heart and tells me every thing will be ok
It whispers through its tubes a strong message of peace to my body
I can feel my lungs rejuvenate, release, and recharge as if the electricity from the wall-plug is directly transferred into my bloodstream
I breathe in, I breathe out, I am new for the day.

Only morning, only morning

In the morning, I am active
I swallow
I take
I spit
I ache
I smile
I wake
Only morning
I eat
I drink
I push away sleep
I panic
I breathe
Only morning
I cry
I sweat
I ignore regret
I run
I am done

GRACE KNIGHT

Only morning
I tire
I groan
I cough
I'm alone
Except for a drone
Only morning
I clear
I move
I'm loud
I breakthrough
I commit
I do
Only morning

Only morning, only morning

In the morning, I have the most hope I will have throughout the entire day.
The release of mucus, mimics the release of my fears, the letting go of the things that hold me back

I am stronger in the morning, ready to face the world,
With the swell of my lungs inhaling medications, follows the swell of my spirit
I take in more air now, I breathe in more hope, I fill my lungs with the toughest, hardest, brightest smoke possible so when something goes wrong, and life gets hard, I can handle it.

Only morning, only morning

Morning is when I prepare for battle

I go to the chest that holds my armor
The chest that saves my chest
I open the jars that give me life
The jars that open my lungs
I pick my potions
I mix my medicines
This is what saves me
This is what protects me
When I face my enemies
And bacteria crashes against my cell walls
They cringe at my power
And scream at my arsenal
It is comical really that they
Think they can get to me
When I have my secret weapon
When I have the morning.

Only morning, only morning

Sadly, morning cannot be all fun and games. It is in the morning that dread creeps in.
I am washed in a sea of doubt
Can I keep fighting? Can I keep breathing? Can I survive this storm?
This storm, the storm that lasts for a long long lifetime
I wake up and I am reminded that it didn't leave my body as I had dreamed, or walked out of my life like so many things have
It sticks to me like glue, like tape, like that one friend who needs everything but gives nothing back
It takes from me, It feeds off me, It is part of me.

In the morning, I feel this the greatest. I have not had time to forget its existence
Or to shrug off its whispers

In the morning it clings to me like the last remnants of sleep,
It tangles around my throat like the sheets around my body,
It holds me down, daring me to get up
To rise and face its teeth, its claws, its lifeless expression, its eyes a void of darkness.

In the morning, every morning, I awake to this monster
And in the morning, every morning, I stand up to its terror.

Because

The sun rises in the morning
The birds sing in the morning
The day breaks in the morning
And like the world around me
I shine bright, blare loud, and break boundaries in the morning
Against all odds
Against all pain
Against all demons
And hurt
And sickness
And death
And defeat
And sadness
And disease
And heartache

Against everything I stand tall

And in the morning, I am a winner, the champion, the best of the best

Only morning is the place where I come up on top
Because in the morning, I rise. ▲

Grace is 20 and has CF. She is a college student from Tyler, TX. You may contact her at: graceknight7@gmail.com.

Meet The New Director—Jessika Biele

I am 32 years alive and absolutely ecstatic to be able to contribute to the mission of USACFA. I have grown up watching the CF community drastically change for the better, and I am excited to finally be a part of that change. I was diagnosed with CF at the age of four and at the age of 13 I was diagnosed with CF-related diabetes. I have been through major ups and major downs in terms of my CF, but have found that mindful and positive thinking, in association with friends and family, exercise, healthy eating and staying organized, keep me grounded in mediocrity in an otherwise chaotic world. I have learned to embrace both.

I am currently a freelance ghostwriter specializing in self-care, relationships, health and wellness, personal finance, motivation and organizational happiness. I am also a professional editor and copywriter. I have loved and continue to love being a ghostwriter mostly because of the flexibility and the varying nature of the work. I have a bachelor's degree in accounting, am currently pursuing a master's in organizational leadership and plan on directly pursuing a Ph.D. in organizational psychology. While my current academic background aids in my current career, there are plans on implementing my knowledge into the CF community.

JESSIKA BIELE

I live on the east coast of Florida in a small beach town on a barrier island with my significant other, Paul, and our new puppy, Bhodi. Paul is a recreational surfer, so we chase the swell around the world (thanks honey). He surfs and I beach. We just adopted Bhodi, an American bulldog-great Dane mix from a rescue down in Key Largo, FL. He is currently being trained to be my diabetes medical alert dog. I love a good book, a good movie, good music, good food and a good cup of coffee. I live for the sun, the sand and the ocean. I am an artist. I absolutely love to cook. I am a bit obsessed with health and wellness. I don't have a Facebook. Cleaning is therapeutic for me. My favorite kind of sushi is tuna. I love talking to strangers. Bullet journaling has been a blessing to my productivity. And I probably do drink way too much coffee. ▲

TILLMAN continued from page 15

against Pseudomonas aeruginosa. The treatment not only showed it could significantly reduce P. aeruginosa burden, but it could do it faster and at a lower concentration than that reported for M. abscessus.

http://tinyurl.com/y9zk334z

Award Granted To Neem Biotech To Develop Antimicrobial Intervention For Chronic Lung Infections

Neem is working on a novel approach to antimicrobial resistance, which involves preventing the formation of and disrupting mature biofilms that are produced by bacteria such as Pseudomonas aeruginosa and Staphylococcus aureus. In lung biofilm communities, bacteria are both protected against antibiotics and the host immune system. Elements within the biofilm also impact upon the level of hydration of the lungs in cystic fibrosis. Any compound developed using this approach has a different mechanism of action to conventional antibiotics and is designed to be used alongside antibiotics in the treatment of persistent bacterial lung infections. By disrupting the chemical signaling used by these bacteria to form biofilms, it will allow antibiotics to work more effectively and can minimize the impact of phenotypic antimicrobial resistance.

http://tinyurl.com/y9bxsjpn

Pancreatic Cell Transplant Program Offers New Hope To Some CF Patients

It is relatively common for patients with cystic fibrosis (CF) to develop small cysts in the pancreas (pancreatic cystosis), as a result of impaired hydration of the organ, high protein content secretions and inflammation. There is no treatment for this condition, and only management of the symptoms or surgical removal of the cysts are possible therapeutic options. The Chronic Pancreatitis and Autologous Islet Cell Transplant Program at the University of North Carolina was established in 2016. The treatment consists of removing the pancreas while saving, to some

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In the Redwoods the Ghosts Have Not Gone

The redwoods still stand
Between older blackened stumps
Their over-hanging, long-lived branches
No longer receive smoke from the annual candles
Lit for those campers who did not return.
Layers of needle-filled shadows supplied sanctuaries
In which those wispy ghosts, hungry for life,
Could circle above the community of campers in love with life.

We could not usually guess
Who would not return
But I knew Santos would die.

Santos had a stronger smile
Than all the other campers' smiles.
Santos of the single name,
Son of hippies living in a trailer in the foothills.

Santos, an eight-year-old light-brown-skinned skeleton,
Too sick to splash for long; let alone to swim in the pool;
Shivering in the sunlight on the concrete bench;
Tiny towel wrapped around smooth thin shoulders
Smiling at his new friends in the water.

Skimpy towel, skimpy medical care, a family with skimpy food,
Could not stop his smiling.
My thick beach towel wrapped above his
Could not stop his shaking.

PHOTO BY STEPHEN BOYER

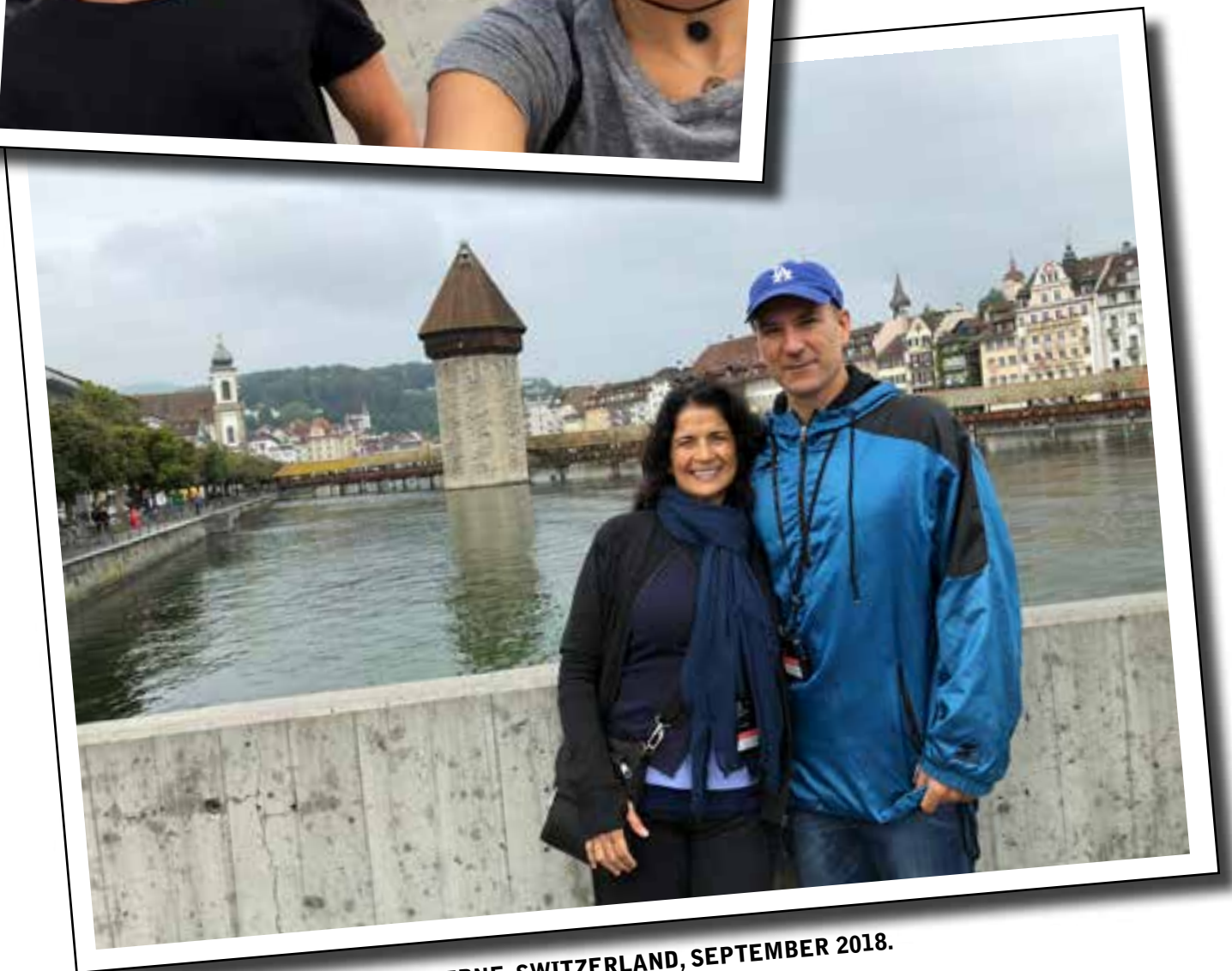
CF camp was a beacon for all to return each year,
Now the camp itself has also died.

I picture myself with a thick beach towel,
Holding it open and empty
At the height of a child
Releasing Santos' memory
Into the trees.

- M. Delano, M.D., 2003

FROM OUR FAMILY PHOTO ALBUM...

JESSIKA BIELE AND PAUL FERRARO AT THE LOUVRE IN PARIS.



JEANIE & JOHN HANLEY IN LUCERNE, SWITZERLAND, SEPTEMBER 2018.

RACHEL STEINMAN AND RAIDEL LAZCANO ON THEIR WEDDING DAY, JUNE 2018.

ELLA BALASA WITH GELATO IN HAND ON A BRIDGE OVER A CANAL IN VENICE, ITALY, JULY 2018.

LAURA MENTCH

FROM OUR FAMILY PHOTO ALBUM CONTINUED...

DAVID TARNOW (LEFT) WITH HIS TWO COUSINS LEXINGTON HEILMANN AND ERIC TARNOW. THE PHOTO WAS TAKEN AT LEXINGTON'S WEDDING ON THE LAKESHORE, IN CHICAGO, IL.

JOHANNA THILL AND ROBERTO J. DE LA NOVAL LATIN DANCING IN SOUTH BEND LATIN DANCE WEDNESDAYS.

ISA STENZEL BYRNES WEARING HER MEDALS AT THE TRANSPLANT GAMES OF AMERICA IN SALT LAKE CITY, UTAH, SUMMER 2018.

Autumn 2018 Recipients Of The USACFA Lauren Melissa Kelly Scholarship Announced

The US Adult CF Association (USACFA) is pleased to announce the recipients of the Lauren Melissa Kelly Scholarship.

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

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

We are pleased to announce **Roberto De La Noval** and **Julia Ruggirello** as the recipients of the scholarships for this semester. Congratulations to them! They will be awarded $2,500 each.

Roberto De La Noval is a motivated young man who is completing his Ph.D. in Theology at the University of Notre Dame. In the span of his graduate studies he has published three peer-reviewed articles in professional academic journals, and had six papers accepted at academic conferences, only one of which he was unable to present due to CF complications. With his Cuban heritage and impressive command of multiple languages, he helped take a group of undergraduate students

ROBERTO J. DE LA NOVAL

on a class trip to Havana, Cuba, where he served as a translator for them. He has raised over $4,000 for CF research and has given talks on behalf of his local CF center. Through his scholarly achievements and community involvement, his ultimate goal is to serve both the academy and the broader public as a professor and a public intellectual. When he is not working on his degree or writing for periodicals, he enjoys Latin dancing, playing chess and learning languages. To read Roberto's article turn to page 1.

Julia Ruggirello is an accomplished young woman pursuing her master's degree in Educational Administration at Michigan State University. She is building upon her leadership skills in hopes of becoming an administrator within the realm of education who would work on improvement and optimization of policies regarding inclusion for students with special needs. She currently enjoys her role as a special education teacher in Grosse Pointe, Michigan, as well as being a proud new homeowner. During her undergraduate five-year program, which she completed in four years, she was on the President's

JULIA RUGGIRELLO

Advisory Committee for Disability Issues at Michigan State, along with being an advocate for the CF community where she raised $30,000 at her first speaking engagement.

Both Roberto De La Noval and Julia Ruggirello demonstrated the leadership, intelligence and drive of Lauren Melissa Kelly. We at USACFA look forward to seeing them further develop their leadership and advocacy in the cystic fibrosis community.

We are happy to announce more scholarship opportunities coming soon! Please stay tuned for more information. For questions, please contact us at scholarships@usacfa.org. ▲

The CFRI National CF Family Education Conference

Report by Laura Mentch

Connecting The Dots: Bridging The Gap Between Isolation And Social Connection

By Chelsea Toth, DSW, MSW, LSW

CHELSEA TOTH

Dr. Chelsea Toth, a pediatric social worker, shared her research about isolation and social connection in the CF community through the experience and voices of young adults with CF, ages 18-25.

The familiar feelings of living with an invisible illness was shared by many young adults. The desire to feel normal, fit in and be accepted is challenged by the responsibilities of living with CF. This invisibility fades when we stand out as being sick, by wearing a mask or coughing in public, and when activities with our friends are postponed or cancelled by the need to do treatments or be in the hospital. Many young adults shared the loneliness of living with a disease that often means being isolated from others.

"It would be nice, though, to have a CF friend that you could hang out with because they would understand."

This wish of having a friend who can relate to the experience of living with CF was echoed by young adults interviewed for Dr. Toth's research. Not feeling isolated and having a social connection with your peers supports mental health, yet we seek to avoid cross-infection by limiting personal contact in the CF community.

> "The desire to feel normal, fit in and be accepted is challenged by the responsibilities of living with CF."

While listening to Chelsea Toth describe the experiences of feeling separate or different from others because of CF, I felt fortunate. I have, for another year, passed the sputum test and agreed to adhere to the CFRI Infection Control Policy, so that I may participate in this conference with several of my peers, adults living with cystic fibrosis.

How do you overcome the barriers to intimacy and create community while observing cross-infection guidelines?

At my first CF clinic visit, after my late diagnosis, I asked about support groups for adults with CF. A newbie, I did not yet understand there would be few, if any, adults with CF in my community and that guidelines restricting our contact with one another had recently been released by the Cystic Fibrosis Foundation. After a few years I found *CF Roundtable* and a CF chat group. The wisdom and knowledge shared by others helped me at the beginning of my journey of living with CF.

Technology brings opportunities to create community with attention to infection control precautions. New initiatives for those living with CF include:

- Virtual education events and social media groups given by CF centers for patients and their families.
- The Cystic Fibrosis Foundation has established CF Peer Connect, a friendship and mentoring program, and BreatheCon, a virtual discussion event guided by adults with CF.
- Cystic Fibrosis Research Institute (CFRI) offers support and education for adults and families living with CF that include support groups led by a social worker, programs on mindfulness-based stress reduction, physical therapy and yoga for CF, the annual family education conference and retreats for families and adults with CF.

These resources are designed to address the concerns brought forward during Chelsea Toth's research. Take-away messages from her presentation include: 1) people with CF have unique experiences and perceptions, 2) it is important to identify barriers to isolation and discuss the importance of connection and 3) encouragement to develop networks for individuals living with CF and their families. Together we can begin to address the gap between social isolation and connection expressed by young adults with CF. ▲

Laura is 65 and has CF. She is a Director of USACFA. Her contact information is on page 2.

A Report From The 2018 CFRI National CF Family Education Conference

By Jeanie Hanley

The 32nd CFRI National CF Family Education Conference took place in Redwood City, California, on the weekend of August 3, 4 and 5, 2018. This year the conference opened up with an outstanding talk by our very own USACFA Director Reid D'Amico.

Reid just recently received his Ph.D. and he shared his young (he's only 25!) and many life experiences during a fun-filled and fascinating weekend of many such inspirational speakers and sessions. There was never a dull moment and the CFRI folks (shout out to Siri Vaeth Dunn and Mary Convento!) change it up every year so it's always a new experience.

Many new concepts and research advances were introduced. Since all sessions can be seen online at www.cfri.org (via YouTube) you can watch them in the comfort of your home. Reid's presentation provided an excellent framework for the rest of the conference just as his title suggested: "Science, Advocacy and Life with CF." That just about encompasses the crux of this conference.

Reid's thought-provoking presentation (a must-watch!) created discussions of the changes I, and many others with CF, have experienced as the years fly by. Many of these changes are noted above. While there are ups and downs, I've found most downs are accompanied by an upside (making lemons out of lemonade!) and sometimes vice versa. For example, I've often gone to clinic nearly certain that my lung function is holding steady only to find that it has declined. Other times I've felt that my lung function must be lower, but it was not changed or if I was fortunate, it even had improved.

Here are a couple of last notes about the CFRI conference. Although our *CF Roundtable* newsletter is focused on adults, I must say that I enjoyed seeing parents who have children with CF present there. Many were newcomers to the conference. They saw firsthand, and possibly for the first time, adults with CF who are thriving in life and active members of the CF community. Also, the organizers stressed infection prevention (how could they not with the daily reminders of infection control precautions and practices!) and how not to instill fear of others and of life in their children. Instead, emphasis was on helping these parents and their children to be active and engaged, yet practice healthy and practical precautions.

Some adults with CF wore masks at the conference, which is an extra precaution that I applaud. Only elbow bumps were allowed to greet others. It would be wise for people, in general, to greet each other in the same way rather than shaking hands (a very automatic, but unsanitary practice). It also wouldn't hurt to wear masks at gatherings when anyone is actively sniffling, sneezy or coughing; or better yet, not even attend. Fortunately, I didn't have any setbacks after the conference. The organizers certainly helped to encourage good hygiene practices with many bottles of hand sanitizer, gloves and masks available at the conference. I hope you plan to attend the conference next year. There is much to learn and the benefits are many. ▲

Jeanie is 56 and is a physician who has CF. She is a Director of USACFA and is the President. She lives in Los Angeles with her family. Her contact information can be found on page 2.

Pay It Forward

BRONZE

Anonymous
Michelle Allen
William Beck (in memory of Wendy Beck)
Doug Booten
Marie Henry (in honor of Anne Henry)
Barbara Jenkins (in honor of Steve Jenkins)
Ted Kowalski (in memory of Joseph Kowalski)
Adam Levy (in honor of USACFA Directors)
Virginia C. O'Brien (in memory of Ken O'Brien)
Richard Quinn
Arvey Stone, MD

SILVER

Leslie Sternlieb (in memory of Helen Eisenman)

GOLD

Phyllis Koscoff (in memory of Stephanie Koscoff)

SUSTAINING PARTNERS

Horizon Pharma USA, Inc.

WELLNESS

An Unintended Consequence Of Aging With CF: Survivor Guilt

By Julie Desch

I have probably waxed poetic in previous articles regarding the high incidence of depression and anxiety in our population. As a review, a recent (TIDES) study was a nine-country epidemiological study to screen both patients and parent caregivers for symptoms of depression and anxiety. Unsurprisingly, there were elevations in symptoms of both depression and anxiety. Elevated symptoms of depression were found in 10% of adolescents and 19% of adults with CF. Anxiety was also a significant issue; among adolescents it was about 20% and adults again were a little bit higher at about 30%. These rates are about two to three times what we would see in a normal (I hate that word) population.

There is no evidence that CF directly causes depression or anxiety. But it doesn't take great imagination to speculate on causes of this increased incidence. Living with an illness that can strike with volcanic force regardless of how diligent you are with treatments, rest, pills, exercise and nutrition could possibly (sarcasm) lead to a smidge of anxiety. I personally have been diagnosed with both depression and anxiety. Thankfully, both are relatively controlled, although recently I have had a bit of a resurgence of generalized anxiety. In fact, the article I was *going* to write was about this, and about how I am using mindfulness techniques to work with it.

But then Claire died.

For those of you not familiar with Claire Wineland, she was a 21-year-old YouTube phenomenon who used her social media platform to offer inspirational and, frankly, badass advice not just to those of us with CF, but to everyone. "Death is inevitable. Living a life we can be proud of is something we can control." Who doesn't need to hear that?

I didn't know Claire, but since I heard of her death, I can't stop watching her videos. Her energy, her humor, her beyond-her-years wisdom jumps out from the computer screen, making me literally sick to my stomach.

Twenty-one years is the age of my oldest son.

Of course, young people die of CF way too often. Claire is not the only one who was robbed of adulthood. But for some reason (perhaps her fame? her spirit? her obvious maturity and wisdom?) her death has hit me hard. I can't write about anxiety. I'm too sad.

But maybe there is a relationship between what I am feeling and our high incidence of both anxiety and depression. I feel grief of course. This is so unfair. Her family and friends don't deserve the grief they must be going through right now. Grief seems like a normal response.

JULIE DESCH, MD

But I noticed that very quickly this grief is supplanted by guilt. I have lived well over twice as long, and for some reason am still going strong. It's the same guilt I feel when I think of my sister who died at 31, or my brother who died when he was my current age. It's the same guilt I feel every time I see a Facebook post alerting us all that someone else had died of cystic fibrosis. It's relentless.

Survivor guilt is no joke. It's not a terribly rational response to loss, but then, emotion often is not rational. I'm fairly certain that none of those whom we have lost to CF would actually wish that people who are alive feel guilty. In my case, I think that guilt is an easier emotion to deal with than sadness, so that is where my mind goes. To me, guilt feels more manageable than grief. There is something I can "do" with guilt. I can try harder. I can get another certification. I can write articles. I can coach people or teach mindfulness. I can try to improve my chances of feeling "worthy" of this life.

Grief is a whole different animal. I have historically avoided feeling grief at all costs, because it just seemed too much to bear. When I was very young I hid in my room covering my ears when I heard my sister's body wracked with coughing. I even avoided attending to my brother when it was so clear that CF was devastating his body. I avoided CF camps. I spent a large portion of my life avoiding getting close to people who I knew might die of the disease that I carry in my genes. I feel ashamed admitting this, because so many people with CF that I now know and love have spent their lives forging close relationships with others with the disease. Their lives are richer for this

practice. I, on the other hand, lost a lot of time and a lot of potential friends.

There is much sadness to bear when living with CF. Indeed, sometimes it seems overwhelming. I know I'm not the only one who has been bingeing on Claire Wineland YouTube videos with tears streaming down my face. So what is different now? Why am I allowing myself to feel the grief instead of becoming more driven by guilt?

One of my mindfulness teachers likes to quote the Buddha:

> "Praise and blame
> gain and loss,
> pleasure and pain,
>
> fame and disrepute
> are the eight worldly winds.
> They ceaselessly change.
>
> As a mountain is
> unshaken by the wind,
> so the heart of a person
> is unmoved
> by all the changes on this earth."

Grief is part of life. It is the natural reaction to loss, just as happiness is the natural reaction when we get something that we really want (like that triple drug combination that Vertex is working so hard on as I write this). Going through life only willing to be with pleasure gain, pleasure and fame and not with blame, loss, pain and disrepute leads to a life half-lived. And here is where depression and anxiety fit in. When you spend energy suppressing unpleasant emotions, such as grief, depression and anxiety can be the unwelcome side effects. Imagine that you are a mother trying to ignore her crying baby in another room. You can put on headphones and blast rap music all you want, but you still know what's happening nearby. Immediately you feel anxious, but over time this can easily transform into depression.

"When you spend energy suppressing unpleasant emotions, such as grief, depression and anxiety can be the unwelcome side effects."

As I've gotten old(er), my meditation practice has taught me the good news/bad news aspect of life's changing weather. Everything, and I mean *everything*, is constantly in flux. Of course we know this rationally, but in meditation you come to know it as direct experience. The good news is that this means that pain, loss, blame and disrepute don't last. They eventually change. Pain either resolves or changes into something more or less manageable. Loss is grieved and fades into fond and loving memories. Blame is forgotten or forgiven. Disrepute even dissolves as amends are made or the truth becomes known.

Of course, the flip side is also true. Praise is blotted out by a single negative commentary. Gains are eventually lost (entropy always wins). Pleasures dull over time as we accommodate to them. And fame, as they say, lasts about 15 minutes.

Knowing the truth of this undeniable fact, that everything is impermanent, makes it easier to stay with something as uncomfortable as grief and allow it to make its mark, to teach its lesson and to blow away making room for what is next.

There is a quote from Geshe Kelsang Gyatso that comes to mind as we sit between the grief of ever-present losses and the excitement/anticipation of new therapies, "Endure both, whichever arises." Taking this to heart, I can bear the grief (and guilt) that arises when heartbreaking news flashes across my computer screen and then later enjoy the beautiful day as my lungs carry me up the steep hill to walk my dog in the open space above my house. As Claire put it so well, "*Go enjoy your life. Really. I mean that seriously. Go enjoy it. 'Cause there are people fighting like hell for it.*" ▲

Julie is 56 and is a physician who has CF. You may reach her at jdesch@usacfa.org.

USACFA Speakers Bureau

The U.S. Adult CF Association, Inc. (USACFA), now has speakers who will come to speak at fundraisers, education days and other CF-related events. All the speakers are adults who have CF and can speak with experience on living with CF and what is happening in the CF world. USACFA has budgeted for travel and lodging costs for the speakers, so there is no cost to the hosting organization.

If your organization is interested in having a CF speaker present at your event, please contact rdamico@usacfa.org

SAVORING SERENDIPITY

Bayou Blessings: The Pace Of Grace

NEW COLUMN

By David Tarnow

It was a hot, delicious and superstitious day in February on St. Charles Street in New Orleans. The year was 2009. Purple, green and gold shiny colored beads, doubloons and trinkets of all shapes, sizes and kind littered the street. We had had our fair share of celebrating during the preceding day, so when it came time to leave, I did not object.

The spirit of the holiday, called "Carnival" in some parts of the world and "Mardi Gras" in South Louisiana, can be summarized by the Cajun French expression: "Laissez les bon temps rouler," which in English translates to, "Let the good times roll." By tradition, the following day is one of stark contrast to Fat Tuesday whereby participants and members of the Roman Catholic Church ritually pledge to realign their focus from that of debauchery and excess (in the days prior) to one of solemn remembrance and austerity at the start of the Lenten season.

The sun, high in the sky, was shimmering down through the magnolia leaves onto the street filled with crowds of people pursuing the limit of hedonism. A cacophony of sounds came from all directions as the floats and marching bands paraded down the street. My tired body longed for rest and I was actually looking forward to returning to school and work the following day.

As I wished farewell to the rest of the group, there came, out of nowhere, a loud terrifying series of bangs similar to that of fireworks on the fourth of July. These sounds were, in fact, bullets flying through the crowd at blistering speed. My sense of awareness heightened as my adrenal glands pumped massive amounts of adrenaline into my bloodstream. My perception of time slowed to a crawl. Matrix time, for those of you who have seen the film, is real.

In the midst of the panic and pandemonium, to avoid being shot ourselves and following the crowd away from the source of gunfire, my girlfriend and I took cover behind a large ice chest left abandoned at a tailgate which seconds earlier was filled with people, music and merriment. Before I was able to process the situation and after the gun shots had stopped, I glanced down at my body to see if I had been hit.

To my surprise, I had been shot in the abdomen as evidenced by the hole in my t-shirt and the sintered and bloody wound beneath it. I had not even felt the bullet penetrate through my body. As I looked around me to get my bearings, I noticed another man who had seemingly been shot also, recoiled in pain, lying down in the middle of the street. That's when it hit me. I've been shot, and I have a choice to make right now. I can either panic and let my consciousness succumb to a state of shock (which would have been productive for no one) or relax and trust that I would be taken care of.

> *"For a period of time after being shot, I was able to find peace in the simple, dreadful, yet beautiful fact that I was still alive. I did not die on that day."*

DAVID TARNOW

My girlfriend, thankfully unscathed by the incident, trembled in shock as she looked at her cell phone and struggled to recall and subsequently dial the number 911. Fortunately, there were first responders nearby who swiftly came to my aid. I was carried and laid down on the front porch of a private residence just off of St. Charles Street.

A group of women opened the front door to see what all of the commotion was about. As serendipity would have it, it was quickly made known that I was lying on the front porch of a surgeon's house (where a Mardi Gras party was in progress), and many of the women who came to my aid were nurses.

> *Through the practice of mindfulness, I experienced the transformational power of presence and gratitude in daily living.*

As I lay there on the porch, God and I had a private moment while I gazed up and into the blue abyss. I was surprisingly calm. Before I knew it, I was moved from the porch onto a stretcher and put in an ambulance. The sirens were deafening and everything seemed so surreal as I attempted to process the present moment. Next to me was the young man whom I had seen lying in the street just after the gunmen ceased fire. Kneeling between us was an EMT holding pressure on both of our wounds. I recall the guttural agony in the young man's voice as he transcribed unto the airwaves the magnitude of his pain.

When we arrived at the hospital, known for its high performing trauma center, I was stripped of all of my clothes, poked, prodded and questioned to assess my state of conscious awareness. I later came to find out that the woman questioning me was the same woman from whom my mom received the call saying that I had been shot. Needless to say, I know my mother must have felt like dying at that moment herself. After it was determined that there was no exit wound, I was rolled away to imaging to assess the position and location of the bullet and whether or not surgical intervention was necessary.

To my relief, the radiologist was quick to review the imaging and quick to snap me back to reality by telling me just how fortunate I was to be alive. There was no necessity to remove the bullet, however, as its location was benign and would become encapsulated with tissue over time anyway. Attempting to remove it carried the potential risk of doing more harm than good.

As I waited to be discharged several hours later, the last thing on my mind was my cell phone. However, by the time that my girlfriend and her mother were allowed to leave the waiting room and come see me, word of what had happened had quickly spread to friends and family. I was in disbelief at just how many people were genuinely concerned for my well-being and had attempted to contact me as a result. You never know just how loved you are until you are dwelling on the threshold of life and death.

Why do I share this story with you, you may ask? I share it because whether I realize it or not, getting shot on that fateful February day not only wounded my body but my spirit just the same. One of the great Mystics, Rumi, once said that, "The wound is the place where the Light enters you." Boy, oh boy, was he right.

My professors and employer were gracious enough to give me autonomy in determining when I would be strong enough to return to school and work. In the following weeks and months, I undertook an earnest pursuit of healing and truth. I began to meditate. I took comfort in and basked in the wonder of nature all around me at any given moment and the gravitas of the gift that I had been granted in the arena of life. I was given a second chance to play. The game was still going whether I knew what for or not, so I was going to take advantage of the opportunity.

For a period of time after being shot, I was able to find peace in the simple, dreadful, yet beautiful fact that I was still alive. I did not die on that day. As I write this, it seems serendipitous that after such a traumatic incident I was able to carry on and do so well, nonetheless. I was in a magnanimous trance that was critical to my emotional ability to make peace with my survival.

In time, this sense of magnanimity faded as other significant life events took place that scarred my spirit. I was still standing though. Better vertical than horizontal.

Through the practice of mindfulness, I experienced the transformational power of presence and gratitude in daily living. How could I ever be in want if I am grateful for what I have? This seemed to me as somewhat of a philosophical paradox. More and more, I began to recognize and appreciate the opportunity to create my own joy and happiness. For me, this observation transformed into the most beautiful of revelations — freedom to choose whether I would continue to suffer, and be bound by fear, or live life with expectancy rather than expectation.

We all are at liberty to choose an overarching worldview of joy rooted in gratitude and expectancy. Often, if not always, as is the case when living with CF, my experience is that life can and will in time (as a consequence of the progressive, genetic disease for which there is no cure) lead toward more suffering. We (CFers) got the short straws in the genetic lottery; there's no rational doubt about it. This, however, is where the irrational comes into play. It may seem a little odd to take joy in suffering, but my experience of living is that reality is perspective and perspective is reality. You simply cannot come to know the highs without also knowing the lows in this life. That is to say, we all, ultimately, have a choice between joy and suffering, and it can make a

Continued on page 30

TARNOW continued from page 29

world (pun intended) of difference in one's experience of the gift of life, henceforth, quality of life.

It is for this reason, that I am passionate about savoring serendipity. One ounce of gratitude can go a long way in experiencing a more joy-filled life. Serendipity, by definition, is the faculty or phenomenon of finding valuable or agreeable things not sought for. Sounds like gratitude to me.

What great fortune we have before us then to be alive and to be presented once again with a new day. Another day in which I know that I will do my best in choosing to savor serendipity rather than to suffer in suffering. I encourage you all to try it out for yourself and make of it what you will. We all are unique. We all are grateful for different things. Let us be grateful, then, that by the singular nature of our existence (the phenomenon of being alive) we may all suffer less and savor more.

As our bodies waste, our spirits strengthen. Energy is neither created nor destroyed, only transferred. It is, therefore, the miracle of living, the ability to choose our own reality.

I hope you all join me and continue to subscribe to *CF Roundtable* and encourage others in the CF community to do so as well. There is strength in numbers. We are warriors with a cause, and our cause is living. With each insufferable breath, with each serendipitous inspiration, let us as a CF community stand to empower and encourage others toward the pursuit of joy against all odds.

In Love and Light,

Dave ▲

David is 30 and has CF. He was born in the Rocky Mountains and lived in various places throughout his adolescent life including Louisiana. He has currently come full circle and is back in his native state of Colorado. He graduated with a BS from LSU. He continues to relish the pursuit of self-actualization by taking roads less traveled. You may contact him at: dtarnow@usacfa.org.

TILLMAN continued from page 18

extent, the function of secretory cells. After the pancreatectomy, the clinical team collects healthy cells from the harvested organ and infuses them into the patient's liver. Implanted in the liver, these cells are able to survive and continue to produce the necessary enzymes and hormones as they do in the pancreas, namely insulin. With this cell transplant approach, it becomes possible to achieve permanent pain relief, while it also can help prevent the development of brittle diabetes.
http://tinyurl.com/y7jfkt5k

In CF Patients, Addressing Mental Health Key, Experts Say

Anxiety and depression among cystic fibrosis (CF) patients is far more common than most people realize. The prevalence of depression ranges from eight to 29 percent among children and adolescents with CF, and from 13 to 33 percent among adult CF patients. There is now a focus on efforts to implement screening programs among CF patients worldwide.
http://tinyurl.com/ycmv98je

Cinnamon Oil Compound Might Block Bacteria Like P. aeruginosa From Forming Biofilms, Researchers Say

A natural component found in cinnamon oil, known as cinnamaldehyde or CAD, may be able to prevent Pseudomonas aeruginosa bacteria from spreading in an organism and inhibit their ability to form antibiotic-resistant biofilms. Cinnamaldehyde, one of the major components of cinnamon oil, is responsible for its characteristic flavor. This compound is known to have antimicrobial activity against many bacteria, including P. aeruginosa. The research team conducted several experiments to evaluate the impact of different concentrations of cinnamaldehyde on P. aeruginosa biofilms. They found that nonlethal amounts of the essential oil compound could disrupt by 75.6% antibiotic-resistant, preformed P. aeruginosa biofilms. Cinnamaldehyde was found to prevent the production of a bacterial-signaling protein essential for bacteria communication and biofilm formation. In a concentration-dependent manner, cinnamaldehyde also could reduce the motility of the bacteria, preventing them from spreading elsewhere.
http://tinyurl.com/ybfo4h5v

MRSA Difficult To Eradicate, Even With Multimodal Antibiotics, CF Study Finds

The prevalence of MRSA in CF patients has increased in the past two decades, and it is estimated that currently 26 percent of CF patients in the U.S. are infected with MRSA. Staphylococcus aureus is the most prevalent bacteria in the airways of CF patients. MRSA is a particular challenge in CF due to its resistance to standard antibiotics used in the clinic. Persistent MRSA infection is associated with worse outcomes in CF; 43 percent of the patients had a faster decline in lung function, which led to a lower survival, relative to CF patients without MRSA or to those in whom the infection was eradicated. Previous studies suggested that aggressive antibiotic therapy is a potential approach to eradicate MRSA in CF patients, although these studies lacked appropriate controls. Thus, researchers performed a randomized, double-blind, placebo-controlled Phase

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

Michelle Allen
Portland, OR
67 on September 14, 2018

Beth Sufian
Houston, TX
53 on August 13, 2018

Linda Stratton
Louisville, CO
64 on July 23, 2018

Kathryn Yoder
Portland, OR
56 on August 16, 2018

Wedding

Michelle and **Gary Allen**
Portland, OR
22 years on April 27, 2018

Jeanie and **John Hanley**
Los Angeles, CA
32 years on June 28, 2018

Beth Sufian and **James Passamano**
Houston, TX
30 years on July 2, 2018

NEW BEGINNINGS

Wedding

Rachel Stein and **Raidel Lazcano**
Married in New York, NY
On June 21, 2018

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2 trial (NCT01594827; the Persistent MRSA Eradication Protocol, or PMEP) to evaluate the safety and effectiveness of a 28-day eradication strategy — a combination of oral and topical antibiotics, with or without vancomycin — in CF patients with persistent MRSA infection. The study's primary objective was to assess the percentage of patients who became MRSA-free one month after completion of treatment. Additional goals included changes in lung function from the beginning of the study up to the three months of follow-up, and changes on the Cystic Fibrosis Questionnaire-Revised (CFQR) focusing on the respiratory score. At the end of the 28-day treatment, 50% of patients treated with inhaled vancomycin plus antibiotics were negative for MRSA, compared to 40% in the placebo-antibiotics group. One month after treatment completion, there was no difference between the two groups. Three months after completing treatment, there was still no statistically significant difference in MRSA eradication rate between the two groups. Additionally, there was no difference in lung function and in the CFQ-R questionnaire between the groups at any time point analyzed. Overall, the study supports how persistent MRSA infection is difficult to eradicate, even with multimodal antibiotics.
http://tinyurl.com/ydgtxruo

Telavancin Promising Potential Treatment Option For MRSA In Cystic Fibrosis Patients

Investigators have found that telavancin (TLV)—a drug that is currently used to treat skin infections and hospital-acquired pneumonia—has potent in vitro activity and low resistance development potential when used against S. aureus isolates in patients with cystic fibrosis, making it a promising potential treatment option. The investigators screened a total of 333 strains of CF patient-derived S. aureus of the wild-type or small-colony-variant phenotype, collected from both adults and children at three different cystic fibrosis centers: TLV was found to display activity against all 333 strains collected.
http://tinyurl.com/y8cec8g5

Early Diagnosis, Treatment Of NTM Infections Important In CF, Study Finds

An earlier diagnosis and treatment of nontuberculous mycobacteria (NTM) infection in patients with cystic fibrosis (CF) may positively affect the patient's lung function. Researchers analyzed the prevalence of NTM infections in CF patients to identify factors associated with these infections, as well as monitored current treatments. They determined that an earlier CF diagnosis was associated with a higher isolation of slow-growing NTM and greater antimicrobial use after infection and that NTM acquisition is associated with a worsening of lung function. Thus, both the early diagnosis and treatment of an

Continued on page 32

NTM infection in patients with CF may positively impact lung function.
http://tinyurl.com/yd8vvqs2

Study Finds Bacteria Adapted To The Lungs Are Easier To Kill With Antibiotics

A new study finds evidence that as bacteria adapt to the human body, they can sometimes become more susceptible to antibiotics and therefore easier to kill. Using models of long-term lung infection, the researchers identified mutations that appeared in the bacterial DNA over the course of infection and which led to changes in the ability of Pseudomonas to survive in the lungs. The mutations enabled the bacteria to attach to lung cells more effectively and to resist defense molecules produced by the host immune system. The same changes also made Pseudomonas more susceptible to antibiotics, raising hopes that even bacteria that are well adapted to the lung environment could be combatted with conventional antibiotics. The new study sheds new light on how bacterial pathogens can change over the course of a single infection within a patient. The mutations identified in Pseudomonas in this study were important in allowing the bacteria to resist an anti-bacterial enzyme of the host immune system called lysozyme. However, bacteria carrying the mutation were very readily killed by antibiotics. The team hopes the findings may lead to better diagnosis and treatment of Pseudomonas aeruginosa infection.
http://tinyurl.com/ya6jds34

CF Patients Have More Harmful Gut Bacteria Than Healthy People

Cystic fibrosis (CF) patients have a higher amount of harmful gut bacteria and increased levels of intestinal inflammation than healthy people. CF predominantly affects the lungs, but it can also cause gastrointestinal complications. The CFTR protein defect is abundant in the gastrointestinal tract of patients and affects the normal structure of the intestine. This defect could influence the diversity of the bacteria present in the gut (gut microbiome).

Previous studies have shown that CF patients have a less diverse gut microbiome than healthy individuals. Now, researchers investigated the impact of antibiotic use on gut inflammation and bacterial composition in CF patients. Researchers noted a significant decrease in the Bacteroides, Eubacterium rectale, Faecalibacterium prausnitzii and Firmicutes bacteria in CF patients compared to healthy controls. These bacteria are usually considered markers of a healthy intestinal microbiome. Clostridium difficile and Escherichia coli were found in markedly higher numbers in CF patients. Similarly, a high number of Pseudomonas aeruginosa was also detected in CF patients. A similar change in microbiome diversity was observed in all CF patients regardless of antibiotic use. Although frequent use of antibiotics did not appear to influence the gut microbiome in CF, there was a noticeable shift in the gut population from beneficial to harmful bacteria.
http://tinyurl.com/ybehxzgv

Technique May Improve Lung Delivery Of Bacteria-Killing Phage

A new delivery system for bacteriophages—viruses that selectively attack harmful bacteria—could help give doctors a new way to battle lung infections. Phage therapy is a promising alternative to antibiotics because it attacks specific pathogens, does not harm the body's normal contingent of bacteria and won't contribute to multi-drug resistance. However, therapeutic bacteriophages can be difficult to purify and challenging to deliver to the site of an infection, especially when that location is the lungs. A research team has demonstrated a new delivery technique that uses dry, porous microparticles coated with phages. In animal testing, the phage-coated polymer particles successfully treated pneumonia in infected mice and dramatically reduced bacterial levels in an animal model of cystic fibrosis. The technique might one day allow delivery of the dry-powder phage using a device similar to a common inhaler. Specific bacteriophages can target bacteria such as Pseudomonas aeruginosa without affecting other bacteria. Phage activity propagates beyond the coated particles, but is limited by the host population, so once the targeted bacteria are eliminated, the phage disappears. The phage-coated microparticles were more effective at clearing bacteria than dried phage particles by themselves. The polymer material is biodegradable and was cleared from the animals within a few days. The technique was successful in attacking different strains of bacteria within biofilms, and the researchers did not see evidence that the bacteria were developing resistance.
http://tinyurl.com/y7hb9p8u

AND

http://tinyurl.com/y8zn8pg9

Stem Cell Research For Cystic Fibrosis Leaps Forward

The research, which applies stem cell transplantation, involves harvesting adult stem cells from the lungs of CF patients, correcting them with gene therapy and then reintroducing those cells back into the patient. The new transplanted adult stem cells pass on their healthy genes to their "daughter cells" providing a constant means to replenish the airways with healthy cells and thereby combatting the onset of cystic fibrosis airway disease. The key to these successful transplantations was an innovative method: the existing surface cells were first eliminated, which then created the space required to introduce the new cells. This pioneering research demonstrates that, in principle, human airway stem cells can be transplanted into the lining of the lungs.
http://tinyurl.com/yb8oa6d3

Giving Tuesday Is Almost Here!

Mark your calendars for **Tuesday, November 27th**! The holidays are upon us. Please consider donating to United States Adult Cystic Fibrosis Association (USACFA) and *CF Roundtable* on Giving Tuesday, a national day of giving and our last fundraising campaign for the year.

Your past contributions have been essential in helping those with cystic fibrosis find support, medical information and resources through *CF Roundtable.*

Because of your donations to USACFA:

- All of our readers receive *CF Roundtable* free of charge and have access to the latest research, legal and critical knowledge that has maximized their medical care.
- We can support scholarships for students with CF striving for higher education.
- We can provide presenters with CF via our Speakers Bureau to travel to your special events and speak about CF.
- We can continue all of this and more!

USACFA is run by 14 adults who have CF, an amazing board of directors. Our time is 100% voluntary. Together, we create the *CF Roundtable* publication, website and numerous programs. We do this for you – the vital CF community.

We hope we can count on your support this year. Ultimately, your gift would keep *CF Roundtable* and the miracles coming! We hope to hear from you soon. Please contact us anytime at cfroundtable@usacfa.org or go to our website at www.cfroundtable.com and donate!

Thank you and Happy Holidays!

The Board of Directors
United States Adult Cystic Fibrosis Association (USACFA)
Publishers of *CF Roundtable* ▲

Newfound Airway Cells May Breathe Life Into Tackling Cystic Fibrosis

The newly discovered cell, ionocyte, may be the star of future cystic fibrosis therapies. Researchers have found that the gene tied to the disease is very active in the cells that line the air passages of the lungs. While the cells are rare, making up only one to two percent of cells that line the airways, they seem to play an outsized role in keeping lungs clear. The identification of the ionocyte provides key information for targeting treatments as it regulates fluid movement at surfaces where air and water meet. Researchers had suspected CFTR was most active in ciliated cells. But the new work found very little gene activity in those cells, compared with the ionocytes.

http://tinyurl.com/y7d43aug

AND

http://tinyurl.com/y889e3lc

AND

http://tinyurl.com/ybz6lclz

Celtaxsys Announces Results Of Phase 2 Trial Showing Clinically Meaningful Improvement In Pulmonary Exacerbations In Cystic Fibrosis Patients

Acebilustat demonstrated clinically meaningful improvements in pulmonary exacerbations, both reducing the frequency of pulmonary exacerbations (PEx) and increasing time to next exacerbation over 48 weeks of therapy. Patients in key prospectively identified sub-groups, including those with mild obstruction at baseline or taking CFTR modulator therapy, derived the most benefit in pulmonary exacerbations. That benefit, when used in combination with a CFTR modulator, is an important consideration given the likelihood of an increase in the number of CF patients who are eligible to be treated with new CFTR modulators over the coming years. This supports the unmet need to address lung inflammation adequately for the optimal treatment of patients with cystic fibrosis. On a per protocol assessment, acebilustat-treated patients exhibited a 19% reduction in PEx and a 22% reduced risk in progressing to first PEx versus placebo. Additionally, over 40% of patients treated with acebilustat completed the study without experiencing a PEx, an increase of 32% as compared with patients treated with placebo.

The benefits of acebilustat on pulmonary exacerbations were apparent as early as four months after start of treatment and persisted throughout the 48 weeks of the study. No difference in lung function, as measured by the primary endpoint of FEV_1 percent predicted (FEV_1pp), was observed in acebilustat-treated patients compared to pla-

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cebo-treated patients over 48 weeks of treatment. Additionally, FEV_1pp response did not correlate with PEx rates. Patients with less severe impairment of lung function (FEV_1pp >75) achieved the largest benefit from acebilustat treatment, achieving a 34% reduction in PEx rate, a 43% reduction in risk of experiencing their first exacerbation and a 96% increased likelihood of being exacerbation-free after 48 weeks of treatment. Furthermore, patients concomitantly treated with CFTR modulator therapy exhibited a clinically meaningful 20% reduction in PEx, a 29% increased time to first exacerbation and a 47% higher likelihood of no exacerbations compared to patients treated with CFTR modulators and placebo. Acebilustat was well tolerated with no increased risk of infection.

http://tinyurl.com/y8tgax2a
AND
http://tinyurl.com/ybbubfgd
AND
http://tinyurl.com/ybnjdzk3
AND
http://tinyurl.com/ybs8fsd4

Does Cystic Fibrosis Increase The Risk Of Gastrointestinal Cancer?

The defective protein that is coded by the CFTR does not allow for the proper transport of chloride ions across epithelial cells that line our respiratory tracts. This disrupts the ion balance across the cell causing defects in the transport of water and sodium. A disrupted transport of water across the respiratory tissues increases the thickness of the mucus fluid that bathes the interior of our lungs. This leads to severe obstruction in the respiratory airway and causes breathing difficulties and chronic respiratory distress.

In a recent study researchers wanted to determine the probability of gastrointestinal cancer in patients who have cystic fibrosis. To do so, they conducted a meta-analysis of previous studies of intestinal cancer. Overall they found that patients with cystic fibrosis have a statistically significant higher risk of developing gastrointestinal cancer in comparison to the general population. The most common cancers associated with cystic fibrosis included cancers of bowel, colon, biliary tract and pancreas. The authors suggest that regular gastrointestinal and colorectal screening of cystic fibrosis patients using endoscopic methods should be done to facilitate the early detection of cancers that arise in the affected population.

http://tinyurl.com/ybqkebcd

Airway Clearance Vests Fail To Show Measurable Short-term Lung Benefits In Study

A clinical study into high-frequency chest wall oscillation vests — assessing their short-term impact on standard measures of lung function before and during use — challenges the view that these devices work through airflow bias in the lungs, the process responsible for mucus movement when breathing. Findings, using established tests that include forced vital capacity (FVC), forced expiratory volume (FEV_1) and forced expiratory flow (FEF25%-75%), determined that none of the vest groups showed statistically significant increased airflow in the lungs. Declines in lung function were actually recorded but were transient.

http://tinyurl.com/ycwwvvr6

CF Patients At Higher Risk For Retinal Vein Occlusions, Case Report Says

Patients with cystic fibrosis (CF) have a greater risk of developing abnormalities in their retinal veins, which can impair visual acuity. Researchers claim the higher risk is not due to one single factor, but to several contributing risk factors for vascular events. Ocular symptoms are common in cystic fibrosis patients, usually as a result of vitamin A deficiency. They usually appear as an irritation in the eye, night-blindness or optic nerve abnormalities. Recent studies, however, have described two cases of CF patients who developed occlusions in their retinal veins.

In these studies, one patient had high fibrinogen levels and the other had increased homocysteine, leading researchers to speculate that those were the primary causes for the occlusions. But a third case has challenged this hypothesis, suggesting retinal vein occlusions in these patients are multifactorial and not due to a single risk factor. Patients with cystic fibrosis are at an increased risk of developing retinal vein occlusions likely due to a variety of systemic thrombogenic factors [that support formation of blood clots] rather than a single risk factor.

http://tinyurl.com/ybphlzpm

Predictors For Survival Rates For Adult-Diagnosed Cystic Fibrosis

Researchers of this study examined demographic and clinical data on patients diagnosed with CF as adults to create a guideline of characteristics for predicting outcomes and survival rates of this population. Clinical data included symptoms, genotype, chronic conditions, lung function and body mass index. The researchers concluded that survival rates of individuals who receive diagnoses of CF as adults can be predicted by factors that include their age at diagnosis, lung function and the presence of diabetes. Younger patients with higher lung function had more years of survival, and those without diabetes were less likely to require lung transplants.

http://tinyurl.com/yachnvyd

Mutations In Genes Regulating Digestion Prevalent In CF Patients With Pancreatitis

In addition to mutations in the CFTR gene, cystic fibrosis (CF) patients with pancreatitis also have a high prevalence of mutations in genes regulating pancreatic function, according to researchers. Pancreatic insufficiency

An Invitation To Participate In The Cystic Fibrosis Reproductive And Sexual Health Collaborative (CFReSHC)

The Cystic Fibrosis Reproductive and Sexual Health Collaborative (CFReSHC) is a patient engagement project that provides women with CF the opportunity to be equal partners with healthcare professionals to shape the future of CF research. CFReSHC offers women with CF the opportunity to participate on the Governance Board, Research Advisory Panel and Patient Task Force as patient partners to develop research projects that directly respond to the questions faced by women with CF. CFReSHC holds monthly virtual meetings on topics that impact the lives of women with CF such as: contraception, family building, hormone influences on CF, incontinence and menopause. As we enter our third year, CFReSHC is looking for women with CF who are passionate about sexual and reproductive health research to join our collaborative and who will commit four to six hours a month. We offer a small honorarium for your time commitment.

CFReSHC

Cystic Fibrosis Reproductive and Sexual Health Collaborative

CFReSHC is looking for a:

Social Media Strategist who has connections in the CF community or who is willing to engage with the CF community on our behalf. CFReSHC currently has a social media presence on Facebook, Twitter and Instagram and the applicant would need to be able to post two to three times per week as well as check the platforms regularly and make responses as needed. The applicant would need to attend three paid monthly meetings often held during work hours.

Grant Writer and Coordinator who has connections in the CF community or is willing to connect with the CF community with experience researching funding opportunities, preparing grant applications and receiving project funding. The applicant would need to attend three paid monthly meetings often held during work hours.

Women with CF are welcome to send a letter of interest to: cfreprohealth@gmail.com. ▲

due to a lack of digestive enzymes causes malnutrition in about 80% of CF patients. Genes involved in the pancreatic secretion pathway (PSP) regulate the availability of these digestive enzymes. Trypsin is one of the potent digestive enzymes synthesized by the pancreas. It is produced in an inactive form that is activated by a set of genes involved in intra-pancreatic activation of trypsin (IPAT). Premature activation of trypsin triggers inflammation of the pancreas, called pancreatitis. According to the authors, about 17-22% of CF patients are estimated to have recurrent pancreatitis or chronic pancreatitis (RP/CP). Due to the frequency of pancreatic insufficiency (PI) and pancreatitis in CF patients, the research team evaluated the presence of mutations in the genes involved in IPAT and PSP in CF patients. They found that these patients had multiple mutations involving CFTR and other genes from IPAT and PSP.

http://tinyurl.com/y6ux8roc

Potential CF Therapy SNSP113 Able To Kill Drug-resistant Bacteria In Lab Study

The active ingredient of Synspira's SNSP113, an inhaled investigational treatment for chronic lung infections, demonstrated an ability to completely eliminate antibiotic-resistant Pseudomonas aeruginosa bacteria. These latest findings, together with its proven ability to kill other dangerous bacteria that affect CF patients, make SNSP113 a possible game-changer for the treatment of chronic lung infections. To determine the antibacterial effect of the active component of SNSP113, a complex sugar polymer called PAAG (polycationic glycopolymerpoly acetyl arginyl glucosamine), researchers simulated the natural forma-

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tion of persisters by two methods: exposing Pseudomonas to commonly used antibiotics, or exposing them to a compound called carbonyl cyanide m-chlorophenylhydrazone (CCCP). Treatment with PAAG was able to rapidly kill both types of induced antibiotic-resistant P. aeruginosa. When exposed to PAAG, antibiotic-tolerant P. aeruginosa bacteria numbers were 1 million to 10 million times lower within two to four hours, and the bacteria were completely eliminated within 24 hours. PAAG was also more effective at eliminating CCCP-induced persister bacteria than antibiotics currently used to treat chronic infections, such as tobramycin, aztreonam and macrolides. Complete eradication of CCCP-induced persister bacteria was seen within 24 hours after treatment. Importantly, PAAG was able to eliminate P. aeruginosa persisters at concentrations not toxic to human lung cells grown in the lab. The strong killing activity of the compound was linked to its capacity to disrupt the cell membrane of bacteria within minutes. Recently, the therapy was seen to boost the ability of antibiotics against Burkholderia cepacia and multidrug-resistant Staphylococcus aureus.
http://tinyurl.com/y8mu37jz

Machine Learning To Help Cystic Fibrosis Decision-making

New research claims to have demonstrated that machine-learning techniques can predict with a 35% improvement in accuracy – in comparison to existing statistical methods – whether a cystic fibrosis patient should be referred for a lung transplant. The new algorithmic model, called AutoPrognosis, is capable of achieving a positive predictive value of 65%. Existing practice results in only 48% of individuals being correctly referred, so AutoPrognosis yields a 35% increase in accuracy overall in comparison to current methods.
http://tinyurl.com/yb22nz4f

TREATMENT

SPX-101 Is Stable In And Retains Function After Exposure To Cystic Fibrosis Sputum Juliana I. Sesma, Bryant Wu, Timothy J. Stuhlmiller, David W. Scott. Journal Of Cystic Fibrosis. Article in Press

In healthy lungs, the epithelial sodium channel (ENaC) is regulated by short, palate, lung and nasal clone 1 (SPLUNC1). In cystic fibrosis (CF), ENaC is hyperactivated in part due to a loss of SPLUNC1 function. The authors developed SPX-101 to replace the lost function of SPLUNC1 in the CF lung. The results demonstrate that SPX-101, but not SPLUNC1, is stable in CF sputum. These results support the therapeutic development of SPX-101 for the treatment of cystic fibrosis.
http://tinyurl.com/y79b4dof

Lumacaftor/Ivacaftor Reduces Pulmonary Exacerbations In Patients Irrespective Of Initial Changes In FEV_1 Susanna A. McColley, Michael W. Konstan, Bonnie W. Ramsey, J. Stuart Elborn, Michael P. Boyle, Claire E. Wainwright, David Waltz, Montserrat Vera-Llonch, Gautham Marigowda, John G. Jiang, Jaime L. Rubin. Journal Of Cystic Fibrosis. Article in Press

Improved lung function and fewer pulmonary exacerbations (PEx) were observed with lumacaftor/ivacaftor (LUM/IVA) in patients with cystic fibrosis homozygous for F508del. It is unknown whether PEx reduction extends to patients without early lung function improvement. The researchers found that LUM/IVA significantly reduced PEx, even in patients without early lung function improvement.
http://tinyurl.com/y8vfjx4u

Investigating The Effects Of Long-term Dornase Alfa Use On Lung Function Using Registry Data S.J.

Newsome, R.M. Daniel, S.B. Carr, D. Bilton, R.H. Keogh. Journal Of Cystic Fibrosis. Article in Press

Dornase alfa (DNase) is one of the most common cystic fibrosis (CF) treatments and is often used for many years. However, studies have not evaluated the effectiveness of its long-term use. The authors investigated the effects of one-, two-, three-, four- and five-years of DNase use on lung function to see if the benefits of short-term treatment use are sustained long term. Treatment was estimated to be more beneficial in people with lower lung function; those with $ppFEV_1$ <70% at treatment initiation showed an increase in lung function over one year that was sustained out to five years. The estimated effect of DNase did not depend on age. It was concluded that DNase improved lung function in individuals with reduced lung function but no change in the slope of decline. There was no evidence for a benefit in lung function in those initiating treatment with $ppFEV_1$ >70%.
http://tinyurl.com/yas8d97x

Absorption And Safety With Sustained Use Of RELiZORB Evaluation (ASSURE) Study In Patients With CF Receiving Enteral Feeding Freedman, Steven D.; Wyatt, Colby; Stevens, John; Patel, Dhiren; Gallotto, Robert; Grujic, Danica; Brown, Perry. Journal of Pediatric Gastroenterology and Nutrition: August 1, 2018

Pancreatic insufficiency (PI) and malabsorption of fats lead to reduced caloric intake, inability to maintain weight and increased gastrointestinal symptoms. Thus, enteral nutrition (EN) is used in patients with cystic fibrosis (CF) and poor nutritional status. This study evaluated safety, tolerability and improvement of fatty acid (FA) status in red blood cell (RBC) membranes, a marker of long-term FA absorption, with an in-line digestive cartridge (RELiZORB®) that hydrolyzes fat in enteral formula. RELiZORB use was found to be safe, well tolerated and resulted in increased levels of FAs in RBCs and plasma. This is the first prospective study to show EN can improve FA abnormalities in CF. Since improvement in omega-3 levels has been shown to help pulmonary and inflammatory status as well as anthropometric parameters in CF, RELiZORB may have important long-term therapeutic benefits in patients with CF.
http://tinyurl.com/ycafnmvq

FYI

Italian And North American Dietary Intake After Ivacaftor Treatment For CF Gating Mutations Nina N. Sainath, Joan Schall, Chiara Bertolaso, Carolyn McAnlis, Virginia A. Stallings. Journal Of Cystic Fibrosis. Article in Press

In patients with cystic fibrosis (CF), ivacaftor treatment results in significant weight gain and the impact on diet has not been explored. The authors found that fat intake increased with treatment, possibly due to the recommendation to take ivacaftor with high fat meals. Increased energy and fat intake correlated with weight gain.
http://tinyurl.com/ycc22sxr

Resistin Is Elevated In Cystic Fibrosis Sputum And Correlates Negatively With Lung Function Osric A. Forrest, Daniel M. Chopyk, Yael Gernez, Milton R. Brown, Carol K. Conrad, Richard B. Moss, Vin Tangpricha, Limin Peng, Rabindra Tirouvanziam. Journal Of Cystic Fibrosis. Article in Press

Researchers assessed the association of resistin (an immunometabolic mediator that is elevated in several inflammatory disorders) with cystic fibrosis (CF) lung disease. Resistin modulates the recruitment and activation of myeloid cells, notably neutrophils. They found that plasma resistin levels were only marginally higher in CF than in healthy control subjects. By contrast, sputum resistin levels were very high in CF. Higher plasma resistin levels were associated with allergic bronchopulmonary aspergillosis, and higher sputum resistin levels were associated with CF-related diabetes in CF patients. Overall, the observed links between resistin levels in the plasma and sputum of CF patients showed a correlation with disease status.
http://tinyurl.com/yaef85al

Anaerobic Bacteria Cultured From CF Airways Correlate To Milder Disease – A Multisite Study Marianne S. Muhlebach, Joseph E. Hatch, Gisli G.

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IMPORTANT RESOURCES

Partnership for Prescription Assistance: Phone: 1-888-477-2669 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, 2100 M Street NW, #170-353, Washington, DC 20037-1233 or e-mail them at: info@trioweb.org.

American Organ Transplant Association (AOTA): Phone: 1-713-344-2402 http://aotaonline.org/default.aspx
Helps defray out-of-pocket travel expenses for transplant recipients. Helps to set up trust funds. For more information, write to: AOTA, 21175 Tomball Parkway #194, Houston, TX 77070-1655.

ADA: To learn how the Americans with Disabilities Act (ADA) applies to you, contact the Disability Rights Education and Defense Fund (DREDF): Phone: 1-800-348-4232 http://www.dredf.org/.

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Einarsson, Stef J. McGrath, Deirdre F. Gilipin, Gillian Lavelle, Bojana Mirkovic, Michelle A. Murray, Paul McNally, Nathan Gotman, Sonia Davis Thomas, Matthew C. Wolfgang, Peter H. Gilligan, Noel G. McElvaney, J. Stuart Elborn, Richard C. Boucher, Michael M. Tunney. European Respiratory Journal. 2018

Researchers investigated the correlations of anaerobic and aerobic bacteria cultured from cystic fibrosis (CF) airways. Findings showed the occurrence of anaerobic bacteria at an early age. An association of anaerobic bacteria with milder disease was observed in sputum-producing subjects. This finding indicated that targeted eradication of anaerobes may not be warranted in sputum-producing CF subjects.
http://tinyurl.com/y86ml2yn

Poor Recovery From A Pulmonary Exacerbation Does Not Lead To Accelerated FEV_1 Decline Don B. Sanders, Zhanhai Li, Qianqian Zhao, Philip M. Farrell. Journal Of Cystic Fibrosis. July 2018Volume 17, Issue 4, Pages 492–495

Patients with CF treated for pulmonary exacerbations (PEx) may experience faster subsequent declines in FEV_1. Additionally, incomplete recovery to baseline FEV_1 occurs frequently following PEx treatment. The authors found that accelerated declines in FEV_1 are more likely to precede a PEx with poor recovery than to occur in the following year. Preventing or halting declines in FEV_1 may also have the benefit of preventing PEx episodes.
http://tinyurl.com/yca4umgd

Pulmonary Exacerbations And Acute Declines In Lung Function In Patients With Cystic Fibrosis Jeffrey S. Wagener, Michael J. Williams, Stefanie J. Millar, Wayne J. Morgan, David J. Pasta, Michael W. Konstan. Journal Of Cystic Fibrosis. July 2018Volume 17, Issue 4, Pages 496–502

Patients with cystic fibrosis (CF) who experience acute declines in percent predicted FEV_1 ($ppFEV_1$ decreased ≥10% relative to baseline) are often not treated with antibiotics for pulmonary exacerbations (PEx), whereas other patients are treated even when they have not experienced a decline in lung function. Researchers determined that a clinician's decision to diagnose a PEx and treat with antibiotics often is not defined by measured lung function: a ≥10% FEV_1 decline is not considered an absolute indication of a PEx and the lack of a decline does not contraindicate a PEx. Clinicians appear to use the history of prior PEx plus other variables as factors for diagnosing PEx.

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CFRD

Continuous Glucose Monitoring Guided Insulin Therapy Is Associated With Improved Clinical Outcomes In Cystic Fibrosis-related Diabetes Freddy Frost, Paula Dyce, Dilip Nazareth, Victoria Malone, Martin J. Walshaw. Journal Of Cystic Fibrosis. Article in Press

Researchers determined the impact of continuous glucose monitoring (CGM) guided insulin initiation on the outcomes (including weight and pulmonary function changes over 12 months) in adults without a prior diagnosis of cystic fibrosis-related diabetes (CFRD). They analyzed CGM profiles for 59 patients and started insulin therapy in 37 patients who had evidence of hyperglycemia on CGM. Findings demonstrated that improvements in lung function (improved forced expiratory volume in 1 s) and weight with subsequent reduced pulmonary function decline was observed in association with insulin treatment targeted towards glycemic excursions seen on CGM.
http://tinyurl.com/y9w2te3q ▲

Laura Tillman is 70 and has CF. She is a former Director and President of USACFA. She and her husband, Lew, live in Northville, MI.

REMINDERS

- Please notify us immediately of any address changes. Returned mail wastes money and delays mailings.
- We would like to act as a referral source for active adult support groups. Please send us your group name, leader's name and phone number, number and age range of your members and geographical area covered, and we will add you to our referral list.
- Please let us know of the major occurrences in your life (e.g., marriages, births, completion of educational degrees or training, career advancement, transplants, anniversaries, birthdays), and we will print your information in **Milestones**.
- Share your ideas for **Focus Topics**, feature articles or any suggestions for improvements you may have to help make *CF Roundtable* more relevant and interesting to you.
- You can reach **USACFA** and ***CF Roundtable*** at anytime 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., 712 Main, Suite 2130, Houston, Texas 77005. E-mail: CFLegal@sufianpassamano.com.**
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