CF Roundtable®
A NEWSLETTER FOR ADULTS WHO HAVE CYSTIC FIBROSIS
SPRING 2019

My Journey Through College

By Rebecca Cedillo

I was diagnosed with CF at the age of 6 and spent most of my youth well aware of my differences. Although it was never a general topic of discussion, I was constantly in a state of anxiety or depression thinking about my future, or lack thereof. I was convinced that I would succumb to CF at a young age and, therefore, didn't plan much for my future. Instead, I sulked in sadness and wrote a will, at the age of 12, on Hello Kitty stationery. I was regularly sick during my high school years, missing as many as 73 days just in my sophomore year. How could I plan on going to college when I could barely finish high school?

The year I turned 18, I lost a few friends to CF. That was my turning point. I could either continue to be depressed or I could build a life of which I was proud. That summer, I started running as a way to control my health and build muscle. I was working part time at Sonic, which kept me

REBECCA CEDILLO

on my feet, but I knew it was time to enroll in college. After a brief time off, I enrolled in community college; this allowed me to stay close to home and my medical team, and was the most cost-effective option. As a first generation college student, I had NO idea what I was doing! It took me seven years of intermittent college attendance to obtain my bachelor's degree.

Inevitably, almost every autumn semester, I would get pneumonia or the flu, which forced me to withdraw from classes. I did have some professors who went above and beyond accommodating me — for which I am extremely thankful. I was also working full time at the community college as a way to save money. I was able to take advantage of two free classes per year. I also didn't have to worry about commuting. My supervisor was encouraging me to take classes, and I could flex my work schedule around my class schedule. When I graduated with my bachelor's degree, I was overwhelmed with joy but also felt a bit ashamed that it had taken seven years to complete. Looking back, I did a lot of growing up in those years, and I learned a lot about myself. I realized I had perseverance in the face of hardships, something I've used to propel myself forward when things get difficult.

Since graduating, I've enrolled in graduate school — something I never

Continued on page 10

INSIDE THIS ISSUE

CF ROUNDTABLE
FOUNDED 1990
Vol. XXIX, No. 2

CF Roundtable (ISSN 1057-4220) is published quarterly by the United States Adult Cystic Fibrosis Association, Inc. (USACFA), a totally independent, 501(c)(3) tax exempt, nonprofit corporation whose Board of Directors all have CF. Articles in *CF Roundtable* may be reprinted only with advance written permission from USACFA. All submissions to *CF Roundtable* become the property of USACFA and should include the author's full name, address and phone number. Submissions are subject to editing as needed and we reserve the right to edit any comments that disparage another person either by name or situation. Requests for anonymity will be honored.

CF Roundtable now is free for everyone. Tax-deductible donations are gratefully accepted to help defray the costs of printing, production and mailing of the newsletter as well as the costs of website maintenance. Please subscribe online at: www.cfroundtable.com or by mailing in the subscription form at the back of this newsletter.

CF Roundtable does not give medical advice. Any medical opinions represented in these articles are those of the writer and do not represent the views of USACFA, any of our Sustaining Partners or any other group or individual. We strongly suggest you consult your doctor regarding any medical references and before altering your medical regimen in any way. USACFA does not endorse any products or procedures.

United States Adult Cystic Fibrosis Assn., Inc.
P.O. Box 1618
Gresham, OR 97030-0519
E-mail: cfroundtable@usacfa.org
www.cfroundtable.com

EDITOR'S NOTES

We hope you enjoyed all of the information that was in the Winter 2019 issue of *CF Roundtable*. We were delighted to have so many writers participate. (By the way, we apologize for our error on the printed cover of that issue. Even though three of us checked the copy, we all missed that we had Autumn 2018 instead of Winter 2019 on the front page. It was, however, correct everywhere else in the print copy and all through the online copy.)

This issue has many new writers. Eighteen people tackled the Focus topic – Planning For Education And Careers. They start on the front page with **Rebecca Cedillo,** and go on with: **Ella Balasa, Holly Beasley, Aidan Biggar, Stacy Carmona, Kelsey Doughton, Tess Dunn, Todd Geibenhain, Nicole Kowal, James Lawlor, Fred Merz, Xan Nowakowski, Kylie Ortity, Kailee Scott-Unger, Betsy Sullivan, Devon Wakefield, Anna Ward** and **Emily Woodward.** These writers range in age from 19 to 53 and offer some good information on choosing schools and careers. The topics they cover include part time versus full time work; two- or four-year schools, vocational, trade-based, or on the job learning; scholarships, loans, and other ways of paying for training; and what kind of work is most compatible with CF.

Beth Sufian does something a little different in "Ask The Attorney" for this issue. She reviews a movie, rather than writing about legal issues. Be sure to read her comments on *Five Feet Apart.*

As usual, **Laura Tillman** gives us the links to all the latest "Information From The Internet."

In "Family Matters" **Cindy Baldwin** talks of her experience with pregnancy while taking Kalydeco.

Isabel Stenzel Byrnes writes of the resilience of people in "Spirit Medicine."

In "Speeding Past 50," I write of the value of volunteering.

"In The Spotlight" features an interview with **Paul Quinton**, who is a superstar in our community. I think you will enjoy his sense of humor.

I hope you find much to interest you in this issue. Be sure to check out "Looking Ahead" on page 3 to see if there is a topic that interests you. You don't have to write on the Focus topic. You may write about anything that is CF-related and hasn't been published previously. We would love to have you submit some of your writing or original poetry or art works.

Until next time, please 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, Two Hawks Foundation in Memory of Dr. Lisa Marino and Vertex Pharmaceuticals; 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

IPRESS RELEASES

Cystic Fibrosis May Be a Risk Factor for Hemiplegic Migraine and Stroke

Cystic fibrosis (CF) may be a risk factor in the occurrence of cortical spreading depression, which is associated with auras and hemiplegic migraine attacks. Patients with both CF and hemiplegic migraine risk developing migrainous infarction. The investigators suggest that mild hypoxia (oxygen deficiency) facilitates cortical spreading depression and that extended periods of hypoxia may lead to migrainous infarction. Patients with CF may be more vulnerable to cortical spreading depression and therefore may be considered at higher risk for experiencing hemiplegic migraine attacks with aura; patients with both CF and hemiplegic migraine have a greater chance of developing migrainous infarction.

LAURA TILLMAN

https://tinyurl.com/y4ftokdo

Increased Pulmonary Flares Raise Risk Of Joint Disease In CF, Study Says

Joint disease, or arthropathy, is common among people with cystic fibrosis (CF) and more likely to occur in older and female patients, and those who have more pulmonary flares or elevated blood levels of IgG antibodies. The higher number of flares and increased antibody concentration may reflect a state of chronic inflammation at the root of this clinically distinct type of joint disease, termed CF arthropathy (CFA). Chronic lung infections with Aspergillus spp. fungi also correlate with a higher risk of joint problems. So far, a general definition and systematic data on predisposing risk factors and treatment for CFA are not established. In this observational study, researchers wanted to assess the clinical manifestations, frequency and risk factors of CFA, and to better understand whether

Continued on page 10

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, USACFA, P.O. Box 151676, Austin, TX 78715-1676,** or e-mail to: **cfroundtable@usacfa.org**

Spring (current) 2019: Planning For Education And Careers.

Summer (August) 2019: ENT Problems And Sinus Disease. (Submissions due June 15, 2019.) Have you been plagued with sinus disease? Have you found anything that helps? If you have any secrets for managing your sinuses, please tell us about them.

Autumn (November) 2019: CFTR Modulators. (Submissions due September 15, 2019.) Have you tried any of the CFTR modulators? Are you currently using any of them? What results have you had? Do you have any suggestions for others? Please tell us of your experiences.

Winter (February) 2020: Insurance Issues. (Submissions due December 15, 2019.)

ASK THE ATTORNEY

A Review Of Five Feet Apart:

A Movie Embracing Life With Cystic Fibrosis

By Beth Sufian

I saw the movie *Five Feet Apart* on opening day and LOVED it. And then I saw it again six days later and LOVED it.

The movie has a beautiful message of hope, the importance of friendship, of accepting yourself as you are, and how important it is to reach out to others who are in need of help.

I thought for this issue I would take a break from writing about the law and write about the movie. This article is a longer version of the *CF Roundtable* blog posted the week of March 25, 2019.

By the time this issue of *CF Roundtable* is published, I expect it will be a few months since the movie premiered. When you read this, the movie probably is available online, and some readers may still be deciding if they want to watch the movie.

I encourage you to try it. Wipe movie reviews and online comments out of your mind. Create your own opinions about the film. Allow yourself to feel the incredible experience of watching a major motion picture with three main characters who have CF.

I had such a good feeling at the beginning of the movie when the female character, Stella, started doing breathing treatments. Viewers were given an accurate view of what it means to have CF. The director took the time to show scenes with the characters doing breathing treatments, using the Vest, accessing their g-tubes, and wearing masks and oxygen. I liked how the three different characters with CF dealt with their disease in different ways. I could see parts of myself in each character.

Watch the movie and honor the memory of Claire Wineland, who inspired the director to make the film, consulted on the movie and influenced the actors by sharing her philosophy on living with a chronic illness. I did not know Claire but learned so much from her videos posted on her YouTube channel. I felt the spirit of Claire whenever the character Stella was on the screen. When Stella crossed things off her daily list, I smiled. Just like the character, I get a good feeling when I cross things off my daily list — especially medical treatments I have completed.

> *"I thought I would cry a lot during the movie but, actually, I was so engrossed in the story, I think my brain didn't want any tears to block my view."*

BETH SUFIAN

The other character with CF, Poe, expresses his fears about medical costs and being a burden on his family. These are fears many of us can relate to. His strong bond with Stella mimics the strong bonds I have with so many of my friends with CF.

The male lead character, Will, reminded me of so many teens I met when hospitalized in the 1990s. Will starts out sarcastic and rather hopeless due to a decline in his health. When he connects with others who have CF, they encourage him to do his treatments and he becomes hopeful.

The actor who played Will has spoken in interviews about how the movie has been life changing for him. In an interview he said, "Justin (director), Haley (Stella) and I had our mission statement from the beginning to bring light to CF and represent it with accuracy, passion and care." I think they were all successful in their mission.

I thought I would cry a lot during the movie but, actually, I was so engrossed in the story, I think my brain didn't want any tears to block my view. Also, I think a lot of the movie is not sad but strongly validates many of the feelings I have experienced living with CF. At the end, when a picture of Claire came up on the screen dedicating the movie to her, the tears started to flow.

Six days later, I saw the movie a second time with my staff who work tirelessly on the CF Legal Information Hotline. My staff had red noses and puffy eyes at the end of the movie. Many of the people with CF we help face innumerable health challenges, and my staff saw many similarities between the

characters in the movie and our clients. Everyone was deeply moved.

My mother joined us, and I wondered what she would think of the movie. It took her a while to get up from her seat after the credits had ended. She is still talking about how beautiful the film is and the important messages it conveys.

My mother is known far and wide for her ability to reach out to help others in need, which is an important theme in the movie. She felt the movie showed characters who did their best to face each day with courage and determination, even in the worst of circumstances.

People with CF have always served as great role models for me. We should be happy when others learn from the way we live our lives. It is a good thing if a depiction of characters living with CF inspires, others to value their own lives and to decide to help others in need.

My mother raised two daughters with CF at a time when there were no medications to treat this disease and the prospect of us living to become adults was extremely low. She did not let any of this keep her from encouraging us to do our best. "If you do your best, that is all that matters," she would say. We spent most nights trying to sleep for a few hours. When we could not stop coughing we went to the kitchen at around 2 a.m. for hot tea and honey. My mother turned it into a special time for my sister and me, giving us hope, strength, and words of wisdom.

Growing up there were no cross-infection rules to follow. Doctors did not know that people with CF could cross-infect each other with bacteria. This allowed me the incredible opportunity to make friends with amazing people while hospitalized. Watching *Five Feet Apart* brought back so many good memories of all the people with CF who taught me how I could live life while dealing with all the health challenges CF was putting in my way.

Probably the reality of filmmaking necessitated the characters having masks on for only short periods of time so the audiences could see their faces. But I thought the characters taking off their masks at different points in the movie was very realistic. Many people with CF do not want to wear masks in hospitals or CF clinics. This is a reality at many CF centers. Some people with CF follow cross-infection rules set by their hospital or CF center, but others do not.

> *"Allow yourself to feel the incredible experience of watching a major motion picture with three main characters who have CF."*

I think the movie raises many important questions about the effect of cross-infection rules and concerns within our community. I know some people tell me they are terrified they may come in contact with another person with CF in a store or restaurant. I think it is good to be cautious, but for some people cross-infection concerns have caused significant anxiety. I hope we, as a community, can reduce the fear and come up with more ways for people to connect safely with each other.

I am grateful for the efforts of those who told a story that is sometimes hidden — the story of those with CF who face significant health challenges. Some people in the CF community who are doing well tell me they do not want to hear about those who are facing serious health challenges. Those with serious health issues often tell me they feel forgotten. The movie reminds our community there are still people struggling with CF. We should not forget them.

Our CF community is made up of all types of people with varying levels of health. We need to share the stories of those who have serious health problems and not just stories of people who run marathons, climb mountains, and run companies.

As Claire Wineland said in videos and speeches, a person who lives with a chronic and serious illness can still have a meaningful and joyous life. *Five Feet Apart* clearly conveys her message. The CF community should make sure our definition of success includes all of us.

Every day I speak to people with CF who call the CF Legal Information Hotline and tell me they think their life will be worthless if they stop working. I tell them their life is valuable regardless of their ability to be employed. **Being employed should not be the only way we measure success in the CF community.** I think those who retire from employment to spend more time taking care of themselves are extremely successful! There are many ways a person with CF can help others. For example, you can join us at *CF Roundtable* and help us do great things. Volunteer work can be both rewarding and have a big impact on others who need help.

As 2019 continues, let us join together, support each other, and embrace our differences. ▲

Beth is 53 and has CF. She is an attorney who specializes in disability law and is a Director and current Vice President of USACFA. Her contact information is on page 2. You may contact her with your legal questions about CF-related issues at CFLegal@sufianpassamano.com.

SPIRIT MEDICINE

A Resilient Spirit

By Isabel Stenzel Byrnes

At the CF *Roundtable* dinner during a CF Conference last October, I shared a top-ten list of traits needed for a resilient life. Given that cultivating a resilient mind is a spiritual exercise, I thought I'd share my list in this newsletter's "Spirit Medicine."

Resilience is a buzz word that we've often heard. Resilience is the capacity to respond to pressures quickly, adaptively, and effectively. Imagine a strong wind against a tree. Rather than those branches snapping and breaking with the wind, resilience means the ability for a branch to bend with the pressures of life. Resilience is about living with challenges and hardship, experiencing the range of emotions that accompanies those hardships, and finding ways to cope with and grow from them. We become stronger and better after encountering difficulties.

We are not born with resilience; rather, we learn it along the way. It takes time and experience to gain resilience. And I believe living with CF gives us ample opportunity to learn resilience.

So what is needed to live a resilient life with CF?

1. Resilient people accept change as part of life

Charles Darwin said, "It is not the strongest of the species that survives, nor the most intelligent that survives. It is the one that is the most adaptive to change." One thing is for sure in life: change happens. When things are the same, there is predictability and security. When things change, there is uncertainty and anxiety. Change requires coping skills.

With CF, any change can happen — we might need a transplant, get a port, go to college, drop out of college, get married, go on disability, face end of life, or become a parent. We have to be open to the possibility of anything. There could be both positive and negative changes ahead.

How do we react to change? Do we resist or feel anxious? Do we experience fear, get depressed, or accept or deny change? And is the way we react to change working for us? If not, what are some healthier ways to react to change?

2. Resilient people have positive attitude/optimism

No one with CF can live without the most important medicine — HOPE.

> *"No one with CF can live without the most important medicine — HOPE."*

ISABEL STENZEL BYRNES

As a child, I suffered from serious anxiety. No matter how many treatments I did, I kept getting sicker. I knew I would die young. Every hospital stay felt like I was closer to the end. Yet deep inside, I always stayed positive. I believed everything would be okay in the end. Or that I'd be rewarded someday for all my efforts. And these days, there is more reason for optimism than ever. We need the light of hope to pave the way.

3. Resilient people have positive self-confidence and self-efficacy

Having CF is different. I bet most of you are like me: you just want to be normal. CF can rob us of healthy self-esteem, and, instead, we have to work extra hard to find what we are good at and passionate about. We must find compensatory gifts that outweigh the burden of CF. For me, it was academics and writing. Post-transplant, it was athletics. Now all I've learned living with CF has made me a talented grief counselor. My greatest burden became my greatest strength.

CF gives us plenty of opportunity to gain self-efficacy. When I was 5 years old, I protested against doing my treatments. My father sat my sister and me down and said, "Every treatment you miss is one day less of your life." The more I did my treatments, the longer I could stay out of the hospital. Of course, there is a limit to CF and self-efficacy. I still needed a transplant despite all my compliance. We need to accept that this disease is brutal and not always in our control. All we can do is our best.

4. Resilient people have the ability

to regulate difficult emotions and thoughts

Newsflash: resilient people still have plenty of difficult thoughts and feelings. But they learn how to manage them. This is a skill learned over years, with increased stressors giving us practice to manage our difficult emotions. I believe CF is just as much an emotional disease as it is a physical disease. CF invites grief, anger, guilt, resentment, shame, fear, and self-loathing. What we need most are healthy parents who can teach us to stay calm in distressing situations. I had no greater teacher than hemoptysis and needing to calm myself down so the bleeding would stop. With end-stage disease, I had to spend my energy wisely, and learn not to waste it on expensive emotions.

Anxiety and depression are part of living with CF; they're a normal grief reaction. One really important way to cope with difficult feelings is to take turns *confronting* and *avoiding* the feeling. Somedays we have to pretend to be normal and other days, we have to prepare for a shorter life. We have to grieve the hardships before we can see the positive. We need permission to be a victim, complain or whine about the disappointments, pain and discomfort. But we can't get stuck there. We have to talk ourselves back up. Depression is only adaptive when it tells us to stop, rest, reflect, and reach out for support. It's a message to change inside. Most of all, we must be self-compassionate. We must talk to ourselves in supportive, kind ways, like we would to a friend.

5. Resilient people have the capacity to set realistic goals and carry them out.

We all need something to look forward to. Goals are the best motivators to take care of our health. I wanted to be like my healthy peers, so I pursued college just like them. But I had to be realistic and go to a college with an excellent CF center. I wanted to be a park ranger, but I knew my CF would get in the way. So I pursued a job that could be part time…medical social work.

Sometimes, our goals cannot be reached. We must always have Plan B. After my transplant, I thought my hospital career was over. After many years, I found a perfect job as a bereavement counselor. Reaching goals may take a very, very long time and we need to be patient.

6. Resilient people see difficulties in life as challenges.

No one ever promised that life would be easy. If we expect life to be easy, we'll be sorely disappointed. If we see CF as a curse, a burden, and an enemy, it will suck our energy and keep us from our full vitality. This will blunt our passion and it will block our personhood. If we see CF as an opportunity for growth, then we can also recognize its most precious gifts. At the end of my sister's life, she wrote, "By living alongside death for so long, I have truly lived. By being aware of limited time, I have not wasted any time, and my life has been better for it… There are no regrets…" CF is a teacher.

7. Resilient people have strong problem-solving skills.

CF creates problems in our lives. It steals our time away from doing things we want to do. So we get creative. If I don't have time, I find a way to do my nebulizers in the car. I found clever ways to stick an IV pole out the car window during a long drive. We are constantly being forced to find creative ways to avoid embarrassment, like using the staff bathroom at school so we wouldn't be teased. And to follow our healthcare regimen — by doing a treatment in a storage room at work during lunch hour. We become masters of problem solving, to fit it all in. These skills reach other areas of our lives as well.

8. Resilient people have a strong ability to nurture themselves

As a survival strategy, we must nurture ourselves. We are wired to monitor our bodies and minds and know when we're reaching our limit. It's such a gift to take care of oneself — through eating well, sleeping well, exercising, and finding balance. If we don't do that, the price to pay is getting sick. I also believe having CF taught me to enjoy life. This means truly seeking out beauty, joy, laughter, humor, and adventure. These things make life worth living. I feel like I deserve these things after working so hard to stay alive.

9. Resilient people have a sense of purpose.

We all need a reason to keep the fire inside of us lit, especially when times get tough. One purpose for living is that we need to be needed. Sometimes, when you have CF, it's hard to feel needed. We receive but don't feel like we give back. But we do. It took me a long time to realize this. My purpose is the most fundamental thing- to love and be loved: that's all.

I am grateful for our book and film, *The Power of Two.* Our story could be used to educate others about CF and transplantation. It feels so good to give back. Why am I still here? My sister Ana died of cancer. She was treated at Mission Hospice. I was hired there. There is purpose to all I've been through. What is your purpose?

10. Resilient people have a strong social network.

A will to live requires a "*who.*" To do all our treatments, we need people who love us to motivate us. Whom do we live for? Who gets us up in the morning to keep us going? People with CF need to learn to cultivate healthy relationships. This requires social skills, communication, and boundaries. In ancient times, being alone, or being

Continued on page 11

SPEEDING PAST 50

Volunteering Has Great Value

By Kathy Russell

My experiences with education beyond high school are so out of date that I fear they would not be of much use to anyone today. There were no laws to protect those who had disabilities and there were no special plans for them, either. Believe me, things definitely are better now.

I would hope that if I were young and looking toward a career, there would be counselors with whom I could talk and from whom I could get some decent guidance. I know that I wouldn't go into hospital nursing as I did back in the '60s. The potential for getting an infection is way too high. I did it then because it was training that I could afford without having to borrow any money. It was important to me that I pay my own way.

I might be more interested in some type of computer work. The machines prefer a pleasant atmosphere without too much fluctuation in temperature or humidity. That sounds good to me, too. It is possible that I would prefer some kind of work that I haven't yet heard of. At any rate, my ideas are old and probably very out-of-date.

I know that many people measure worth by how much money a person makes. I don't feel that this is a good way to assess someone's value. Many people never made much money but still gave much to their communities and world. A better measure of worth is how much help, care, or comfort a person has given (or gives) to others and to our planet.

A person may volunteer with one or many organizations that aren't able to pay a salary but can provide care and comfort or education. Is that person any less valuable than someone who makes millions or billions of dollars and doesn't do anything to help or protect people or our planet? I think not. I value people by their kindness and concern. That does not mean that I think it is wrong to earn money. I find it quite convenient to have a little money in reserve.

I have a special place in my heart for volunteers. For instance USACFA, which publishes *CF Roundtable,* is all volunteer. All the members of the board of directors and all the officers serve with no monetary incentive. Each person is dedicated to producing a good newsletter and blog and works hard to see that accomplished. Not to mention the work on the speaker bureau or scholarships, which give back to the community. Although each volunteer has CF, they still all are willing to give their time to all of us in this community.

We have many contributors who write for the newsletter or who give money to keep all of USACFA's projects going. These people have my respect and appreciation. Each one does what she or he can. I call that very valuable.

> *"I feel that my nearly 30 years of volunteering with USACFA has been some of the most fulfilling of my life."*

KATHY RUSSELL

For nearly 30 years, USACFA has grown because of such generous people. About this time of year in 1990, we were working on our first issue of *CF Roundtable.* It was a whopping 12 pages long. It seemed like a real accomplishment to those of us who were working on it. We were so delighted to be able to reach out to other adults, like ourselves, who had CF.

We were fortunate to have the assistance of Lisa McDonough, who had begun a newsletter for adults with CF and called it *Roundtable.* When she realized that producing a newsletter was way too much work for one person, she asked for volunteers to take over the production of a newsletter for adults who have CF. Those who contacted her took up the challenge and formed the United States Adult CF Association, Inc., which is better known as USACFA, the publishers of *CF Roundtable.*

Lisa was kind enough to share her mailing list with us, which made it possible to send out our first mailing of letters to 300 addresses. These letters explained what our goals were and asked for help to start a newsletter. We had a terrific response and, after months of hard work and donations of equipment and money, were able to get *CF Roundtable* going.

There were six of us who were on the board of directors or were officers who began all of this. We knew it would take many years — we guessed it would be about 20 — to get established. We knew that we would have to earn the trust and respect of our readers. We also knew that it would take many hours of work to put out a decent publication. We all were willing to dedicate hours every week to complete this task.

After almost 30 years, I feel that much of what we hoped for has been accomplished. All of this has been done by volunteers who have CF. When we were unable to do a task, we paid a professional to do it. In all of these years, none of us has received any payment, except the good feeling of helping our CF community along with a sense of accomplishment. Believe me, there has been a lot of that kind of payment.

In these 29-plus years, I have been fortunate enough to become friends with people who had just had a child diagnosed with CF and were calling for information. Those children have grown up, gone to school, high school, and some have gone to college or trade school. Some of them have married and some have become parents. It is amazing to me that I have known some of the parents from when they were young married people who were new parents of a child with CF, and now they are grandparents. I have known some of our readers since they were teenagers and now they are nearing middle age!

Seeing them approaching the time for them to take over the work on *CF Roundtable* is a truly satisfying state for me. I feel that my nearly 30 years of volunteering with USACFA has been some of the most fulfilling of my life. I am so happy to have been a part of this organization.

Volunteering has been a part of my life since I was only about three. My mother was a longtime volunteer. She always had been active with the "TB and Health Association" that now is known as the American Lung Association, and she "stuffed" Christmas seals. She took me with her and, since I was able to follow the instructions, I stuffed Christmas seals, too. I kept volunteering for them until I went to nursing school and no longer had free time.

After I graduated, I volunteered for our local chapter of the CFF. My husband was a member of the board of directors and I also volunteered. We had the pleasure of obtaining food for the week of CF summer camp. I volunteered to be the camp nurse. It was great to get to see the kids in a more normal setting than a hospital. I loved getting to go to camp, as I never had been able to go to camp when I was a child.

I also volunteered for the local lighting district, which provided street lights for our community; for the Neighborhood Accountability Board, which kept juvenile offenders out of the court system; the Crime Prevention Advisory Committee, which worked with the police department in my city; and with my husband's co-workers to provide Fantasy Flights for underprivileged children. By that time, I was running out of extra energy and had to give up my volunteer work, except for USACFA. I will give that up in the not too distant future because it is time to let others have the enjoyment of doing for our community.

So, my dear friends, you can see that you are important and have a contribution to make even if you don't have a lot of money or a special gift for writing. Your volunteering can keep this newsletter going. Each one can do what they are able to. We welcome the assistance. ▲

Kathy is 74 (75 by the time you read this) and has CF. She and her husband, Paul, live in Gresham, OR. She is a former director of USACFA and is the Managing Editor of CF Roundtable. *Her contact information is on page 2.*

Encourage Family and Friends to Sign Donor Cards

Give the gift of life that lives after you.

To receive donor cards, call:

United Network For Organ Sharing 1-800-355-7427

CEDILLO continued from page 1

thought I would have the opportunity to do. I'm studying Human Development and Childhood Disorders at UT Dallas and anticipate graduating in December 2019. I've chosen to study this because, as a child, my chronic illness impacted me in many ways — socially, emotionally, and mentally. Doctors often looked at my physical well-being but rarely tuned into my mental well-being. A 12-year-old should never have a written will.

It is my goal to positively impact the lives of those in high-risk situations. I plan on doing this by launching my nonprofit: Hope Through Help Foundation. I just submitted the formal paperwork and am excited to help those in need. The goal of the foundation is to provide families of children with CF and adults with CF financial support during hospitalizations and, more importantly, access to mental health services. My first-hand experience, combined with my degree, will help me succeed in helping those in need.

My journey isn't like that of others. I didn't ship off to a school far from home after high school. I didn't graduate in four years. I've never lived on campus. But I did learn independence, perseverance, and the importance of planning ahead. If your journey isn't like that of others, or isn't what you expected, that's okay. Every one walks a different path — they can learn from you and you can learn from them!

> "I enrolled in community college; this allowed me to stay close to home and my medical team, and was the most cost-effective option."

I've created a list of a few things I learned the hard way, in the hope that those of you who are just starting your journey can get a head start.

• Be your own advocate. All colleges have an office for students with disabilities. Meet with them. Provide them with your diagnosis verification and make a list of accommodations you'll need. These might include: snacks in class; leaving for frequent bathroom breaks; administering IVs in class; an assigned note taker; Skype lessons during peak flu season; extended due dates during hospital admissions; and not having absences count against your final grade.

• Own your CF. It sounds silly, but owning my CF is what allowed me to finally take control over it. Everyone has something that brings them down or causes complications. The only difference is yours might be a bit more visible. I didn't tell people about CF for a while. Then I realized that by telling them, I could learn to ask for and accept their help when I needed it.

• Don't let a fear of failure hold you back. In my experience, the only real failure is never being brave enough to try in the first place.

• Build a support system. Find people with common goals, common personality traits, and let them into your life. Foster healthy relationships because you'll need them and they'll need you. Life is hard but it's a lot harder when you're alone.

Good luck. Don't be so hard on yourself. And enjoy it! ▲

Rebecca is 29 years old and has CF. She lives in Dallas, TX, where she attends graduate school and works full time. She enjoys volunteering for a variety of organizations and just submitted the paperwork to launch her own nonprofit: Hope Through Help Foundation (under construction: hopethroughhelpfoundation.org) which aims to alleviate financial burdens and provide mental health services to those living with cystic fibrosis. In her free time, she enjoys traveling, playing board games with her boyfriend and family, and hanging out with her mom.

TILLMAN continued from page 3

CFA is a clinically different disease from reactive arthritis — a painful and inflammatory form of joint disease (arthritis) caused by bacterial infections. CFA was defined in the study as having at least one symptom of joint pain, swelling, reddening, or limitation of movement. Results showed that a number of risk factors were associated with a higher probability of developing CFA, namely: older age, female gender, high blood concentrations of IgG antibodies, increased number of pulmonary exacerbations, and chronic lung infections with Aspergillus spp. fungi. Although the underlying cause of CFA remains unclear, the study supports a link between CFA and lung inflammation. Based on the clinical manifestations observed, researchers believe that CFA is a separate entity from reactive arthritis.

https://tinyurl.com/y6pza5ls

Sound Pharmaceuticals Advances Phase 2 Hearing Loss Clinical Trial In Cystic Fibrosis

The STOP Ototoxicity Phase 2 clinical trial with SPI-1005 is aimed to prevent and treat ototoxicity in cystic fibrosis (CF) patients undergoing intravenous (IV) tobramycin for the treatment of acute pulmonary exacerbation. Ototoxicity (hearing loss, tinnitus, vertigo or dizziness) is a leading side effect of aminoglycoside antibiotics (tobramy-

BYRNES continued from page 7

abandoned, meant death. For those with CF, connection means survival. We need more than people around us. We need genuine, authentic relationships. To be resilient, we need to be emotionally vulnerable, to express ourselves honestly with, "I think, I feel, I need, I want," and to share what is hard and scary. To be understood is incredibly powerful medicine. Also, as a twin, I could've relied only on my sister for all my needs. But my sister died. I firmly believe we cannot put all our emotional eggs into one basket. We need a village.

We all need a place of belonging. I strongly believe people with CF need other people with CF. We are each other's best teachers. The most effective way to learn resilience is by interacting with other resilient human beings. I would not be alive if I didn't have my resilient CF friends. Thanks to the Internet, there are safe ways to connect with each other. The cost is that we can love and lose our friends with CF. This can cause anxiety... we may wonder when is it our turn? I find solace knowing we all have our own unique stories to live out in this life.

Finally, gratitude is vital for resilience. Meister Eckhart, says "If the only prayer you say in your whole life is 'Thank you,' that would suffice." My parents were told my twin and I'd be lucky to live 10 years when we were diagnosed at birth. And now I'm 47. Though my mother lost one of her daughters, she felt Ana lived four times her prognosis. She had 13 extra years of post-transplant life. It makes this all okay.

The winds of change will come to all of us with CF. Some of these changes will be positive. Some will be difficult. We have choices on how to cope with change. We can be like a tree, withstanding harsh wind, either staying upright, bending with the wind, or at some point, breaking branches or being blown down. I believe we all have deep-seated resilience inside of us, waiting to be discovered. I hope you can recognize your own resilience in your CF path. ▲

Isabel is 47 and has CF. She lives in San Mateo, CA, with her husband of 20-plus years, Andrew. She can be reached at isabear27@hotmail.com.

cin) commonly used to treat respiratory infections involving gram negative bacteria (Pseudomonas). Currently, there are no FDA approved therapies for the prevention or treatment of ototoxicity or any other type of sensorineural hearing loss, tinnitus, or dizziness.
https://tinyurl.com/ydhnrboe
AND
https://tinyurl.com/y5o9ycyd

AR-501 Enrolls First Subject In Phase 1/2a Clinical Trial For Treatment Of Chronic Lung Infections In Cystic Fibrosis Patients

Aridis Pharmaceuticals announced that the first subject has been enrolled in a Phase 1/2a clinical study evaluating the investigational candidate AR-501 for treatment of chronic bacterial lung infections in patients with cystic fibrosis (CF). The study is a randomized, double-blinded, placebo controlled single and multiple dose-ascending Phase 1/2a clinical trial investigating the safety and pharmacokinetics of inhaled AR-501 (gallium citrate) in healthy volunteers and CF patients with chronic bacterial lung infections. AR-501 is an inhaled formulation of gallium citrate that is being developed to treat chronic lung infections in CF patients. Its anti-infective mechanism of action is different from antibiotics. AR-501 acts as an iron analog and is believed to disrupt multiple iron dependent pathways in microbes, leading to growth inhibition. AR-501 has antimicrobial activities against a number of gram-negative and gram-positive bacteria, including antibiotic resistant strains. Preclinical studies have shown that mice infected with P. aeruginosa bacteria can be rescued from mortality with a single inhalation exposure of aerosolized AR-501. This drug candidate is being developed as a once-per-week dosing regimen that is self-administered using a hand-held nebulizer device.
https://tinyurl.com/y6ampvt6
AND
https://tinyurl.com/yy3x9ceq

For Use In CF, New 2-in-1 Powder More Effective Against Resistant Bacteria

Researchers have invented a new way of delivering two antibiotics (colistin and ciprofloxacin) deep into the lungs of cystic fibrosis (CF) patients, enabling more effective killing of antibiotic-resistant bacteria without exposing patients to high systemic doses of these therapies. They have succeeded in combining these two antibiotics into a single particle that can be delivered as a dry powder. According to the team, with this new formulation, more than 60 percent of the antibiotics reach the lungs, as opposed to only 10 percent with a jet nebulizer. In addition, the dry formulation offers improved chemical stability, and it is easier to use than conventional inhalation products.
https://tinyurl.com/y2jvaovc

Tobramycin Fights P. aeruginosa Bacteria That Impair CFTR Function, Study Says

Tobramycin, an antibiotic commonly used against Pseudomonas aeruginosa bacteria, reduces the bacteria's load of virulence factors that impair the

Continued on page 13

FOCUS TOPIC

PLANNING FOR EDUCATION AND CAREERS

One Step Behind

By Aidan Biggar

The Golden Gate Bridge is one of San Francisco's most beautiful and recognizable landmarks; an impressive blend of architecture and engineering. But, from my perspective, it just seemed long. An impressive 1.7 miles long, in fact. Rather worryingly, from where I stood admiring the bridge, its length was nothing compared to the 65km (41 miles) of running I was about to undertake from San Francisco to my dorm on Stanford's campus in Palo Alto. Thankfully, I wasn't alone standing there, feeling completely overwhelmed by the (many) hours of discomfort ahead of me. I had friends and fellow runners waiting every 5 miles of the southbound journey, ready to hop on the train with words of encouragement and smiles on their faces (a nice counterpoint to my pained grimace). If I didn't have those supportive friends joining to run legs with me, I might have been tempted to call an Uber instead; too overwhelmed by the immensity of the challenge.

But such is life with CF in general. Everything can be difficult. Moving cities (or countries) for school or work involves many more unknowns than it does for most everyone else. Will my university have a good CF center? Will my employer provide good enough health insurance? Will it be okay to take time off for a hospital admission?

My parents were understandably nervous when I applied to study in the U.S. It's a very long flight from where I grew up in beautiful Melbourne, Australia, so popping home to pick up a spare container of enzymes was not going to be an option. One of the keys to success for me in making this transition, and the transition from university life into the working world, and navigating through emergency rooms and exacerbations and PICC lines, was my supportive community of friends.

AIDAN BIGGAR

When I arrived at Stanford, I didn't know a soul. I barely even knew which side of the road to drive on or how to not call my flip flops by their Aussie name, "thongs." I also grew up in a country with universal healthcare and no football, so the world of deductibles and copays and out-of-pocket maximums was about as familiar as the difference between a quarterback and fullback and cornerback.

The one thing I did have was my running. During the first week of classes, I made sure to attend the first workout with the running club, which turned out to be one of the best decisions I made as a student. We were coached by a former Olympian, who also served as a second mother when I had missed a few days of medications or was attempting to conceal a cough. We travelled to Kentucky, Indiana, Illinois, and beyond to compete in track and cross country. I met people who inspired me to study mechanical engineering instead of economics. I ran with Olympians and committed once-a-week runners. But, most importantly, I gained friends who were unconditionally willing to be there when I really needed it, even when it was totally inconvenient for them.

Just last week (the first week of March), after a long day of work, I came home and started coughing up blood for the first time in several years. A few hours later, I found myself staring at the off-white ceiling of the ER at 3 in the morning, waiting for a bed to open up on the ward. But I wasn't alone. I had the immense good fortune that my first running club friend was somehow willing and able to drop everything to sit and listen quietly while the doctors talked, and to make jokes, and make the abnormal seem normal (even at 3 a.m.).

These types of setbacks, which are all-too-familiar to us in the CF community, are the reason that my mom decided to begin organizing a 65km (41 mile) walk in Australia to raise money for the cause that is so dear to our hearts (and lungs and intestines). The event has raised over $500k since beginning in 2013, but I had never really engaged with it since I was so far away studying in America.

I decided in January 2018 that rais-

ing money myself and bringing the spirit of her event to the U.S. would be a fun and exciting challenge. I was enjoying a long stretch of good health and was nervous but eager to try. Then, perhaps predictably for someone living with CF, less than three weeks out from the day of the run I found myself sitting in a hospital bed at Stanford with a bad case of the flu, watching with dismay as the nurse began the IV antibiotics. I was released from the hospital a week later, but the IV and antibiotics came home with me, with a course lasting past the day of the run.

Logically then, I was stuck. I'm fairly certain there have been no placebo-controlled medical trials on the risks and benefits of running 41 miles with an IV in your arm and little running training in the weeks prior. Thankfully, the absence of any research gave me plausible deniability to go ahead and run anyway. It seemed almost fitting that for this very public event — I had already raised over $10,000 for CF from more than 75 supporters — my invisible illness was made painfully visible by running with an IV.

The 65km run was in the spring of my senior year at Stanford, making it a fitting close to an incredible chapter of my life. A challenging but life-changing time. Each year seemed to bring a new and different health challenge — new bacteria, exacerbations, seizures and even a medical emergency ending up with major intestinal surgery, not to mention problem sets and exams and group projects. What a whirlwind — and all with my family on the other side of the world. It took a great deal of kindness from a great number of people to make it through that chaos. I am forever grateful to every person who sat by my bedside and chatted about college football or helped me catch up on lectures, because we just can't do it alone.

Being sick in college or while working full time is hard. Folks with CF need to be part of a community that is supportive and available and loving. As much as I would love to be a fully independent person who isn't reliant on anyone else, the truth is that I wouldn't have been able to graduate, or find an incredibly exciting job, or navigate a bewildering American healthcare and insurance world, or run 65km with an IV without selfless friends there every step of the way. ▲

Aidan is 23 and was diagnosed with CF at 6 weeks of age. One of his younger brothers also has CF. He lives in Palo Alto, CA with his three roommates and, hopefully, a dog in the near future. He works on flying cars at a company called Kitty Hawk and he graduated in 2018 with a mechanical engineering degree from Stanford.

TILLMAN continued from page 11

function of the CFTR (cystic fibrosis transmembrane conductance regulator) protein. The bacteria have developed several strategies to halt the immune system's response, including secreting membrane-enclosed bubbles, called outer membrane vesicles, that deliver virulence factors into the lung's epithelial cells to modify the immune response. Despite its therapeutic benefits, tobramycin has a modest impact on reducing P. aeruginosa presence in the lungs. This led to the idea that some of the clinical benefit of tobramycin may be related to anti-inflammatory effects and/or a reduction in the production of virulence factors by P. aeruginosa. So researchers investigated whether the mechanisms behind tobramycin's beneficial effects were linked to a decrease in virulence factors present in outer membrane vesicles released by P. aeruginosa. Treatment with tobramycin resulted in a statistically significant reduction in protein load of P. aeruginosa's outer membrane vesicles. It also significantly decreased the load of 165 proteins, and increased that of 17 proteins in outer membrane vesicles. Several virulence factors, including AprA, a protein that promotes P. aeruginosa survival in the lung, were among those significantly reduced. The team suggested that by reducing AprA, among other virulence factors in outer membrane vesicles, tobramycin may reduce lung damage and improve lung function, thereby providing a positive clinical benefit with only a modest reduction in bacterial load.

https://tinyurl.com/y4rsthcc

AzurRx BioPharma Announces Initiation Of Phase II OPTION Clinical Trial Of MS1819-SD In Cystic Fibrosis Patients

AzurRx BioPharma, Inc. announced that it has initiated the Company's Phase II OPTION study to investigate MS1819-SD in cystic fibrosis (CF) patients with exocrine pancreatic insufficiency (EPI). The Phase II multi-center study is designed to investigate the safety, tolerability and efficacy of MS1819-SD in a head-to-head comparison against the current porcine enzyme replacement therapy standard of care. MS1819-SD, supplied as an oral non-systemic biologic capsule, is a recombinant enzyme that is derived from the yarrowia lipolytica lipase and does not contain any animal products.

https://tinyurl.com/y4jqh4rx

Continued on page 17

How I Found Success Without Having A Full-Time Job

By Ella Balasa

Three years ago, I wrote my first piece for *CF Roundtable*. It was a pained and confused reflection on turning down a lucrative full-time position at an engineering firm, when I was fresh out of college and ready to climb the corporate ladder.

But due to cystic fibrosis (CF), my life had other plans in store. Keeping up with my peers, in that sense, wasn't in the plan. At the time, with my FEV already down to 30 %, realistically, I knew that I wouldn't be able to maintain 8-hour-plus work days and do what I needed to do to take care of my health.

In our society, great emphasis is placed on the origin of our success stemming from our careers. I felt this pressure, too, despite being aware that I was someone with a chronic illness who didn't know what even a few months might bring. It was one of the hardest decisions in my life. I felt like I was giving up a part of my identity before it had even begun. I credit Beth Sufian, an amazing woman with CF, who is a friend, and an advocate. At the time, Beth was a guiding light who encouraged me to continue part-time work, and not risk losing Medicaid benefits (which has proved to be absolutely vital) rather than pursue a job that I imagined would provide me the success I needed — a job that I think many of us yearn for but that truly doesn't define at all who we truly are.

After making the decision to remain with very flexible part-time employment at a university microbiology lab, I vowed to find my success in other ways. I didn't know what this would look like — whether I really would find it or just feel like I was floating through life. Little did I know, that the first article I ever wrote for *CF Roundtable* (when all I wanted to do was vent frustrations and reach others who may have been going through the same things) would ignite a passion for writing that has provided me much fulfillment. Writing has afforded me opportunities to use my skills and biology degree; writing has inspired me to explore and explain my thoughts. I've now been published on multiple websites and even occasionally get paid for it!

ELLA BALASA ON A PIER AT SUNSET IN THE TOWN OF MINORI ON THE AMALFI COAST, ITALY, JULY 2018.

As time has passed, some of the microbiology work that I have done in the lab, has intersected with aspects of CF. For example, common infections in CF lungs and the antibiotic resistances they acquire are also often found and studied in river and water systems. Having this background knowledge and understanding has allowed me to explore and understand CF research and be involved in various research-related advisory committees tasked with providing patient perspectives on research questions and methods.

I've also participated in research virtual-event conference planning and, through many video conferences with doctors, researchers, and those interested in understanding the patient experience, I've cultivated an interest in science communications. This involves working with researchers and professionals and helping them and the general public communicate and relate to each other. Optimal communication of this kind helps with trial participation, community organization involvement, and just raising public awareness about science research topics. Even if I do get the opportunity to work full time — perhaps after a transplant — I still will want to further pursue science communications as a volunteer in my spare time.

Needless to say, the last three years have inspired me, but they have also given me quite a few health setbacks, including three collapsed lungs with subsequent surgeries; months in the hospital and at home recovering, many exacerbations and rounds of IV antibiotics lasting weeks at a time; and, most recently, getting evaluated for a lung transplant. With all the time I would have missed from work at this point, I would have long been fired, having to

take disability, and wouldn't have been able to get back on Medicaid. I also wouldn't have had the kind of time to take care of my health, pursue other interests, and travel.

So, through this message, I want to reassure those who are on the fence about employment full-time or stressing about not having the career they wish they could have because of their health; there *are* other ways to fulfill that need for success and identity. CF makes some of us unable to plan for our careers; although this can be viewed as a negative thing, it can also allow us to pursue avenues we never thought possible. Things in life work out. As long as we have the drive, determination, and ambition to put our best into whatever we do, and to gain fulfillment from that task or accomplishment, we are successful, each in our own way. ▲

> "After making the decision to remain with very flexible part-time employment at a university microbiology lab, I vowed to find my success in other ways."

Ella is 26 and has CF. She is a Director of USACFA. Her contact information is on page 2.

Finding Balance

By Holly Beasley

Simply being a human being in this world comes with struggles and questions of how to make it all work. Throw in being a college student, coming of age, and living with cystic fibrosis (CF) to top it all off, and even more unique challenges present themselves. All of these issues have forced me to find balance, which is something I'm constantly working on and learning about. I recall one time when I saw a therapist, she continued to bring up CF and how it must be extremely difficult even though that was not my particular struggle at the moment or where I wanted to direct the conversation. I left feeling that outsiders thought I couldn't have "normal people" issues because I had CF — that this must always be my most pressing matter and concern from the perspective of many people surrounding me. I am grateful for this experience because it brought me to the realization that I am allowed to have feelings and concerns unrelated to CF, just like everyone else does. For me, this must come with the understanding that a delicate balance is needed.

I would never want to muffle the uniqueness that CF has brought me. I would not even be close to the woman I am today without this aspect of my life. Therefore, I need balance. I am a woman with CF and this is a major part of my identity. But, I am also a friend, a student, a lover of all things science, a 21-year-old, a sunny-day enthusiast, and much more. There are so many things that work together to mold the person I've become, and each comes with its own experiences and feelings. This has never been more true than life in college. As I grow in this setting, I learn more and more about who I am and what matters to me. New adventures, people, and challenges constantly present themselves as I dive into college life. I think that, as people with CF, we must acknowledge that we are abundantly special, while also being ever so normal in many ways. Our hearts can get broken, or we can be down about a test grade, or any other thing everyday life presents us. We aren't stricken to be eternally sad, either, because of our circumstances. We can celebrate and have good days and find joy in the simple things life offers.

> "The way I find balance with all of this chaos buzzing around me is to first remind myself that I must take care of my health if I want to be able to have fun."

Finding balance permeates other aspects of my life, as well, which reinforces how important it is. Most people have a plethora of tasks to complete each day, while also trying to enjoy life and make the most of it. This may never be more relevant than my time in college while managing CF. I am doing everything on my own that I once had help with. I spend hours on therapy, pill organizers, and sterilizing medical equipment each week. I often think about a healthy diet and meal preparation. I'm tasked with cleaning, laundry, groceries, homework, and more every week. A lot of these things are chores that many people will never have to think about, or that I never thought about before entering college. These new duties, along with trying to be a normal college student and having fun, require me to balance my schedule in order to get the most out of my time — and be an effective student.

Every day I see my peers getting coffee, spending time with friends, going to school events, and all of the other good activities that come along with college life. Many of my peers have the ability to spontaneously go do these things. Of course, I would like to be a part of all of these experiences and have the typical college life that many others seem to have. In the back of my mind, I worry when someone asks me to do something fun without much notice. I know it might not be possible because it

HOLLY BEASLEY

could mean missing a dose of an antibiotic or my chest physical therapy. In these circumstances and many other occasions, I have to evaluate what is most important and feasible.

The way I find balance with all of this chaos buzzing around me is to first remind myself that I must take care of my health if I want to be able to have fun. Multitasking is another way that I make my life run more smoothly. For example, I'm doing my chest therapy as I write this, so that I can relax afterwards and enjoy tomorrow with friends. Most importantly in my journey to a balanced college life is that I have to give myself a break to do the "normal" things I want to do. Unlike many others, people with CF fight each day for our lives. I want to ensure that I'm actually living the life I'm fighting for instead of just fighting. If I've been good about doing my therapy all week, I believe I deserve to go out and have fun. Everything else can wait, especially since I've found that all of my professors have been willing to extend due dates and work with me in any way possible to help me manage my health. In my opinion, managing my health constitutes more than just doing treatments and taking medicines. It is a balance of mental well-being (time to digress from all we do) and physical health.

I will graduate, and life will work out the way it's meant to so, in the meantime, I want to create a life worth fighting for. All aspects of my life must find harmony in order to do this, and I will continue to work on furthering this reality. I have much to learn, but I feel that what I have gathered so far is of great value to my journey. I struggled trying to come to one conclusive statement of how to make everything work and be balanced. What I've realized is that I'm not completely sure if there's one right way to do it, or that I know exactly how I do it. What I do know is that it all works, out and that somehow things get done. CF instills in us the ability to power through life. If one thing seems to fall through the cracks in the midst of trying to make everything else work, that is okay. Having a balanced life comes with an acceptance that some things might not get done, but life will go on and so will we. Whether you are a person with CF, in college or simply a human being in this world, I hope that you will give yourself a chance to breathe. ▲

Holly is 22 and has CF. She is a college student at the University of North Carolina at Chapel Hill. She is pursuing a major in biology and a minor in health and society. She has a science-loving side and a writing/English-loving side. Both help her to learn and connect with the CF community in unique ways. She also enjoys staying active in the gym and going on walks with her little dog that lives with her in Chapel Hill. She has a blog that you can find at glitteredwithgrit.com that explores her life with CF while in college and all other things she is enjoying at the moment. Please feel free to follow her there and join her on this journey.

TILLMAN continued from page 13

Phase 3 CF Trial On AeroVanc For MRSA Infection Updates Enrollment Info

Enrollment for a Phase 3 clinical trial testing AeroVanc in patients with cystic fibrosis with methicillin-resistant Staphylococcus aureus (MRSA) lung infections is expected to be completed in the third quarter of this year. AeroVanc is an inhaled formulation of the antibiotic vancomycin hydrochloride. The multicenter Phase 3 trial (NCT03181932) called AVAIL was designed to test the efficacy of AeroVanc in clearing MRSA infections in cystic fibrosis patients and improving lung function.

https://tinyurl.com/y5mnn6rn

Translate Bio Advances Programs In Cystic Fibrosis (CF) And Ornithine Transcarbamylase (OTC) Deficiency

A Phase 1/2 clinical trial of MRT5005, the Company's lead mRNA product candidate, is currently ongoing. MRT5005 is the first clinical-stage mRNA product candidate designed to address the underlying cause of CF by delivering mRNA encoding fully functional cystic fibrosis transmembrane conductance regulator (CFTR) protein to the lung epithelial cells through nebulization. MRT5005 is being developed to treat all patients with CF, regardless of the underlying genetic mutation, including those with limited or no CFTR protein. The primary endpoint of the trial will be the safety and tolerability of single and multiple escalating doses of MRT5005 administered by nebulization.

https://tinyurl.com/y3ovocel

CF Patients With F508del Mutations Show Improvement With Eluforsen, Trial Finds

An exploratory Phase 1 trial showed that repeated intranasal treatment with the therapy eluforsen improved CFTR protein activity in adults with cystic fibrosis (CF) carrying F508del mutations in both CFTR gene copies. Eluforsen (previously known as QR-010) is an RNA oligonucleotide developed by ProQR Therapeutics to bind to the messenger RNA (mRNA) region around the F508 deletion, and to normalize CFTR protein function in the airway epithelium. Messenger RNA is generat-

Continued on page 33

Keep Hope Alive

By Todd Giebenhain

I've never really publicly told the story of my first acting job and how I moved to LA. I've been effectively undercover about my CF for so long that it sorta got lost in the shuffle. But maybe this crowd will appreciate it.

I grew up in a small Oregon town south of Portland called Newberg. After my second year of college at Western Oregon State College (WOSC), my mom told a friend's son who was a photographer about me, and I got some headshots taken. He referred me to his agents and through them I eventually met my future friend, mentor, and teacher, Irene Cagen.

That summer after my sophomore year, I had decided I would quit school to move to California and see what happened. So I didn't enroll for my junior year. Then I got sick from a family vacation in Minnesota and lost a lot of weight. My doctor, for the first time in my life, ordered me into the hospital for the dreaded ritual I'd always heard about but never experienced: a CF clean-out with two weeks of IV antibiotics.

As an athlete who tried to avoid antibiotics for fear of developing resistance, this was a devastating line to cross. Looking back, it was a pivotal event in coming to terms with my condition and my functional denial up to that point. It was demoralizing at the time. A scary defeat. I wasn't the freakishly lucky and healthy punk with CF anymore. I actually had the disease now. The one that had already killed many of the kids I'd gone to camp with.

I still remember sitting and talking with one of my dearest CF camp friends, Chelsea, as I was being discharged. We'd been sweet on each other for years but had fallen out of contact. Her hands were in pain because medical staff had to keep using them for IVs. Her arm veins were getting scarred from PICC lines. I felt so helpless for her, but relieved for myself to be going home. This was in Bess Kaiser Hospital in Portland, Oregon, August 1995. It would be the last time I saw her.

TODD GIEBENHAIN WITH LUCAS NEFF ON RAISING HOPE IN "THE MEN OF NEW NATESVILLE" EPISODE.

Being sick put an end to the half-baked idea of moving to Hollywood, and I ended up staying in Oregon that winter. Since I hadn't enrolled at WOSC, I stayed at my parents' place and took part-time classes at Portland State University (PSU) — tennis, Spanish, and philosophy.

That's also when I started taking acting classes with Irene. She was a casting director in Los Angeles living part of the year in Portland with her boyfriend. Her office was casting the "Must See TV" shows at NBC at the time. I'd been given a stack of fliers for classes when I auditioned for the agents, and when I called her we ended up talking for hours. A truly unique and generous lady. After I got the hang of things in her class, she encouraged me to think seriously about actually moving down south. She even helped convince my parents that I could get work and survive there.

> "I'd booked my first professional acting job. A commercial for Kaiser Permanente, of all the advertisers in all the world!"

By the next summer, I'd signed with the agency and started auditioning in Portland. I'd made friends with another guy from Irene's class and we decided to move down together as roommates. We bought tickets to fly down and stay with another friend to look for an apartment. But when it came time to go, on that day, he had an anxiety attack at the thought of leaving home and backed out. He didn't show up to the airport. I went, anyway.

I applied to move into a small studio apartment in the same building as the LA friend. And in the next few weeks, I was ready to load a truck and set out on my own. Then I got the call that I'd booked my first professional acting job. A commercial for Kaiser Permanente, of all the advertisers in all the world! It would shoot the day or two before I was planning to move away.

When I arrived at the shoot, it was the Bess Kaiser Hospital — where I had been admitted, exactly one year before. We filmed in the same wing where I'd been a patient. I actually stashed my belongings in my old hospital room.

One year after I had imagined that my dreams had been crushed, after I had confronted the seriousness of my disease, after I had become a college dropout, — here I was in exactly the position I'd hoped to be. Filming my first job. Magically, for the company that had taken care of my CF my whole life. With my U-Haul loaded for LA. One year later. Healthy.

I moved down that weekend in '96. And by the end of '97, I'd booked my first TV roles, signed with a local agent, and joined the unions. Couple decades later, I'm still here in LA, living in an apartment with a parrot but, so far, managing to do what I love. Nowadays I check into the hospital twice a year on purpose, just to stay ahead of the infections.

But you never forget your first. For those of you who are interested, the Kaiser Permanente commercial is searchable on my Facebook profile!

I wrote this poem circa 2011. By the end of 2015, my PFTs were down to 32% and I was struggling. Then Orkambi came. Now I'm on Symdeko. Just one pill, twice a day. My PFTs have topped out at 51% so far. We're not exactly cured — yet — but we're finally gaining back some ground. It's coming. ▲

Todd is 44 and has CF. He lives in Los Angeles, CA, with a mouthy 17-year-old African Grey parrot named Bruce. He has been a screen actor for 23 years. He played Frank on Raising Hope *(originally titled* Keep Hope Alive*). He was featured recently with a third eye in an Expensify commercial/2Chainz music video for the Super Bowl and as a guest star on the penultimate episode of "The Big Bang Theory" on May 9th. He is an organizer for CFRI's annual CF Retreat in Menlo Park, CA (come join us in July!). And a board member for the Humanist nonprofit Sunday Assembly Los Angeles.*

2020

I used to have CF. But they cured me.

I used to have a leaden chest but I ain't burdened no mo.
Just 1 pill, twice-a-day, keeps the fresh lungs clean that I grew last season.
A reversal of genes that were put on wrong.

I used to have denial, shouting that these diseases didn't define me. I was my own
person. Watch me play ball, hit schticks like a real boy.
"See me beat the prog."
So it rolled in steady and bogged me down. Calmly gripping my throat.
Shadow casting my boxed-out brain.

Look up. Now I can see the fight. Because it's finally. f**king. over.
Dust is settling behind me but not inside me anymore. No more insulin teasing —
come here sugar and let me stick you.
So, who am I? I am a soldier who made it home,
not a martyr of war.
But like my million twins, the chrome seven samurai,
I so could have been. Would have been
if not for the chip on my brick shoulder leaning against a competitive, blinded world.
Hate kept me alive for Hope. And now I love it all.
My trophy is fearlessness.

I once had scars, blew lots of tissue. Hello there world ;)
my elastic ribbed cage is floating lightly these days.
Come here sugar and let me stick u,
look out sweetness cuz I'm loaded for bearing
mighty weight.
Stack it up, dragons,
& brings it.
I'ma breathe fire and knock ya damn socks off.

--Future Todd

You Can Do Anything: 24 Years In The Making

By Tess Dunn

Cystic fibrosis. It's been my life for 24 years. Treatments are like brushing my teeth. Downing enzymes is like swallowing candy. I've grown used to what it requires, the highs, the lows. College and a career were always in sight — even though numbers didn't indicate much of a future when I was born. I was determined to prove everyone — and statistics — wrong.

After graduating a year early from high school, I chose to attend California State University, Monterey Bay. It was close to home, but not right next door.

> "I was thrilled to find out that there was an actual disability advisor whose job was to take care of students with disabilities."

Multiple humorous situations occurred because of my CF. The second day of school, my shower overflowed, seeping into the carpet. Two weeks later, while talking with a friend, we found something that looked like mold. I had experienced a significant drop in lung function from allergic bronchopulmonary aspergillosis (ABPA) in high school. We called the residential advisor (RA) on call. I was told that I had to leave the room immediately so, at midnight, eight RAs helped me move everything in my room to a temporary room across campus. Even though what I saw wasn't mold, the entire bathroom did, indeed, have black mold in the walls. I ended up living in the temporary dorm for two weeks while the wall and carpeting were removed and replaced. I had half-royal blue, half-denim blue carpet for the remainder of the year.

Second semester, I was up late studying when my fire alarm started going off. I left my room, thinking it was the building. Nope, just my room. The UPD came and turned the alarm off, but it came back on 15 seconds later. It was hypothesized that the build-up of crystallization from my nebulizers was most likely what caused it to go off, since there was no smoke and I didn't own a heater. The annoying chirping eventually went away, but it was quite the nuisance at 2 in the morning.

After two years at CSUMB, I transferred to a different college. I planned on studying audio engineering; I'm a musician, and I wanted to have the ability to record myself and others.

I pushed myself extensively at the secondary campus, spending up to 40 hours a week working on projects for multiple terms. It was stressful, I wasn't sleeping and I wasn't eating. I began napping at 11 p.m., only to wake up at 3 a.m. and realize I still needed to do my treatment. On very rare occasions, I simply went back to sleep. My lung function was still fairly high. I felt extreme guilt over this, confessing to my social worker in tears. I felt like a hypocrite, but I was 19 and sick of living with the disease.

On top of that, school was difficult. The registrar was also our disability advisor. She had no formal training, and there was no formal documentation. I was expected to act as though I had no illness whatsoever. I remember a higher-up from the school called me into his office after I had mentioned that early mornings were out of the question. When I said I needed to sleep late, he asked: "What are you going to do in the real world?"

I wanted to say: "The real world? This is my real world. And you have no idea what it's like living it." But it was the push I needed to transfer to their main campus.

TESS DUNN

I was thrilled to find out that there was an actual disability advisor whose job was to take care of students with disabilities. I felt heard and we established a positive relationship where I felt cared for.

At the end of the first term, however, I received a 0 for participation in one of my classes. I e-mailed the professor, reminding them that I had a disability and they told me that, since I had missed a certain amount of classes, it resulted in a 0. I called my disability advisor and told him what happened. Instead of saying, "You are absolutely

right," he said "So you *feel* as though you shouldn't have a 0."

I shouldn't have had a 0. We had talked about this.

It was all sorted out eventually, but it left a bitter taste in my mouth. I eventually dropped out of this college, where there was no disability awareness or "official" disability support. This even after facing severe suicidal thoughts (I also have bipolar I disorder), and the headaches that were thought to be faux disability.

Following a semester off, I started at Sonoma State University in the fall of 2016. I immediately felt safe with Maggie, my amazing disability advisor. She was a complete advocate for me and made my life significantly easier. It was a far cry from my second college and I was beyond grateful for that.

Most teachers were extremely understanding. But there was a teacher who was one of the least empathetic professors I ever had met. On our first day of class, he said, and I quote, "You get two absences. I don't care if your mother dies. You get two absences." When I explained that I had a disability and presented him with the disability office's papers, he was kind enough. But when I said I would be missing a third class due to a doctor's appointment, he told me that there was no excuse and that my grade would drop.

I argued that I was protected by the ADA and the disability office, but he would not budge on his stance. I eventually ended up rushing from my doctor's appointment two hours north to make it in time for class. I got an A at the end of the semester, but it made me wonder: what had happened to any previous students with disabilities who couldn't make it back? I made sure to report back to Maggie, in case another student ever ended up in that situation.

During my final semester at Sonoma State, I had an internship at a magazine company. With commute traffic, I was up at 6:30 a.m. and doing my Aerobika and nebulizers as I drove. I did this twice a week for eight weeks. I did not tell anyone about my disability. Normally outspoken, I decided to keep it to myself. After the eight weeks, they offered to take me on for the finality of the semester. I explained my situation then, and they allowed me to come in early only once a week.

> "While the industry is disappointing, I couldn't stay away from the healthy outlet I've used to channel all emotions through."

I was a prolific writer in my time there. This led to them hiring me full-time. I moved 10 minutes away and got the go-ahead to come in half an hour later, so I was able to wake up at 8 in the morning, do my full treatment and then head to work. I enjoyed my time there, and my team was absolutely incredible and supportive of me and my health.

Unfortunately, I had to go on temporary disability a few months into my job. I tried oral antibiotics, then IV antibiotics when my lung function dropped further. I was weak and unable to do much of anything. I eventually chose to freelance, giving me time to rest and work from home. Thankfully, the IV antibiotics worked and my lung function increased again.

This also gave me time to refocus on my music.

I had released my first album when I was 14. Since then, I've gone on multiple tours of the West Coast, played Vans Warped Tour seven times; been featured on Vans' Pass the Bucket series; filmed countless videos; fundraised for a plethora of causes and met some of the kindest, smartest, funniest, most talented people I know. I've been lucky to have a "yes" team around me — people who will keep me in check, but make sure my dreams come true.

I had been recording my upcoming album while working at the company, a year after I had decided to quit music to focus on my writing. While the industry is disappointing, I couldn't stay away from the healthy outlet I've used to channel all emotions through. As I write this, I am looking forward to March 15, 2019, when the album comes out. I am beyond proud of it.

Now, I have the ability to teach myself how to produce my own music, and it's a challenge I adore. I have an incredible producer who has become a close friend. And I recently acquired a musical mentor as a manager — I am so lucky! Despite CF, my life has been amazing.

So, cystic fibrosis can be a burden. But it's not always lonely. Find people who understand you and help you and don't be afraid to lean on them. Find an outlet that lets you express yourself in a healthy way. And don't ever let someone tell you that you can't do something because of your illness.

You are more than CF. And I believe in you. ▲

Tess is 24 and has CF. She is an electropop singer-songwriter from Santa Cruz, CA. At the time of submission, her next album of music, "Lightswitch," was awaiting release on March 15. In her spare time, she enjoys writing poetry and lyrics, eating sushi, and playing with her cat. You can follow her music at iamt3tra.com.

Embracing The Nonlinear Path

By James Lawlor

For those of us with cystic fibrosis (CF), our relationships to our careers are multifaceted: rewarding, challenging, anxiety inducing, prone to unexpected changes, fragmented, typical, or completely non-traditional. For many of us, our careers are not only how we provide for ourselves like everyone else, but represent our key to accessing the complex care critical to our survival. That access to care, in turn, is critical to our ability to maintain a career. Our careers and our health can have huge effects on one another — and just like our healthcare stories, sharing our career stories, aspirations, and challenges can be hugely impactful for ourselves, our CF peers, and our community.

As it happens, this issue's theme of careers and education is a subject that I've been thinking about a lot in recent months. Perhaps I've hit that point in my career where I almost feel like I know what I'm doing. Also, I've recently have been fortunate to connect with a handful of fellow CF adults in science careers during the CFF's first ResearchCon, which has given me fertile ground for considering how I got here, what I've learned, and what I have that I can share with the community. I've wanted to be more involved in mentorship, and that experience taught me that sharing my story as both an adult with CF and a professional scientist can be transformational for each of those communities.

So, here's a part of my story. It's not linear, but I've realized...career paths are not linear. I think this is true for nearly everyone, but it's especially apt when CF is involved.

So, where am I? In brief, I've spent the last two and a half years working as a computational biologist in a genetics

JAMES LAWLOR

research lab with the HudsonAlpha Institute for Biotechnology. There I am part of a team of scientists using whole genome sequencing to diagnose pediatric developmental diseases (think intellectual disability, seizures, congenital heart disease, and the like). It's a role that is part programmer, part data scientist and part geneticist. I feel extraordinarily privileged to have wandered into a field that is technically challenging and full of things for me to learn. It is also personally meaningful because I'm able to offer other CF patients the critical link between gene and disease that is the foundation of so much success in treating our disease. Before that, I spent several years in the highly unrelated field of cost estimation for the U.S. government; in college, I did a wide range of tutoring and led peer study sessions for a number of classes. I've been, and continue to be, a jack-of-all-trades, and I wouldn't have it any other way.

In retrospect, it's easy for me to line up the steps I took through my education and early career that brought me to my current job: how I took a wide range of classes across my favorite branches of science (chemistry, biology, and math); how I really dug in to learn from some of the general education classes that I took (composition, programming, public speaking); how I built up my resume with my first professional job and used the skills and experience I gained, along with some strategic education, to execute a career change. That nice, orderly path is, of course, largely a lie.

I didn't ever plan to end up in the particular role I have today. I think it's more accurate to say that I planned to end up somewhere. Throughout the process, I certainly had a lot of anxiety over whether I was "on the right path" and which way I should go next. It took me a while to really absorb the idea that there was more than one path that could be realistic and fulfilling for me.

My undergraduate career was successful, challenging, and full of joy: a double major in chemistry and math, three years' of collegiate choir — where I met many of my closest friends to this day, and got to tour Italy — and a tutoring reputation of which I was proud. Happily, my health remained quite stable during these years as well (something I've always been blessed with). I had research experience from rotations in protein biochemistry labs. But those accomplishments certainly did not stop me from thinking, "Now what?!?"

I wanted to advance science, which points toward graduate school. But then the daunting choice: where, what programs, and to what end? I remember a conversation with one of my favorite professors, during which I told

him that I was interested in pursuing a master's degree in his lab, and the first thing he said in response was: why? It was a question I was not at all prepared to answer. He gave me a lot of advice in the next half-hour: that I should expand my horizons elsewhere and to be ready to put all my passion and energy into the path, 12 hours a day, six days a week, as he put it. It was good advice and intended to prod me out of my comfort zone but it brought up mixed feelings. I valued my variety of interests and hobbies, felt at home in Huntsville, and was intimidated about the length of the career path toward becoming an academic investigator: several years in graduate school, a post-doc and the high-stakes process of seeking university tenure. I worried about health insurance. I also worried that when I left school, expecting to finally hit career stability, I would have to deal with declining health.

I decided that, despite these concerns, I should apply to Ph.D. programs and go from there. I thought that, surely, with my transcript and strong recommendations, I would easily get into the top-tier programs. But I didn't. In retrospect, I had not appreciated one of the lessons I thought I had learned out of my undergraduate experience: that I was much more successful when I slowed down and thought about why I was taking that next step.

Instead, I ended up being offered a long-term contract position as a cost analyst in the defense industry, not what I was planning on. They wanted to hire math majors, and I was eager to get some real-world experience (not to mention health insurance and a 401K!). It was different, and I learned a lot, but I'd be lying if I said I didn't sometimes have second thoughts: was I wasting my potential? Is this a diversion from what I really want to do, or is this a career path I should stay in? However, I learned a lot that was valuable to my future. I had an amazing boss and mentor who taught me how to navigate professional interactions, evaluate my own accomplishments and weaknesses, own and fix my mistakes, and recognize when I was letting work get in the way of my health. There were, of course, bumps in the road with CF: anxiety disorders, influenza, pneumonia, missed business trips, IV schedules, difficult decisions about when and where to disclose CF, insurance changes, and so forth. People covered for me; I covered for them.

> "It took me a while to really absorb the idea that there was more than one path that could be realistic and fulfilling for me."

After a couple years, I began working on my master's degree (paid for by my job!) and decided to use that as a pivot back to a science career. My breakthrough came in reaching out to biology professors I had known from my undergraduate work to say, "Hey, I'm interested in applying statistics and analysis back to biology. Are you looking for students?" I found an advisor but, before long, I landed on a months-long IV treatment for NTM. I hoped my advisor wouldn't have a problem with how little research work I had gotten done over the summer. His response? "I've got a couple of CF genetics projects in the works. We should talk about them!" And there was a path to my thesis project. That project, and his recommendation, led directly to my current computational biology role.

There are a lot of things I've learned along the path so far, but I'll focus on three:

1. Nothing has to be a distraction

Art, music, theatre, science, writing, yoga, family, gaming, CF, advocacy — these things, and all of our other interests and passions and baggage, are part of our lives and allow us to bring more diversity and innovation to whatever task is before us.

2. Network and make relationships

Don't get me wrong, a lot of "networking events" make me bored and uncomfortable. I'm almost certainly going to lose your embossed business card. But the truth is, I've never ever had a job or major experience in my life that didn't start with a personal connection. After we slogged through homework sets together, a classmate introduced me to my manager for my contracting job. Before the holidays, I reconnected with an old friend from my chemistry days and now she's a graduate student working for my boss. Professors took note when I asked questions and were happy to offer me positions or connections. I e-mailed a fellow researcher to introduce myself as another scientist with CF and now we're collaborating on multiple projects. Networking isn't about being a social butterfly, about selling something or being sold, or being a raging extrovert. It's about making connections, asking for what you need, and offering what you have to share.

3. Prepare to be flexible

I'd imagine that after dealing with CF, we all qualify for a Ph.D. in flexibility and expecting the unexpected. We should take that experience we have managing uncertainty and let it

Continued on page 37

Cystic Fibrosis: More Than A Disease

By Kailee Scott-Unger

> "I found a renewed determination to complete my degree and prove that despite CF's ups and downs, I could accomplish anything."

Every morning when I wake up, doing my respiratory therapy and taking my medications is the first thing on my mind. I was diagnosed with CF in 2011 when I was 14 years old and though it may seem strange, learning I had this disease wasn't as significant as what came after. It took several years for me to change my perspective on CF and how it could impact my life positively in many ways. Immediately following my diagnosis, I was bombarded with doctors, medications, therapies, and surgery. It was a difficult time in my life but also a relief to finally know why I was sick all the time.

Surgery is a terrifying experience, especially when it pertains to a lifelong illness. But it also inspired me towards a career that I have been pursuing ever since. It was on the day of my surgery that a nurse spearheaded my decision. For confidentiality purposes, I'll refer to him as Sam. Sam was there when I received my diagnosis, and he happened to stop by on the day of my surgery. He could tell immediately that I was very anxious, and while my parents were talking with the doctors, he sat with me and reassured me. It seems like such a small thing — to sit, hold my hand, and talk about my classes or how my dog Theo is doing. But it was the best thing he could have done. From then on, Sam became my support through my surgery, my recovery, doctor's visits, respiratory infections, and my first call if I had questions or concerns. He helped me accept my new reality with CF and, as equally important, he taught me how nurses play a crucial role in patient care.

Once I knew I wanted to pursue a career in nursing, I had to decide where I was going to complete my program. Access to my CF clinic was the primary factor in my decision of where I would attend school. In addition to its proximity to my CF team, the school I am attending has a highly competitive curriculum and a high NCLEX pass rate for its graduating classes. I wanted to be in a program where I felt confident my faculty would provide me with the tools to succeed.

KAILEE SCOTT-UNGER

Adjusting to college life while also managing my CF is an ongoing challenge and it has put many obstacles in my path. During my second year in the program, I developed a Pseudomonas infection and I received intensive antibiotics for over a month. The infection strongly impacted my physical health and made it difficult to keep up with my studies. Fortunately, I was not hospitalized, but I fell behind and I struggled to finish the semester. I also dealt with extreme fatigue that no amount of sleep could fix and, as a result, my ability to exercise regularly dwindled. I saw my lowest PFT scores that year and I was afraid they would never improve, and my dream of becoming a nurse would be halted. Fortunately, that summer, I focused heavily on getting healthy again and I was able to bring up my PFT scores to what they were before my bout of Pseudomonas. During this experience, I found a renewed determination to complete my degree and prove that despite CF's ups and downs, I could accomplish anything.

During my third year in the program, I experienced only mild respiratory infections, with one to two weeks of antibiotics. However, even to that degree, any infection can determine whether I attend my clinical rotations or not. Clinical experience is critical in nursing school, and missing even one day can affect my completion of the semester. I'll admit, I worry about keep-

ing my health as close to 100 percent as possible so that I prevent this unfortunate possibility. I have learned that keeping up with my therapies and medications is crucial to preventing infections, but there is another danger. A career in the healthcare industry has its risks for me but, after researching other nurses with CF and the steps they have taken, I have made it my top priority to make sure I stay healthy.

Limits are self-created and can be overcome only if I choose to believe I am capable of overcoming them. Sam helped me to learn how to live with CF and that I still can be successful in any career I choose. He once told me to be my own lifeline. Being one's own lifeline means not relying on someone else to pull you out of a difficult situation. With the challenges of nursing school and my experiences in the CF community, I have become my own lifeline. When an obstacle presents itself, I tell myself that CF is not my limitation and that anything can be overcome with the right mindset and resources like family, friends, and my CF team. I have realized what matters most in life is that I truly live it. Becoming a nurse is a potential risk, but the opportunity to help others in a career I am passionate about is the best decision for me. I can't think of anything more fulfilling. ▲

Kailee is 22 and has CF. She lives in Portland, OR, on campus where she attends nursing school. She enjoys physical activity such as weight lifting, cycling, and cross-fit. In her free time, she likes to ride the streetcar and get off at new stops to explore more of Portland, an amazing city she calls home. She will be graduating with a Bachelor of Science in Nursing in December, 2019. You may contact her at: ksunger1996@gmail.com.

BOOMER ESIASON FOUNDATION'S PROGRAMS

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

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

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

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

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

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

Career Spotlight: Meet a Medical School Professor With CF!

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

Developing a career in the health sciences while living with CF is both realistic and exciting, and offers amazing opportunities to give back to our community. I'm a professor at a highly selective medical school that focuses on helping marginalized patient populations live and age well. This includes older adults, people with chronic conditions, racial and ethnic minority groups, queer and trans folks, lower-income individuals, families living in rural areas, etc. I like to say that I have the best job in the world, and I mean it! Certainly my concept of my "dream job" has evolved in tandem with my sense of what it means to live and age with CF. But looking back, I would not have done anything differently in my career journey.

My career satisfaction comes first and foremost from how my professional life allows me to center my CF in positive ways. My own experiences with chronic illness have always driven my passion for health equity and care quality improvement. Likewise, I see every day how my own journey with CF gives me a unique perspective as a scholar and advocate. So, rather than stopping me from pursuing a dream career, I'd say that my CF in some ways "became" my dream career. I have to live with it anyway, so why not put it to impactful use and make a good living in the process?

Getting to my current career stage also involved a long process of learning and growing from my own experiences beyond just CF. I grew up in a neurobiology research lab at a different medical school and figured out over time that I wanted to be some kind of health scientist, but I didn't see much appeal in basic science research because what really drives me are the relationship-building and advocacy components of scholarship. I also never seriously considered a career as a medical clinician during my adult life because of the infection control issues involved in that type of work.

ALEXANDRA "XAN" NOWAKOWSKI

I'm a very social person and very passionate about justice in healthcare. So when I got the opportunity to work for a school that centered these aspects of medical education and research, I jumped on it and never looked back. I've been with FSU College of Medicine for almost a decade now, doing medical sociology research and public health program evaluation! I started out in 2010 as a research assistant after finishing my M.P.H. up at Rutgers, then became a project manager and data analyst, earned my M.S. and Ph.D. while working full time in those roles, switched into a research faculty position once I finished the doctorate, and then got promoted into a ranked faculty position after a couple more years, and transferred to our Orlando Regional Campus.

Both moving to Florida and relocating to Orlando were decisions that involved my health to a large degree. The cold weather up in New Jersey was always brutal for me and my health suffered substantially each year because of it. Moving to Florida significantly improved my ability to breathe well year round, but after about a year of living in Tallahassee, I contracted NTM and then lived with intractable pneumonia for six years.

This probably is where I should mention that because genetic testing for CF was so unsophisticated when I was a child and I barely ever sweat, it was never actually confirmed that I had CF until I was 32 years old despite an initial clinical diagnosis being made at age 5. Like many adolescents, I pretty much fell off a cliff services-wise, so I was receiving very minimal clinical management until I moved to Orlando where I'm fortunate to work with an amazing CF center. I'm here for the long haul now because having seen the improvements in my health from a couple years of working with my team at Orlando Health, it would be really hard for me to go back to Tallahassee where there's no adult CF care available.

I would recommend my career to others with CF in a heartbeat, and often have. In fact, I now mentor more junior health scientists with CF in pursuing their own career goals! Being a researcher is a very flexible career; that same flexibility often exists outside

university environments as well as within them. I structure my schedule each week to accommodate all of my various healthcare activities at home and in clinical settings. Likewise, my work always follows me home and I love that. So I'm always working, but I'm also always flexible, and have peace of mind knowing that I can allow my career to evolve as my health does. Being open with my colleagues and students about having CF is key to this. I could never be stealthy about my CF — and I think it would hurt the impact I make in my career to try.

I would also recommend the approach I took to simultaneously continuing my education and building my professional reputation. If you get a chance to work for a university, do it and continue your education on an employee tuition scholarship! Doing this made earning my graduate degrees at FSU extremely affordable. I was also fortunate to have additional support from my parents to pay for research hours, which freed up resources to spend on my healthcare, nutrition, housing, etc., during the time I was in graduate school in Florida. Living on a graduate student stipend is often not realistic for people with CF unless you have a support network with substantial resources, so planning accordingly is key. And absolutely take advantage of scholarships available for people with CF. I didn't do this because I had a substantial income and help from family, but folk in different circumstances should absolutely leverage these resources.

I would say a balance of "can I physically do the work" and "can I make a positive impact for others" has guided my career choices. I'm also pretty achievement oriented, which I suppose goes with the territory of being raised by professors. I want to make the best possible difference for others with the work that I'm doing, and that requires working hard every day while also keeping an eye on my health and making sure I don't run myself into the ground. I will note that I was not always as proficient at that last bit as I am these days, and I paid a harsh price for that overzealousness many times. Prioritizing rest and allowing myself to receive support from my family has been key to getting a better handle on my health and thus having more energy for my work each day.

I definitely had to make those kinds of choices in my first couple of jobs after college. Once I started on the path to becoming a professor and started working full time for universities, this became something of a moot point because the only jobs I was considering always came with full benefits. But without going into too much detail, let's just say my life was very different in my early 20s and that I often made very constrained choices as a result. At one point I was working one full-time job plus two consulting jobs to support my household and maintain health insurance coverage, and trying to go back to school in the middle of all that. I got sicker and sicker over a period of several months, and eventually landed in the ICU with incipient signs of multisystem organ failure.

> "My concept of my "dream job" has evolved in tandem with my sense of what it means to live and age with CF."

Realistically, office- or campus-based careers with at least some flexibility to work remotely were probably going to be my only options. I'm very conscious of the tremendous privilege I now enjoy in both my career and my healthcare, and the fact that a lot of the impact I'm able to make hinges on my work adapting to my healthcare and vice versa. I feel especially thankful that my CF care team sees hospitalization as an absolute last resort, which allows me to stay in the community where I'm least likely to be exposed to additional infectious pathogens and most likely to be reminded of why the fight to age well with this disease matters so much.

Being able to work is vital for my mental health, to be quite frank. I said to my spouse just the other day that I know beyond a shadow of a doubt that my work is my most important motivation in putting up a good fight. If I didn't have that anymore — if I could no longer dedicate myself every day to improving the healthcare landscape for others — I'd see little reason not to let the disease win. So making career choices that minimize the trade-offs I would ever have to make between my physical health and my work is critical for me.

GI issues have been my biggest barrier with regard to work environments. Although I am fortunate to have much better lung function than many of my age peers with CF, thanks to improved medical management and aggressive physical therapy, I have diarrhea on a daily basis and also periodically deal with partial obstructions. And because my CF has impacted my kidneys and bladder substantially, I also sometimes experience renal and urinary issues. All of this makes regular, immediate bathroom access a must in any work environment. This and my lung function issues also impact how I approach my business travel.

Continued on page 33

Music Therapy Journey

By Anna Ward

As a small child, I always knew I wanted to have a career in some sort of human services profession. My dream started out, from as young as I can remember, as being a doctor (since I was so fond of my CF and pediatric doctors) and morphed into wanting to do CF research, which then changed into "just something in a hospital setting."

I had always had a passion for music, but I was not sure how I could combine my love for music with my love for people in the medical setting. As I progressed through high school, I realized how much more I loved music than studying for hours and hours every night. This led me to pursue a college degree in music. I entered into college as a sacred music major at Gardner-Webb University in North Carolina. I still was not sure what I was going to do with this degree, as I did not necessarily want to work in a church, but I thought it would be a foundation I could build on once I figured out the direction of my path.

Around the end of my sophomore year, I heard about music therapy. I began doing research into this field and realized that it offered the perfect combination of my love for music and my love for people (especially those with significant medical needs). Music therapy is the clinical and evidence-based use of music interventions (i.e., active music making, songwriting, lyric discussion, movement to music, relaxation to music, etc.) to promote change and growth in specific areas of an individual's life. The therapeutic relationship (between the client and the therapist) and the use of music are the two dynamic forces of change. I was already too invested in my undergraduate degree to transfer schools and start a new degree program, so I decided to complete my sacred music undergraduate degree and go straight into a masters in music therapy degree at Appalachian State University (also in NC). I intended to work in a medical hospital as a music therapist upon graduation.

MATT WARD (HUSBAND), ANNA WARD AND CHILDREN, JONAH (3), AND SILAS (1).

Although I loved the music therapy program, and was thriving in my academia, my health was suffering. The altitude and cold weather in the Appalachian mountains were tough on my lungs. I was also losing (and eventually lost) my father to cancer during this time, and the emotional stress was taking an additional toll on my lungs. These stressors led to multiple hospitalizations and courses of IV antibiotics, after 22 years of not needing IV antibiotics!

My doctor had a heart-to-heart discussion with me about midway through my program, sharing the dangers of working in a medical facility full time. I was devastated, but knew in my heart he was right. The beauty of music therapy is that it is effective with *many* populations of people; i.e., older adults in a nursing home, early intervention preschool programs, individuals with intellectual development disorders, individuals with autism, individuals with mental health needs, at-risk adolescents and so forth. I opened my heart to the idea of working with a different population and eventually settled on working in an inpatient psychiatric facility.

Although it took time for me to work through the frustration and heartbreak of not being able to practice with the population that most interested me, the rewards of working with individuals with mental illness have been beyond what I could have imagined. I've found that there are so many similarities in the treatment of mental illness and the treatment of chronic, physical illness. I have learned to relate to my patients in a different, but still impactful, way.

Looking back, I know this was God's purpose for me all along. He brought me through all the challenges and struggles to make me a better therapist for my patients, whoever they may be and whatever they may face. CF may have changed my course, but the destination is still the same. ▲

Anna is 31 and has CF. She lives with her husband, Matt, and two boys, Jonah (3) and Silas (1), in North Carolina. She is a board-certified music therapist and creative expressive arts therapies supervisor who works with patients with severe and persistent mental illness in an inpatient psychiatric facility. Anna and her family enjoy taking adventures, trying new foods, and hiking. You may contact her at annaward531@gmail.com

PHOTO BY ROBERT WRIGHT

Hidden Agenda

The invisible motivator

I'm standing in front of fifty architects and engineers
I'm telling the way I want it to be
So we help sustain mankind and our modern lifestyle.
So we'll stop blowing up mountains to find coal.
So we'll prevent climate change progressing.
But it all started with air quality
Me discovering the giant tailpipes in the sky,
spewing coal ash in my air so we can feel cozy indoors.
My audience can't see my core motivation,
my lungs struggling against the bacterial cesspool destroying them from inside.
These lungs crave fresh air, a deep unrestricted breath, healing.
Something these lungs will never experience again.
But their malfunction is your gain, they push me to fight for cleaner air.
The hope our successors will read about our short-sighted lifestyles in history books.
This is my fantastical vision, and these crippled diseased lungs that you don't see,
are really what motivated me, to want to feel like part of the solution.

— E. Hyman, 2010

FROM OUR FAMILY PHOTO ALBUM...

AIDAN BIGGAR (RIGHT) AND AN UNKNOWN RUNNER IN A 1500M EVENT AT THE UC BERKELEY TRACK IN MARCH 2018.

ELLA BALASA AND A FRIEND WHO HAS CF, JACKIE PRICE, STAYING FIVE FEET APART AT ELLA'S 27TH BIRTHDAY PARTY.

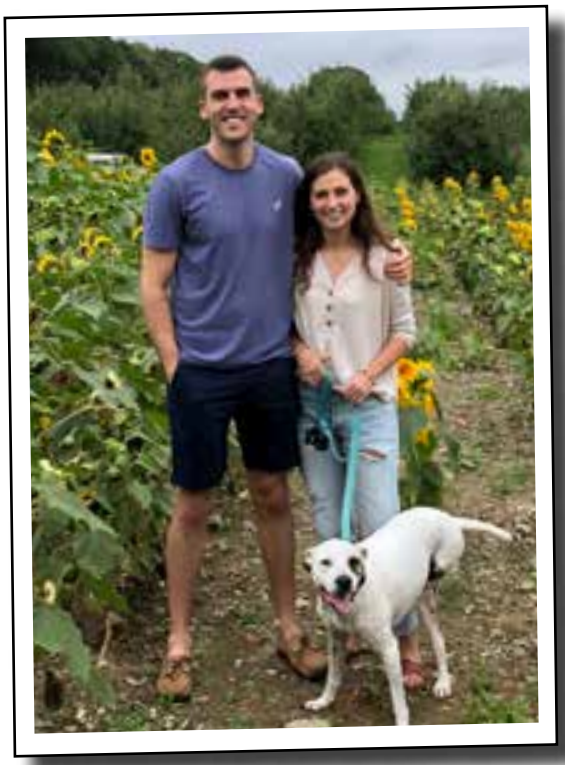
BETSY PREPARING FOR GRADUATION WITH HER FRIENDS. FROM LEFT: DIVINA CARRILLO, MEGAN THOMAS, BETSY SULLIVAN, NATALIE THORNTON, KAT NINO, JOCELYN CASTILLO, AND COURTNEY BARTLETT BEHIND THE CAMERA.

EMILY WOODWARD ON A RECENT TRIP TO NIAGARA FALLS, CANADA, AT A BUTTERFLY SANCTUARY.

HOLLY BEASLEY (CENTER) WITH HER FRIENDS KAYLA RICKMAN AND JORDAN YOUNG ON A VACATION IN ASHEVILLE, NC.

MARINA DAY AND JEANIE HANLEY IN BRENTWOOD, CA.

REBECCA CEDILLO PARTICIPATING IN THE 2015 CYCLE FOR LIFE 60-MILE BIKE RIDE, BENEFITTING THE CYSTIC FIBROSIS FOUNDATION.



KELSEY DOUGHTEN, WITH JAMES MULVEHILL, HER BOYFRIEND, AND HER DOG TEDDY.

FRED MERTZ CUTTING A ROLLED TATAMI WITH A SWORD (KATANA), 2009 WHILE HE PRACTICING IAIDO, WHICH IS THE WAY OF SWORDSMANSHIP.

FROM OUR FAMILY PHOTO ALBUM CONTINUED...

STACY CARMONA WITH HER HUSBAND, DANNY CARMONA, AT THEIR HOME IN CALIFORNIA WITH THEIR TWO DOGS, BRADY AND JULIET.

XAN NOWAKOWSKI PLAYING GUITAR, A FAVORITE HOBBY, ON A SUNNY WINTER DAY IN FLORIDA.

FROM LEFT TO RIGHT: NICOLE KOWAL'S PARENTS ERNIE AND CONNIE MATTHEWS, MICHAEL KOWAL, NICOLE KOWAL, SCOTT BIELER, AND KATHY LASHER.

TESS DUNN PLAYING GUITAR.

TODD GIEBENHAIN WITH MARINA GONZALES AND JESSICA MARTENS AT CF RETREAT 2018.

NOWAKOWSKI continued from page 27

I think the most illuminating example of troubleshooting an obstacle in my career would be getting airway clearance devices that allow me to stay independent when I travel, without having to lug around a ton of weight. I wasn't kidding when I said my GI and exocrine issues are pretty pronounced. I'm 5'4", so not especially short, but I struggle to keep my weight above 80 pounds. Coming from a long line of skinny people genetically doesn't exactly help with this. I'm quite strong for my size and work hard to stay that way. But that goes only so far when you're trying to lift heavy equipment into the overhead bin on an airplane. I also far prefer active methods of airway clearance versus a vest.

What works best for me by far is a little inspiratory/expiratory resistance pipe called The Breather. One of my clinic nurses recommended it to me when my team was mulling over how best to solve my problem of coming back from business trips feeling like garbage after several days of no manual CPT or percussor machine therapy. I have gotten such amazing results from using this device that I now have four of them — one for travel, one on each of the main levels of my house, and one that stays parked right on my desk in my office. This makes doing regular airway clearance a breeze and actually strengthens my lungs further in the process. Best of all, these devices are very affordable out of pocket and do not require a prescription, so they are a huge innovation in access even for people who don't have insurance or a steady income. And I can now spread the word about my own success with this approach to others in the CF community.

Being creative about solving problems is definitely critical in building your dream career. In that spirit, my best advice to others about professional development is to remember that your career journey with CF will be equally about your own unique goals and your own unique experiences with CF. The more aware, thorough, and realistic you are about consciously planning your career development strategy around your health management approach and vice versa, the better your satisfaction will be over time. And the more professionals with CF in your target fields that you connect with and get feedback from, the better you'll be able to make these decisions in your own career journey. ▲

Xan is 35, has CF, and currently is a medical sociologist and public health program evaluator serving as Assistant Professor in the Geriatrics and Behavioral Sciences and Social Medicine departments at Florida State University College of Medicine. Xan, along with spouse, Dr. J. Sumerau, founded the "Write Where It Hurts" project (www.writewhereithurts.net) on scholarship engaging lessons from lived experience of illness and trauma.

TILLMAN continued from page 17

ed from DNA, and contains the information for protein production.
https://tinyurl.com/y4umwtjp

AND
https://tinyurl.com/yd9gwcb4

Eloxx Pharmaceuticals Announces Participation In The HIT-CF Project On Cystic Fibrosis

Eloxx Pharmaceuticals, Inc. announced it joined the consortium agreement of the European HIT-CF project, a European Union funded preclinical and clinical research program evaluating the efficacy and safety of several disease modifying drug candidates in cystic fibrosis (CF) patients with rare genetic mutations. Eloxx's lead investigational drug candidate, ELX-02, a small molecule eukaryotic ribosomal selective glycoside (ERSG), will be evaluated in CF patients with nonsense mutations for whom there are few available treatment options. The goal of the European HIT-CF project is to investigate whether a positive response to therapies in a patient derived mini-intestine (organoid) can be predictive of clinical response in a controlled trial.

Secondly, based on the reaction in the organoids, a smaller group of patients will be assigned to studies (clinical trials) with one of the drug candidates.
https://tinyurl.com/y4scnwdy

Calithera Biosciences Initiates Phase 1 Trial Of Arginase Inhibitor CB-280 For The Treatment Of Cystic Fibrosis

Calithera Biosciences, Inc. announced that it has initiated a Phase 1 clinical trial with CB-280, a novel, orally dosed arginase inhibitor being developed for the treatment of cystic fibrosis (CF). Pre-clinical studies conducted by Calithera and collaborators have validated the potential of arginase inhibition to reduce infection and improve lung function in CF patients. The first in-human Phase 1 trial will evaluate the safety, tolerability and pharmacokinetic profile of oral CB-280 in healthy volunteers. Arginase is an enzyme secreted by immune cells and is thought to play an important role in CF pathophysiology. By decreasing production of nitric oxide (NO) in the airways of CF patients, arginase impairs antimicrobial immune response and airway function. It has been observed that lower NO levels directly correlate with worsened lung function and increased pathogen colonization in CF patients.
https://tinyurl.com/y2aqv8jh

Continued on page 35

Giving Back To Those Who Gave To Me: My Nonprofit Journey

By Kylie Ortity

When I was 36-hours-old my parents were told that I had cystic fibrosis (CF). My mother remembers saying to the doctor, "That's the disease where you have to beat on their backs, right?" My parents didn't know if I would make it out of my adolescence; and frankly, they couldn't have been more terrified. I've had CF my whole life. I've never known a different lifestyle compared to those who have been diagnosed later in their lives. I don't recall my parents ever telling me that I had CF; I just knew I had a lot of doctor appointments and it was a way of life.

Since CF is labeled as a life-threatening illness, I was given a Make-A-Wish when I was 8 years old. I received the trip of a lifetime to go to Disney World. I stayed at Give Kids The World in Kissimmee, FL, which is truly a magical place. Give Kids The World Village is described as "an 84-acre, nonprofit resort in Central Florida that provides weeklong, cost-free vacations to children with critical illnesses and their families." My family stayed in our own condominium at the village. Every week, Santa visits. I got to make my own pillow, and a star with my name on it was placed on the ceiling. I believe this is how my story began.

I always wanted to make a difference in people's lives. I wanted them to see that every day is a gift, and you never know what the next day is going to bring. I know that I can always have it worse, so I constantly focus on making sure others are happy. I believe that my Make-A-Wish trip lit a fire inside me that I've been unable to put out. I've become so involved with philanthropy, I can't imagine my life going in a better direction.

Through the ages of 11-13, I began fundraising for a local organization, Pennsylvania Cystic Fibrosis, Inc. (PaCFI). This organization makes sure people with CF in our area have the necessary equipment in order to live their lives to the fullest. With the support of the organization and the help of family and friends, I was able to raise more than $11,000 for the organization. But my passion didn't stop there.

During my time at college, I went on to accept an internship with the Mid-Atlantic Make-A-Wish. It was a truly rewarding experience to be able to say I was a wish kid, and then to have the opportunity to make sure other children also received wishes.

I was able to see early on that nonprofit work is something I want to devote my life to. All nonprofits have missions that touch different people in different ways. My passion for helping all nonprofits succeed is how I got to where I am today. I went on to receive my bachelor's degree in English with a minor in professional writing from Mansfield University of Pennsylvania. I also took the leap to get my Master's Degree in Nonprofit Management from the University of Central Florida in order to understand every aspect of a nonprofit organization. I will graduate in the spring of 2020.

> "I am very excited to put all of my passion into making sure this organization is able to give other children the magical memories I once received."

KYLIE ORTITY

I have recently been able to accept my dream job with Give Kids The World as their Digital and Virtual Fundraising Manager. I am very excited to put all of my passion into making sure this organization is able to give other children the magical memories I once received.

Only 20 years ago, most people with CF weren't making it into their adult lives. The life expectancy was low, and people couldn't even think about having their own families. I am truly

thankful for how healthy I have remained. Since there are new medications that specifically target CF genes, more and more people are living into their 50s and 60s. They're getting married and starting their own families. No one is thankful that they have CF, but I am thankful for the opportunities it has allowed me to have. I am thankful it has taught me I can reach anything I put my mind to, and I am thankful it has taught me that every day is truly a gift.

CF has allowed my life to come full circle — not just once, but twice. I always dreamed of being involved with Make-A-Wish and Give Kids The World since I stayed there in 2003. There's no better feeling than being able to give back to both organizations that were able to give me magical memories when I was so young. ▲

Kylie is 24 and has CF. She was born and raised in the small town of Lewisburg, PA, where she spent most of her time swimming, hiking, and reading outside. She absolutely adores her bunny named Willow. She recently moved to the Orlando, FL, area. She is the Digital and Virtual Fundraising Manager for Give Kids The World in Kissimmee, FL. If you'd like to learn more, please e-mail her at kylieortity@hotmail.com.

TILLMAN continued from page 33

CF Patients May Benefit From Recida's Goal To Treat Bacterial Infections

RC-01 is Recida's lead therapy candidate for multidrug-resistant Gram-negative bacteria, including the highly resistant Pseudomonas aeruginosa. RC-01 inhibits the bacterial LpxC enzyme, a key protein involved in the production of a fat molecule, called lipid A, a component of the bacteria's outer membrane and essential for their survival. In in vitro (in the lab) assays and in animal models infected with different Gram-negative bacteria, RC-01 showed a sustained and fast anti-bacterial activity against multi-drug resistant strains. RC-01 showed excellent preclinical safety and tolerability, and a favorable pharmacokinetics profile (referring to the processing of the therapy into, through, and out of the body), even when administered at high doses.
https://tinyurl.com/y2ezkd5p

P. aeruginosa Infections In CF May Respond to Antibiotics Better With Oxygen Treatment, Study Says

Exposing cystic fibrosis (CF) patients to high levels of oxygen through hyperbaric oxygen treatment may make Pseudomonas aeruginosa bacterial aggregates more sensitive to antibiotics like tobramycin and easier to eradicate from the lungs. Bacteria slow their metabolism in an oxygen-deprived environment, lessening the effectiveness of antibiotics like tobramycin that are dependent on metabolic activity to work. The mucus of CF airways is known to have very low oxygen levels (anoxic conditions). Hyperbaric oxygen treatment (HBOT) can be used both as a primary therapy or in combination with other therapies like antibiotics to enhance their efficacy. The procedure is performed in a hyperbaric oxygen chamber, and exposes patients to oxygen levels higher than atmospheric levels. HBOT increases tobramycin sensitivity in vitro by re-oxygenating and subsequently re-activating the bacteria's metabolism and growth.
https://tinyurl.com/yyvksgyg

Correct Antibiotic Dosing Could Preserve Lung Microbial Diversity In Cystic Fibrosis

Patients with cystic fibrosis (CF) whose lung infections were treated with suboptimal doses of antibiotics had fewer changes in lung microbial diversity during the IV treatment, and their microbial diversity levels were higher 30 days later. By contrast, patients who were treated with therapeutic doses had greater decreases in lung microbial diversity and significantly lower diversity levels when the antibiotic treatment ended as well as 30 days later. These findings establish the importance of the interplay between baseline microbial diversity and lung function and have the potential to improve clinical practices. Worsening lung function in CF patients has been linked to decreased microbial diversity in their lungs, a factor thought to be caused by repeated dosing of antibiotics. Although it's well known that patients frequently don't achieve therapeutic doses of antibiotics that effectively clear their infections, it's been unclear how microbial diversity changes in patients who receive sub-therapeutic doses compared with patients who receive therapeutic doses. The researchers suspect that patients who often don't achieve therapeutic blood levels of antibiotics may be genetically predisposed to metabolize beta-lactam antibiotics quickly. Repeated sub-therapeutic courses of antibiotics could significantly knock down microbial diversity without effectively clearing infections, leading to more lung damage that negatively impacts lung function over time.
https://tinyurl.com/y3mbjyxp
AND
https://tinyurl.com/yxho333v

WHO Approves Ensifentrine As Name For Potential Inhaled Treatment Now In Clinical Trials

The World Health Organization

Continued on page 41

Going Home For College

By Betsy Sullivan

Transitioning to college evokes a huge mix of emotions, and CF only adds to the process.

Throughout my many hospitalizations in high school, I had to put a ton of extra effort into my school work. Because of my determination, I ended up on honor roll every semester despite never making a full week of school out of my four years; and despite some cranky teachers, like the one who called CF-related diabetes (CFRD) fake. Even though CF was an obstacle in my education, I was able to overcome it and go away to college, something I was never sure I would have the opportunity to do. I wanted to go to a small school in Austin, TX. It seemed perfect for me because it was only an hour away from home, so I could still be independent but could come home as needed. I earned many scholarships to be able to go to my "dream school," only to be faced with extreme disappointment the moment I moved in.

During the summer, my mother and I had been working with the disability office to make sure I would have the accommodations I needed to be successful in college. The most important accommodation I requested was a single room (which I would highly recommend to all CF patients). I requested this to make sure I would have the appropriate amount of room for my medical equipment and the privacy I needed to take care of all medical needs, and the school said this was no issue, so I was super excited! My mistake here was that I didn't request to look at the room before moving in to figure out how I would be able to set up my medical equipment, which I HIGHLY recommend to all CF patients, even with a roommate. On move-in day, I was SO excited. I

BETSY SULLIVAN

couldn't wait to meet new people and start a new chapter of my life. But as soon as I walked in my room my face dropped. Of course, all dorm rooms are small and I was prepared for that, but I'm not exaggerating when I say I had a shoebox for my room. I was supposed to get a normal-sized room but instead got less than half of that. It was impossible to fit all of my medical stuff into this tiny room, so I had to sacrifice proper sterilization for my nebs. There were no other empty rooms, so I had no choice, and it was awful.

The other issue with my accommodations was with flexible attendance. With CF, you're bound to miss class more than the average student; there's no way around it. I met with my professors to speak about this need, and most of them were not accommodating at all. They didn't understand why I needed this and basically said that every time I miss class after the second time I would lose a whole point from my final grade, and be forced to drop the class after missing the equivalent of two weeks of class. I totally understand why this rule is in place for students who skip class for no reason, but if I bring a doctor's note and am keeping up with the work, how is it fair to punish me so harshly for having an illness? This, combined with the tiny room, caused me extreme anxiety like I had never experienced before. I felt trapped and like I could not take proper care of my health because of these teachers' rules. If I have to be on IVs, I'm going to need to miss class for a few days, but is it worth needing to drop classes and falling way behind in my graduation plan? Of course, health comes first, but I was so scared that something like this would happen and my grades would suffer, I had panic attacks about it day and night.

During the first week of classes, my CFRD started acting up because CF always has to make an appearance during the most important times of your life. I was experiencing constant lows, and it wouldn't stop. I was eating well-balanced meals and snacks and changed my insulin dose, and still nothing. These lows caused my anxiety to skyrocket to a level I can't even fathom. I couldn't sleep at night because I was so anxious about everything going on, from the accommodations, diabetes, normal CF stuff, and normal college stuff. I felt like I was going crazy. Even if I was doing something else, it was at the back of my mind. I avoided going out with my friends because I feared a diabetes scare or picking up an infection/virus that would land me in the hospital. My anxiety confined me to my tiny dorm room.

Around this time, I started considering transferring. It was all becoming so much. It wasn't so much handling

> I felt trapped and like I could not take proper care of my health because of these teachers' rules.

CF on my own physically; it was more the mental aspect of needing to be around people who at least partially understood it and be at an accommodating school. I had good friends my first semester of college who were understanding towards my CF, but I still felt that I needed more support around me. I had that support at home from my parents, family, and friends, which resulted in me coming home almost every weekend, and it was so good to be home. I was so much more relaxed than at school. I so vividly remember Sundays being the worst days of the week because I had to go back, and being in such a good mood on Fridays when I got to come home.

During all of this, I genuinely don't believe I would have gotten through my first semester without the support of my best friends from home. They are the most incredibly supportive group of people I have ever met, and I'm constantly astounded at the amazing people they are. My friends and I were so far apart, yet I still talked to them every single day. Just opening a funny text from them or Face Timing them lowers my anxiety so much. Days when I Face Timed or hung out with them, I was less likely to need to take medicine for panic attacks. I think every person with CF needs someone like that in her or his life — someone to vent to and someone to make you laugh when you are at your lowest point.

By October, I had decided I wanted to transfer to the University of Texas at San Antonio (UTSA), but I was so embarrassed. I felt like I was giving up, something so uncharacteristic of me. It took so much support from my friends and family to realize this was the right decision for my physical and mental health. Eventually I was so excited to be transferring. One of my best friends was already attending school there and she loved it! I finished my semester in Austin on the Dean's List and shortly after started school at UTSA, and I LOVE it! I am so glad I chose to transfer instead of staying at a school where I wasn't happy or healthy. I definitely have a better social life here, and professors acknowledge my accommodations. I know lots of people feel ashamed when they have to move back home, but there is really no reason to be. Since moving home my PFTs have gone up, plus I get to spend more time with my family and adorable niece who is also my namesake. Getting to watch a child grow up is a huge inspiration to take care of myself so I can continue to see her grow.

Anxiety has had a huge toll on my freshman year. I am so grateful that my mother is so understanding and hasn't made me feel embarrassed about my anxiety. Instead, she helped me find ways to manage, and encouraged me to ask my care team about medication. Although I was reluctant at first, I am so thankful they worked with me to find a regimen to control anxiety, a totally normal thing with CF.

I did everything I was supposed to for my health and accommodations, but some things just aren't right for everyone. Although it didn't work out, I'm glad I went away. I think it's important everyone makes the right decision for her or his health with school, even if it means you have to come home. ▲

Betsy Sullivan is 19 and has CF. She is a student from San Antonio, Texas. Her Instagram is @betsysulli and her e-mail is betsysulli12@gmail.com.

LAWLOR continued from page 23

teach us to evaluate what tools and information are available to solve problems in new ways, to evaluate contingencies and plan for change, and to stay grounded and make sure our careers — whatever they are — serve both ourselves and others.

In the context of chronic illness, I think it's important for us to carefully consider how we contextualize the meaning of "career." After all, some of us may have careers cut short or changed due to CF, or face difficult decisions regarding feasibility or stability. I've not faced the choice of whether to continue working, and I hope not to for a long while. But I do worry about how I would approach that situation and how it would affect me, especially since I find my work so meaningful. I try to remember that my career is more than any one particular job or experience, but the total of the ideas, efforts, and experiences that I contribute to my community and the world at large. Over the past year or two, what I consider my career has expanded from the role of the scientist, the part that pays my salary, to include more activism and involvement with the CF community. Now I find myself serving on advisory councils and focus groups, reviewing proposals and protocols, and reflecting more on how my experiences as a CF patient intersect with my professional experience. If my path so far is any indication, all of these roles and activities are part of an evolutionary journey that is sure to take me to plenty of unexpected places along the way. ▲

James Lawlor is 32 and has CF. He works as a computational biologist and spends his free time as a choral singer, gamer, advocate, and fiction-lover. He lives in Huntsville, AL, with his disreputable dog, Kibeth.

A Day In The Life — Working Full Time With CF

By Devon Wakefield

I am a 27-year-old man with CF. CF sucks. I live in Seattle, Washington. I work for Microsoft and, last year, my insurance paid over $180,000 in medical bills. In high school, I originally thought I would become a historian or biologist. I love science and history; I am utterly fascinated by the stories we tell ourselves about our past and figuring out what happened, and I have endless curiosity for learning how things work. But I took an intro to computer science class just before applying to colleges. I gained a profound respect for the power I quickly gained over my computer—for making it useful *and* for breaking it completely. I wanted to know how people have used computers before me; I wanted to tell computers what to do; and I wanted very good health insurance coverage. So, I changed course and applied for computer science and engineering majors in my college applications. I enrolled in Santa Clara University's School of Engineering in 2009 and graduated in 2013. I got a job later that year, took a break from working in 2014 while my sinuses got infected and I still could be on my parents' health insurance. In 2015, I got another job, although I was quickly disappointed in the sort of work they had me doing. In 2016, I landed a much more satisfying job at Microsoft (better work, better pay, better insurance...), so I left California for Washington. Here is a day in my life:

5:38 a.m.: Alarm goes off, playing Lovin' (Clarixx Mix) by CRW.

5:38 a.m.: A loud screech from the bed fills the room.

5:39: The screech subsides into groans.

5:42: I crawl out of bed. Groans of

DEVON WAKEFIELD

resistance continue to flit about the air.

5:44: I put together albuterol sulfate + ipratropium bromide solution, DNase, and colistin nebulizers.

5:48: Start inhaling all the meds.

5:49: Open a book.

5:49: Immediately close the book to cough out a loogie into a sputum cup.

5:50: Where was I in my book?

5:53: Yay! I got out a mucus plug! The little bastard.

5:57: I must poop.

5:59: Spit a loogie into the toilet & wipe my tush.

6:02: Wash hands. Switch to *warm* water, thank you.

6:03: Continue cleansing my lungs of filth.

6:11: Switch to DNase. Chop up that DNA!

6:22: Switch to colistin. Kill those germs with your terrible smell!

6:33: Feed me! Time for Greek yogurt, cottage cheese, a cup of thawed frozen berries, and some granola! Delicious, nutritious, and I am still hungry: how about donuts? It's OK to have way too many carbs, because soon I'll be at the gym to slap my blood sugars back down.

6:52: Gotta go soon! Hurl together a turkey and cheddar sandwich for lunch! Put a cup of frozen peas and carrots and a slab of butter into a container! Fill another with nuts and peanut-butter-filled salted pretzels!

7:02: Pack backpack and don't forget portable compressor and gym clothes!

7:11: Off to the gym!

7:19: Walk past someone pooping next to the sidewalk. Try not to make eye contact... (Author's note: this happened to me only once, but I was very taken aback.)

7:30: Time to get sweaty! Get ready to squirt, pores!

8:30: I'm very sweaty.

8:35: Post-workout run to catch the bus!

8:37: Ooo, Grindr tells me there are several hunky guys *desperate* for some sensual attention. This topless guy is only 645 feet away! Don't you just love apps?

8:53: Switch buses!

8:56: Wow, according to Scruff, this bus route has even more hunky men on it!

9:16: At work! First, drink some chocolate milk, then shower.

9:29: Time to work. What was I even working on, again? Oh, that's right...

11:31: I am so hungry. Where is my lunch buddy? Let's eat!

11:35: Microwave veggies. Toast sandwich.

11:40: Shove it all in my face!!

12:10 p.m.: Do a treatment. I use the meditation room that has a frosted window-door (all the others are very transparent) so I have some privacy. The office administrator helped me find this room. Duoneb is meditative, right?

12:40 p.m.: Back to work! Work, computer, twerk!

5:15: Time to catch my bus again! Bye everyone!

6:30: I am home, STARVING, and need a treatment. I'll microwave the food I cooked Sunday for the week. Today it is butternut squash, chicken thighs, broccoli, and caramelized onions.

6:40: Food is steaming, I have my glass of whole milk, and my stomach demands justice.

7:10: My appetites are sated. Just need to put my dishes in the sink — never mind I'll do a treatment first.

7:15: Time to purge my lungs of muck, yet again! Sheesh, lungs, how would you survive without me?

7:43: Ugh, I am HUNGRY again, time for dessert. Let's see, I have some gelato, and I have a spoon...

7:52: Now I am cold. Let's have some tea.

8:03: What a refreshing glass of tea! Oh, shoot, it's 8 p.m. already?? I got to get ready for bed! No time for dishes, weekend-Devin can handle that. Weekend-Devin is so helpful that way. (Weekend-Devin, who is writing this, would like to chime in to say that weekday-Devin is a lazy son of a...)

8:43: OK, nebs are washed, face is washed, I actually washed a few dishes (no thanks to Weekend-Devin), I did my sinus flush, I took my evening pills, I am in pajamas, I brushed and flossed my teeth, I jettisoned my bowels' artifacts, and am ready to snore!

8:45: Let's stretch a little before hopping into bed.

9:10: That was awesome! Time to fall asleep.

9:15: Why aren't I asleep yet??

9:26: GO TO SLEEP! I NEED MY REST!

9:38: zzz... zzz...

A few notes about my day:

• Times used are general estimates, except, yes, I do set my alarm to 5:38. I like to set it to non-round numbers.

• I haven't told anyone in authority at work that I have cystic fibrosis. Some colleagues know, but I haven't been specific with my boss. My first boss knows I do some medication at work and need a place to do it, and that it can be loud, but I never said "lung disease," "cystic fibrosis," or anything of a diagnostic nature; just what accommodations I need.

• My boss, actually, wasn't very helpful finding a place to do my midday Duoneb. The office administrator knows better where someone can cough up some loogies in private.

• I got switched to a new team last year, and I haven't told my new boss anything. Not that he's a bad person or would abuse the information, but he just hasn't needed to know yet.

• I switch between Cayston and colistin. At the time of this writing, I'm doing neither because recently I coughed up a bunch of blood and went in-patient for six days, two on a weekend. I worked from home while finishing my IVs at home. My lungs had great timing, because during and after my hospitalization, Seattle had the most snow it has had in the past 50-plus years. Everyone else was working from home, too.

• I'm wildly gay, but I only included a few moments throughout my day thinking about hunky men. For those curious, I find I have good luck on Scruff. On Grindr, I'll sometimes get wild messages (scams) from women (bots) saying they'll pay me for sex if only I text them. Scruff is also great when you're in the E.R. and have nothing better to do in between hemoptysis episodes. One guy I messaged turned out to be a hot respiratory therapist. Last Sunday, we went out for pancakes and French toast. We'll see how it goes.

• I try to exercise as much as I can. I go to a CrossFit class about five times a week and will occasionally run on the weekend for 4-6 miles. Obviously, I sometimes take a day off for a doctor's appointment or when I do not feel well (usually after an episode of hemoptysis). ▲

Devon is 27 and has CF. He lives in Seattle, WA.

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 speakers-bureau@cfroundtable.com

Working Through CF

By Nicole Kowal

I don't know about you, but when I was diagnosed with CF, the doctors told my parents I would be lucky to graduate high school. So, when high school graduation came around, I was ecstatic to have the opportunity to go to college. (I wasn't the greatest student, attending summer school three out of the four years in high school. My parents were happy when I finally graduated.) Not only did I attend high school, but I took advantage of BOCES, which stands for Board of Cooperative Educational Services. It is a public organization that was created by the New York State Legislature in 1948 to provide shared educational programs and services to school districts.

I took advantage of a BOCES Animal Care Taking course, and had the opportunity to do an internship at a local dog kennel, which turned into a full-time job. When attending college, I made sure I let the counselor and professors know that I have CF, that coughing isn't contagious, and that I may miss class. During my college years, I went to school with a heart monitor on, because I was being diagnosed for neurological cardiac syncope. I then had to battle parvo — you know the disease your dog gets vaccinated for — yup. I got that and it sucked. I also ended up getting cellulitis really bad in my foot and was on crutches for a few days.

Letting teachers know that I have health issues was one of the best things I did. I taught them about CF and still excelled in their classes. My goal to go to college was to continue working at a kennel, and one day even open my own dog kennel. But with dogs, kennels, and cleaning up after dogs in kennels, I started becoming more ill. I had my first sinus surgery and decided that

MICHAEL AND NICOLE KOWAL.

kennel life wasn't for me. I made my heartfelt good-bye to a place I loved working at so much, and to all the dogs I never got the chance to see again.

But when one door closes, another usually opens. I applied at a local car dealership: West Herr Toyota in Orchard Park (WHTOP), NY. I just thought this would be a nice little job that would help me make some money until I found something else. I graduated college while working there and stayed on. The light bulb went off that this work environment isn't too bad; instead, it's AMAZING!!! I was able to find a job, a career, that actually allowed me to be — me. Working there has been simply a true blessing. I am always upfront with my managers and share my health issues with them (sometimes even a little too much), but they all have a great sense of humor.

I started out as a greeter; moved to a sales assistant; then to a service greeter; from there to filing paperwork; back down to a greeter; and finally to administrative assistant to an amazing boss, one whom I consider a brother-like figure. Justin Rojek, has been my manager at WHTOP for 11 years now. His family has become part of mine. I love babysitting his kids, and his wife, Rachel, is awesome. Having a boss who treats you not only as an employee but a friend and almost a little sister figure has made the battle with CF in the workplace so much easier.

I am able to work full-time hours, leave for doctors' appointments, and know that no one is judging me and my health issues. My work colleagues are HUGE supporters of WNYCFF and have donated more than I could ever ask for. The president of West Herr, Scott Bieler, has a heart of gold and has made a difference in my life more than anyone will ever know. It usually brings tears to my eyes when I talk about how blessed I am to work for a great company, to have the understanding of management, and to have job security even when I'm sick. I do wear a mask sometimes; I sanitize like crazy; and I shoo people away from me when they are sick. I have no problem protecting myself from illnesses in the workplace, and I have my management team to back me up on it.

I also sell Rodan + Fields to help with some of my medical bills as a little side job. I also babysit and dog watch, too. I love keeping my options open to any side job and opportunity to fulfill my life with as much as possible. The great thing with these side jobs is that I can make them fit around my schedule and my life. I work when I want and need, too. Sure, I could fill my schedule with every side job on offer, but if I know I have appointments coming up or that I am getting run down, I don't

> “I was able to find a job, a career, that actually allowed me to be — me.”

have to fill the schedule. All of my clients know that I have CF and understand that when I come over or they come over, that coughing happens along with every other thing that CF carries.

When looking for a career, please make sure you don't settle. Be open about you. Show the world that no matter what is in your way you are able to overcome it. Look at how fast a CFer has to grow up. We are well beyond our years, and have been for a long time. We have responsibilities from such a young age, which is why I also think we tend to be great workers. Having a very sturdy back bone makes us strong in many other areas in life. Just because we are “sick” does not mean that we need to roll over and allow our disease to overrun our life. There are jobs out there that will suit you and turn out to be amazing. You just need to know what works for you. The kennel was not great for me, but being at the dealership has allowed me to grow not only as a worker but as a person. With the workforce and people being more open about things, I truly feel that bosses will just appreciate you trying to work through your hurdles. Find people who won't fight against you in the workplace but will fight with you. They will sometimes tend to be some of your biggest fans. ▲

Nicole is 31 and has CF. She lives in Orchard Park, NY, with her husband. She loves volunteering and fostering animals until they find their “forever” homes. You may contact her at abnormalnicole@westherr.com.

TILLMAN continued from page 35

(WHO) approved ensifentrine as the recommended name to identify the cystic fibrosis (CF) treatment candidate RPL554. Verona Pharma's ensifentrine is an inhaled inhibitor of the enzymes phosphodiesterase 3 and 4, designed to have bronchodilator and anti-inflammatory properties. Preclinical studies also revealed that ensifentrine, either by itself or in combination with other treatments, was able to stimulate different types of CF transmembrane conductance regulator (CFTR) mutants. Ensifentrine was also shown to boost the induced increase in CFTR activity by Orkambi (lumacaftor/ivacaftor, by Vertex Pharmaceuticals) in cells expressing specific CFTR mutants.
https://tinyurl.com/y4gwo8kp

USask Discovery May Help Improve Cystic Fibrosis Treatment

Doctors have treated CF patients with an inhaled concentrated salt solution (hypertonic saline) to increase the volume of airway surface liquid (ASL)—a microscopically thin liquid lining that helps remove infected secretions from the clogged chest of a CF patient. The scientific consensus has been that an osmotic reaction drawing water from the blood was responsible for the beneficial increase in ASL from this saline treatment. But by using synchrotron imaging at the Canadian Light Source (CLS) the investigators have concluded that scientists have not completely understood the body's reaction to the saline treatment. Only about half of the airway surface liquid production is caused by the osmotic process. The other half of the liquid secretion is caused through the saline mist stimulating the airway neurons to act. The researchers found that sensory neurons monitoring the airway perceive the inhaled saline mist and trigger a defense mechanism that prompts cells (epithelia) to produce more airway liquid, which also contains potentially beneficial antimicrobial compounds.
https://tinyurl.com/y6thyo9b
AND
https://tinyurl.com/y2uhc9p2

CF Study Finds Link Between Pseudomonas aeruginosa, Disease Severity

Patients with cystic fibrosis (CF) who have never been exposed to the Pseudomonas aeruginosa bacterium have a milder form of the disease, a retrospective study based on registry data suggests. Previous reports suggest that P. aeruginosa prevalence in CF patients increases with age up to 20-25 years old, and then stabilizes at 50-70 percent after that. This means that 30-50 percent of CF patients do not have P. aeruginosa. These percentages also include patients who have been infected but cleared of the bacterium by antibiotic treatment. To better understand predictive features associated with P. aeruginosa infection history, a research team evaluated differences between CF patients with no clinical history of P. aeruginosa and patients with reported P. aeruginosa exposure. The researchers compared clinical features of CF patients, including age, CFTR mutations, pancreas-related implications, and lung function. They also analyzed associations between P. aeruginosa exposure history and the prevalence of other pathogenic bacteria.

Continued on page 43

College, CF, And Nursing, Making It Work

By Emily Woodward

I go to school at Colby-Sawyer College in New Hampshire, where I am currently a sophomore nursing major. My passion for this career has stemmed from my very personal experiences in the field. For my entire life I have been in and out of hospitals, so I have seen my share of nurses. Let me tell you that they have made the biggest difference in my life and just in the general way I have dealt with my disease. I have built special bonds with them that I could never have with other people, and these are the same bonds that I hope to have with my future patients. I want to be able to help children with life-threatening illnesses realize that you can achieve your dreams and goals. With support and determination, anything is possible.

Transitioning into college life was not really difficult due to the fact that a really close friend went to the same school, and she was able to help guide me through my first year with having a chronic illness. She, amongst my other friends, was a great source of support for me and continually reminded me to do my treatments. She also accompanied me to my various appointments. My friends keep me on track. It is easy to get caught up with homework and other activities, but they let me know that nothing is more important than my health. Having a strong support system of family, friends, and a great care team has been such a blessing. They have helped me every step of the way.

The same time that I was starting college, I also transitioned into an adult clinic. At first I was a little scared to transition to adult care, but it went pretty smoothly because I was coming fresh from an amazing pediatric team to an equally fantastic adult CF team who continued to advocate for my every need. I was also involved in some clinical trials, so I had the added advantage of having a clinical trial team. The opportunity to become involved in trials has been a great experience. The world of research and development has been fascinating and one that has continued to better my health and motivate me to keep pursuing my career.

Colby-Sawyer has been very accommodating to me. Through their disability resources, I was able to get a single room and bathroom. My advisors and professors were all willing to work and accommodate my health needs so I could be successful. It was important to be upfront and honest with all of my professors from the start. They understand if I need to take some time to go to a doctor or a little extra time with assignments. Being a nursing major, my illness has actually opened up a few conversations in some classes. One particular event that occurred was when I was in my health assessment class. We were discussing a pulmonary assessment and one of the features was that of clubbed nails. No other students knew what they looked like and since I have them I was able to be a learning tool, which was pretty neat for my own experience. I enjoy being able to take opportunities to educate others on CF.

> “Let me tell you that [nurses] have made the biggest difference in my life and just in the general way I have dealt with my disease.”

EMILY WOODWARD

The most important thing when it came to planning my education was a program that offered a highly reputable nursing degree as I hope to become a specialized nurse one day. I wanted to be close to home but also far enough away so that I could have a new experience in another state. I am financing my college through scholarships, loans, and the financial aid my school provided for me, as well as working part time as a licensed nursing assistant to help pay my everyday living expenses

while being in school. It has been hard at times to find a balance of health, school, and work. I have a great social worker who has helped me find grants and scholarships to help pay for some expenses so I could work less and focus on school and my heath. Having an open, honest relationship with my care team has been great. They are a valuable source of knowledge and resources that I never would have known about. I also try to apply for as many scholarships as I can. There are many sources of scholarships and the more you apply for, the greater your chances of getting a few. I would advise other students to not get discouraged and to keep searching and applying.

One obstacle I have faced is being the first nursing student with CF to be in my nursing program. Now that I am starting a clinical rotation in the local hospital, new questions come up that the faculty has never had before. There were a lot of unknowns and times when they weren't sure how to pursue some of my needs (like not working with CF patients). However, I shared with them any recent test results and communicated with my adult CF team who liaised between me and my nursing faculty to make sure that I have a safe experience. In this way I was able to get the best nursing education possible. Although navigating these new waters with the nursing program has, at times, been stressful, I am so proud to know that I am helping pave the way for others who have life-threatening illnesses who may want to enter the healthcare field.

One piece of advice I would give is to be an advocate for yourself and surround yourself with those who support and continue to fight for and with you. Never give up on your goals and seek out friends and colleagues who are willing to help achieve those goals with you. One of my team members once told me that no matter what I want to do in nursing, we will figure out a way to do it: and that is the support I needed to help me continue my education. ▲

Emily is 20 years old and has CF. She is a sophomore at Colby Sawyer College, NH. When not at school she lives in St. Albans, VT, with her parents and brother. She is an avid dog lover. Emily can be contacted at erosew101598@gmail.com.

TILLMAN continued from page 41

They found six clinical features that were independently associated with CF patients without documented P. aeruginosa infection: older age at diagnosis, absence of diabetes, absence of aspergillosis (infection by fungi Aspergillus), pancreatic sufficiency, absence of F508del mutations in the CFTR gene, and forced expiratory volume in one second equal or greater than 80 percent, which is considered a normal value, indicating preserved lung function. Analysis of CFTR mutation types showed that a lack of F508del in both alleles was significantly associated with patients with no documented P. aeruginosa infection. F508del is linked to about 70 percent of defective CFTR alleles, and has been shown to predispose CF patients to infections by opportunistic pathogens such as P. aeruginosa. Negative correlations between four preventable or treatable comorbidities (coexisting conditions) and the absence of P. aeruginosa infection were also found. Patients who had never been exposed to P. aeruginosa showed an absence of aspergillosis, diabetes, and pancreatic insufficiency. Furthermore, compared with CF patients with previous P. aeruginosa exposure, these patients also had preserved lung function. When researchers further analyzed the association between bacterial infections and the two patient groups, they found distinct microbiological profiles related to patients without reported P. aeruginosa exposure. Specifically, this patient group was significantly associated with the presence of Haemophilus influenzae, and absence of Stenotrophomonas maltophilia, Achromobacter xylosoxidans, and Aspergillus.

https://tinyurl.com/yy8h852o

Association Between HbA1c And The Development Of Cystic Fibrosis-related Diabetes

In people with cystic fibrosis-related diabetes(CFRD), researchers analyzed HbA1c as a predictor of risk for future development of CFRD and evaluated the correlation with the development of retinopathy. The results indicated that severity of diabetic retinopathy was related to a higher HbA1c and longer duration of CFRD. A connection was found between HbA1c level and the future development of dysglycemia in CF based on oral glucose tolerance test and microvascular results. Although the current guidelines do not advocate the use of HbA1c as a diagnostic tool for CFRD, it can be of clinical use in identifying people at risk of future CFRD development.

https://tinyurl.com/yxluzvuv

AND

https://tinyurl.com/y5onstkx

New Bacterial Signaling Language Offers Pathway To Treat Infections

Scientists discovered that Pseudomonas aeruginosa uses a family of fatty acids, (oxylipins) in a cell-to-cell signaling language critical for its viru-

Continued on page 54

Dare To Dream—A Message To Future College Students

By Stacy Carmona

I still vividly remember the moment when I opened my acceptance letter to my dream school. After the shock wore off, I jumped up and down screaming for what felt like hours. It doesn't get much better than living on the beach in beautiful Santa Barbara, California. When I look back on my college experience, those were some of the best years of my life. I learned so much about myself and had many memorable experiences. But, like everything else with CF, there were challenges I encountered during that time that threatened my potential to succeed.

At the age of 18, when I went away to school at the University of California Santa Barbara, I was struck by the realization that it was the first time in my life when I was solely responsible for taking care of my health. I also realized that I would need to prioritize my health while surrounded by so many exciting distractions around me. Unfortunately, I learned the hard way that I needed to find a way to manage my time in order to maintain my health and my grades.

Fast forward to my sophomore year of college: I had to take a semester off when I was hospitalized with pneumonia. I was devastated to leave my friends and my independent lifestyle for a sterile hospital room. After several weeks on IV antibiotics, I moved back home with my parents to recover for the rest of the semester. That experience was my much-needed wake-up call to make better choices and put my health first. At the end of the day, if I didn't feel well, I wasn't able to enjoy anything.

Despite my setbacks, I was determined to graduate in four years with my classmates. I had to work harder and smarter than the average college student to manage my studies and my health, but it was worth the extra effort to graduate on that sunny June afternoon and accept my diploma. That day, I graduated with honors and with pride in my heart.

STACY CARMONA

Having been through the highs and lows of college life, I learned some valuable strategies that I would like to pass on to future CF college students to help them in their journey. The following recommendations helped me tremendously and I hope that they will help you, too.

Utilize the Disability Center

Every university has a disability center, and there are many valuable services offered through this program. The disability services that helped me the most during my time in school were: note takers for my classes in case I missed class, a parking pass to park my car on campus, and a private room in the dorms during my freshman year.

Talk to Your Professors

Let your professors know about CF at the start of each semester so if something comes up with your health, they are more likely to accommodate you and give you extra time to complete assignments. I think you will find that most professors are understanding if they know up front about the possible accommodations you may need. You may need to provide them with a doctor's note.

Find a Good Living Situation

If you are planning to live in the dorms, work with the university to get a single room, if possible. This will allow you to be able to get the sleep you need, take your treatments in privacy and properly clean and disinfect your equipment. This will also allow you to stay healthy by avoiding sick roommates. You can design your room so that it is welcoming for people to come socialize. Even though I had a single room, there were always visitors due to my TV, dish chair, and excellent selection of snacks. If it is not possible to get a single room, either financially or otherwise, make sure to connect with your roommate(s) early and let them know how your special circumstances could impact their college experience. Make sure in advance that your roommate(s) are willing to be accommodating and flexible.

Utilize Teaching Assistants and Office Hours

Because of the nature of CF, there were days when I wasn't well enough to attend class. By going to office hours I was able to learn the material I missed and get extra help (similar to tutoring).

This will help you maintain your grade point average despite any absences.

Take a Lighter Class Load

I took the minimum number of units each semester so that I could allow myself the time to still take care of my health and to pace myself. It allowed for less pressure and stress each semester. I did have to take summer school during two summers so that I could still graduate on time.

Create Your Own Schedule

One of the great things about college is that you get to choose your own classes and schedule. I never signed up for an early morning class to allow myself plenty of sleep and enough time to take all of my treatments. I also had Fridays off to allow for time to go to doctors, pick up prescriptions, etc.

Pick a Convenient Disinfecting Method

There are several different methods for cleaning and disinfecting nebulizers and other equipment. My advice is to pick the one that is most convenient and sustainable for you with your living conditions. In my dorm I had access to a kitchen where I could boil and clean my treatment pieces.

Find a Local CF Center

For those of you who are going to school away from home, it will be important to find a local CF center where you can go in case of emergency or if you get sick. If you do not have a CF center near you, find a local hospital and a general pulmonologist in case of emergency.

Have Transportation Options

There are some days when we don't feel well enough to walk or bike to class. I had plenty of those days during my four years in college. I was fortunate to be able to take my car with me and park on campus. This allowed me to drive to class if I needed to. For those who do not have access to a car, I recommend scoping out the local transportation options (bus, train, campus shuttle, etc.) for the times when you may not want to make the trek to class or if you go to the doctor.

Everything in Moderation

There are a lot of temptations in college and I am not going to tell you that you shouldn't enjoy yourself. But I will say that it is important for things to be in moderation. Try to limit how much smoke you expose yourself to as that can be very harmful to your lungs, and talk to your CF doctors about alcohol and your specific health circumstances.

Carry Snacks

Maintaining our weight is an important part of CF care, so I recommend having snacks and supplements in your dorm room or apartment, and take them with you to class in case you get hungry.

Stay Active

Some people, when they go to college, become less physically active than they were in high school. Make sure to find a way to stay active, whether that's joining an intramural sports team or working out at the gym or in your dorm room. Exercise is an important part of CF care and you should try to make the time for it in college.

Study Abroad

I had the pleasure of studying abroad in college and it was one of the best experiences of my life. Please know that it is certainly possible to study abroad; it just takes more planning and preparation.

Share CF with Others

Before college, I was very secretive about having CF. I never wanted anyone to know and I kept it very private. When I went to college, I found it very helpful to be open about CF so that new friends could keep me company during my treatments and support my healthy habits. They became my family away from home and even had to take me to the ER a few times. I know that it can be difficult to share such personal aspects of our lives with those around us, but I found that to be the most beneficial thing I did in college and beyond. It can be lonely going through life without people who truly know you, so if you are comfortable sharing, I highly recommend it. As the saying goes, "Those that mind, don't matter and those that matter, don't mind." I have found this to be very true. It took me a long time to learn that having CF is nothing to be ashamed of. It is just part of who we are. Hiding that from others only makes the burden greater on us.

Above all, try not to let CF limit your life or dreams. If your health and finances permit, allow yourself the opportunity to go away to school and have the college experience. Even though it takes more careful planning, it is definitely doable and well worth it. I can't imagine my life if I hadn't gone away to school. I did not have the greatest high school experience and I found that in college, people were more open minded, less cliquey, and more diverse. The truth is, I learned more outside the classroom than I did inside and that was perhaps the most rewarding part of those four years of my life. I found myself in college along with many of my lifelong friends. I acknowledge that college isn't the path for everyone, so whatever it is that you hope for yourself, don't let CF stop you! ▲

Stacy is 32 years old and has CF. She lives in Laguna Hills, CA, with her husband, Danny, and two dogs, Brady and Juliet. She works as the Director of Patient Advocacy at Kroger Specialty Pharmacy, where she advocates for patients and families with CF. Stacy loves to travel and spent a year living in Australia. She can be reached at stacylcarmona@gmail.com.

Planning For Education And Careers

By Fred M. Merz

Due to the sophistication of medicines and treatment, more and more people with cystic fibrosis (CF) are reaching adulthood. Along with that comes the challenge of entering into the workforce.

Looking back, I see myself as a pioneer within the CF community — I belonged to one of the first generations of young adults with CF who explored college and careers. We asked ourselves what our "calling" was, what hopefully might be a satisfying career, and how we could contribute to the community.

When I was in my sophomore year of high school, my parents asked what I wanted to do (I guess there was no career in building 1/24 scale model cars back then), and it was the same question many others would ask me. My image of a career went from a Formula-one (F-1) driver to truck driver, then auto mechanic, to being on an F-1 team as an auto mechanic...I love cars!! Although the truck driver career was a potential reality (after seeing many truck-driving- school commercials on my UHF channels, I was convinced I could do that), I realized the idea of becoming an F-1 driver was a bit far-fetched!

I wanted to get into something technical, and my dad had suggested electronics. He then proposed I look into the future, to a time, he said, when automobiles and electronics would merge. Electronic skills would prove beneficial. My dad also pointed out that an electronics work environment would be cleaner than a garage or a factory. I did, however, get a brief taste of the automotive field when I worked in a research and development department at one of the big three (auto manufacturers).

During my sophomore year, I was accepted into a special class in which we toured various job locations and got some feedback from the employees. By my junior year, I had signed up for vocational electronic classes, which actually started to shape my future.

But as I look back now, I really think that if CF hadn't been a deterrent to a lot of opportunities, or if a lack of financial resources hadn't been an issue, I might have had the potential to achieve more or to do something else. Back in the '80s, it was beneficial to join the military because the government offered education advancement once you were honorably discharged from the service. Because a few siblings had joined the military, I was visited by recruiters, but since CF was among the list of health issues that the military wouldn't accept, I had to choose between going to college or trade school.

FRED MERZ PRACTICING HIS GUITAR, 2018.

So, yes, my choices might have been different. I might have even gone into the psychology or psychiatry fields, but I was still under the impression that CF was deadly and not something that allowed me to plan big for the future. I might have even gotten more into music back then, but maybe that choice wasn't really CF related. Right after high school graduation, however, I also had my first hospitalization for IV treatment; I was 18 then. As I sat in the hospital that week, I had decided to join a martial arts club. I was determined to do something I would enjoy and that would help me stay healthy and out of the hospital (but this is a story for another time).

My high school counselor had helped me by looking at grants and loans that I could apply for. Some were helpful sources of income, but others had to be paid back. Some that I already applied for didn't give me enough time to acquire a stable income before they had to be paid off.

Finally, I was given two choices; an offer to a university half-way across the state for which the State would pay a large portion of the tuition, or a community college just an hour's drive down the freeway, for which the State would also pick up part of the tuition if I went full-time. There was a third option: come up with a better plan.

I chose the community college because I wasn't comfortable with living so far from home where I would not know anyone, I did not know much about that particular university but I believed it would not guide me to the career I wanted. I was an introverted person in those days, so felt more

comfortable with family and familiar doctors nearby. While staying near home, I worked part-time jobs here and there at the minimum wage for that era ($3.35/hour) and even managed to get just above that in some workplaces. This helped pay for books and general expenditures, and my parents helped with the other college fees that the grants and loans didn't cover.

And so choice of an electronics career made in high school allowed me to transition seamlessly to the Henry Ford Community College (HFCC) in Dearborn, MI. HFCC was recognized in the electronics field, so job searching wasn't as much of a challenge. Years later, however I wonder what would've happened if I had made the decision to go to university instead, or was even able to join the military.

Care centers really didn't factor at all in my decision to stay near home. At that time, the CF community had started to create CF adult clinics. I was 17 when I started going from a children's hospital to an adult hospital for the first time.

The Internet wasn't fully developed then and resources were limited when searching for student loans and grants. Now, in this age of technology, it is easier to research financial aid, courses, and careers. It's easier for people to decide between two- and four-year degrees. More in-depth "soul searching" is therefore possible. There are also online colleges that are worth investigating. I cannot offer any other information and although I did almost take a course or two, I eventually decided against joining an online college course due to financial circumstances.

I would recommend the electronics field to CF patients because, in most cases, the jobs are either in an office, in front of a computer, or possibly in some sort of laboratory, all of which offer fairly clean environments.

> "My job choices were never based on time commitment, health maintenance, or even socializing, etc., becoming successful was my goal."

Electronics is also a fairly stable career choice. I have to add, however, that finding a specialty leads to better opportunities.

I had two major concerns when choosing a job or career: would I enjoy my work and would I make enough money to stay healthy and have access to insurance that was good enough so I could stay out of debt. I didn't examine in detail the health insurance offered by the various companies to which I applied. After all, I wasn't interviewing for health insurance but for a position! Even though I came across some jobs in which health insurance was expensive or insufficient, my CF was mostly digestion related, and these issues were mostly covered.

As I started on my career, I considered the specialties within electronics. I had considered biotechnology as one field, but the college I went to had a very poor curriculum and the field was already saturated by then. I had gotten a few jobs in the printer repair field, and another was eventually in automotive companies, and I ended up in one of the big three.

My job choices were never based on time commitment, health maintenance, or even socializing, etc. Becoming successful was my goal. While working in the automotive field, I had come up with this crazy idea to take on a foreign language, and use that to my advantage. Since the area I was living and working in was starting to see an increase in Japanese workers coming from Japan, I thought that learning Japanese, might be a great career move. I could then, with my background in electronics, work as either translator or liaison between two automotive companies (or liaison to an American and Japanese company). That idea, 25 years ago, led to marriage and a move to Japan.

My misinformed wife (we are now divorced) assured me of the availability of Japanese doctors who could manage my health issues. Not true. But I am doing very well for a 53-year-old person with CF, and I'm still in the workforce in Japan.

I ended up being a self-advocate, learning the special terminology to communicate with doctors and probably even encouraged them to study up more about CF, thereby increasing their CF awareness. I can also attest to the fact that Japan's social health insurance is actually better than any insurance coverage I would have had access to in the USA.

Of course, I wouldn't recommend such a crazy idea to leave the country and start a career just anywhere, unless you end up in one of the EU countries, or somewhere where they not only have decent social health insurance, but are also aware of CF.

And, as a final point, depending on your health situation, you should go for what you feel you can achieve. When a person finds a job that they enjoy so much they would do it for free, that is the ultimate job! ▲

Fred is 53 and has CF. He lives in Osaka, Japan. He has been in martial arts, electronics and advertising. He enjoys playing guitar and racing RC cars. He also likes to travel in Japan in his car.

The Right Time

By Kelsey Doughten

I was diagnosed with cystic fibrosis (CF) at the age of three. For years, my parents struggled to understand what was wrong with their brown-eyed baby girl. I was malnourished and very ill, but doctors could not determine the cause. My parents' wonder faded to grief when they received my diagnosis.

"She will probably not live to be 18." That was the fact of the matter according to pulmonologists. CF had always been considered a childhood disease because patients rarely lived to see adulthood. My parents pored over books, articles, and expert opinions, searching for answers and insight into the disease that gripped their daughter.

My diagnosis came around the time that my family was considering moving. My mom and dad were trying to move into a house that they could barely afford, in an amazing school district, so that my brother and I could have the best life possible. My parents wondered if this was the "right time" to move because of my recent diagnosis. They worried about the financial burden that comes with a chronic illness. They wondered if I would even be able to attend the local school or if I would have to be home schooled.

My grandfather gave my mother some advice during that difficult time. "If you treat Kelsey like she is sick all the time, then neither of you will ever be happy."

My parents decided they would not let CF limit my opportunities. They didn't wait for the "right time" to move because there never would have been one. There will always be a reason to stay stationary. I have found throughout my education and career that if you wait for the "right time," you will miss out on some of the best prospects that life has to offer.

In May 2018, I graduated from the University of Kentucky with a degree in mechanical engineering. This was not an easy feat, considering I was hospitalized 10 times in my last two years of college. Having a PICC line in my arm during class became a regular ordeal. In November of my senior year, I interviewed for an operations leadership development program at BAE Systems in Nashua, NH. I was absolutely in love with the position and wanted nothing more than to work for the aerospace and defense industry.

KELSEY DOUGHTEN

When I got a call about my acceptance into the program I was thrilled, but New Hampshire is a long way from Kentucky. It was completely uncharted territory for me. I had always lived in KY, never more than an hour from my parents (with the exception of a few summer internships). I had a decision to make.

Due to all my recent hospitalizations, I questioned if it was the "right time" to move across the country. I could think of a hundred reasons to stay with my family in Kentucky. What if I got really ill so far from home? I didn't know anyone in the Northeast. I didn't know much about the CF center in New Hampshire. What if I couldn't work and had to go part time? Those thoughts quickly fled from my mind because I have never been one to turn down a challenge. I was absolutely positive that this job was perfect for me. After graduation, I packed my bags and my three-legged dog Teddy, and was off to Nashua.

Moving to Nashua to work for BAE Systems was the best decision I could have made. If I had waited for the "right time" to move, then the opportunity would have passed. I would not be working on electronic warfare. I would not be networking with the highest ranking executives in the company. I would not have developed into the person I am today.

If you want to float through life, waiting for the right time to take a leap, then you will never find it. There will always be reasons that now is not a good time. Go on the trip you've always planned on. Go back to school and get your dream degree. Run that marathon. Move to the location you've always wanted to. Go skydiving. Go ask your boss about a promotion. There will always be an excuse. If you have CF, then you have countless excuses to wait until you feel better to do something.

As our disease progresses, it only becomes harder to do the things we've always hoped to do. Life does not slow down for us, so don't wait for the "right time." The right time is now. ▲

Kelsey is 25 and has CF. She grew up in Anchorage, KY, with an older brother and her mom and dad. She recently moved to Nashua, NH, for a job in the aerospace and defense Industry. She graduated from the University of Kentucky, Magna Cum Laude, in Mechanical Engineering. She has a three-legged dog named Teddy. She loves to play sports, especially soccer. She also loves to read and listen to classic rock records.

SIX Ways To PAY IT FORWARD To CF ROUNDTABLE!

May is CF Awareness month. What better way to pay it forward than by supporting *CF Roundtable*, a vital organization in the CF community. Please consider making a tax-deductible donation today.

With your help, our mission to support the CF community can be kept strong through this newsletter, social media, blogs, scholarships, awards and free community speaking engagements.

Please consider Paying It Forward in these six ways:

1. **Unrestricted Gifts**.
2. **Milestone Celebrations**. There is no greater reward than celebrating YOU and special events in your life.
3. **Tribute Gifts**. Donate to honor or remember those you love.
4. ***Endowment Fund.** These donations help keep *CF Roundtable* sustainable in order to continue our mission to you and the CF community.
5. **Matching Gifts** by your employer are always greatly appreciated.
6. ***Bequest** – a consideration for your estate planning.

*For contributions regarding Endowment and Bequests or for any questions, please contact us at cfroundtable@usacfa.org.

To make a donation **ONLINE**, go to www.cfroundtable.com and click on DONATE NOW. Or **MAIL** us with the information below.

USACFA, Inc., proudly produces *CF Roundtable,* a newsletter for adults who have cystic fibrosis.
www.cfroundtable.com ▲ cfroundtable@usacfa.org

CF Roundtable

Cut at dotted line, fill out form below and mail to: USACFA, Inc., PO Box 151676, Austin, TX 78715

This donation is for: In Honor/In Memory/Milestone** Name of person

Age Address City/State Zip Ccde

If applies – date and #years of milestone** Person/family to notify if "In Memory"

**Please specify your Milestone: includes Birthday, Anniversary of Wedding or Transplant, Marriage, Birth, Other.

Address City/State Zip Code

Donation Amount: Comments:

Please make checks payable to USACFA

FAMILY MATTERS

A Kalydeco Pregnancy

By Cindy Baldwin

This article is not meant as medical advice. Every person's case is different. It is extremely important to discuss the best course of action with your CF team, if you become pregnant while taking Kalydeco, Orkambi or Symdeko.

As a CF adult with G551D and ΔF508 mutations, I was one of the first patients to get access to Kalydeco when it received FDA approval in early 2012. My doctors and I expected Kalydeco to boost my lung function numbers and stabilize my health — what we didn't expect was that it would also end my infertility!

My husband and I had been trying to get pregnant for about a year and a half at that point, without success. In addition to thick cervical mucus (common in women with CF), I have never had regular cycles and frequently went three months or more without a period. At age 23, the year before Kalydeco was released, my primary care physician tested my estrogen and progesterone and found that they were so low I was almost in perimenopause. To everyone's surprise, beginning Kalydeco was enough of a correction for all of these issues, so that within the first three months of starting the drug, I became pregnant twice (the first was a very early miscarriage).

As soon as I got the positive test, my first call was to my CF care team. Although they knew I'd been trying to get pregnant for a while, and although we'd discussed a possible future where I might become pregnant while taking Kalydeco, none of us had expected it to happen immediately. At that time, nobody in the world had given birth while taking Kalydeco; I'd only heard of one woman who had already become pregnant while on it, and she was only a few months along. (Later, I discovered two more women who had been pregnant on Kalydeco around the same time as I had, but I didn't connect with any of them until after we had all delivered.)

Because of my experience as one of the first women in the world to be pregnant on a Vertex small-molecule corrector, and because I've written about it both on my blog (blog.cindybaldwinbooks.com) and CFMothers.com, I get regular e-mails from women who have found out they're pregnant while on Kalydeco or one of its successors. As a CF patient myself, I'm not in any place to give medical advice! However, for today's column, I thought I'd share how I made my decision and why I ultimately decided to remain on Kalydeco through my pregnancy — and later for two years of breastfeeding.

> *"I thought I'd share the things that helped me come to the decision to remain on Kalydeco through my pregnancy and breastfeeding."*

CINDY BALSWIN

As I mentioned, as soon as I'd gotten a positive pregnancy test, I called my CF team. Within a few hours, my doctor had called me back. My doctor at the time was fairly conservative when it came to medication, and was extremely uncomfortable with the idea of me remaining on Kalydeco, but he was equally uncomfortable with me coming off it. There is a growing body of belief both among CF doctors and CF patients that going off Kalydeco after you've started the therapy can lead to very fast declines and sometimes permanent damage, something that neither of us wanted me to experience at the start of a pregnancy. In addition, because my hormone levels had been too low to sustain a pregnancy before starting Kalydeco, we both thought that there was a fairly high chance that stopping it could lead to a miscarriage.

We had a long conversation about all the potential risks with either choice. Ultimately, my doctor didn't feel comfortable advising me either way; he said that it was my choice to make, and he'd support me in whatever I decided. (We also briefly considered stopping Kalydeco just for the first trimester and then resuming it, but agreed that it would still carry the risk of serious lung illness or miscarriage.)

In the end, after talking to my doctor, talking with my husband, and reading the scant amount of literature available about Kalydeco and pregnancy, I decided to remain on it for the duration of my pregnancy. While each case is different, and any CF woman who becomes pregnant on a new medication will have her own set of pros and cons to weigh, I thought I'd share the things that helped me come to the decision to remain on Kalydeco through my pregnancy and breastfeeding.

• One of the first places I turned when I found I was pregnant was to the Kalydeco package insert, which contained the only existing pregnancy information at that time. Kalydeco and Orkambi were both placed in the FDA pregnancy risk category B, indicating that while there was no research on humans, animal pregnancy and lactation studies showed no evidence of fetal abnormalities even at many times the recommended human dose. (Currently the FDA is in the process of phasing out the use of pregnancy risk categories, and Symdeko has not been assigned a risk category. However, the Symdeko animal study results are similar to that of Kalydeco and Orkambi, and can be seen in Section 8 of the Symdeko package insert.) Although rodent studies aren't always indicative of how a drug will perform in a human pregnancy, it was comforting to know that there had been no teratogenic effects in rodents even at very high doses of Kalydeco.

• With CF pregnancy, every medication is a risk/benefit analysis, and many CF women don't realize that some of our must-have medications — like digestive enzymes — are actually potentially less safe for a developing fetus. Most pancreatic enzymes were previously classed in pregnancy category C, due to a lack of animal testing altogether. Other medications, like intravenous tobramycin, have shown a positive evidence of fetal harm. In staying on Kalydeco, I hoped that a less-risky drug might help me avoid a more-risky drug, like tobramycin, later on.

• There is increasing research on "ivacaftor withdrawal syndrome" in patients who have previously been taking Kalydeco and then have to stop. Although much of this was anecdotal in 2012 when I became pregnant, there is now real research on the phenomenon, with at least one paper published in 2018 documenting the effect on patients coming off Kalydeco quickly. Each patient described in the paper immediately and rapidly deteriorated. Knowing this was a possibility, my doctors and I both felt like such a fast and steep decline would be a bad start to a pregnancy.

In my case — though this is somewhat unique — because my extremely low hormone levels were thought to have resulted from CF, and because they had risen enough for me to get pregnant upon starting Kalydeco, my care team and I both felt there was a high chance of miscarriage if I were to stop Kalydeco treatment. (Even with Kalydeco, I required supplemental progesterone through the first trimester to maintain my pregnancy.) I haven't heard of any other cases of CF infertility that were quite like mine, but there is now a fairly clear link between Vertex correctors and a boost in female fertility, so it's possible this might be a factor for other women as well.

Many years after my 2012 pregnancy, there is now a small community of women who have successfully been pregnant on Kalydeco and Orkambi (I don't know of anyone who has yet been pregnant on Symdeko), and a fair amount of anecdotal evidence for the possibility of their safe use while pregnant and breastfeeding. My daughter, now six, was a healthy and happy baby who weighed a whopping nine and a half pounds! In addition, I was able to avoid IV antibiotics and hospitalization for the duration of my pregnancy, which was the longest I'd stayed off IVs in years. While choosing to be pregnant on a Vertex corrector may not be the right answer in all cases, and while any CF woman who finds herself in that situation will need to consult her doctor, I'm very grateful that Kalydeco helped me to have a stable, healthy pregnancy! ▲

Cindy is 30 and has CF. She lives just outside Portland, OR, with her husband and spitfire of a daughter (who never makes CF care easy!). Her debut novel, Where the Watermelons Grow, *was published by HarperCollins Children's Books in 2018. You may contact her at cbaldwin@usacfa.org.*

Cystic Fibrosis in the Wild

Has your child ever wondered what it would be like to move like an animal? To hop like a kangaroo or swim like a fish? To crawl like a crab or climb like a monkey? In Jerry Cahill's latest children's book, the young CF patients learn how to stay active and exercise by mimicking the movements of animals… they get to see *Cystic Fibrosis in the Wild.*

IN THE SPOTLIGHT
With Paul Quinton, Ph.D.

By Andrea Eisenman and Jeanie Hanley

By guest interviewer Kathy Russell

Paul Quinton is an icon in the world of adults who have CF. He is responsible for discovering one of the major advances in understanding the basic defect in CF.

Until he officially retired two years ago, he held the Nancy Olmsted Chair in Pediatric Respiratory Medicine and is Professor of Pediatrics at the University of California at San Diego and Professor of Biomedical Sciences at the University of California at Riverside, Emeritus.

His specialty is epithelial fluid and electrolyte transport physiology. Paul, who has CF himself, discovered that the basic defect in CF is due to anion impermeability and not defective anion exchange.

Paul has a wicked sense of humor and is a very interesting person. While recovering from the effects of a stroke, he agreed to answer some questions for us.

Meet Paul Quinton. Spotlight, please.

Age: I am 74.498 years of age.

Hometown: Southeast Texas, near Houston.

Are you married? If so, how long?

Twice — first for 20 years; second for 25 years.

How was your CF diagnosed?

Girls!!! As a sophomore at the University of Texas (UT) Austin, I fell for a young lady and began to question why my lungs were messed up and how they might affect my marriage prospects. I went to the UT Research Library and began reading about lung diseases, until I came across a footnote that referred to "Cystic Fibrosis." This was 1964. I told my pulmonologist I had CF and he didn't believe me but referred me to a CF clinic. I went and, three sweat tests later, I had CF!

How is your health regarding CF?

Fairly stable. Kalydeco and Symdeco have really helped suppress infection and airway inflammation.

What other health issues do you have?

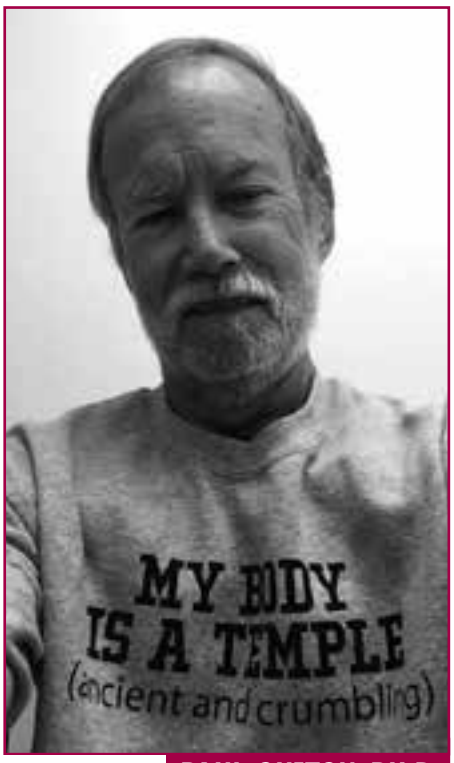
Chronic sinusitis, atrial fibrillation, post-stroke, essential hypertension, half deaf, BPH, Old Age!!! ... maybe dementia and tiny bladder for sure ... Shall I go on?

Any other health history of interest?

I've been through two upper lobectomies. 50-plus years ago it was the treatment for bronchiectasis.

What made you decide to have the Watchman procedure done?

I had an embolic stroke in the central cerebral artery. Later, it was determined that the most likely cause was atrial fibrillation with a PFO (patent foramen ovale). The left atrium has an appendage that is a small pocket in which blood can stagnate and clot when the atria fibrillate. Clots so formed can escape the left atrium via the PFO into the right atrium and then enter the arterial circulation directly causing an embolism to go somewhere such as the brain to cause a stroke.



PAUL QUITON, PH.D.

Tell us about your Watchman?

The Watchman is like a little umbrella placed in the mouth of the appendage to block it off, after which the endothelium should grow over it and completely seal it so that no blood can enter and stagnate. The alternative is to stay on anti-coagulants and not install the Watchman, but in the case of CF with a history of hemoptysis, anti-coagulants are a menace. Hemoptysis that doesn't clot and continues to hemorrhage makes it difficult to continue breathing.

Where do you work and what do you do?

I work at the University of California at San Diego, Pediatrics Department. I do some medical school teaching, but I do mostly academic basic research on sweat glands, bicarbonate secretion and interaction with mucins, fluid secretion and absorption in small airways.

How long have you been doing it?

Since about 1967.

Did you know what you wanted to be when you were growing up?

I said I wanted to be a rancher; still a wannabe cowboy.

What do you like most about what you do?

Discovering how it works. ("It" has a lot of definitions.) Discovery probably gives the greatest of all joy...watch a

two- or three-year-old.

What do you like least?

Administrative forms and asking for money.

You hold a Ph.D. How was it to go to school for so long?

It didn't seem so long. It was over quickly and comprised some of the best years of my life.

Did it affect your health?

I don't think it had much effect on my health; it more likely was the other way around. Understanding CF health was a reason to go to school.

Do you exercise? If so, what do you do?

I ride my bike to my office/lab about two miles each way with a couple of hills. I try to keep "chinning" (pull ups) at least four times in the morning to keep my biceps from becoming too spindly.

Are you religious or spiritual?

No, I've already been sent to Hell.

Have you ever considered having a transplant, if you needed one?

Not seriously. Not sure it is worth it at my age.

Do you know your genetic mutations?

DF508 and R117H/5T.

Are new correctors, such as the Vertex drugs, available to you?

Yes, I have been in trials for about five years, and yes, they have greatly stabilized my lung function.

Did you work on the research for any of these?

I did not work on developing these drugs per se, but we discovered earlier that the basic defect in CF is due to an impermeability to chloride. Correcting that defect was the test used to screen thousands of compounds as potential drugs, a few of which were effective and are now used or being tested clinically.

What do you do for fun?

Not much. I like to play with my grandson and granddaughter. I like to make people laugh, but not so good at it.

Do you have pets and why?

I always have had a dog and I love horses. Dogs teach to play and horses to work. I suspect animals teach us more about life and death than humans.

What or who helps you cope?

My wife, Liesbet, especially — more so when I'm not driving her crazy. Some friends, especially in the CF community, but one of the most significant problems I have experienced in this regard is the CF Foundation's mandate that people with CF stay away from their meetings, which not only labels me and others as unacceptable, but keeps us away from friends and support. It is a harmful policy that must receive more thought and consideration for well-being, and not just a hasty conclusion presumed to avoid infection. CFRI has adopted attendance policies that are compatible with health and well-being for all.

AUDREY RAE QUINTON CELEBRATING HER NUMBER TWO BIRTHDAY WITH HER SILLY GRANPAPA, PAUL QUINTON.

What is a perfect day for you?

A clear blue sky away from the city, followed by a clear silent night.

Do you have hobbies?

I like to try to write a poem now and then or make note of a new insight.

Do you have a funny story about CF?

This was the most difficult of the questions. Not much funny about CF. Maybe...when I was in elementary school, they began sending mobile x-ray machines to all the schools to screen for tuberculosis (TB). I had lung damage from chronic bronchitis and bronchiectasis, but we knew nothing of CF in those days. After I went through the x-ray scan, my parents received notice that I likely had TB, which was alarming in those days. After many tests, I proved not to have TB. So, after that, every time they announced that the mobile x-ray machine was coming to our school, I made sure to have a stomach ache on that day and stay home...not so funny!

Is there a motto by which you live?

"Believe none of what you hear and only half of what you see." — Benjamin Franklin. ▲

Kathy Russell is 74 (75, by the time you read this) and has CF. She is a former Director of USACFA and is Managing Editor of CF Roundtable. *Her contact information is on page 2.*

Jeanie Hanley is 56 and is a physician who has CF. She is a Director and the President of USACFA. Her contact information is on page 2.

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

TILLMAN continued from page 43

lence. While bacteria use other chemical molecules in signaling systems, the scientists say this is the first time that oxylipins have been shown to play a part in cell-to-cell communication in any bacterium. The researcher's findings suggest that disrupting oxylipin production by P. aeruginosa will defend against bacterial infection.

Bacteria mainly communicate via quorum sensing, which involves the production of various small molecules that function as "words" of the bacterial language. In P. aeruginosa, oxylipins function as new "words" in a novel quorum sensing system. This system controls the changing roles of genes in a bacteria community, turning some on and others off. In P. aeruginosa, the oxylipins are synthesized from oleic acid, a specific fatty acid, which is abundant in the tissue of a host organism. The scientists previously found that these oxylipin molecules play a key role in the formation of the biofilm that acts as a shield to protect P. aeruginosa from attack by the human immune system and from antibiotics.
https://tinyurl.com/y565yvmb

Can Vitamin D Supplements Help Cystic Fibrosis Patients?

While it's long been established that vitamin D deficiency is a common problem in patients with cystic fibrosis (CF), there has been little success following implementation of a number of recommended protocols for supplementation. A new study examines existing guidelines and what else can be done to improve treatment of vitamin D deficiency and reduce respiratory exacerbations. The study reviewed guidelines for vitamin D supplementation published by the North American Cystic Fibrosis Foundation (NACFF) in 2012 and found they were still insufficient to increase vitamin D to clinically beneficial levels in CF patients. The lead author of the report states that vitamin D deficiency is undertreated in patients with CF, even with current guidelines that already recommend high doses of supplementation. This study is the first to test the efficacy of the NACFF protocol and supports previous research suggesting much higher doses of vitamin D are needed for a clinical benefit in CF patients. Increased serum vitamin D levels are associated with fewer pulmonary exacerbations. This report is unique in that it shows the benefit of administering active vitamin D over inactive vitamin D, and correlates serum vitamin D levels to the number of days a CF patient may stay in the hospital. Researchers note that the role of vitamin D in preventing autoimmune disease, enhancing antimicrobial protection, and fighting viral infections has opened the door to more research that could further help CF patients avoid exacerbations. The study also suggests that malabsorption and a need for megadoses of vitamin D could be partly to blame for the lack of clinical efficacy of the NACFF protocol, as well as a lack of data on patterns of vitamin D metabolism in patients with CF compared with control groups.
https://tinyurl.com/y469tapu

Dual β-Lactam Combinations Highly Active Against Mycobacterium Abscessus Complex In Vitro

There has been a dramatic increase in chronic infections caused by Mycobacterium abscessus complex (MABC) strains that are usually recalcitrant to effective antibiotic therapy. The recent rise of macrolide resistance in MABC has further complicated this clinical dilemma. In this study, investigators have discovered that dual βlactams, and specifically the combination of ceftazidime with either ceftaroline or imipenem, are synergistic and have clinically relevant activities against clinical MABC isolates. Similar synergy was observed in time-kill studies against the M. abscessus ATCC 19977 strain using clinically achievable concentrations of either imipenem or ceftaroline as the addition of ceftazidime showed a persistent bactericidal effect over five days. This study's finding that there is synergy between certain β-lactam combinations against M. abscessus infection provides optimism toward identifying an optimum dual β-lactam treatment regimen.
https://tinyurl.com/y4wtza8c

Antifibrinolytic Agents For Hemoptysis Management In Adults With Cystic Fibrosis

Researchers created a clinical treatment pathway for inpatient and outpatient use that involved the use of antifibrinolytic agents to manage hemoptysis in cystic fibrosis (CF). They compared the rates of admission for bleeding before and after implementation of this pathway. They analyzed all adult CF patients who received systemic antifibrinolytic agents over a 54-month period in adherence with the treatment pathway. They obtained information regarding demographics, baseline CF-related characteristics, and bleeding and treatment parameters. Hospital admissions were reduced as a result of the implementation of a pathway utilizing systemic antifibrinolytic therapy to treat hemoptysis in CF patients. Findings revealed no serious adverse events.
https://tinyurl.com/y47d897y

Risk Of CF-Related Diabetes Not Linked To S. Maltophilia Infection, Study Says

Despite being commonly found in patients with cystic fibrosis-related diabetes (CFRD), the bacteria Stenotrophomonas maltophilia cannot be considered as an independent risk factor for developing CFRD. In recent years, the spectrum of microbes causing pulmonary infections in patients with cystic fibrosis (CF) has shifted dramatically. S. maltophilia (previously known as Pseudomonas maltophilia and Xanthomonas maltophilia) is a Gram-

negative qne multi-drug resistant bacteria that is estimated to affect up to 31 percent of all CF patients. The clinical significance of S. maltophilia remains controversial as no clear evidence for more rapid pulmonary decline following S. maltophilia acquisition has been found. It is therefore of interest to understand which factors affect the acquisition of S. maltophilia, and recently CFRD has been implicated. Based on the results of their study, the researchers concluded that although S. maltophilia is more common in people with CFRD, it is not an independent risk-factor for S. maltophilia acquisition.
https://tinyurl.com/y2n9wzn6

Non-Invasive Ventilation And Clinical Outcomes In Cystic Fibrosis

A reassessment of U.K. medical records showed that in cystic fibrosis (CF) patients, breathing support provided by gas inhalation, through a face or nasal mask, helps improve lung function but not survival, researchers report.
https://tinyurl.com/y2cowmw5

SOCIAL/EMOTIONAL

Social Support Is Associated With Fewer Reported Symptoms And Decreased Treatment Burden In Adults With Cystic Fibrosis. Kassie D. Flewelling, Deborah E. Sellers, Gregory S. Sawicki, Walter M. Robinson, Edward J. Dill. Journal of Cystic Fibrosis. Published online: February 13, 2019

The current study explored the relationships between social support, mental health, physical health, treatment activity, and disease-specific quality of life in a sample of adults with CF. The findings indicate that social support was associated with fewer mental and physical health symptoms; those with more social support reported less treatment burden; more social support was related to better emotional, social, and role functioning; and social support predicted improved vitality, body image, and health perceptions.
https://tinyurl.com/yy5zzd8z

Breathlessness Catastrophizing Relates To Poorer Quality Of Life In Adults With Cystic Fibrosis. Danijela Marasa, Louise Balfoura, Giorgio A. Tascaa, Ena Gaudetb, Shawn D. Aaronb, William D. Cameronb, Smita Pakhale. Journal of Cystic Fibrosis. January 2019 Volume 18, Issue 1, Pages 150–157

Treatment advances have resulted in increased life expectancy and a subsequent need to better understand psychosocial issues associated with quality of life in adults living with CF. Emerging research suggests that anxiety and depression are related to poorer health-related quality of life (HRQoL) in CF patients, but there is little research examining cognitive processes, such as breathlessness catastrophizing. The present study addresses this gap. Results indicate that increased breathlessness catastrophizing was related to poorer HRQoL, after controlling for lung function, depression, anxiety, and pain. Depression, pain, and breathlessness catastrophizing all emerged as significant unique predictors of HRQoL. Thus, breathlessness catastrophizing is a potential target for clinical intervention and might impact HRQoL. Further research on breathlessness catastrophizing in CF is warranted including longitudinal studies to examine the mechanisms by which breathlessness catastrophizing relates to HRQoL and treatment outcomes in CF.
https://tinyurl.com/y4fs8btz

Measuring Recovery In Health-related Quality Of Life During And After Pulmonary Exacerbations In Patients With Cystic Fibrosis. Patrick A. Flume, Ellison D. Suthoff, Mark Kosinski, Gautham Marigowda, Alexandra L. Quittner. Journal of Cystic Fibrosis. Published online: December 23, 2018

Continued on page 56

TILLMAN continued from page 55

The time-dependent impact of pulmonary exacerbations (PEx) on health-related quality of life (HRQoL) using Cystic Fibrosis Questionnaire–Revised (CFQ-R) data from two large cystic fibrosis (CF) trials was explored. Findings show that pulmonary exacerbations have significant effects on multiple domains of HRQoL, and recovery across multiple domains post-PEx can take several weeks. These findings provide insight into the impact of PEx on patient HRQoL and recovery post-PEx.
https://tinyurl.com/y3msyor8

NUTRITION

A Survey Identifying Nutritional Needs In A Contemporary Adult Cystic Fibrosis Cohort. Siddhartha G. Kapnadak, Kathleen J. Ramos, Andrea M. Lopriore, Christopher H. Goss and Moira L. Aitken. BMC Nutrition. 20195:4

Malnutrition in the CF population has been considered a critical problem requiring clinical intervention and care. More recently, excess weight among CF patients has also become a matter of concern, in particular in patients with less severe disease. This change in the nutritional status of CF patients may be due to aggressive caloric supplementation, suboptimal food choices, lack of exercise, and medications. Maintaining a healthy weight may be a challenge for CF patients due to their bodies' impaired absorption of nutrients. Patients with higher BMIs were more concerned about preventing weight gain, while patients with lower BMIs were more focused on strategies to prevent weight loss. Additionally, CF patients' primary concerns were different between males and females. The preferred choice for health-improvement programs was online access to CF nutrition and fitness information, and the second choice was financial assistance for food and supplements. Overall, the study revealed a wide variety of nutritional needs in CF adults including a high prevalence of overweight status, many patients desiring weight loss, and many seeking financial resources. These findings support the individualization of modern-day CF nutrition programs and development of online resources, in an effort to address the heterogeneous barriers that exist in the contemporary CF population and improve outcomes in patients with the disease.
https://tinyurl.com/yxnkeltm
AND
https://tinyurl.com/yce3gkt3

Resistin Is Elevated In Cystic Fibrosis Sputum And Correlates Negatively With Lung Function. Osric A. Forresta, Daniel M. Chopykc, Yael Gernezd, Milton R. Browna, Carol K. Conradd, Richard B. Mossd, Vin Tangprichac, Limin Pengf, Rabindra Tirouvanziam. Journal of Cystic Fibrosis. January 2019 Volume 18, Issue 1, Pages 64–70

Resistin is an immunometabolic mediator that is elevated in several inflammatory disorders. Resistin modulates the recruitment and activation of myeloid cells, notably neutrophils. Neutrophils are major drivers of cystic fibrosis (CF) lung disease, in part due to the release of human neutrophil elastase- and myeloperoxidase-rich primary granules, leading to tissue damage. Researchers assessed the relationship of resistin to CF lung disease. Plasma resistin levels were only marginally higher in CF than in healthy control subjects. By contrast, sputum resistin levels were very high in CF. Among CF patients, higher plasma resistin levels were associated with allergic bronchopulmonary aspergillosis, and higher sputum resistin levels were associated with CF-related diabetes. Overall, sputum resistin levels were negatively correlated with CF lung function, independently of other variables (age, sex, and genotype).

https://tinyurl.com/y2ls2xk8

TREATMENT

Antimicrobial Susceptibility Testing (AST) And Associated Clinical Outcomes In Individuals With Cystic Fibrosis: A Systematic Review. Ranjani Somayajia, Michael D. Parkinsb, Anand Shahc'd, Stacey L. Martinianoe, Michael M. Tunneyf, Jennifer S. Kahleg, Valerie J. Watersh, J. Stuart Elborndi, Scott C. Belli, Patrick A. Flumej, Donald R. VanDevanter. Journal of Cystic Fibrosis. Published online: January 29, 2019

Antimicrobial susceptibility testing (AST) is a cornerstone of infection management. Cystic fibrosis (CF) treatment guidelines recommend AST to select antimicrobial treatments for CF airway infection, but its utility in this setting has never been objectively demonstrated. Researchers conducted a systematic review of primary published articles designed to address two PICO (patient, intervention, comparator, outcome) questions: 1) "For individuals with CF, is clinical response to antimicrobial treatment of bacterial airways infection predictable from AST results available at treatment initiation?" and 2) "For individuals with CF, is clinical response to antimicrobial treatment of bacterial airways infection affected by the method used to guide antimicrobial selection?" Relationships between AST results and clinical response (changes in pulmonary function, weight, signs and symptoms of respiratory tract infection, and time to next event) were assessed for each article and results were compared across articles when possible. They discovered little evidence that AST predicts the clinical outcome of CF antimicrobial treatment, suggesting a need for careful consideration of current AST use by the CF community.
https://tinyurl.com/y3cahbvt
AND
https://tinyurl.com/y5en4hhy

Effectiveness Of Ivacaftor In Cystic Fibrosis Patients With Non-G551D Gating Mutations. Jennifer Guimbellota,b,1, George M. Solomona,c,1, Arthur Bainesd, Sonya L. Heltshed,e, Jill VanDalfsend, Elizabeth Joselofff, Scott D. Sagelg, Steven M. Rowe. Journal of Cystic Fibrosis. January 2019 Volume 18, Issue 1, Pages 102–109

The cystic fibrosis transmembrane conductance regulator (CFTR) potentiator ivacaftor is approved for patients with CF with gating and residual function CFTR mutations. The results of an observational study investigating its effects in CF patients with non-G551D gating mutations indicate that these patients experienced improved lung function, nutritional status, and quality of life. This study supports ongoing use of ivacaftor for patients with these mutations.
https://tinyurl.com/y2u3yrjv

Investigating The Effects Of Long-term Dornase Alfa Use On Lung Function Using Registry Data. S.J. Newsome, R.M. Danielb, S.B. Carrc, D. Biltond, R.H. Keogh. Journal of Cystic Fibrosis. January 2019 Volume 18, Issue 1, Pages 110–117

Dornase alfa (DNase) is one of the

Continued on page 58

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

Janie Davies
Arlington, TX
72 on January 16, 2019

Andrea Eisenman
New York, NY
54 on November 28, 2018

Lynn Pancoast
Allentown, PA
60 on October 26, 2018

Patricia Spadafora
Baldwin, NY
64 on December 4, 2018

TILLMAN continued from page 57

commonest cystic fibrosis (CF) treatments and is often used for many years. However, studies have not evaluated the effectiveness of its long-term use. Findings indicate that DNase improved lung function in individuals with reduced lung function, bringing a step-change in 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 FEV1 > 70%.
https://tinyurl.com/y4tttlv2

Repeated Hot Water And Steam Disinfection Of Pari LC Plus® Nebulizers Alter Nebulizer Output. Melanie SueCollins, MatthewO'Brien, Craig M.Schramm, Thomas S.Murray. Journal of Cystic Fibrosis. Published online 14 September 2018

Currently, cystic fibrosis patients require daily nebulized treatments to achieve optimal lung health. Growth of pathogenic bacteria in patient nebulizers is well known, and disinfection guidelines have been established. The authors sought to discover what effect, if any, repeated nebulization/disinfection cycles had on nebulizer output. Saline was nebulized repeatedly after exposure to boiling water, steam, and alcohol disinfection methods. While alcohol disinfection did not affect nebulizer output, boiling water and steam significantly decreased nebulizer output from baseline. This decrease in nebulizer output could significantly increase the duration of nebulizer treatment time and negatively impact the burden of care on patients with cystic fibrosis.
https://tinyurl.com/y3vh55kw

SINUS

Outcomes Of Endoscopic Sinus Surgery In Adult Lung Transplant Patients With Cystic Fibrosis. Paolo Luparello, Maria S. Lazio, Luca Voltolini, Beatrice Borchi, Giovanni Taccetti, Giandomenico Maggiore. European Archives of Oto-Rhino-Laryngology. pp 1–7. First Online: 28 January 2019

Chronic rhino sinusitis (CRS) associated with or without nasal polyposis usually occurs in adults and affects close to one-half of all CF patients. The goal of this study was to evaluate the impact of Endoscopic Sinus Surgery (ESS) in the quality of life (QoL) of the CF patients and demonstrate an improvement of the functional outcomes in the patients who underwent the surgical procedure rather than in those not treated, particularly in lung transplant patients. The investigators found that ESS represents the best option to improve clinical QoL of CF patients who do not respond to conventional medical therapy, with a stabilization of respiratory function after transplantation.
https://tinyurl.com/yy2n48nk

DIABETES

Association Between HbA 1c And The Development Of Cystic Fibrosis-related Diabetes. Choudhury M, Taylor P, Morgan P, Duckers J, Lau D, George L, Ketchell R, Wong F; Diabetic Medicine (Jan 2019)

This study aimed to examine HbA1c as a predictor of risk for future development of cystic fibrosis-related diabetes (CFRD) and to assess the association with the development of retinopathy in people with CFRD. A 7-year retrospective longitudinal study was conducted in 50 adults with cystic fibrosis, comparing oral glucose tolerance test results with HbA1c values in predicting the development of CFRD. Retinal screening data were also compared with HbA1c measurements to assess microvascular outcome. The researchers concluded that there is a link between HbA1c level and the future development of dysglycaemia in cystic fibrosis based on the oral glucose tolerance test, as well as microvascular outcomes. Although current guidance does not advocate the use of HbA1c as a diagnostic tool in CFRD, it may be of clinical use in determining individuals at risk of future development of CFRD.
https://tinyurl.com/y3cmvmf8
AND
https://tinyurl.com/y5o4ya32
AND
https://tinyurl.com/yxozbvuv

FYI

Clinical Manifestations And Risk Factors of Arthropathy In Cystic Fibrosis. Jobst F. Roehmel, Tilmann Kallinich, Doris Staab, Carsten Schwarz. Respiratory Medicine 147 · February 2019

Recurrent joint pain is frequently observed in patients with CF and can lead to reduced activity and quality of life. This observational study was conducted to assess the clinical manifestations, frequency, and risk factors of CF associated arthropathy. Of 186 patients included in the study, 54/186 had experienced joint symptoms. Joint pain and swelling were the most frequent symptoms. The joints of the hands (JOH) followed by the joints of the feet were most affected. No specific pattern of autoantibodies was discovered. Joint symptoms in CF are a frequent and clinically relevant phenomenon with a distinct clinical pattern. Pulmonary exacerbations and elevated levels of total serum IgG may reflect chronic inflammation in patients with CF and may lead to a specific arthropathy associated with this condition. Older age, gender, aspergillus colonization, elevated serum IgG and pulmonary exacerbations were identified as risk factors.
https://tinyurl.com/yxggwslw

Antifibrinolytic Agents For Hemoptysis Management In Adults With Cystic Fibrosis. Hanny Al-Samkari, Kelly Shin, Lauren Cardoni, Emily H. Pighetti, Jean M. Connors. Chest. Available online 18 February

CF ROUNDTABLE SUBSCRIPTION FORM (Please Print Clearly)

PLEASE CHECK ONE: ☐ **NEW SUBSCRIPTION** **OR** ☐ **UPDATE**

NAME _____ **PHONE (** **)** _____

ADDRESS _____

CITY _____ **STATE** _____ **ZIP+4** _____ **+** _____

E-mail address: _____

I am interested in *CF Roundtable* **because I am a:** CF ADULT _____ BIRTHDATE _____

OR I am a: Parent ____ Relative ____ Friend ____ Medical/Research Professional ____ Caregiver ____

CF Roundtable is available at no cost. However, to help defray printing and mailing costs, donations are always welcome.

- Donation to defray costs . . . . . $ _____
- Please send me the back issues I have listed on a separate sheet of paper.
- Please send subscriptions to the names and addresses I have listed on a separate sheet of paper.

Please do not write in this space

SP 19

For donations, make checks payable to USACFA and mail with this completed form to:
USACFA, P.O. Box 151676, Austin, TX 78715-1676
We ask that everyone keep their home and e-mail address information up to date.

OR

You can also subscribe online! Go to www.cfroundtable.com. You can now scan this QR Code with your smart phone to go directly to our online registration page. By subscribing online you have the ability to download a PDF of the latest newsletter, receive the mailed version or you can receive both. The online version will be available two weeks prior to the mailed version.

KEEP YOUR INFORMATION CURRENT

To keep our records up to date, please be sure to complete and return a subscription form, on this page, to us or register online with any changes to your information, www.cfroundtable.com. (Any issue of the newsletter that is returned to us costs us about two-and-a-half times the first-class postage rate for that piece. Currently that runs about $3.73 per returned copy.) **Thank you for helping us with this.**

2019

Hemoptysis is a major cause of morbidity and mortality in patients with cystic fibrosis (CF). Antifibrinolytic agents have demonstrated efficacy in a broad range of bleeding disorders and conditions. The authors examined the use of antifibrinolytic agents to manage hemoptysis in CF. They developed a clinical treatment pathway for inpatient and outpatient use, and report on rates of admission for bleeding before and after implementation. Their findings show that a pathway utilizing systemic antifibrinolytic therapy to treat hemoptysis in CF patients was associated with a reduction in hospital admissions. No serious adverse events were observed. https://tinyurl.com/y62qzaul ▲

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 any time by e-mail at **cfroundtable@usacfa.org**
- Send your questions of a general nature regarding legal issues that relate to CF to our legal advisor: **Beth Sufian, Esq., 712 Main, Suite 2130, Houston, Texas 77005. E-mail: CFLegal@sufianpassamano.com.**
- You may subscribe at www.cfroundtable.com

PRINTED WITH SOY INK

Published by the United States Adult Cystic Fibrosis Association, Inc.
CF Roundtable is printed on recycled paper.

IMPORTANT RESOURCES

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

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

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

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

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