CF Roundtable
A NEWSLETTER FOR ADULTS WHO HAVE CYSTIC FIBROSIS
AUTUMN 2017

CF Gastrointestinal Myths Debunked

By Michelle Stroebe, M.S., R.D.


MICHELLE STROEBE

I don't know about you, but I can get lost in complicated language or procedure talk with my physician. The field of medicine is complex and often we are pressed for time during an appointment; therefore, clarification of medical jargon may be on the bottom of your list. I try my best each day to make sure my CF patients have the opportunity to ask questions, and to make sure they understand the what and why of everything I ask of them. I have found there are a lot of misconceptions and myths about gastrointestinal (GI) related symptoms, and that these misconceptions may lead to trouble down the line. Having a better idea of how CF affects the GI tract is the first step to understanding what you can – and cannot – do to prevent discomfort, pain or embarrassing situations from happening.

As a cystic fibrosis dietitian, most of my workday consists of talk about poop. While I recognize this is quite different from the typical Silicon Valley office talk, in my office, bowel habits and GI symptoms may be our first glance at what is happening internally and are extremely important to discuss. At first, new CF patients at our center are shy about sharing their bathroom habits with a stranger. However, once they understand why I am asking ten plus questions on bathroom habits, they are eager to share it all: the good, the bad and the ugly of their GI tract.

Dietitians and other clinicians talk a lot about the GI tract. You may be asking yourself, what exactly is this?! The GI tract is the digestive tract, which starts from your mouth through your anus and includes all accessory organs needed to digest food and process waste. Essentially, it is where food is processed and turned into nutrients your body needs to do the things you want to do, from walking and talking to pumping the blood through your veins and keeping your heart beating. Several organs in the GI tract are affected by CF, including the pancreas, liver and colon. Dysfunction in these organs can lead to issues including pancreatic insufficiency, CF-related diabetes, CF liver disease or constipation. Careful monitoring of GI symptoms (including, you guessed it! your bowel movements) let us know if these organs are function-

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EDITOR'S NOTES

I hope that each of you survived the weather that has been plaguing much of our country. I know that many of you have had difficulties with getting your meds and treatments. I wish you a winter that is better.

This issue is packed with many good articles right from the front page where **Michelle Stroebe** debunks some myths about our gastrointestinal systems. **Mark Schroeder** continues that discussion in "Voices From The Roundtable." Other "Voices" include **Barbara Harison** telling of aging with CF, **Leah Sands** who relates two years on a new drug and **Linda Stratton** who talks about changes in her own care.

Beth Sufian discusses both short- and long-term disability in "Ask The Attorney." "Active For Life" finds **Aimee LeCointre** writing of her workouts with a partner, **KC Velez**. As always, **Laura Tillman** has compiled a list of "Information From The Internet." "Searching For The Cure" has **Meranda Honaker** answering FAQs about clinical trials, and **Reid D'Amico** offers a list of some clinical trials that may interest you.

The Focus topic of this issue is Dating And Relationships With CF. We are fortunate to have many excellent articles: **Sarah Albright** writes of finding love when you have a chronic illness; **Ella Balasa** asks if we are that different; **Tabby Caldwell** tells us of growing with CF; **Reid D'Amico** says it takes time to figure out dating and CF; **Nicole Kowal** writes of dating care free; and **Sydna Marshall** says to love me or not.

The "Conversation Corner" has a suggestion by **Paul Quinton** about a safer way to greet people. **Joan Finnegan Brooks** tells us of another way to connect with others who have CF. **Colleen Veitengruber** talks of embracing life.

Caleigh Haber is "In The Spotlight." **Dana Giacci Rogers** shares thoughts from **Kaeti R.** in "Parenting." In "Spirit Medicine" **Isabel Stenzel Byrnes** speaks of our relationships with life interruptions. In "Wellness" **Julie Desch** reminds us to be kind to ourselves.

We have two reviews from the CFRI Conference. **Jeanie Hanley** talks of making new friends and renewing old friendships, and summarizes several presentations. Physical Therapy and its importance to CF is the presentation that **Laura Mentch** writes about.

Look at page 26 to see **Jacob Greene** and **Elizabeth Shea**, who are the two recipients of the autumn 2017 USACFA scholarships. Congratulations to them!

Be sure to check out the "Looking Ahead" box on the next page. See if there is an upcoming topic that is of interest to you. If so, please write about it; we love to hear from you.

Stay healthy and happy,

Publication of *CF Roundtable* is made possible by donations from our readers and grants from Sustaining Partners - AbbVie, Boomer Esiason Foundation, Cystic Fibrosis Foundation, Cystic Fibrosis Services/Walgreens, Foundation Care, Gilead Sciences, Hill-Rom, Kroger Specialty Pharmacy and Two Hawks Foundation in Memory of Dr. Lisa Marino; Pearl Sustaining Partners - Marshall & Margherite McComb Foundation; Diamond Sustaining Partners - Nancy Wech (in memory of daughter, Lauren Melissa Kelly & in honor of son, Scott Kelly).

Information From The Internet...

Compiled by Laura Tillman

PRESS RELEASES

Novoteris, LLC, Receives FDA and Health Canada Clearance to Start a Phase 2 Clinical Trial of Its Thiolanox® Nitric Oxide for the Treatment of Cystic Fibrosis

Novoteris will begin recruiting cystic fibrosis (CF) patients for a Phase 2 clinical trial of its inhaled nitric oxide product Thiolanox. Nitric oxide is a naturally occurring gas that can reduce lung infections, leading to improved respiratory function in people with CF. The gas can counter gram-positive and gram-negative bacteria, viruses, fungi and yeast. Gram-positive bacteria are more susceptible to antibiotics than gram-negative bacteria. And gram-negative bacteria are more likely to develop resistance to the drugs. Patients will be treated with either Thiolanox nitric oxide or a placebo. Novoteris will deliver both with its computerized trace-gas mixing system. The primary goal of the trial is to compare the treated and placebo groups' change in lung function between the start of the therapy and the 15th day. The broad-spectrum antimicrobial property of nitric oxide widens the enrollment eligibility of subjects in this Phase 2 trial with the only exclusion being for Non-Tuberculosis Mycobacterium (NTM), which will be studied in separately planned trials. Gaseous nitric oxide's potent antimicrobial properties, lack of bacterial resistance and small molecule penetration capabilities could provide a promising alternative, non-antibiotic approach to treating infections in people living with the disease.
http://tinyurl.com/y9ybernq

Synspira's CF Therapy May Be Game-Changer for Drug-Resistant Bacteria

Chronic pulmonary infections are a hallmark of lung disease in patients with cystic fibrosis. Due to the variability in bacteria that can colonize the lung and the specific antibiotic treatments that may be effective, treatment strategies are often tailored to individual patients. But some bacterial infections are often difficult to treat across the CF population. Burkholderia cepacia complex (Bcc) is notoriously difficult to attack due to its natural multi-drug resistant state and its quick ability to develop resistance to antibiotics. Infections arising from Bcc can lead to permanent loss of lung function and early death in patients with CF. Synspira

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LOOKING AHEAD

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

Autumn (current) 2017: Dating And Relationships With CF.

Winter (February) 2018: Becoming A Parent With CF. (Submissions due December 15, 2017.) How is your CF affecting your decisions about becoming a parent? Whether you already have children or are wanting to start a family, tell us about your journey to parenthood.

Spring (May) 2018: Maintaining Mental Health With CF. (Submissions due March 15, 2018.) Does your CF affect your mental state? What do you do to deal with it? Do you have any information to share with our readers on how to deal with depression or other mental conditions that are caused by having CF?

Summer (August) 2018: What Is Most Important To You When A Disaster Strikes? (Submissions due June 15, 2018.)

ASK THE ATTORNEY

Short Term and Long Term Disability Questions Answered

By Beth Sufian, J.D.

Many readers have asked questions related to Short Term Disability (STD) and Long Term Disability (LTD).

The following column provides information on these two types of benefits offered by private companies. Nothing in this column is meant to be legal advice about your specific situation but is meant to be only information.

The CF Legal Information Hotline can answer questions related to insurance, SSA benefits, employment and education. E-mail CFLegal@sufianpassamano.com to schedule a time to speak with a member of the CFLIH staff.

While individuals who have no health problems may be able to purchase an individual short term or long term disability policy it is unlikely a person with CF will be able to purchase a policy on his or her own. There is no law that requires an insurance company to sell a short term or long term disability policy, to any person. The lack of legal mandate to sell a policy to anyone makes it likely an insurance company will reject applications for the purchase of policies by a person with CF.

Therefore, almost all people with CF who have an STD policy or LTD policy have such policies as an employee benefit offered by their employer. Occasionally a person who was diagnosed late in life may have purchased an STD or LTD individual policy before the person was diagnosed with CF.

I. General Disability Benefit Information

As more people with CF live longer, more people are able to work at full-time jobs. Sometimes an adult with CF finds he or she must stop full-time work to focus more time on his or her health.

People with CF are not alone in their need to stop work. More than one quarter of people entering the workforce today will become disabled before they reach age 65.

In the context of disability insurance benefits, the term "disabled" has a narrower meaning focusing on the individual's ability to work. In fact, the meaning of "disability" in the context of private disability insurance benefits is different from the meaning of "disability" in the context of Social Security.

When a person with CF considers stopping full-time work, the person is concerned with replacing his or her lost income and maintaining his or her health insurance coverage.

II. Short Term Disability Insurance Benefits

The first potential form of assistance is short term disability insurance (STD). Short term disability insurance pays a benefit when an employee is unable to perform the material and substantial duties of his or her job due to an illness or injury. Short term disability insurance is offered by some employers. Some employers pay the premiums for STD benefits, but often the employee has to elect STD or LTD benefits and pay the premiums for the STD or LTD policy. If the person with CF does not elect to purchase the STD or LTD policy and later becomes unable to work due to a decline in health, the person will NOT be entitled to a benefit.

BETH SUFIAN

Many people with CF say they did not think they would ever have to stop work and so they did not want to pay the $10 or $20 it cost to purchase STD or LTD benefits. These individuals understand they made the wrong choice after they become unable to work due to their health but have no STD or LTD benefits to help them replace lost income while they are unable to work due to illness. Short term disability policies typically pay a benefit that ranges from 40% to 60% of the covered employee's weekly income, but the amount of the benefit depends on the policy.

Short term disability policies are designed to assist a worker who may recover from his or her illness or injury in the short term and return to work. Because a return to work is anticipated, the worker typically remains an employee during the short term disability leave period. While on STD leave, the employee's health benefits (if any) usually remain effective because the worker remains an employee. However, the employer has no legal obligation to maintain employment or maintain the

> *"In the context of disability insurance benefits, the term "disabled" has a narrower meaning focusing on the individual's ability to work."*

worker's employee benefits, unless the employer is covered by the Family and Medical Leave Act.

III. Long Term Disability Insurance Coverage

If the worker is still unable to return to work after an STD benefit ends, then the person can apply for long term disability benefits if such benefit is offered by the employer. Long term disability insurance pays a benefit equal to a percentage of the worker's pre-disability earnings. While each policy has its own definition of disability, these policies generally require evidence that the worker is unable to perform the usual and customary duties of his or her own job because of an illness or injury. The policy may require the person applying for the benefit to show he or she cannot perform any job due to his or her illness.

In most cases, a worker cannot apply for long term disability until the short term disability benefit is exhausted and after an elimination period has expired. An elimination period is the period of time during which the individual is disabled but not receiving long term disability benefits. The actual policy is the best source of details on the terms and conditions of the policy. Even when a worker is enrolled in an LTD policy or plan, the worker must still meet all other conditions to receive a benefit, such as completing any elimination period.

Because long term disability policies have an elimination period, many employees fear that if employment is terminated during the elimination period, before they have an opportunity to apply for benefits, then they will lose the opportunity to apply for long term disability benefits. However, most policies only require the employee to become disabled while employed. If the individual is terminated during the elimination period, he or she is not precluded from applying for LTD benefits as long as he or she became disabled while employed.

Most LTD policies offset the monthly LTD benefit by other income replacement benefits received by the claimant, such as Social Security benefits, worker's compensation, railroad disability income benefits etc. A person with a benefit of 60% of pre-disability income and an annual income of $40,000 will have a monthly benefit of $2,000 before taxes and about $1,600 after taxes. When this amount is offset by a $1,200 monthly Social Security benefit, the net LTD benefit would be only $400 in this example.

Conclusion

Replacing lost income and maintaining health benefit coverage are the two things a person with CF wants to do when he or she stops work due to declining health. Short term disability and long term disability benefits can help supply the funds needed to pay living expenses and medical expenses after employment stops. ▲

Beth is 52 and has CF. She is an attorney who specializes in disability law and is a Director of USACFA. Her contact information is on page 2. You may contact her with your legal questions about CF-related issues.

YOU CANNOT FAIL

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

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

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

SPIRIT MEDICINE

The Spirit Of Interruptions

By Isabel Stenzel Byrnes

This issue's theme is about relationships. Since this newsletter is chock full of good stories about human relationships, I'd like to muse about our relationship with life's interruptions.

The inspiration for this topic comes from Dr. Martin Luther King's little known sermon called "Interruptions: The Man from Porlock." He preached this sermon at Ebenezer Baptist Church on January 21, 1968, in Atlanta, Georgia. I have used the transcript of this sermon for my writing through loss groups at work for several years. The story starts with how a man was writing a great piece of literature, and a man from Porlock knocked at his door. This man needed something and distracted the writer. The writer lost his train of thought; his creative inspiration was gone forever. He was interrupted.

Dr. King shares that to live a human life means we will experience interruptions to our lives. How we choose to deal with them will determine the quality of our lives. Dr. King shares examples of people who are broken by their interruptions. Some people become bitter and resentful for these uninvited changes. Others withdraw and become introverts in an effort to hide from others because of the pain of life's interruptions.

Living with cystic fibrosis means we will face interruptions on a regular basis. We are tested constantly on how we choose to deal with those interruptions. I'm blessed to be quite well and functional 13 years post-transplant. But this year alone, I was training hard for the Transplant Games and got hit hard by the flu. I was deeply immersed in work, music, my new hobby of cycling and had to have significant surgery on my eye for skin cancer, impacting my vision. Blockages, sugar lows, injuries, dehydration, infections... these are the normal interruptions of CF life. I have to pause, lay low, recover and then resume. One after another, my CF friends are also interrupted with health crises—many much more serious than mine. And we have to pick up and start up where we left off. It's easy to become depressed and helpless when there are so many interruptions. Why bother getting back on track if something else is around the corner?

> *"People just bounce back from an interruption in different ways, at different paces. But ultimately, how we deal with interruptions shapes our character."*

ISABEL STENZEL BYRNES

Sometimes, I get really frustrated with these interruptions. Perhaps it's our culture of control and self-determination... and I forget to be open to the natural unfolding of inevitable situations that arise that get in the way of what "I" want to do. Perhaps it is the dogged desire developed by a lifetime of health challenges that makes me just want to live my life, unencumbered, that makes yet another setback annoy the hell out of me. To deal with the anger, I'll push myself really hard to ignore or endure through the difficulty... until the health issue becomes serious enough and I have to address it. Sometimes, I throw up my hands and ask, "What's next?" Or, like in the case of my skin cancer, I'll resign with laughter, and joke, "Guess this is one more thing to die from!"

Dr. King preaches, "The way to deal with the interruptions of life is to face them as realities but then develop something on the inside of you that gives you the power to endure them and thereby transcend them." Ah, yes. This is the magic bullet.

How do we do that? How do we

change our relationship with interruptions?

After my initial reaction of disappointment, frustration, profanity and grief, I settle into myself. I look at the big picture. Here are some ways that help me change my relationship with interruptions:

1. I expect them, so I'm not caught off guard. I know my job, health and security are all temporary. I try to live in the moment, to enjoy what is good, now.

2. I'm grateful when they don't come. Phew. I acknowledge the good runs.

3. I notice positive interruptions. They're not all bad.

4. I stay calm. That's hard to do. I made the mistake of Googling my type of skin cancer and had a physical stress reaction. It took time and intention to relax my body.

5. I tell the truth. I remain emotionally authentic, and tell someone how the interruptions impact me. More accurately, I complain to those I trust.

6. I relax into them. The more I resist, the more exhausted and frustrated I get. Accepting them doesn't mean I want them. But this is just life.

7. I fall into proactive problem-solving mode. I do what I have to do to minimize the impact of the interruption. This includes communicating to those impacted, like co-workers.

8. I surrender to the interruptions. I give them up to God, the Universe, to my life story that's unfolding, and just allow what's not in my control to emerge.

9. And finally, I look inside to see what kind of person I want this challenge to make me. Do I want to waste energy protesting or save my energy to cultivate peace, strength, meaning, growth and wisdom?

Dr. King, back in 1968, preached about a trendy word these days: resilience. Resilience is the ability to bend with flexibility at forces that come at us, like a tree blown by strong winds. If the branches weren't flexible, they'd snap and break. They have to spring back to their upright nature after the winds die down. Dr. King knew that "interruptions are part of the scenery of life. Storms are part of the normal climate, like the ever flowing waters of the river, life has its moments of flood and its moments of drought." So every normal life will be faced with storms, and will be faced with ways to adapt. Everyone is resilient in their own way. People just bounce back from an interruption in different ways, at different paces. But ultimately, how we deal with interruptions shapes our character.

I love how Dr. King encourages all of us to use our internal force to cope with the storms of interruptions that come our way. He says, "You have something in there which says that I have a determination, an in-spite-of quality." As a preacher, he says this fire to keep going is "touched by the power of God" ... "a God who can transform dark yesterdays into bright tomorrows." A drive. A life force. It's in there.

I encourage you to listen to Dr. King's sermon, at this website below. To hear his powerful voice is what really moves the Spirit. Interruptions will continue to arise in our lives with CF. How will you change your relationship with them to deal more effectively with them? Where does your Spirit to keep going come from?

https://archive.org/details/cueth_000023

9:35"- 27" of Part 4 of 5, "Interruptions": A sermon delivered by Rev. Martin Luther King, Jr., at Ebenezer Baptist Church, Atlanta, Georgia. ("Martin Luther King Speaks") ▲

Isabel is 45 and has CF. She lives in Redwood City, California, with her husband, Andrew. She would like to thank her friend Olin Dodson, author of Melissa's Gift, for introducing her to "Interruptions."

TILLMAN continued from page 3

announced the results of a combination study using its inhaled polycationic glycopolymer drug candidate called PAAG and conventional antibiotics as a potential treatment for pulmonary infection caused by the Burkholderia cepacia complex in patients with CF. The investigative treatment, SNSP113, is a glycopolymer-based therapy being developed as an inhaled treatment to improve lung function in patients with pulmonary bacterial infections. SNSP113 is known to interact with the biofilms that make treating bacteria difficult in CF, and is thought to break the biofilms apart and reduce the viscosity and adhesion of mucus in the lungs. Biofilms and mucus lead to pulmonary exacerbations in CF, and SNSP113 may address both issues in cystic fibrosis. If approved, the treatment would not only open up treatment options for Bcc, but may also have positive outcomes when treating other common infections in CF, like Pseudomonas.

http://tinyurl.com/y8vuz5or

AND

http://tinyurl.com/ybjg7tef

Cystic Fibrosis Drug Bronchitol Succeeds in Phase 3 Study

An international Phase 3 trial of Bronchitol (mannitol, Pharmaxis Ltd.) in adults with cystic fibrosis (CF) has met its primary endpoint. Bronchitol is

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SPEEDING PAST 50

We Can Handle Embarrassment

By Kathy Russell

At last, after almost 100 days of heat, we have rain. Since June 15, we have had only about .005 inch of rain. Now we received almost half an inch in 24 hours. I am grateful. The rain cleared the air and helped to calm the wildfires. I had to stay inside for nearly four-and-a-half weeks because of the smoke from those fires. Our air quality in the Portland area was the worst in the entire country. Downtown Portland looked like Beijing! Not only is it raining at my elevation, it is snowing on Mt. Hood. That could bode well for an early ski season. Hooray.

Our Focus topic in this issue is Dating And Relationships With CF. Since I have been married for more than 52 years, my dating experience may not be of interest to most of our readers. I'll mention some of it, anyway. I didn't date much in high school. Mostly I went out with longtime friends. We didn't really talk about ourselves. We weren't looking for life partners, we were just going to school dances or games, seeing movies or visiting with other couples.

When I dated after I was out of high school, I didn't say anything about CF until I felt that we had some kind of relationship. Often I would go out with someone for only one or two dates. I felt there was no reason to mention CF to those people.

On the other hand, when I became more serious about a date, I would mention that I had CF and give a short explanation of what it was. If he was interested, he might ask questions and I would answer them to the best of my ability. There were only two men who showed real interest. I was engaged to each of them, eventually. I married the right one.

We have been selective about telling people about my CF. My standard answer, when anyone would comment on my cough was that I had allergies. Since I do have allergies, I wasn't lying. That would satisfy most people. Some of our friends with whom we've been close for decades are quite well educated on CF. Other friends know that I have it, but I doubt that they really understand about it and I am sure they don't actually care about it.

> *"Speaking of perspiration reminds me about dancing with a fella. I had to keep a handkerchief in my hand so that the guy's hand wouldn't get saturated with my sweat."*

My relationships with friends are fairly normal, except when it comes to health issues. Most of our family and friends know that they need to stay away from me when they are ill or are just getting over being ill. Also, they need to stay away when I am feeling less than great. I don't want to expend energy on trying to be social, if I am not feeling well. If truth be told, I am not big on being social. I am very choosy about being with anyone other than my husband. He doesn't need me to socialize and is content to let me sit quietly, which is my preference.

KATHY RUSSELL

The friends that I do see understand about why I don't shake hands. The good ones get elbow bumps. There are a few who still get a hug. Even my physician and I give elbow bumps. It just seems so much cleaner than handshakes. Maybe we could get everybody to change from handshakes to elbow bumps. That would be ideal. Although I must say that some people look at me with a strange expression when I say that I don't shake hands. Some people just don't get the whole cross-infection idea.

Which reminds me...when I would apply for a new job, I was right up front about my CF. I didn't want to have it come back and bite me in the rear-end

chassis because I failed to disclose a potential problem. It was surprising to me that so many educated healthcare people just discounted my statement of having CF because I was "too old" to have it. (Shows how much they know!)

In all fairness I should point out that even the CFF perpetuated the idea that CF was only for little children. There are many of us who have passed 50 and are doing okay. I understand the thinking behind the idea of using little kids in ads. Little children, especially cute little children, are much more apt to get donors to open their wallets and give generously to a charity. Who wants to give money to a charity that is looking for a cure for a disease that affects old people? As it happens, the families and friends of those old people want to give money to help find a cure for the ones they love. Even people who don't know anyone who has CF can still be willing to make a donation. I am so happy the CFF has come into the 21st century and has realized that CF isn't just for children.

Okay, so I got way off the subject, didn't I? In a way, I didn't. The perception that people have of CF depends largely on what the CFF says about it. So, when I tell someone that I have CF, if they believe only what they have read in information from the CFF in past years, I am too old to have it. That immediately colors their understanding of what I am saying. They know that I am too old to "really" have CF, so they doubt anything that I say. Usually, I am able to clear up their confusion and help them to understand the realities of CF.

CF care has changed a lot over my lifetime. When I was young, nighttime mist tents were standard operating procedure. We used glass nebulizers that had to be sterilized after every use. Our compressors were big, cumbersome things that took up a lot of space and they were noisy. People slept in those damp mist tents and it amazes me that we didn't all die of pneumonia from that. It was not easy to disguise our old machines. The little nebulizer compressors that we use now are so compact, they are easy to cover up. By not having all of our equipment out in plain sight, it is easier to seem normal. I think this helps with having more normal relationships. These new ones aren't like a gorilla in the room. Instead they are only a monkey in the works!

I just thought of another aspect of our lives that can have an impact on relationships. Our bathrooms, especially the toilet, come to mind. I think that most of us would prefer to have a bathroom that is for only our use. I realize that this may not be possible, but if it is possible it really is nice. My bathroom is just for me. I don't have to worry about being "offensive" to anyone else. In case someone does have to use my bathroom, it has a hands-free soap dispenser and a single handle faucet so that things are touched as little as possible. I also keep a box of Kleenex paper towels in there for others to use. I try not to use public restrooms and also try to avoid using the bathroom at the homes of others. This takes some advance planning, but it usually works out okay. If the researchers ever get a good fix for our GI woes, that will be a banner day.

Another thing that can get in the way of normal relationships is our perspiration. Or should I say SWEAT? My perspiration could melt a pair of good leather shoes in a few weeks when I was young. Not only was that costly, it was embarrassing. I remember the Mother General at one hospital where I worked was always nagging me about my shoes. This was in the time when nurses wore white shoes. My shoes would turn a kind of greenish color from my sweat. So, I got Corfam shoes. (Corfam was the first type of artificial leather which came out in the early '60s.) They were some kind of plastic material that didn't change color from my sweat. They didn't breathe like leather does so they were way too hot and the paint would come off of the eyelets. I would try to get white shoe polish to stick to them, but it didn't stick very well. She would snap at me that I needed to clean those eyelets. When I told her that they were clean, she was not happy with me. I did the best that I could. This was long before the Americans with Disabilities Act, so there was no accommodation made. Oh, well.

Speaking of perspiration reminds me about dancing with a fella. I had to keep a handkerchief in my hand so that the guy's hand wouldn't get saturated with my sweat. How embarrassing. I used to keep my hands in my pockets a lot of the time so that I wouldn't leave damp handprints all over.

I had one really embarrassing time with sweaty hands. My parents and I were traveling to southern Oregon for a convention. We were riding with a friend in his black auto. I was sitting in the front passenger seat and had my hand out the window, resting on the shoulder or hip of the car door. After a few miles, I moved my hand and saw that I had left a salt mark in that beautiful black paint job. Fortunately, he was a kind of car buff and he was able to repair my handprint. Still, it was very embarrassing.

Fortunately for me, embarrassment doesn't kill anybody. If anything, it makes us stronger. We know how to face adversity and still make our way in this world. A little less angst would be okay with most of us, though.

Till next time, stay healthy and happy,

Kathy ▲

Kathy is 73 and has CF. She is Managing Editor of CF Roundtable. *Her contact information is on page 2.*

WELLNESS

Be Kind To Yourself

By Julie Desch

I've thought about writing this article for a long time, but have not wanted to sound like an old curmudgeon. But, in less than two months I will be 57, and since that qualifies as an old curmudgeon, here you go.

Once upon a time, at least 15 years ago, I had a PICC line and was doing home IVs for an exacerbation. I was probably in my second or third week and must have been feeling better because I thought it was time to start jogging again. So naturally, I went to my gym and got on the treadmill. I had the Intermate device in my fanny pack and didn't think anyone would even notice. But sure enough, somebody who knew me pretty well came up to me and asked if I had hurt my arm. I had a mesh stocking over the PICC dressing, but it did look pretty suspicious.

I responded, "Oh, my arm is fine, I'm just infusing some antibiotics."

"Are you okay?" she responded quite naturally.

"Yes, I just have a touch of pneumonia."

Incredulous, she asked, "Why aren't you home in bed?"

Indeed. What was I thinking? At the time, I probably thought that this woman didn't understand that I could not afford to let up on my fitness program just because I was infusing meds. She didn't get that this was WAR...and I was not going to give any ground. This was life or death. People without CF just didn't understand.

I tell this story just to emphasize that what I'm about to say is not about judgment. I've been there. I've acted the same way. But since people are now very open on social media about what they are doing for exercise (which is great), it is clear to me that there is an elephant in the room when it comes to staying "fit" when you have CF.

Sometimes more is not better. Sometimes high intensity exercise is actually the last thing your body needs. Maybe 20 box jumps while in the hospital infusing a fluoroquinolone is a bad idea. Often, the thing we are least compelled to offer ourselves is what is needed the most—compassion.

We all know our own bodies best, of course. I hate it when somebody gives me advice about how hard, or even if, I should exercise. Historically, I have ignored such advice. So if you want to ignore me, I would understand. But, I'm going to give advice anyway, so you young kids listen now.

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a spray-dried form of mannitol, delivered to the lungs by a portable inhaler. The 26-week, randomized, double-blind, parallel-group study assessed improvements in lung function and other parameters, as well as safety.

The trial demonstrated the superiority of Bronchitol versus the comparator on the change from baseline in the forced expiratory volume in one second (FEV1) during the 26-week treatment period. The improvement in lung function was less than that seen in the adult CF population in previous Phase 3 studies. The investigators observed no statistically significant differences between the two treatment groups on secondary endpoints, although a trend was observed in favor of Bronchitol for the change in forced vital capacity (FVC), another lung-function parameter.

http://tinyurl.com/y9quq5lq

Theravance Biopharma (TBPH) Announces Presentation of Positive New Data from Studies of VIBATIV at ASM Microbe

Theravance Biopharma, Inc., announced the presentation of positive new data from multiple studies of VIBATIV® (telavancin), the company's proprietary FDA-approved antibiotic. Study results highlighted greater in vitro potency for VIBATIV against difficult-to-treat Staphylococcus aureus (S. aureus) pathogens, including those considered to be multidrug resistant (MDR), as compared to other commercialized antibiotics. Researchers collected and analyzed MRSA and MSSA cystic fibrosis clinical strains from three different CF centers in the U.S. Results from the study demonstrated that VIBATIV possessed potent in vitro activity against both MRSA and MSSA CF clinical strains, including those resistant to ceftaroline. When compared to daptomycin, ceftaroline and vancomycin, VIBATV showed the greatest in vitro potency.
http://tinyurl.com/y74z9vvb

Proteostasis Therapeutics Reports Preliminary Data from Phase 1 Study of PTI-428 in Patients with Cystic Fibrosis

Proteostasis Therapeutics, Inc., announced preliminary data from the Multiple Ascending Dose (MAD) cohort of its Phase 1 trial designed to evaluate the safety and pharmacokinetics of PTI-428 in CF subjects. PTI-428 is the Company's cystic fibrosis transmembrane conductance regulator

When you are sick with a fever, obviously you shouldn't exercise. Fortunately, when you are that sick you usually don't feel like exercising and that helps. This is time for water, food and bed. I can't think of too many people who would disagree with me on this.

It gets trickier when there is no fever, but you are either trying to stave off an exacerbation or are at the end of one, still on meds but feeling better. Then what do you do? Past Julie would have done whatever she could. If a PICC was still accessed, then probably she would have skipped the push-ups but definitely squats were called for. Kettlebell swings? Sure. Kettlebell clean and press? Sure, but only on the side without the PICC. Past Julie thought that being a badass was always the right choice.

Current Julie knows that Past Julie was sort of mean to herself.

When fighting off an infection, the body needs all available resources. Every bit of energy that the body makes is needed by the immune system to

JULIE DESCH, MD

regain homeostasis and get you back on your feet again. Any energy you put into that workout is energy that is not available to fight infection. It is quite possible that springing back to your usual workout routine too soon might actually delay the recovery process. This does not mean you should sit around like a slug. In fact, even while on IVs, I still walk every day. But these are shorter walks, with the simple goals of getting fresh air and moving and stretching a bit. Fatigue is not the goal. In fact, if I don't feel better after the walk than I did before, I went too far or too fast. Current Julie waits until all IV antibiotics are discontinued before she goes back to lifting or doing anything other than these walks.

Basically my attitude has shifted a bit. Yes, I still feel like I'm at war. But I'm not at war with my body. I don't need to whip it into shape the minute I'm afebrile. Rather my body is now my ally, and we are at war with pseudomonas. With this small mental shift, I don't feel strange about napping rather than hopping on the elliptical. My body tells me what it needs and now, finally, I listen. ▲

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

(CFTR) amplifier. This Phase 2 portion of the study is expected to inform researchers on a dose level for the triple combination proof-of-concept study combining the company's three CFTR modulators, PTI-428, PTI-801 and PTI-808. PTI-428 is unique among CFTR modulators, in that in vitro studies suggest activity across CFTR genotypes, enabling a potential treatment paradigm where therapy for the majority of CF patients could be based on combinations anchored by PTI-428. Proteostasis is currently conducting a 14-day MAD Phase 1 study of PTI-801 (corrector) in healthy volunteers in the U.S., to be followed by dosing in CF patients. If positive results are achieved in the Company's PTI-428 and PTI-801 programs, the Company intends to initiate a triple combination Proof of Concept (POC) study, which will combine PTI-428, PTI-801 and PTI-808 (also known as PTI-NC-733), in an F508del homozygous population who are not taking Orkambi®, at the end of 2017. The study will explore different doses of PTI-808 with fixed dose combination of PTI-428 and PTI-801.
http://tinyurl.com/y7wbnrcv

Cystic Fibrosis Therapy Developer Proteostasis Asks FDA to Help It Advance PTI-808

Proteostasis Therapeutics has filed an Investigational New Drug application with the U.S. Food and Drug Administration for PTI-808 as a treatment for cystic fibrosis. PTI-808 promotes the activity of the cystic fibrosis transmembrane conductance regulator, or CFTR, the protein that is faulty in CF. Scientists call the drug a potentiator because it's designed to bolster CFTR's activity. Proteostasis Therapeutics developed PTI-808 as a component of a triple combination treatment for CF.
http://tinyurl.com/yaldlnnw

CFF Therapeutics to Give Up to $5M to Spyryx for Phase 2 SPX-101 Trial

Cystic Fibrosis Foundation Therapeutics (CFFT) will increase funding to support Spyryx Biosciences' Phase 2 trial assessing SPX-101 in people with cystic fibrosis, named HOPE-1.

SPX-101 is a drug designed to increase the amount of liquid on the airway surfaces (airway hydration), which makes clearing mucus easier. The drug acts on epithelial sodium channels

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FOCUS TOPIC

DATING AND RELATIONSHIPS WITH CF

Growing Into Cystic Fibrosis

By Tabby Caldwell

I attend a large school, Brigham Young University in Utah, and so the chances of encountering someone with cystic fibrosis are much larger for me here. As everyone with CF knows and has likely heard, we don't actually look sick, and so I never know when I could be sitting next to, walking by, or even having a conversation with someone else with CF. This has been one of the strangest phenomena I have experienced here at BYU, and it often makes me feel strangely vulnerable and exposed. Since starting back here this fall, I've been thinking about this more, and how I communicate my disease with the world around me, especially to my fellow students.

I am sometimes so hesitant to share my health conditions with others because I fear the judgment that may follow and fear being treated differently because of my CF. I realized that I have these fears because in general many people do not know much about CF, or understand any of its complications, and end up asking many questions that inevitably make you feel like a lab rat being prodded at. When I tell someone I have cystic fibrosis and they respond by asking, "What is that?" I simply explain a basic version to them that I have committed to memory. In my mind, however, my insecurities take over, and I really want to tell them that this conversation does not give them permission to treat me any differently than before. Ideally, I would say this:

> I do not want to be treated like I am fragile. My CF has made me strong, not weak, and my scars have made my skin thick. I am brave and I am not simply the product of an unfortunate genetic mishap; I am more than my disease and I deserve to be treated as such.

"I am sometimes so hesitant to share my health conditions with others because I fear the judgment that may follow and fear being treated differently because of my CF."

TABBY CALDWELL

While I do know that most of the people I talk to about my CF aren't actually judging me, having a chronic illness your whole life tends to make you skeptical. However, since starting college last year, I am getting better at handling situations like those described above every day. I have started to see what the other person might be thinking and how they are probably just trying their best to be sensitive about my health and my feelings. I am gaining confidence in myself all the time and, each time I talk about my CF, I feel more comfortable with it. I can see myself becoming very open about CF in the future and hopefully I can live to see a day when the whole world is educated about it, and the negative stigma surrounding chronic illness is replaced with love and positivity for those affected by it.

This semester has also got me thinking about how I relate to other students and how my CF can sometimes prevent me from doing so. One area where I often feel like I got the short end of the stick is in prioritizing while in school. Not only do I have to choose things like whether to study for an extra hour or spend some time with friends, but I also have to decide between things that both seem necessary, like whether to sleep for an extra hour so I won't get a cold which could lead to a lung infection or to spend that extra time working out and exercising my lungs so I can keep breathing easy. See the dilemma? Often these choices are just a catch 22: if I choose to exercise, I will likely still feel congested in the morning because of the little sleep I got, or if I choose to sleep, my lungs might feel extra weak

and tight from not exercising them.

These decisions have come to define a part of my college experience, as I constantly have to prioritize health over everything else, and then further try to choose which elements of my health to prioritize above the others.

Something else odd I have experienced for the first time while in college is feeling jealous of someone for being able to be so careless with their sleep. Even if they get sick, it will just be a cold or something small and, if it doesn't go away within the week, they can probably sleep it off this weekend. This is probably a jealousy many adults with CF have felt and now I'm experiencing it to its full extent!

Overall, my time in college has been filled with new experiences and memories I will never forget; I have met the most amazing people and made incredible friends who respect my health and help take care of me when I forget to. I know that cystic fibrosis is a challenge, but it is one that I am willing to fight each day and am growing stronger from it all the time. I hope that while I am here, I can continue to spread the word about CF and create an environment of learning and understanding around it, so that everyone with CF can feel comfortable sharing their health with others.

As a runner-up for the USACFA Lauren Melissa Kelly Scholarship, I am extremely grateful to have been chosen and to be able to use this scholarship to keep working and keep fighting. Without the continuous support and love from USACFA and the entire CF community, I would be nowhere. I am excited to see where my education takes me and hope to continue learning more about myself and my CF along the way. ▲

Tabby is 19 and has CF. She is a sophomore at Brigham Young University in Utah where she is studying chemistry. She loves learning, eating (especially peanut butter pretzels) and spending time with her friends and roommates.

Love Me, Love Me Not?

By Sydna S. Marshall

Navigating the murky waters of a new relationship is challenging, to say the least. Do I call him? How long should I wait before calling him back? Should I text him? Can I ask him out first? Let's not forget the age-old dilemma between the sexy panties at the crucial moment versus "...chances of reaching crucial moment greatly increased by wearing these scary stomach-holding-in pants very popular with grannies the world over" as Bridget Jones notes. These and a myriad of other insecurities are inevitable in every newly budding relationship. However, take all of those jitters and raise them with a chronic terminal illness and you're playing in the big league now.

I was diagnosed with cystic fibrosis (CF) at age 11 after polyps were found in the first of many routine sinus procedures. Most of us with CF are diagnosed early in life, often before the 12-month mark. I had 11 years of being a normal kid without treatments and without an entire box of pills at each meal. Taking time out of school only to return with a condition that requires nebulizer treatments in the nurse's office during lunch isn't exactly first choice for any kid. To make the pills more fun, my parents created a game and whoever had the most pills to take each night won. We still play this game and I'm still winning!

CF research back then hadn't taken off, and the treatment options were fewer and less time consuming. When I started high school, my nebulizer treatments were still short and sweet, lasting maybe 10 minutes each time: one vial of Xopenex nebulized twice daily to open up my lungs. Back then, my CF wasn't a daily struggle, but it wasn't absent from my relationships either. I remember asking one of my first boyfriends to participate in the CF walk with me and the overwhelming shock, followed by sadness, when my dad and I went to pick him up on the walk day and not only was he not joining, he broke up with me.

College dating was a bit trickier, and the little signs that CF was taking its toll on me started creeping into the picture. If I'm being honest, I wasn't the best at taking care of my health during that time either. It's so easy to leave

SYDNA S. MARSHALL

home and no longer prioritize treatments. Who wants to say no to everything because treatments and pills are more important? My last two semesters of college I worked two part-time jobs on top of my course work. In the summer, I'd work at my dad's law office for six hours, sometimes more, then head over to the bookstore (where I met my future husband) and work the closing shift until 11 p.m. Many nights I wasn't home until 2 a.m., only to get up at 5 a.m. and do it all over again. Rinse and repeat, day after day and you've got a CFer whose understanding of work-life balance is dim, at best.

After nine months of back-to-back oral antibiotics and multiple visits to my CF doctor, I found myself in the hospital for my first two-week tune-up only four months after my college graduation. My lung functions were 30 percent lower than my baseline and the realities of CF hit close to home. Right or wrong, one of the main reasons I married so young was my need for health insurance. When you grow up as an attorney's daughter you quickly remember the logical life lessons, like the loss of health insurance and the group policy loopholes.

My first husband had 15 years on me, had decent insurance and was a safe bet. We didn't have that much in common, but we didn't argue and I mistakenly assumed that we'd grow together with time. That safe bet, that age gap and our fundamental ideological differences would feel isolating and larger than life as my CF declined. For him, I was a dying woman who needed handling. Being a detail-oriented person, he had researched the cost of managing CF and what that would entail before I even moved into his home. He handled me and he managed all aspects of my CF, including all the pharmacy trips, the treatments, the doctor appointments and the health insurance. It didn't take long for me to realize I was codependent on someone who loved me despite my CF.

I was more consistent with my treatments than I had been in college and the easy nebulizer treatments from high school were now 40-plus minutes followed by percussion therapy twice daily. During my first marriage, I had two hospitalizations and my sputum cultures grew *Pseudomonas aeruginosa* for the first time, which eventually colonized in my lungs permanently. My

second hospitalization was eye-opening as I quickly realized that visiting me in the hospital was too much for my husband, both because of time constraints, and more importantly, emotional desire to support me. He rarely visited me and when he did, his discomfort was evident. On one occasion, he opted out of visiting me, saying he was tired and it was later brought to my attention that he was out drinking with friends.

I was gifted a camera from my husband the day I got the call from my CF doctor about my culture. His words to me as I opened the gift still stand out in my mind: "So you can start documenting your memories." In hindsight, that was the moment we started unraveling permanently. I realized I was simply the pet-project wife who was dying before him, and it was his job to take care of me, not out of love, but obligation. He no longer saw me as a spirited young woman, and perhaps, never did. Toward the end of our marriage, we argued about my ability to have children and whether I could even last nine months without my medications. I finally had the courage to leave him in the summer of 2007. When I calculated the costs of living on my own, I intentionally left out my healthcare expenses as I knew I'd lose my courage and stay in an unhappy marriage.

That same summer, I got to know Adam, who worked in a different department at the same office. I asked him out on our first date and soon enough we were dating and spending most of our time together. Unlike my first husband, Adam and I are akin to Forrest and Jenny from Forrest Gump – peas and carrots. Adam is five years younger than I am and he brings to our relationship an open willingness to experience everything life has to offer.

Regarding CF, I knew I had to handle that conversation differently than I had in the past. Thirty days into our new relationship, I still hadn't told him about my CF. I wasn't doing any treatments so he had no reason to suspect I would have CF, or so I thought. I analyzed this constantly. Do I tell him now? What if he runs for the hills? What if he looks at me like the dying woman my first husband saw? What if this is way beyond what he bargained for in a relationship? If I don't tell him, will he be hurt that I'm withholding something so central to me? Is this a trust issue? Would I want to know now or later about a chronic illness? I finally told him and to my surprise, he already knew about my CF based on my online screen names and was simply waiting for me to tell him when I was ready. Adam went to high school with two siblings who not only both had CF, both of them passed away during high school. His experience with CF landed on the other side of the bell curve and I failed to recognize then that he chose me early on even knowing some of the forks in the road ahead.

"I finally told him and to my surprise, he already knew about my CF based on my online screen names and was simply waiting for me to tell him when I was ready."

We've been together for ten years now and married for nearly four of those years. My health has declined since then, especially in the last two years. I used to count the number of scars on my arm from my peripherally inserted central catheter (PICC) lines, which are used for the 14-day courses of intravenous (IV) antibiotics I need for a CF tune-up. I used to count my sinus surgeries, in the beginning, when I mistakenly believed I would only need one or two at the most. Twelve PICC lines and seven or more sinus surgeries in, I've stopped counting. I spent a lot of time soul searching in the beginning of our relationship. I didn't want to be the woman who couldn't and wouldn't give Adam a family. As the older woman, I didn't want to strip Adam of his life choices, as my first husband had done to me.

After my first marriage, I struggled with leaning into Adam for the support I desperately needed for fear of losing my independence. Having a chronic terminal illness adds more challenges to the equation. He's seen me in the most humiliating and vulnerable times: blindfolded with drain tubes, a catheter and a bedpan. Adam has been my rock through the most debilitating and embarrassing CF digestive issues, he's slept for days on end in the chair next to me in the hospital and he's driven an hour each way to pick up meds I've forgotten during Christmas holidays. We have a running joke that no vacation or trip is complete without a trip to a pharmacy, so we now plan accordingly. After my frontal sinuses were removed, he spent hours taming the tangled mess and combing the gelled knots out of my hair. Little things like that and accessing my port for IV antibiotics or carrying all of my nebulizer equipment at the airport and subsequently putting them in the overhead bin mean the world.

In the ten years of surgeries, hospitalizations, tune-ups and additional health challenges, Adam has been my partner, my person, my soul mate, my support, my cheerleader and my best friend. He continues to handle the rapid-fire, almost-weekly health challenges life has thrown at me in the past year with a confident serenity. This

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Dating And CF: It Takes Time To Figure Out

By Reid D'Amico

> "As I started to grow into my CF identity, I would slowly introduce it into my relationships, as I felt comfortable."

Relationships are hard. CF is hard. So unsurprisingly, combining the two of them together leads to some unique and difficult challenges. CF is a disease that doesn't have the same name recognition as other more common diseases such as diabetes and cancer. So opening up to others about our disease often leads to "teaching moments" where we have to build the foundation so someone can understand what it's like to have CF. But here's the challenging thing: we learn over years what it's like to have CF; we learn how to do our treatments, stay healthy and manage our insurance, but we don't truly learn what it's like to date with CF until we are adults.

When I was younger, I thought I had my CF journey all figured out. I adapted my treatments to my hectic schedules and made time for a social life. But when I really started dating in college, I found myself having to learn how to weave CF into my identity with strangers. In the following, I want to highlight the three types of relationships or dates I've had and how they relate to CF. I do, however, note that I am only 24 years old. I am still learning how to integrate CF into my relationships and learn more every year. But for now, I want to step back and look at my first real attempts at dating during my late teens and early twenties.

1. When I first started to date, I never talked about having CF. It was a secret and I wanted to cover it up as much as possible. CF was a part of my personal life and it wouldn't affect my relationships with another person. I remember having dates over to my dorm in college and remember being asked, "What's that machine in the corner?" I used to change the subject quickly. The last thing I was going to do was talk about a piece of equipment I use during my therapies. These relationships rarely lasted long for me. At the time, I didn't yet know how much CF had influenced my passions for science and advocacy. Since I dodged all aspects of it, conversations ran shallow and these dates quickly died out.

REID D'AMICO

2. As I started to grow into my CF identity, I would slowly introduce it into my relationships, as I felt comfortable. It would start off as a "lung thing" and would slowly build as the weeks went by. However, I always found myself building CF in a different manner. When I finally revealed the name of the disease, I would immediately say that my symptoms weren't that bad and would recommend my date not read about it online. I wanted to build my own CF identity in our relationship. Looking back, the reason this attempt failed was that I was still having trouble with accepting my disease myself. I covered it up and wrote it off. I remember doing treatments before leaving to spend the night with my now ex. I never would do those things in front of people at that time in college. I was embarrassed and wanted people to overlook the flaw that was CF. These people I dated knew I had CF but never got a real window into my life.

3. As I've gotten older, I've created a strong foundation for my CF identity. I've woven it into my career, my advocacy and my common discussions. For this reason, I sometimes "nuke" my dates. By this I mean I drop the CF bomb on them early. I've adopted the mentality of "this is who I am, take it or leave it." However, this mentality is still a bit flawed because I do get worried and upset that someone may not want to date me because of CF. I remember my first time doing my treatments in front of a significant other. I had to immediately turn them off because I started sobbing. I couldn't believe that I had let someone into my life like that. It was embarrassing, intimate and the repercussions of those personal walls falling were immense. However, that was only the beginning of opening up with CF. I still don't know how to feel when a significant other is "worried about me" or "hates"

CF as much as I do. It sometimes makes me feel powerless, even if it's from a place of love. I'm hoping that with years and maturity, I'll better learn to embrace these views. They are likely an inevitable part of being with someone who has any disease.

It's funny, as I was writing this piece I realized how I still am uncertain and unstable in how I present my CF to others. I can think of many examples where I would present my CF in a different way or at a different time. Looking back, I don't regret the ways I approached having CF. For those who weren't able to handle it, good riddance, and for those who stuck around and supported it, I thank you for allowing me to have my time at whatever stage of life we met. Being an adult with CF is largely unchartered territory. What an honor and scary thing to be the ones figuring it out. ▲

Reid is 24 and has CF. He is a Director of USACFA. His contact information is on page 2.

In Memory

Donovan Couture, 49
Milton, VT
Died on December 30, 2016

Immediate family members may send in the names of CF adults who have died within the previous year for inclusion in "In Memory." Please send: name, age, address and date of death.

Send to:
CF Roundtable
PO Box 1618,
Gresham, OR 97030-0519
E-mail to:
cfroundtable@usacfa.org

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(ENaC), and so SPX-101 acts independently of the underlying genetic mutations that cause cystic fibrosis. Participants in the Phase 1 study did not experience changes in lung function. They also did not experience abnormal serum potassium levels or bronchoconstriction (airway constriction) and no serious adverse events. SPX-101 is currently undergoing a small Phase 1b trial (NCT03056989) in patients with cystic fibrosis. In preclinical testing, the drug improved survival and mucus clearance in a mouse model of cystic fibrosis.
http://tinyurl.com/yckvszr7

AIT Therapeutics Announces Enrollment of First Patient into Its NO-NTM Abscessus Phase 2 Trial in Nontuberculous Mycobacteria (NTM)

AIT Therapeutics Inc., a clinical-stage anti-microbial therapeutic company treating respiratory diseases with nitric oxide (NO), announced the dosing of the first patient in its Phase 2 trial targeting NTM patients with Mycobacterium Abscessus Complex (MABSC). Intensive NO treatment for just 21 days may obviate the need for long term, toxic and costly antibiotic therapy. AIT has completed a Phase 1 clinical safety trial on healthy subjects, and has acquired positive human clinical data on over 50 patients with cystic fibrosis, bronchiolitis and NTM. These data show that inhaled NO is safe with strong evidence of activity. AIT's device is designed to safely and effectively deliver 160 ppm NO to patients for the treatment of severe lung infections. Nitric oxide (NO) is a powerful molecule proven to play a critical role in a broad array of biological functions. In the airways, NO is believed to play a key role in the innate immune system at concentrations of approximately 200 ppm. In vitro studies suggest that NO possesses anti-microbial activity not only against common bacteria, both gram positive and gram negative, but also against other diverse organisms including mycobacteria, fungi, yeast and parasites, and has the potential to eliminate their multi-drug resistant strains.
http://tinyurl.com/y9uccyql
AND
http://tinyurl.com/y9lmx4tq

Two of Proteostasis's Three Cystic Fibrosis Therapies Are in Clinical Trials, and the Third Is Heading for Trials

Two of the three therapies that Proteostasis Therapeutics has developed to correct the underlying problem in cystic fibrosis are going through clinical trials and a third is heading for trials. Proteostasis's therapies are PTI-428, which is a CFTR amplifier; PTI-801, a new-generation CFTR corrector; and PTI-808, a CFTR potentiator. An amplifier increases the amount of an immature form of CFTR protein, so there is more material for other agents to act on. A corrector addresses problems in the protein. And a potentiator improves the performance of another therapy. The three therapies are designed to improve the activity of cells with a mutated CFTR gene and restore the cells' normal transport of chloride. A Phase 1/2 study (NCT02718495) is assessing PTI-428's safety, stability and ability to function in the body as a therapy. The Phase 1 part of the trial demonstrated that PTI-428 is safe and well-tolerated. All patients' lung function remained stable, and adverse events were mild or moderate. The trial also showed that PTI-428 did not significantly interfere with Orkambi's activity. The Phase 1 results were good enough for Proteostasis to start the Phase 2 component, which will look at the effec-

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Finding Love With A Chronic Illness

By Sarah Albright

> "Your partner may not know exactly what you're going through, but the support is crucial."

Relationships can be complex when you're healthy, but finding love while living with a chronic illness, now that is complicated! We all want to love and be loved, that is one of the glorious parts of being human. But how do you love someone who suffers from CF, this illness that affects them every day in so many ways? Because dating isn't complicated enough, let's add in the medication regimen, treatments during weekend getaways, doctor appointments, hospital stays and the overall emotional burden of watching your partner gasp for every breath, knowing you can do little to help.

In high school my lung function was fairly "normal" and there was little indication that I had a chronic illness, so dating was easy. As I grew older, I became sicker and it became harder to date. For a while it was still easy to keep my illness hidden. I would do my breathing treatments before I went out, I would take my enzymes in the bathroom after dinner and I was able to walk away or use the restroom to cough if I felt a coughing fit coming on. As I got older, I felt more pressure to hide the fact that I was sick in fear of scaring away my partner. However, it became increasingly difficult to keep up with the active people I dated. While rock climbing, pulling myself up the rocks, I had to hang on tightly while trying to suppress my cough. At concerts, trying to jump around and sing loudly left me breathless. Even nights out at a bar enjoying drinks, dancing and laughing into the night left me totally exhausted.

Now that I have come to terms with my disease and all the aggravating things that accompany it, I have stopped hiding my illness in the dating world and tried out online dating. Social media and the internet is everywhere now, so why not use it to my advantage? My dating profile did not mention CF; after all there is a lot more to me that just CF, so I left that discussion for future conversations. Determining when to explain to your partner that you have a chronic illness is difficult to time. Do you tell them right in the beginning before they get emotionally attached? Do you wait until it becomes a little more serious and then break the news? Or do you wait until they fall completely in love with you? It's not an easy decision! For me, I decided that from that first conversation, I was going to be open about my illness, making sure to be upfront about it from the beginning. I felt, in this way, I gave any potential partner the opportunity to make a decision on whether they wanted to proceed with getting to know me better or opt out.

SARAH ALBRIGHT

Most of the reactions I received from previous dates were filled with concern and questions. Most asked what CF was and I'm sure once they got home, they did their own Googling. Some asked more in depth questions, others didn't speak any more about it after I told them. Not everyone's journey and illness is the same, even people with the same illness, so questions are always welcome. As I now go through this transplant journey, I have a boyfriend I can lean on for support and guidance. You need a support system while living with a chronic illness; it can be draining, not only physically but mentally and emotionally as well. We all need to vent from time to time, especially when living with a disease. Your significant other should make you feel you have a safe place to be vulnerable and comfortable while discussing and coping with your illness. Your partner may not know exactly what you're going through, but the support is crucial. Having a supportive person with you while you sit in a waiting room, at a doctor appointment or in a hospital is so comforting. It can be extremely lonely and boring lying in a hospital all by yourself. It also helps to have your partner around to debrief after difficult clinic visits. Sometimes I'm so overwhelmed by what the doctor is saying, I can miss important details. So it's helpful to have someone there

to process the information with me once it's over. What exactly did the doctor say? Sometimes the focus is on one piece of information and I end up missing others all together.

It's not always easy dating when you're chronically ill. I've found that not everyone has the emotional capacity to love someone who is sick and not everyone can be emotionally supportive of a chronic illness. This has taken me almost my whole life to figure out. It takes a lot to sit with someone for hours in the ER, or listen to the difficult news brought by your illness progressing. I know it's not easy waiting around for me to complete my medications before we can go do something fun; I know it's not easy not being able to run around with me or chase me or to cancel plans because I don't feel well. Because my lung capacity is so diminished, my boyfriend now needs to carry me up inclines and stairs. I know it's not easy seeing someone you love struggle to take every single breath. I salute my boyfriend, family and friends, who stick by my side while I struggle to do pretty much everything. I could never thank them enough for all the patience, love and care they have given me. And even for the ones who didn't stay; there were plenty of people who just couldn't handle it and to them I say thank you as well, for teaching me that not everyone can love someone who is sick and that's okay. I respect their honesty. I thank them for teaching me that whoever falls in love with me and whoever I end up with will love me for everything that I am, mucus and all. ▲

Sarah is 25 and has CF. She lives in Manhattan Beach, CA. She is currently awaiting a bilateral lung transplant, but in her free time she works for a pulmonary rehabilitation center, is applying to graduate school for her Ph.D., is writing a book on living with a chronic illness and does some modeling for a local clothing company.

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ing as they should.

Constipation: This is a word you probably hear a lot when you visit your CF clinic. We like to talk about poop. You may have even been asked to compare your bowel movements against those of a "poop chart." I know this is a favorite of many of my CF patients in clinic! (If you haven't seen this yet, check out the Bristol Stool chart.) Constipation is common in CF and, when poorly managed, it can lead to more serious medical conditions such as distal intestinal obstruction syndrome (DIOS), rectal prolapse or hemorrhoids. To avoid these more complex conditions, the goal is to prevent constipation through assessment of stooling behaviors, bowel and enzyme regimens. Constipation is stool impaction in the colon that is usually associated with a reduced number of bowel movements that are often smaller in volume or hard, pebbly stools. Constipation in CF, however, may be characterized by an increased number of liquid stools, known as overflow diarrhea. Constipation often leads to gas, bloating, mild abdominal pain and general discomfort; it occurs in both pancreatic insufficient and sufficient patients. Your CF team may recommend laxatives to help achieve the goal of two to three daily bowel movements that are formed, sinking and easy to eliminate.

What can you do to prevent constipation? First, with the help of your CF team, find a bowel regimen that works for you and stick to it! Second, maintain strict adherence to an enzyme regimen that helps you absorb all your nutrients. Finally, make sure you hydrate well, especially when you are exercising, in summer months or during a CF exacerbation. Talk to your team about these steps to get further details about your regimen and needs.

Constipation myth: "When I take my enzymes, I get constipated. A little grease in the system helps keep me regular!" FALSE. In fact, not taking enzymes – or, taking a lower dose of enzymes than recommended – leads to malabsorption of nutrients (which you may experience as greasy, floating, smelly stools) and can increase the risk for constipation or DIOS due to sticky intestinal mucus. The most common cause of constipation in CF is malabsorption. Keep in mind that taking the proper dose of enzymes may result in fewer and more formed bowel movements due to proper absorption and decreased excretion of undigested nutrients in stools.

Approximately 85-90% of CF patients are pancreatic insufficient (PI), requiring digestive enzymes when eating. Despite affecting most of the CF population, the importance of appropriate dosing, proper timing and with which foods to take enzymes is confusing and sometimes misunderstood. Pancreatic enzymes help a person digest and absorb carbohydrates, proteins, fats (the nutrients in food that provide our calories), vitamins and minerals. As such, they are vital for achieving a healthy weight and to prevent vitamin deficiencies. They are also important for control of GI symptoms like constipation, abdominal pain, bloating, gas, urgent bowel movements or diarrhea that may be the result of poorly digested food in the intestines.

Enzyme myth: "It doesn't matter when I take my enzymes, as long as I take them at some point with my meals. But I don't need them with my milk/smoothie/milkshake." FALSE. With pancreatic enzymes, timing is key! It is important to take enzymes at the beginning OR the beginning and the middle of meals so that they mix with the food in your stomach before passing into the small intestine where they are active. Take your enzymes with all foods (liquid/solid) that have pro-

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Dating Care Free (CF)

By Nicole Kowal

In April 2011, I broke up with a guy I had been dating since high school. Five years of dating someone who couldn't be in the same room with you while you do your treatments, who when fighting would say that there is no future and even his friends stated how they could never imagine dating someone "sick." So I made the choice to NEVER go back to that lifestyle and to be even more open about my CF and my hurdles that I simply jumped over, no matter what they were. I began the wonderful world of online dating, OkCupid was the choice of this adventure and it ended amazingly!

In the beginning I started out with not telling anyone anything until I was on the second date. I had some guys who were curious, but then after they asked their questions, went home probably to research, I never heard from them again. I had other guys who left right after I told them, and I had some guys who were down right jerks about me dating while "sick." So after feeling "defeated," I picked myself back up and I turned a new page. I laid it all out there, everything from my CF to my gluten allergy. I even said on my dating profile, "If you can't handle what I have, then I can't handle you." I had some extremely rude messages and some guys that said good luck and keep going. After a few dates with a couple of guys I was seeming to find no luck, until I saw this picture of a guy next to a plane...I messaged him first.

After a month of back and forth messaging, we met in person. His name is Michael. I thought he was the biggest nerd, not the cutest, but he was simply different. But I believe in second chances. Our second date was to Niagara Falls; he picked me up and thought the guy in my front yard doing

> "I even said on my dating profile, "If you can't handle what I have, then I can't handle you."

NICOLE KOWAL

yard work was our gardener. Nope, it was my dad. Our date was great and it led us to schedule a next one. That was all in July. By August 28, 2011, we were driving down to see my sister at college and he asked if he could switch my Claddagh ring around to say I was taken. Of course I was beaming and said sure.

On October 5, not only did my parents put my family dog down, but I was told I can't see my dying grandmother because I was so sick. By 9 p.m. I had a knock on the door. Michael came to the house to bring me Reese's and a shoulder to cry on. It wasn't his day to be there, but he showed up anyway. A surprise visit and a shoulder to cry on. That night, although it was a horrible night, I said I love you to him. By October 30, 2011, we signed the lease on our first apartment, an 1820 farmhouse apartment that we LOVED. In March of 2013 we bought our first home, and in September of 2014 he asked the question any girl madly in love would want to hear come out of her boyfriend's mouth, "Nicky, will you marry me?", all while standing in my family's blueberry patch, during labor day weekend at our cabin. He picked the perfect time — all my family was there, all my friends and loved ones. I walked back to the cabin crying and beyond excited to tell everyone what happened, and the wedding planning began. He had the ring made especially for me, the side two diamonds from his mom's engagement ring, the metal being palladium (stronger than platinum) because he knows I am far from gentle on things.

A year and a half went by, we planned our perfect day and on March 5, 2016, we said "I do." Now, we sold our first home, bought my grandfather's home and are simply enjoying life together. I find it truly amazing that I was able to not only find the man of my dreams and have him capture my heart but by doing everything with a smile. He goes with me to CF Clinic, he laughs with me at the people who make weird faces while I travel wearing a mask and he sits up with me while I

am coughing during the night. He cups my back while I am gasping for air, he brings light to some of my darkest thoughts and he simply loves me throughout all my illnesses.

I guess what I would have to say is tell someone when you, yourself are ready to share it. I am very open about my CF and my other health problems. I believe in sharing word of this disease and showing others that I have no problem trying my hardest to live every day to the very fullest and to do "normal" things, from dating to marriage and buying houses. We may have complex bodies, but we are human just like everyone else, and sometimes that may be the most normal thing about us.

Go with the punches while dating. There will be ups and downs just like life, but we are fighters and choose to never give up. Have fun with dating and your relationships. I truly think finding Michael was the best thing to happen to me. I have taken control over my health again, and I have learned to love myself even more. He has brought out the very best and all while laughing with me, through my coughing spells because of laughing and smiling at what life throws our way. Not only did he marry me but my diseases and he is okay with that, just like I am okay with sharing to the world about them. Be open, be honest and be yourself. Don't ever let anyone stomp your fire or your fight out. ▲

Nicole is 29 and has CF. She and her husband, Michael, live in Buffalo, NY. They have a dog, Dory, two Russian tortoises, Boris and Natasha, two red-eared slider turtles, Pookie and Pancake, and two cats, Sophie and Menssah. Nicole teaches ice skating to children with handicaps and is an active EMT for the local volunteer fire company. You may contact her at: nkowal@westherr.com, 716-480-6953 or even on Facebook!

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tein, fat or carbohydrates, including beverages such as milk, formula, smoothies and shakes. Simple carbohydrate foods (fruit snacks, juice, soda, hard candy) may be consumed without enzymes. Expired or heat-damaged enzymes lose their effectiveness; so, if you're unsure about when you got those enzymes or where they have been stored, throw them out!

CF "gut" myth: "My stomach bothers me most of the time, but that's just my 'CF gut'; it can't be fixed." FALSE. From my experience, CF patients are some of the most resilient, toughest and kindest people I know. I have found that many simply deal with stomach discomfort or pain rather than talk to their providers about it; they "accept" that discomfort is part of having CF and move on with their lives. Don't accept pain or discomfort as the norm! Talk to your CF team about what you are experiencing. It may be as simple as an adjustment to your enzyme or bowel regimen that may alleviate these symptoms. When in doubt, talk to your CF team. We're here to help!

Michelle is the clinical dietitian at the adult CF Center at Stanford Health Care in Stanford, CA. In her spare time, she enjoys rock climbing, hiking, yoga and cooking. Her e-mail is: mstroebe@stanfordhealthcare.org

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tiveness of a combination of PTI-428 and Orkambi. This stage will involve patients receiving Orkambi and either PTI-428 or a placebo for 28 days. The company is planning another Phase 1/2 study for its CFTR corrector PTI-801 — by the end of 2017. The U.S. Food and Drug Administration has approved the company's investigational new drug application for the third therapy in its pipeline, PTI-808. That prompted Proteostasis to start a Phase 1 trial of its safety and pharmacokinetics in healthy volunteers. If PTI-428 and PTI-801 prove effective, the company will start a trial that evaluates combinations of all three agents by the end of 2017. That study will cover CF patients with the F508del mutation who are not receiving Orkambi.
http://tinyurl.com/y9tb9aqh

H2-Pharma launches multivitamin line for those with malabsorption conditions

H2-Pharma launched their MultiVitamin ABDEK family of vitamin offerings. MultiVitamin ABDEKs are specifically formulated for individuals with malabsorption conditions. H2's ABDEK product line provides key fat-soluble vitamins (A, D, E and K) in a water-miscible form, as well as essential vitamins from the water-soluble group like vitamin C and the B-complex vitamins. MultiVitamin ABDEKs are gluten-free, and are available in three dosage forms: chewable tablets, pediatric drops and softgel capsules. In addition, H2's MultiVitamin ABDEKs meet the daily recommended vitamin supplementation suggested by the Cystic Fibrosis Foundation when taken as directed.
http://tinyurl.com/ydz6xkmw

CF Foundation Opens Door to Speedier Trial of Anthera's Digestion Therapy Sollpura

Sollpura is a pancreatic enzyme replacement therapy, or PERT. It contains the enzymes lipase, protease and amylase in the same concentrations that are found in the stomach. Most PERTs are derived from pigs. Sollpura is not,

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Are We That Different?

By Ella Balasa

I think many of us with CF have built up emotional walls around ourselves. They are our shields blocking the unwanted invasions from people's judgment, perhaps even people's concerns. These walls are built from the stones of fear and uncertainty, about being different, being unattractive, being unwanted. These walls make it difficult and usually impossible for those wanting to get close to us to break down. For this reason, dating having cystic fibrosis or any other chronic condition can be very daunting, frightening and viewed as more challenging than "normal" dating.

"Normal" people don't have the health issues we have to deal with. "Normal" people don't always cough. They don't have to deal with taking medications on a date in front of someone they don't quite know yet. They don't have to deal with that first tune-up hospitalization after they have been seeing someone new for two months and they feel uneasy or even panic thinking there is something deathly wrong.

No, the average person doesn't deal with these things. But, everyone, every single person has his or her own battles that they face. I like the reference, "Everyone bears their own cross." We all struggle with issues that we as individuals have to deal with in our own ways. Everyone has insecurities, so is anyone "normal"? Aren't we all normal? Though the battles of someone with CF are far greater than those of some others, it is our perspective that determines the effects they can have on our relationships with others, in this context, on our relationships with potential partners.

Perspective, the viewpoint from which we approach a situation, can be in either a positive or negative light. If we view our disease as a negative and think about how it prevents us from doing things, such as going all day to a festival with a date because we don't want to have them see us doing treatments or taking a hiking trip to the mountains because we simply cannot walk those inclines, we limit ourselves from potential possibilities. The possibility to have a wonderful time. The possibility to build a connection with someone. But those inhibitions, those fears about their judgment of us doing breathing treatments or coughing, it's a struggle within ourselves.

> "When we are transparent about our disease, what we have to do and why we have to do it, people feel more comfortable."

ELLA BALASA ON A BOAT CRUISE OFF MLJET ISLAND, CROATIA.

I believe once we accept ourselves for the way our bodies are and have been created, we convey authenticity about ourselves. If we are being authentic in a relationship, we show our true selves. People don't doubt or question knowns, only unknowns. When we are transparent about our disease, what we have to do and why we have to do it, people feel more comfortable. They understand better. It's there for them to see and most people accept it. They really do.

When we aren't afraid to be ourselves, we aren't afraid of others. We aren't afraid to speak our minds, and people like that. They will want to be in your company. Everyone is attracted to confidence. Potential partners get to know you and, sure, there is an element of CF in there, but it doesn't define the whole relationship. It's about how our personalities and qualities align with someone else's.

So I think dating with CF isn't all that different from the way other people date. Everyone has insecurities that they have to deal with and having self-doubts, worries and barriers against rejection certainly makes it more difficult. Yes, it does take someone who is strong and capable of handling stressful circumstances, someone who is compassionate, helpful and understanding to stand by you, but I do

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believe when we know ourselves and our needs, we find someone that fits and complements us. The connection will be the focus. CF shouldn't be the focus of a relationship, it's just a part that has be dealt with as well.

I say don't turn down that date because you think you will be judged or because you don't want to be seen doing necessary treatments. Our normal includes a slightly different set of parameters and that is all right. Everyone accepts it if you do. That includes people who want to date us. So go hike up those hills. Take your oxygen tank if you require it. Take breaks, let him carry you part of the way, if that's an option. And stop when you really can't go farther and don't feel bad about it. It's circumstance and people value your effort and your drive to want to do everything you possibly can. It's honestly admirable.

I recently went on a trip to Europe for three weeks with a guy I've been dating who lives in Amsterdam. If I let fear of the unknown get to me I never would have been able to do it—fear of traveling, fear of his rejection from seeing me coughing lots of mucus and fear of doing treatments in public places where strangers could stare. Don't let the fear of other people's thoughts grip you. It's their thoughts, not yours, so does it matter to you anyway?

Maybe this has just been my experience with dating, but I absolutely think when we can be ourselves and show people our personalities, talk about our goals, dreams and passions, CF isn't a barrier to finding the person that fits for us. It's part of who we are and that's it. It's an extra challenge for us and for our partners, but CF isn't a negative factor with the right partner. ▲

Ella is 24 and has CF. She is a director of USACFA. You may find her contact information on page 2.

which means it overcomes risks associated with obtaining enzymes from the animals. Those risks include viral contamination and limited supply. The Phase 3 RESULT clinical trial (NCT-03051490) that Anthera has just started will build on results of the Phase 3 SOLUTION trial (NCT02279498). SOLUTION demonstrated that Sollpura was as effective at treating exocrine pancreatic insufficiency as Pancreaze (pancrelipase), a pig-derived PERT. A key finding of the SOLUTION trial was that both Sollpura and Pancreaze maintained patients' height and weight, an indication their bodies were getting enough nutrients.

The RESULT trial will further assess Sollpura capsules' ability to overcome CF patients' exocrine pancreatic insufficiency, compared with Pancreaze. Researchers will adjust doses individually to try to achieve the best results.

http://tinyurl.com/yaz6a4bk

Anthera Achieves Half of Target Enrollment for Trial of Sollpura as Treatment for CF-related Digestive Disorder

Anthera Pharmaceuticals has enrolled half the patients it is seeking for a Phase 3 clinical trial of Sollpura (lipromatase) as treatment for exocrine pancreatic insufficiency, or EPI. The company expects to release the key results of the RESULT trial in late 2017 or early 2018. Many pancreatic enzyme replacement therapies, or PERTs, are derived from pigs. Sollpura consists of non-pig-derived lipase, protease and amylase enzymes. It helps people digest food better, which leads to them having better nutrition. The therapy's synthetic origin reduces the risk of a user developing a pig-related disease or conditions such as gout and kidney impairment. RESULT (NCT03051490) will compare Sollpura's effectiveness with that of approved pig-derived PERT treatment, such as Pancreaze, in CF patients with EPI. Researchers will look at Sollpura and Pancreaze's ability to help patients do a better job of absorbing fat and nitrogen after four weeks. The team will follow patients who were randomly assigned to Sollpura for 20 weeks after the treatment, checking their overall health and the therapy's long-term safety. Because Sollpura's formulation is more compact than pig-derived PERTs, it may also reduce the size and number of pills that CF patients need to take each day.

http://tinyurl.com/ydfp5zcn

Vertex Continues to Pursue Broad Range of Therapies for Cystic Fibrosis Patients

Vertex Pharmaceuticals is continuing to develop treatments for all people with cystic fibrosis, with clinical trials exploring combinations of its potentiator and corrector therapies. At the same time, efforts to expand the use of already-approved treatments are ongoing. In addition, the company recently completed the purchase of Concert's CTP-656, yet another CFTR potentiator in development for use as part of future combination regimens. As reported earlier, the company also has worked with the FDA to get Kalydeco approved for people with more rare CFTR mutations. The number of mutations approved for treatment with Kalydeco is now 33, and Vertex continues working with the FDA to include additional rare mutations in the drug's prescription label. Meanwhile, Vertex has submitted a new drug application with the FDA for the combination treatment tezacaftor/ivacaftor. The treatment had been granted orphan drug designation, and is intended for patients

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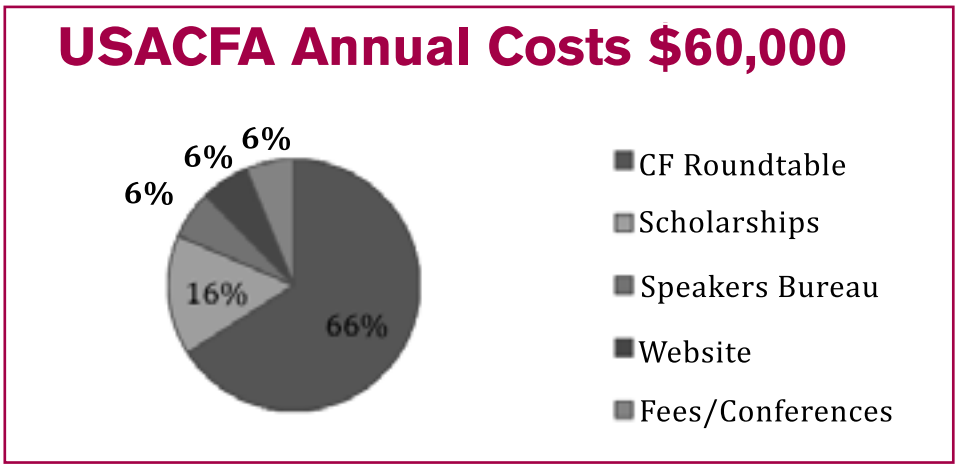
Announcing the USACFA Endowment Fund

The U.S. Adult CF Association (USACFA) is proud to announce the establishment of the Endowment Fund. The endowment was established in May 2017 by a generous donation from Nancy Wech. This endowment is critical to funding the *CF Roundtable* publication, website, blog and related programs.

The semiannual college scholarship is one such program, which was created in memory of Nancy's daughter, Lauren Melissa Kelly, who was an outstanding student and campus leader at the University of Georgia. Although Lauren lost her battle with CF late in her senior year, her memory lives on through the scholarship program that bears her name.

USACFA has partnered with The Oregon Community Foundation (OCF) to create the USACFA Foundation Endowment Fund of OCF. Our endowment fund will be maintained, in perpetuity, to provide a stable stream of income that will support both the *CF Roundtable* and USACFA programs, which the adult CF community utilizes and relies upon to maintain support and nationwide communication.

Our goal is to raise $30,000 this year. Meeting this endowment goal will enable our all-volunteer board of directors, all of whom have CF, to spend less time seeking grants and individual donations and more time advancing programs and disseminating information that benefits our CF community.



Please consider a donation to the USACFA Endowment. Your donation may be designated as a gift to this permanent fund to ensure future funding for our work. You also have the option of creating a life income gift, often called a planned gift. OCF staff works with our staff and donors to create bequests, charitable remainder trusts (CRTs) and charitable gift annuities (CGAs).

Contact Stephanie Rath, Treasurer/Fundraising Chairperson, at cfroundtable@usacfa.org for further information about the USACFA Foundation Endowment Fund.

USACFA is a 501(c)(3) non-profit organization run by an all-volunteer board of directors, all with cystic fibrosis. 100% of your gift is directed toward sustaining *CF Roundtable* and our related programs. Your gifts are fully tax deductible as specified by the IRS. ▲

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12 years old or older. Vertex also is exploring the tezacaftor/ivacaftor combination in a Phase 3 trial in patients with one F508del mutation and a second mutation that causes a gating deficit in the CFTR protein. Finally, the company is running trials exploring three triple combinations — tezacaftor/ivacaftor together with either VX-440, VX-152 or VX-659. They recently provided an update on the trials, demonstrating that all three were effective in patients with difficult-to-treat minimal function mutations.
http://tinyurl.com/yde2p96a

New Combination Therapy for Cystic Fibrosis Being Reviewed by FDA and EMA

Vertex Pharmaceuticals has announced that both the U.S. Food and Drug Administration (FDA) and European Medicines Agency (EMA) have accepted its applications for the use of the tezacaftor/ivacaftor combination treatment in a subset of patients with cystic fibrosis (CF). The application acceptance is for people with CF ages 12 and older who have two copies of the F508del mutation or an F508del mutation and one residual function mutation that is responsive to tezacaftor/ivacaftor. If approved, the tezacaftor/ivacaftor combination treatment would become Vertex's third medicine to treat the underlying cause of cystic fibrosis, offering an important new treatment option for a large group of patients with this rare and life-shortening disease.

Ways To Give

There are several ways you can support the mission of USACFA, which offers support for adults with CF through our *CF Roundtable* publication, website and related programs.

1. Unrestricted Gifts
2. Matching Gifts
3. Tribute Gifts
4. USACFA Endowment Fund
5. Bequests

USACFA is a 501(c)(3) non-profit organization run by an all-volunteer board of adults with cystic fibrosis. As such, 100% of your gift is directed toward sustaining *CF Roundtable* and our related programs. Your gifts are fully tax deductible as specified by the IRS.

Unrestricted Gifts: An unrestricted gift to USACFA/*CF Roundtable* enables us to direct funds where they are needed most. Gifts can be made by cash, check or online at www.cfroundtable.com/newsletter/donate/ using a major credit card or PayPal.

Matching Gifts: Many employers participate in matching gifts programs. This benefit can double or even triple your gift to USACFA/*CF Roundtable*. Check with your Human Resources Department for matching gifts programs and forms.

Tribute Gifts: This is a thoughtful way to honor someone whose life has been impacted by cystic fibrosis. Your gift can also recognize milestones in their life – birthday, marriage, lung transplant etc. or express the sense of loss when someone loses their fight with CF (In Memory). Your gift will be acknowledged by sending a card to the honoree or family. To make a dedicated gift, please contact us at cfroundtable@usacfa.org.

The USACFA Endowment Fund: We have partnered with The Oregon Community Foundation (OCF) to establish this permanent endowment fund. With OCF's stewardship and long-term investment program, our endowment fund will be maintained in perpetuity to secure the future of *CF Roundtable*, the Lauren Melissa Kelly scholarship and our related programs. We hope that through your donation, the Endowment Fund will grow to provide an income stream able to sustain *CF Roundtable* and its many programs in perpetuity. Our long-term funding goal is $1,200,000. We also offer our donors the option of creating a life income gift, often called a planned gift. OCF staff works with our staff and donors to create bequests, charitable remainder trusts (CRTs) and charitable gift annuities (CGAs). For more information on the endowment, please contact us at grants@usacfa.org.

Bequests: A bequest is a simple and easy way to remember USACFA/*CF Roundtable* in your estate plan. Please consider including USACFA as a beneficiary. To assist you and your attorney, the following sample language would allow you to include USACFA in your will or trust:

"I hereby give and bequeath $_____ to the United States Adult Cystic Fibrosis Association, Inc., located at P.O. Box 1618, Gresham, OR 97030-0519, federal tax ID #93-1036770."

If you have already included us in your estate plans, please let us know by e-mail grants@usacfa.org or mail so that we can thank you now for your future support.

Our mailing address for check donations is:
U.S. Adult Cystic Fibrosis Association, Inc.
P.O. Box 68105
Indianapolis, IN 46268-0105 ▲

Tezacaftor is intended to address the processing defect of F508del-CFTR and permit the protein to reach the cell surface, while ivacaftor can further enhance the function.
http://tinyurl.com/y7fg8dlv
AND
http://tinyurl.com/yckcaw2n

Fighting an old enemy in the battle against cystic fibrosis

The biggest problem for CF patients is the mucosal bacteria, which form aggregates of cells that bind together to create a substance that surrounds and protects the harmful bacteria. This often means lifelong infection for the patient. Existing treatments can reduce the bacterial population, but the infection almost always returns, due to a small portion of the bacterial cells, aptly named persisters. The cycle repeats, each time causing further organ damage, until eventually there is not enough tissue left to function. Researchers believe that they can target and destroy persister cells using a combination of two existing drugs working together in a new way. They have found that combining triclosan, a commonly used antimicrobial found in such products as toothpaste, soaps and detergents, enhances the persister-killing ability of tobramycin, an antibiotic commonly used to treat CF bacterial infections. The initial test results have been promising. The researchers found that compared to tobramycin treatment alone, adding tri-

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Recipients Of Autumn 2017 Lauren Melissa Kelly USACFA Scholarship Announced

IN MEMORY OF
LAUREN
MELISSA KELLY

The U.S. Adult CF Association (USACFA) is pleased to announce our autumn 2017 recipients of the Lauren Melissa Kelly Scholarship.

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

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

We are pleased to announce **Jacob Greene** and **Elizabeth Shea** as the recipients of the scholarships for this semester. These two exceptional recipients demonstrate outstanding potential just as Lauren did years ago. Congratulations to them! They will be awarded $2,500 each.

JACOB GREENE

ELIZABETH SHEA

Jacob Greene is a motivated young man who has volunteered his time to public speaking engagements on behalf of cystic fibrosis awareness not only in his community, but internationally as well. He graduated as valedictorian of his high school in Bellevue, Washington. He has won many awards for his scholastic accomplishments including Advanced Placement awards and participation in a summer internship at a cancer institute where he helped develop new research for myeloid leukemia. He will attend Stanford University, in Palo Alto, CA, where he will be a freshman in the autumn of 2017.

Elizabeth Shea is an accomplished dean's list student and musician from Weston, Florida. Not only does she excel academically, she has devoted countless hours playing the flute in symphonic band and orchestra throughout high school and college. She has also volunteered her time to raise awareness and funds for cystic fibrosis research by participating in the Great Strides Walk, being a team leader. Elizabeth demonstrates leadership and drive in every aspect of her life. She attends Stetson University School of Music and will be a junior in the autumn of 2017.

Both Jacob Greene and Elizabeth Shea demonstrate the leadership, intelligence and drive of Lauren Melissa Kelly. We at USACFA look forward to seeing them further develop their leadership and advocacy in the cystic fibrosis community.

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

My Extraordinary Friend

As our high school years are ending,
Remember that our friendship will never end.
I hope you know how important our
extraordinary friendship is to me.
When I needed you the most, you were
always there.

As you already know,
My shoulder will forever be there for you
to cry on.
We've shared so many laughs and cries together,
That every time we talk or laugh, I know it
is for real.

I never believed I would find a friend like you,
But without you in my life,

I don't know where I would be today.
You are certainly my guardian angel from God.
He would want us to watch over each other until
our days have ended.
We must stay strong for each other and keep
on fighting.
Keep on striving for your goals,

Never stop being the wonderful and loving
friend you are, to me.
By far, you are my "bestest" friend.
My gratitude and love cannot be expressed in words.
When our days have come to an end, just
remember this:
I love you!

-K. Pierce, 2004

PHOTO BY STEPHEN BOYER

"Through the Looking Glass: Images of Adults with Cystic Fibrosis" and "Caregiver Stories" are projects of Breathing Room, a non-profit organization. Breathing Room hosts these and other projects to facilitate open and candid communication in the CF community, supports the development of a community of adults with CF and provides education and insight for families, caregivers and medical professionals who impact our lives.

FROM OUR FAMILY PHOTO ALBUM...

LEAH SANDS

MICHAEL AND NICOLE KOWAL ON THEIR WEDDING DAY. PHOTO TAKEN AT THE BRIARWOOD COUNTRY CLUB IN HAMBURG, NY, ON MARCH 5, 2016.

TIM, ANNA AND COLLEEN VEITENGRUBER, IN SEPTEMBER 2017, ON THEIR ANNUAL CAMPING TRIP.

RICH AND BARBARA HARISON CELEBRATED BARBARA'S BIRTHDAY ON THE LINKS AT THE SANDPIPER GOLF COURSE IN MAY 2017.

ADAM KEYS AND SYDNA MARSHALL.

CALEIGH HABER IN HAWAII.

CHRIS GAUTIER (48) AND JEN WEBER (43), BOTH POST-LUNG TRANSPLANT RECIPIENTS WITH CF, RODE WITH TEAM TRANSPLANT — ALONG WITH CHRIS'S WIFE AND TWO OF THEIR TRANSPLANT DOCTORS, IN THE INDIANAPOLIS CFF CYCLE FOR LIFE, AUGUST 26, 2017.

OUR FAMILY PHOTO ALBUM... *Continued*

ELLA BALASA VISITING THE CITY OF DUBROVNIK, CROATIA. WHERE GAME OF THRONES IS FILMED

MARK SCHROEDER WITH EMELIE, HIS WIFE, AND SHANE, THEIR DAUGHTER, IN GINATILAN, CEBU, PHILIPPINES.

JACOB GREENE AND HIS SISTER, KASEY, IN WARWICK, UK, ON TOP OF THE WARWICK CASTLE, DURING THEIR TRIP TO ENGLAND AND PARIS LAST SUMMER.

CF Peer Connect May Help You

By Joan Finnegan Brooks

This year my husband, Peter, and I celebrated 28 years of marriage. Celebrating our anniversary always makes me think of when Peter and I started dating. I remember that prior to meeting him, I experienced lots of angst about new relationships – do I tell him about my CF? When and how? What do I say? I didn't have anyone I could talk to about it since I didn't know anyone older than me with CF. It would have been so helpful to hear about how someone else with CF had handled the same situations and questions.

When you're dealing with CF, you can really feel like you're in this battle alone. In the era of infection control, the sense of isolation can be magnified even more. Since I'm one of the older members of our community at age 57, I've felt like a "lone ranger" in facing many life decisions. It's not easy trying to figure out what choices are best in the context of living with and managing CF. You can feel a bit like a pioneer where everything you want to achieve is something new that's never been done.

Since we now have more adults with CF in our community than children, there are many adults facing these same life decisions. I know many of the questions I've dealt with, like dating and relationships, many other adults with CF have faced and managed, too. While we all have different ways in which we've handled those situations, sharing those experiences is the valuable part.

I feel a great sense of obligation and responsibility to make sure that other adults with CF who face the same situations don't feel alone as they chart their path forward. For the last few years, I've co-chaired a committee for the CF Foundation charged with developing a peer-to-peer mentoring program. This new free program, CF Peer Connect, is a way of leveraging the experiences we have in the adult community and sharing them with each other. The collective experiences of the CF adult community are really broad and offer a rich resource to tap into.

JOAN FINNEGAN BROOKS

CF Peer Connect is focused on adults sharing experiences around life issues (dating and relationships, living away from home, starting a career, transitioning to work, getting listed for transplant, parenting etc.). If you want to talk to someone else with CF about a specific topic like dating, you can request a peer mentor through CF Peer Connect. Hearing someone else's story and being able to ask questions of another adult with CF can be a great source of support.

Once you sign up, CF Peer Connect will match you with another CF adult who has experiences on the topics you're looking to learn more about and who truly understands what it's like to have CF. (Peer mentoring isn't for medical advice and it's not for mental health counseling.) Since all the connections are virtual, mentoring connections can take place over video, phone, e-mail or text — it's up to you and your matched CF adult how often you want to talk and what you talk about. It allows you to tap into expertise where it exists, no matter where you live.

My first experience of understanding how valuable a mentor could be in my life occurred in my mid-20s when I moved to a new city to begin a job. It was a life-changing experience for me to talk to two CF adults who were also volunteering on the board of directors for our local CF Foundation chapter. After I lost my older brother John to CF, they were the first individuals I talked to who were older than I was with CF. Like me, they were working and dealing with adult life, and married (I was single at the time). Their lives served as an example of what might be possible for my life — career success, marriage and family life. It was wonderful to talk to another adult who had gone through some of the same situations I was facing and who could share their experiences and answer my questions. They continue to be important in my life.

If you're wondering if connecting with a peer mentor would be of value to you, I urge you to reach out. CF Peer Connect facilitates the connection between individual CF adults for specific purposes — to share experiences that are relevant to their lives and for other people to learn from. You're not alone. If you're on the fence, inquire about it. I think you'll be really glad you did. For more information and to request a peer mentor, please visit CFF.org/PeerConnect. ▲

Joan is 57 and has CF. She and her husband live in Osterville, MA. You may contact her by e-mail at joanfbrooks@p-fmr.com or by phone at 617-312-0931.

ACTIVE FOR LIFE

Benefits To Working Out With A Partner

By Aimee Lecointre

Aside from the weight training and step aerobics in high school (14 years ago), I used to be a strictly workout solo kind of person. I liked the freedom to work out whenever I wanted, even on a whim, without having to coordinate a time with someone else.

Then a few years back I started going to the gym a couple of times a week with a friend. I was hesitant at first. What if she wasn't reliable? What if she doesn't take this seriously? This list went on and on. I worried for nothing. In fact, I found it fun to have a workout buddy a couple of times a week. And it turns out, there are plenty of benefits to working out with a partner.

Accountability. It's easy to cancel on yourself. It's easy to convince yourself, "not today, tomorrow." But when you have someone else relying on you, it makes it quite a bit harder to cancel, especially last minute.

Motivation and encouragement. Who doesn't feel more motivated when you have someone in your corner? When you have someone who supports you you'll also be more likely to stick to your goals! When the going gets tough and the struggle is real, it's easier to work through it when you have someone cheering you on.

Easier to try new workouts. When you have someone by your side, it's easier to feel less intimidated to try new exercises. How many of you have thought about trying a new gym, new class or new style of exercise but felt intimidated or worried? Find a buddy who wants to try something new and go for it!

Variety. Your workout partner will most likely have different interests, skills and knowledge when it comes to staying active. They can show you new things and you can return the favor. Personally, I love variety. I get bored with the same old routine. Trying new activities is one of my favorite ways to stay active and stay motivated.

FUN! I find it much more enjoyable to go to the gym or try new activities (think, paddle boarding, hiking etc.) with a friend. It makes for a better memory and lots of laughs while trying to figure out what exactly you're doing. And you know what they say? Time flies when you're having fun!

Partner Exercises. There are all sorts of new moves you can try out when you've got a buddy that you can't do solo. I've asked my friend and certified personal trainer, KC Velez, who also has CF, to share some partner exercises with us!

Here's what KC has to share: The exercises below are my favorite total body circuit workouts targeting the major muscle groups of the body. Notice that almost all of the exercises involve a core musculature component. Our core helps us in everyday functional movements, such as bending to get in and out of a car or even walking up a flight of stairs. With people who have cystic fibrosis, in particular, a strong core can help us develop muscles used for deeper breathing and to move mucus out of our airways, helping to prevent infection. Exercise is also important because it promotes a greater quality of life.

KC VELEZ

Editor's note: You should check with your CF physician before doing any exercises, especially those that cause you to touch one another or to be nearer than six feet to each other.

1) Side Medicine Ball Pass

This exercise targets the core and obliques. A strong core is important and can help you develop muscles used for deeper breathing.

How to do it: Sit on the floor with your legs crossed and slightly in the air. Have a partner stand right beside you facing forward and toss you a medicine ball as you catch it from the side. Complete 12-15 repetitions before switching sides. Complete a total of three sets.

2) Pushup to Alternating Handshakes

This workout develops upper body strength and incorporates core stability.

How to do it: Start with proper pushup form (hands directly under shoulders, maintaining a flat back while keeping your butt down and keeping your core engaged and tight). Face your partner as each of you completes a pushup. Once you come up, reach out and touch each other's hands as if you were giving a handshake and alternate hands with each repetition. If

you cannot do a traditional pushup, drop to your knees, but still maintain the same proper form mechanics as described above. Aim to do three sets of 10-12 repetitions.

3) Back-to-Back Squat Hold

Squat holds are a bodyweight exercise and are a great way to work your quads, glutes and hamstrings and are a great way to build into weighted squats.

How to do it: Stand back to back with your partner and assume the squat position. Feet should be hip width apart and facing forward. Draw in your core and keep your body tight. Aim to hold this position as long as possible. If needed, start with 30- to 60-second increments and work your way up. Complete three sets.

4) Burpees to High Five

This is a total body exercise and a great way to mix in some HIIT (High Intensity Interval Training) cardio, which works to strengthen the lungs and circulatory system.

How to do it: Begin in a standing position, facing your partner. Each of you will get down to the ground until you are on your stomachs. As you come up at the same time, jump up and high five your partner. The goal with this particular movement is to complete the repetitions as fast as possible in order to keep the heart rate elevated. Try to do 15-20 repetitions.

Note: When beginning a workout program, you may need to take time to rest between exercises or even do fewer repetitions. That is perfectly okay. Begin at a level with which you feel a light to moderate intensity and work your way up. This should be moderately difficult, but fun! ▲

Aimee Lecointre is 32 and has CF. She lives in Salt Lake City, UT. She is a Certified Nutritional Therapy Practitioner, Certified 21-Day Sugar Detox Coach and Registered Yoga Teacher. She is passionate about helping the CF community through movement and nutrition. In her free time you can find her hiking, doing yoga, cooking, reading, writing, fishing or eating! Find out what she's up to on Facebook at The Nourished Breath or on Instagram @thenourishedbreath.

KC Velez is 30 and has CF. She lives in Virginia. She obtained her Certified Personal Training accreditation through the National Academy of Sports Medicine. She also obtained a Bachelor of Arts in Political Science before she realized that fitness and healthy living were her true passions.

TILLMAN continued from page 25

closan caused a 100-times reduction in persister cells after six hours and, after 24 hours, showed near or complete eradication of the cells. Not only did the combination treatment kill all of the persister cells, it worked on every clinical sample tested.

http://tinyurl.com/y9hflo6j

French Biotech Receives an Award to Develop a Novel Cystic Fibrosis Antibiotic

CARB-X has awarded $8.9M to Antabio for the development of a new antibiotic against resistant Pseudomonas infections in cystic fibrosis patients. The candidate is a small molecule against Pseudomonas aeruginosa infections. Specifically, it targets the LasB elastase virulence factor of P. aeruginosa, which is responsible for the bacteria's ability to evade the immune system.

http://tinyurl.com/ycuj75lh

Cystic fibrosis alters the structure of mucus in airways

Mucus is important for maintaining healthy lungs. Inhaled particles, including bacteria and viruses, get trapped in mucus and then cilia (tiny hair-like projections on the surface of the airway cells) sweep the mucus out of the airway. In lungs affected by cystic fibrosis, the mucus is abnormal and the lung-clearing process breaks down. This deficit may contribute to lung infections and inflammation that cause serious, life-shortening illness in people with CF. Researchers investigated how CF alters the structure of mucus produced in airway passages. The study focused on two gel-forming mucin proteins, MUC5B and MUC5AC, that are the major components of airway mucus. The researchers found that these proteins have distinct and different structures and origins. MUC5B is produced by submucosal glands in the form of strands, while MUC5AC is secreted by goblet cells as wispy threads and thin sheets. The study also showed that once these two types of mucus protein emerge onto the airway surface, they combine so that the MUC5B strands are partly covered with MUC5AC sheets. This overall structure may be helpful for capturing and clearing inhaled particles. However, in airways that are affected by CF, these mucins look different. The strands of MUC5B become tangled, and often fill the submucosal gland ducts and fail to detach properly, and MUC5AC sheets are larger and more abundant. This work reveals that mucus from submucosal glands and mucus from goblet cells have different morphological structures. These structures may serve different purposes in clearing particulates and bacteria from lungs. The study also shows how these structures are abnormal in cystic fibrosis, explaining why patients with this disease have difficulty clearing mucus from their lungs.

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PARENTING

There Are Many Ways To Make A Family

By Dana Giacci Rogers

One of our readers, Kaeti R., wrote to us to share her story of how she and her family are beginning their journey of family planning. There are indeed many ways to make a family. Here is their story:

Ever since I can remember, I have always wanted to be a mom; not just a mom, but carry a child. I wanted to feel its movements and see the ultrasounds. I wanted to know that we had created something so precious and beautiful. Never did I think I wouldn't be able to conceive. Throughout high school and the beginning of college, I was able to maintain a 90-100% PFT score. I was very active in dance, running and cheerleading. I always knew there was a chance I was sterile and couldn't reproduce, but I also didn't think it would happen to me.

Our story began a little over six years ago. After being introduced to each other by mutual friends, my future husband and I started dating and fell in love. A year later we were married and eager to grow our family. We started with a dog, Sadie, and soon after that we started trying to conceive. After a year of trying with no success, we decided to see an OB GYN and have some tests run. Every test came back normal. There was nothing medically wrong with my female organs, but they weren't working right. My husband's tests came back normal, too. We were at a loss for words. It was a mystery. It was at this time that my lungs started to deteriorate and I switched my focus towards improving them. After a few months, my doctors told me that my lungs weren't strong enough to go through with a pregnancy and we should stop trying.

There are many different and unknown reasons as to why women cannot carry/conceive children. Women with CF are no different. Yes, our bodies have endured a lot more than some, but for the most part they all work the same. We CFers are abnormal when it comes to the function of a cell protein called the cystic fibrosis transmembrane regulator (CFTR). With this comes thickened mucus and secretions. We notice this more in our lungs, but it obstructs other areas too.

We took a year or so to grieve what we thought was our chance of having a family. Soon after moving to Colorado, we started attending a wonderful church. During a message, the pastor shared his family's story about how they had adopted their two boys and how they felt extremely blessed. It was as if God knew we both needed to be there and hear that sermon. After church, we went to lunch and both started talking about adoption. We hadn't brought it up before because we were both taking our own time to heal. After that day, we started looking into the process and going to informational meetings. We soon realized that God had a different plan than we originally thought. He was showing us that families come in many different shapes and sizes and that we have so much love to give.

KAETI R.

In Colorado, you have to go through an agency in order to adopt a child; every state is different. We interviewed and researched many agencies and in the end we decided to go with Bethany Christian Agency. We knew there was a chance of us moving again and wanted to make sure the agency we chose was active in other states, too. After being in Colorado for three and a half years, we decided our hearts were back in California and we moved back to be closer to family and friends. Our adoption file and books followed us here and we are now waiting, surrounded by loved ones.

We don't know the rest of our story — only God does — but we do know that there are many children waiting for their forever family. Our story isn't over; I believe it is only beginning. Just know you aren't alone. Everyone has trials and tribulations, and there is always someone else going through something similar or the same. All it takes is having the courage to speak up about it.

Much love,

Kaeti

Thank you Kaeti for sharing your story with us. We all wish you the best of luck.

With Gratitude,

Dana ▲

Dana is 27 and has CF. She lives in Troutdale, OR. You may contact her at: dgiacci@usacfa.org.

CONVERSATION CORNER

Bonding Bumps

By Paul Quinton

PAUL QUINTON

Beyond complete isolation in a "bubble," it is not possible to know with certainty what conditions will protect and what conditions will foment cross-contamination among people with and without preexisting disease. It is a given, however, that isolation and labeling are painful experiences for humans that are also capable of, if not certain to, cause much suffering, and pathology. So how to best manage the uncertain risk of cross-contamination when achieving "Health" requires one human being to recognize and equally respect the presence of another? "Health" is not simply the absence of bacteria or infections or even of pathology. Full "Health" requires self-respect and self-acceptance, which require equal respect from and acceptance by others.

We cannot ignore the ever-present risk that pathogens from one person may be transmitted to and infect another. Neither can we predict when contamination will occur. So, we try to behave to minimize what we have learned about reducing the chances of ill effects. But when we do so, we put at risk and even destroy self-esteem, agency and mutual respect; that is, other essentials for "Health." Is being truly healthy impossible? If we now, with our status quo policies, become isolated to ensure maximal medical protection, we ensure the sacrifice of identity and ability to respect "who we are." If we ignore the medical threats and behave normally, we may well go to the hospital or worse.

The direction to maximizing the possibility for being "Healthy" requires changing "normal." "Normal" is the common, expected behavior of all members of a community. Persons with CF cannot change the fact that they are abnormally susceptible to respiratory infections. Persons without CF cannot change the fact that they are normally susceptible to respiratory infections. But we could change "Normal" to the benefit of everyone staying more "Healthy." Currently, in our community shaking hands is normal. Disregard of distance in face-to-face conversation is normal. Depressed awareness of covering coughs and sneezing is normal. Insisting that one group of the community obey and follow a distinct behavior from the rest of the community may improve physical protection, but it is certain to undermine self-worth with stigmatism and banishment.

Then, could our community come together by adopting the same behavior for everyone to the benefit of each one of its members? If it were abnormal to shake hands, if it were strictly abnormal to cough and sneeze openly, if it were polite to openly insist on keeping a distance in conversation, the community as a whole and not just the more susceptible members would benefit from better health. Granted, it is currently regarded as inappropriate to cough and sneeze and invade space, but it is just as unacceptable to openly reprimand or chide these behaviors. If these new behaviors were the norm for everyone, no one would feel different and strange or odd and excluded for following normal rules of the community.

The conclusion then is that the CF community can accomplish much more, much less painfully, if our whole community adopts a normal practice of not shaking hands. With each threatening viral outbreak, the World Health Organization broadcasts advice against shaking hands and recommends replacing the custom with much safer elbow bumps. No one wipes or blows his nose with or on his elbow. By adopting just this one behavior as the norm, we would realize at least three important benefits. First, not only those with CF, but also those without, will be better protected from cross-contamination without being stigmatized and ostracized. Second, the community will be strengthened by this simple unifying act that signals acceptance, support and mutual respect for all of its members that bonds us and shuns stigmatizing and ostracizing. Third, the CF community could lead and set an example for the rest of the world to promote better respiratory health for all.

Handshaking is a wonderfully bad habit that spreads more illness than any other human behavior. It should be abandoned, and the cystic fibrosis community should be the first to abandon it in favor of a "bonding bump" of elbows. ▲

Paul is 73 and has CF. He is professor of Pediatric Pulmonology in the department of pediatrics at U.C. San Diego School of Medicine and Rady Children's Hospital in La Jolla, CA. You may contact him at: pquinton@ucsd.edu

IN THE SPOTLIGHT

With Caleigh Haber

By Ella Balasa

Caleigh is a young woman with CF who has established quite a presence on social media, having more than 70,000 followers on an Instagram account, @fight2breathe. She documents her life dealing with CF and a bilateral-lung transplant; now more recently dealing with organ rejection from her transplant and the rapid health decline as a result. She is incredibly knowledgeable about the many procedures and tests she and her doctors discuss, and she shares them with her community of followers in a way everyone can understand. Her genuine personality, charisma and strength are all palpable in her words through which she relays her true fears, hopes, insecurities and raw emotions about an unknown future.

Name: Caleigh Haber

Age: 27

Hometown: Orange County, CA.

Currently living: San Francisco, CA.

Siblings with CF: None. I have two older siblings, a brother and sister, neither have CF.

College: Le Cordon Bleu, Culinary School.

When/Where were you transplanted:

October 20, 2015, at Stanford University in Palo Alto, CA.

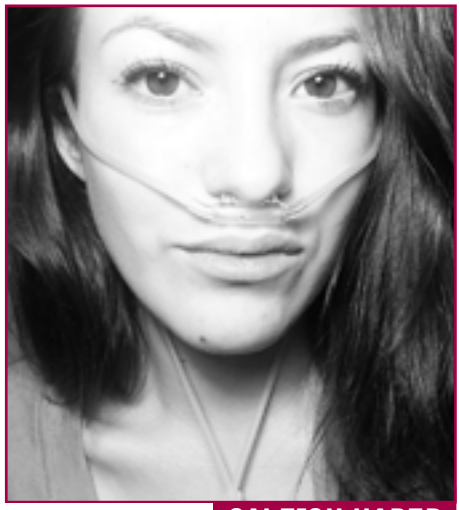
When were you diagnosed?

It's an interesting story actually. My mom was in a car accident while she was pregnant with me. She went to the hospital to get checked, at which point they realized I had meconium ileus, which is a blockage in the intestine of infants. Upon birth I was wheeled into surgery. That's when I was diagnosed. It was very common that newborns with CF were diagnosed because of this intestinal issue at birth. The newborn screening didn't include CF until 2005. Then it wasn't until 2010 that all 50 states in the U.S. included CF.

How did you decide to start sharing your life on social media? How has it evolved?

It all started when I was going through the transplant evaluation process and the doctors encouraged me to start fundraising for expenses for not only the transplant itself, but for other medical costs and living expenses. It was required that I have two full-time caregivers through the process. My caregivers are my mom and my brother. I decided to raise money through a website. At that time, I had a friend who helped make the website for direct donations so all money I raised could go toward my transplant expenses. As a more personal way to reach my friends and family, I began writing and posting more details on the site and other social media outlets. It evolved naturally. I posted short blogs on the site about health updates to keep everyone informed of my condition and status. I started gaining more and more followers as time went on.

What makes you want to be so open with strangers/followers about your struggles and triumphs dealing with CF?



CALEIGH HABER

I've always been very open about my disease. I also have quite an outgoing personality. So it just seems natural for me. Growing up I was raised "normal." I was never not allowed to do something or treated differently because of CF. That helped me feel like there isn't anything to hide. It's just part of my life. All my friends and family always knew everything. I was encouraged by my mom to share about my CF health journey. After my Make-A-Wish trip to New York City my freshman year of high school, I gave a speech in front of my whole high school about CF. I've always shared and educated.

You have more than 70K followers on Instagram now. To what extent has gaining this audience impacted your life? Has it also at times torn you down?

Honestly, it's given me tremendous support. 100 percent. I know I wouldn't be this motivated to live and go on without them. All of the uplifting and encouraging prayers, thoughts and comments make me feel grateful, joyful. In a way I feel responsible to succeed. That drives me. I have all these people looking up to me, I have to give it everything and not let them down by dying. Especially those viewers who have CF and are younger and don't know anyone else's CF story besides mine and their own. I want them to see that there is hope for their future. One thing that does scare me is passing away and leaving hopelessness in the minds of those who thought I could fight forever. Hopeless for perhaps their own fate with fighting CF. Sometimes I get insincere comments

from people who truly just don't understand this disease, my situation and my view of life. Such as why am I trying to still do so many things when I am so sick? And why am I asking for donations, when it looks like I do so many things and eat out a lot? And why am I fighting so hard to live a life if I am not really doing much living now, just existing? There will always be questions. Overall though, like I said, it's been a tremendous blessing to have the support I do.

You have a wonderful family support system, too. You talk about them frequently in your posts. In what ways have they gotten you through the many obstacles you have faced?

My family is my biggest support. They encourage me to keep fighting and never give up. They are always there for me, for every doctor appointment, hospital stay, procedure. My family has always believed that I can do everything and anything. They believe nothing can hold me back and make me believe that also. If there is a concert or event I really want to do or see, they will make it happen. Whatever that entails, oxygen, wheelchair, the whole thing. My brother and I are incredibly close. He's always told me to be myself, to be confident in who I am, that I am cool because of the person I am and that people will like me for being me. This has certainly helped me when I have lost friendships along the way due to being absent in those friends' lives and unable to participate in social things due to my health. He encourages me to keep active, too, in order to remain strong. My family is also very supportive of my social media platforms. During hospital admissions, they will say, "Caleigh come on let's show your viewers that you can do a squat. One more leg lift." Even though they know it's so difficult for me, they push me to be stronger and keep a positive outlook on life.

You've also shared quite a bit about your boyfriend recently. Can you talk about how you met and what his understanding about CF was in the beginning and how he supports you now?

We met at a wedding. He was a fraternity brother of the groom; I am a childhood friend of the bride. We made small talk, and he looked me up on social media after we talked about our "professions." I wasn't really interested in a relationship at the time. I was just focusing on my health. I was still recovering from a major health setback a few months before. I barely had any hair and had recently relearned to walk. Anyway, I caught the bouquet and I have a tradition of drying the flowers out and giving them back to the bride after the wedding. His friends and mine had all hung out at a fire pit and I accidentally left the flowers there. I knew he was still there so I direct messaged him on Instagram, as I remembered he had recently started following me. I asked him to bring them to me. When he handed them to me, he said, "This will be the story we tell all our friends when we get married." Soon after, he flew from his home state of Ohio to visit me. Unfortunately, I was hospitalized during his visit. He extended his four-day visit to two and a half weeks, quickly. I was hesitant at first to include him in all aspects of my health, but we got closer over the next few months. I flew to visit him and he came to see me often. A few months into our relationship, I wasn't able to travel anymore due to my failing health, so he made the move to San Francisco for us to be together. He knew right from the beginning that I have cystic fibrosis and I'm a transplant recipient. He has always been extremely supportive. In the beginning my health definitely scared him, I believe. When I was sick and hospitalized and he was across the country on the East Coast, he felt helpless because there was nothing he could do for me. Now we've both gotten to the point where he helps me with my medical regimen daily. I am so fortunate. We have a very strong relationship.

You are facing a very challenging time right now, going through chronic rejection. You seem to have a good medical understanding of all the treatments and options you have. Do you ask the doctors for in-depth explanations or do you do your own research?

Both. I've always been very interested in medicine. Growing up I wanted to be a doctor and surgeon. I do my own research for sure. I am very proactive about my health, and I have a very good relationship with my care team at Stanford. They know my personality. I ask a lot of questions and they don't sugar coat things for me. I'm a realist and they always tell me exactly what is happening and what my options for treatment are. They lay out my options and we together make the best decisions for my health. As far as relaying the information to my followers, I certainly try to explain in a way that everyone can understand.

Has this recent rejection diagnosis been the hardest point in your life thus far? If not, what was?

No. My most difficult experience was six months post-transplant. I was feeling really great. I could run, had lots of energy. I honestly felt like I was living a "normal" life. My brother, mom and I decided to take a trip to Hawaii for vacation. I never would have thought anything could go wrong since I was in such good condition. During the vacation I got dehydrated, which very easily happens after transplant, and developed an intestinal blockage. While on Oahu I was hospitalized. I got so bloated that my stomach was pressing on my lungs

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and I wasn't able to breathe. They tried to clear the blockage, but after only a couple days I was put on a ventilator. It was made clear to my family that the hospital wasn't going to be able to keep me alive. My mom and brother are my heroes. It was the Fourth of July weekend and there were no life flights available to medically evacuate me back to Stanford. My mom figured out a way to hire a jet, two nurses, two respiratory therapists and two pilots to transport me. After an ambulance ride to the Honolulu airport, a six-hour jet ride to San Jose and another ambulance ride to Stanford in Palo Alto, I made it to the ICU at Stanford. I ended up in a coma and on an ECMO, as well as the ventilator. When I woke up many weeks later I was unable to talk or move. I had developed severe infections in my lungs from aspirating into them. After I finally cleared my infections and gained stability, I was able to come off the vent and ECMO. I had to relearn how to do everything. First to communicate through a letter and signal card. Then to lift my head, swallow, chew, talk, sit, walk, use the restroom and care for myself. It took many months to rehabilitate me, but I am glad I had the support.

What kinds of things do you occupy your time with on a daily basis now that you really aren't feeling well, low energy and such?

I try to do as much as I can. I stay busy with morning doctor appointments and treatments almost on a daily basis. I try to do things around the house, some chores. I'm a social eater, we like to go out for food. And sometimes we just like to drive around and explore. Since I'm really not able to do much on foot, it's a different way to see places I've never been, even around here in San Francisco. I also love puzzles, binge watching TV and answering e-mails from followers.

When your health improves, and I know it will, when you are given the opportunity for another transplant, what do you hope to accomplish in the future? What are your aspirations, big goals?

I'd one day like to have a family. I'd really also just like to be able to go to the grocery store, browse the aisles, shopping for good food to make. Since I went to culinary school, I'm a big foodie. I would like to be able to provide for my partner one day. Have dinner cooked when he gets home from work after a long day. I want to have independence again. I'd also love to have one more amazing, carefree vacation, where no medical issues and obstacles stand in my way. I'd really enjoy that. Going back to the question about some negative comments I receive from viewers about why I'm fighting so hard to live a life when I'm not really doing much living right now. Well, it's because I want to get back to that point. I want to fight through this time to be able to hopefully have that one day. I won't lose hope. I want to be present, I don't want to miss anything. It's really the innate will to live and enjoy it while I'm doing it.

What advice do you have for those facing a very difficult time with their own struggles with CF? What's your motto or philosophy?

First, as a general point, don't allow yourself to be treated differently or to think you are different or less than because of CF. It's very important to have self-confidence, to know who you are. When you know yourself, others accept you and like you the way you are. If they have a problem with it and can't accept it, it's on them not you. Mental strength is very important in battling our obstacles. Second, stay active. Taking deep breaths with our lungs and working them only helps. Be compliant. It's hard to do and sometimes it's easier to give up on treatments and medications because we are just tired of it, but don't, it will help keep you alive for longer. Last, make sure you have good communication with your care team. It's vital to be active and heard in your care process, after all this is your body not anyone else's. When you take an active role in your health, you are more invested in the outcome.

Lastly, can you talk about how you connect with the CF community besides just sharing your story?

I recently started a campaign called Fighting2EmpowerOthers. This is a way for me to connect with others who have CF and to build our community. I have a questionnaire that people can fill out. It is available on my social media platforms. I ask people with CF to share pictures of them being active, cooking a high calorie recipe, doing their daily treatments and really, anything they want to share. And then through my social media platforms, I share examples of these people living their lives to hopefully encourage and motivate more people. It's important for people to know they aren't alone in this fight and we are all doing it in our own ways and succeeding. It's important to have friendships with people going through the same experiences you are. I want as many CF patients as possible to get involved within the CF community and know they have a support system. I never want anyone to feel alone.

If you are interested in following Caleigh's story and having a glimpse into one young woman's life living with CF, then find her on Instagram @fight2breathe, as well as on Facebook and consider donating for her possible second lung transplantation at https://shop.fight2breathe.org/. ▲

Ella Balasa is 24 and has CF. She is a director of USACFA. You may find her contact information on page 2.

Andrea Eisenman is 52 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. Jeanie Hanley is 54 and is a physician who has CF. She is a Director of USACFA and is the President. Her contact information is on page 2. If you would like to be interviewed for "In The Spotlight," please contact either Andrea or Jeanie.*

True Warriors

By Linda Stratton

"American Ninja Warrior," I love this television program! The strength, control and power of the contestants' bodies, men and women, cause me awe and wonder. The warriors are required to go through a course of obstacles testing the strength of upper and lower body, their speed and timing essential. A fall off of an obstacle usually results in a plunge to the cold water below.

During a previous season's city trials, a young man from Pennsylvania prepared to take his turn on the course. During a video introduction, he explained about battling cystic fibrosis (CF) and how it affected his training. As a fellow CF sufferer, my admiration for this hopeful warrior grew while listening to his story. He worked hard to fulfill his dreams, not allowing CF to get in his way. He talked about nutrition, training and nebulizer treatments as tools in his arsenal against the difficult, challenging course as well as the disease. In my eyes he became a true, life warrior.

His words opened a door in my mind to long lost memories of a time, as a young CF adult, when nothing got in the way of my own desires and how I wanted to live. CF was a small part of me and my active life—an annoyance to be tolerated. In those earlier days, having been blessed with less severe symptoms, I could ignore the disease and live freely. My activities included travel, sports, hobbies, dancing and parties; all of this while working a full-time job. Nebulizer treatments were done sporadically, hospital stays almost non-existent and oral medications few. In my view of life, there were no limits! Truthfully, until the need for supplemental oxygen came into the picture, I lived as I pleased with small increments of time and energy devoted to CF, when necessary. Was that time of freedom taken for granted? Most definitely!

Today, looking back with fond memories of those carefree days, I sit through strictly scheduled nebulizer and other medical treatments. My battle with CF has become a serious one, taking priority over all else. Spending several hours a day doing numerous treatments, I have plenty of time to watch my favorite television shows. In fact, during this last holiday season one of the stations ran a "Ninjathon," starting with the very first program aired, several years ago, to the most current—an entire seven days running. Viewing hour after hour and day after day distorted my sense of reality. About halfway through, I began to relate closely to the contestants and felt I, too, could play this game. With visions of a lean, strong body, I pictured jumping and moving through the obstacle course. Leaning left, leaning right and tensing up, I found myself breathing harder with all of the warriors' efforts. It wasn't until I mistakenly walked in front of my bedroom mirror, after a shower one morning, that shock brought me rudely back to my grim reality: I am a sixty-three-year-old woman, fighting a chronic, terminal disease. I'm pale, overweight and out of shape; a combination of the havoc of CF and a few poor choices in caring for my body and health.

Looking forward to each new season of American Ninja Warrior, I no longer have illusions about watching the "Ninjas" deftly conquering the course set before them and my living vicariously through the adventures of their competition and reward—I still find it exciting and exhilarating! During a recent clinic visit, my doctor asked if I'd been exercising. I answered, "Does watching American Ninja Warrior count?" ▲

LINDA STRATTON (RIGHT), CELEBRATING HER 63RD BIRTHDAY WITH SISTER, CHERYL GROSSHANS.

Linda is 63 and has CF. She lives in Denver, CO. Having cared for her parents for the past ten years, she is now looking forward to a new, active life in a senior community located in "quaint" Louisville, CO. She'll be participating in local social gatherings, yoga classes and community gardening.

SEARCHING FOR THE CURE

Clinical Trial FAQs Answered

By Meranda Sue Honaker

Research remains at the forefront of advancing current and future therapies of all people who have cystic fibrosis (CF). A multitude of people are involved in research including but not limited to physicians, scientists, nurses, clinical trial coordinators, pharma companies and, especially, patients. We all play a pivotal role in helping advance research forward. As a veteran of 15-plus years participating in CF research, I have firsthand knowledge of the research process from a patient's perspective. This knowledge has allowed me to better assess clinical trials I find of interest, in addition to sharing my unique knowledge with others interested in participating in CF clinical trials. In this column I am discussing five FAQs I have personally received from others interested in enrolling in CF research. Although the information provided in this column is not extensive, it provides answers to questions I have received from friends and parents of CF children interested in participating in clinical trials.

1. **How do I know if I qualify for a clinical trial?** Qualifying for a clinical trial depends on the study being conducted. Inclusion and exclusion criteria for a trial is a list of factors taken into account to ensure the appropriate group of patients are selected for a study. For example, CFTR studies will state inclusion criteria for a specific genetic mutation as one of the qualifying factors for participation. Other studies, such as an anti-infective study, will have a specific organism listed under inclusion criteria (i.e., MRSA, pseudomonas et cetera). In general, research studies that are nonspecific to an investigational drug do not have restrictions on genetic mutations, infection or age. These types of studies include biomarker studies, early intervention studies and observational studies. One example of a CF study in which I participated is a home spirometer study. This was part of an early intervention study and there was not an extensive list of inclusion criteria to participate. Essentially any person with CF willing to conduct daily home spirometry and answer a few symptom questions could participate.

2. **What are the risks and benefits of a clinical trial?** There are risks and potential benefits associated with participating in investigational drug studies. The U.S. Food and Drug Administration (FDA) has strict rules and protocols in place to help minimize risks associated with participation in drug studies. Specific risks associated with a drug study are unique to each investigational drug. It is impossible to predict how a person will respond to any medication, FDA-approved or not. It is essential to assess potential risks associated with a drug study by discussing it with your CF doctor and the principal investigator overseeing the drug study that you are considering. Benefits of participating in a drug study are not guaranteed but are possible. For example, in recent years I have participated in two CFTR studies that allowed me to gain early access to effective medications years prior to FDA approval. These experiences have been overall positive, despite various side effects, due to improving my overall quality of life.

MERANDA SUE HONAKER

3. **What safety measures are in place when participating in a clinical trial?** Safety remains a top priority in clinical trial participation. Prior to a medication being tested in humans it must be approved for testing by the FDA. This diligent process begins with a detailed review of the drug company's FDA application to the laboratory studies as well as risk assessment based upon the data available. Moreover, the FDA continues to review information as it becomes available in drug studies to ensure patient safety is prioritized. The FDA has strict guidelines in place designed to protect those partipating in research. The rules and protocol set in place for a clinical trial must be followed by all those involved in the clinical trial process from the study sponsor to the physician overseeing the study. Lastly, when participating in a drug study there are a multitude of tests being conducted to monitor the safety of an investigational drug. Some of these tests may include: blood tests, EKGs and spirometry.

4. **What if I decide I no longer want to participate in a clinical trial?** Participation in clinical trials is voluntary. This means you are able to withdraw from a study at any time. Withdrawing from a study will not affect your care at your CF center. Making informed decisions is part of

the clinical trial process from enrollment to completion of a study. Although I have not withdrawn from a clinical trial to date, I know others who have, feeling it was in their best interest. Their physicians remained supportive and continued providing consistent quality healthcare.

5. How will I know if I receive drug or placebo in a blinded clinical trial? This is the question every study participant, myself included, ponders. Blinded studies are necessary to adequately determine how effective a drug is in a patient group. Our disease symptoms can vary from day to day and for that reason are challenging to follow. When we are "blinded" to what we are receiving we cannot, with entire confidence, know if our symptoms are related to an investigational drug. Researchers need to compare patients on placebo with those on the drug to look at both safety and efficacy of that drug.

I find it quite exciting to play the "placebo guessing game" after enrolling in a drug study. I always hope for active drug (don't we all?) but always consider that I may be on placebo. In my personal experience I realized a few months into the Orkambi Phase 3 blinded study that I was in fact on the drug. This was due to persistent side effects and changes in my quality of life that persisted for the duration of the study. However, I enrolled in a different double-blinded CF study over a year ago where I played the "placebo guessing game" the entire duration of the blinded phase of the study. Once transitioning into the open label phase of the study I became fairly confident I did, in fact, have placebo during the blinded portion of the study.

Participating in a clinical trial is a personal choice that is made after learning all the information available about a specific study. I encourage all our readers to consider partipating in CF research as best you are able. There are great opportunities available to play an active role in future advancements affecting everyone who has cystic fibrosis. With every new approved medication we are one step closer to our ultimate dream: a cure. I am confident with the phenomenal group of dedicated people involved in CF research our dream will come true. If you have any questions, concerns or would like to write to me about clinical trials, please contact me at mhonaker@usacfa.org. ▲

Meranda is 33 and has CF. She is a Director of USACFA and is the Vice President. Her contact information is on page 2.

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past year alone I've been diagnosed with Graves disease and CF liver disease in addition to my existing diagnoses of osteopenia, asthma, chronic sinusitis and, of course, CF. Having a partner with the right kind of support, the right amount of patience and the constant hopefulness makes the difference between a good day and a bad day. It takes a special person to both choose and commit to a spouse with CF. I learned the hard way that compliance means nothing when you're treated like the sick, dying person implicated by your diagnosis. CF, after all, is merely a diagnosis and life is only 10 percent what happens to you. The other 90 percent is how you respond. Don't settle in your relationships or with your health. You can have both if both of you work together through sickness *and* health. ▲

Sydna is 36 and has CF. She is a Director of USACFA and is the current Secretary. Her contact information is on page 2.

Clinical Trials

Compiled by Reid D'Amico

1. Phase 2 study of VX-152 combination drug in people with cystic fibrosis (Vertex VX-152-102)

https://www.clinicaltrials.gov/ct2/show/NCT02951195

https://www.cff.org/Trials/Finder/details/458/Phase-2-study-of-VX-152-combination-drug-in-people-with-cystic-fibrosis

2. Study Assessing PTI-428 Safety, Tolerability and Pharmacokinetics in Subjects With Cystic Fibrosis

https://clinicaltrials.gov/show/NCT02718495

3. Phase 2 study of inhaled nitric oxide in people with CF (Novoteris NO-CF-02E)

https://clinicaltrials.gov/ct2/show/NCT02498535

https://www.cff.org/Trials/Finder/details/377/Phase-2-study-of-inhaled-nitric-oxide-in-people-with-CF

4. Phase 1b study of inhaled AZD5634 in adults with CF (AstraZeneca D6600C00002)

https://clinicaltrials.gov/ct2/show/NCT02950805?term=astrazeneca+cystic+fibrosis&rank=2

https://www.cff.org/Trials/Finder/details/471/Phase-1b-study-of-inhaled-AZD5634-in-adults-with-CF

5. The Impact of Insulin Therapy on Protein Turnover in Pre-Diabetic Cystic Fibrosis Patients

https://clinicaltrials.gov/show/NCT02496780 ▲

Reid is 24 and has CF. He is a Director of USACFA. His contact information is on page 2.

Two Years Ago

By Leah Sands

LEAH SANDS

I'll never forget the emotions I was feeling that morning. I had just gotten to work and was sitting in my cubicle. Everyone was going about their normal day at work, and I had this crazy moment happening to me. I was anxious, excited and scared. I felt like I was holding my future in my hands — a future that could be amazing or one that could be very disappointing. And I had absolutely no control over what would happen. The only control I had was whether or not I wanted to take the risk. If I chose not to take the risk, I would continue on my normal path where my world was predictable. But as safe as I wanted to feel" with predictability, the expected was scary as well. I felt a tear fall down my cheek, and I quickly wiped it dry so no one could see my feelings seeping out. This was really a moment that I had been waiting for my entire life, and that moment was today, August 14, 2015. I looked at these two oval-shaped, pink pills in my hand with some random letters on one side. I shook them around like a pair of dice, made sure no one was watching, and then swung them into my mouth. No strange taste, not too big, not too small, and a quick sip of water sent them down. Okay, done. I did it. Now I wait. Wait for what?

I spent the next hour thinking. I couldn't even focus on my work; I had much more important things to think about. Was I noticing anything different yet? Nothing yet. I tried to focus on other things, but it was difficult. A couple more hours passed. I started to notice something. I was feeling a bit of tightness in my chest, which was making me cough. Some people warned me that would happen. And for some people it was enough of an uncomfortable side effect to make them stop the drug. Part of me was confident and strong, telling myself that I was going to push through it. But the other part of me started to get a little scared, because I didn't want to lose any ground with my health. The way I was suddenly feeling almost made me feel like I had made a mistake, that I shouldn't have taken the risk. My emotions kept swaying back and forth all day and continued throughout the night. As I lay in bed on the first night of this new day, I realized I had made it through what was quite possibly the first day of a "new" rest of my life. I gently drifted off to sleep...until I was rudely awakened by coughing fits and sweating. Something was happening to my body. It didn't feel like my own. I liked it and I hated it all at once.

Over the next several days, I noticed more subtle changes. Things were feeling different, but in a good way. I was sharing my experiences with others on social media, who were also going through the same things. We shared our emotions, our symptoms, our bodies. We were trying to make sense of what was happening to us. We were part of a clump of people with our disease that was given a possibility - the possibility of living a better life. This was one of the first drugs ever to treat our condition at a cellular level. This was something we had been waiting for our entire lives. Being born with a grim prognosis of living only into childhood or teenage years was an existence most of us had gotten used to. It was our reality. And now our reality had been skewed. We had potential to go from our health progressively getting worse, to perhaps our health remaining somewhat stable. So together, we struggled to understand ourselves.

As we shared our experiences together, I noticed something terrible. My friends, my fellow fighters, my inspirers...were getting left behind. This new chance at life wasn't available for all of us with the disease. It was meant to only treat a fraction of us, and I felt their agony. I felt like I was hiking on a trail with my friends. Suddenly, some of us had the power to see in the distance and the trail looked beautiful ahead - sun rays shining through the leaves, new vibrant colors, and the obstacles on the trail became easier and less. Something wonderful was possibly at the end. And as I looked behind me on the trail, I was leaving many friends back there to walk alone. Where they were still walking, it was a little less bright and the barriers were many. They were struggling and fighting for each step on the path. I wanted them to see what we were seeing, and to see what wonderful things might be at the end. I wanted so badly to turn back and help them, but I couldn't. And that's not me. So I wanted to just stop the walking until everyone had the power to join me. I didn't want an unfair advantage, because I have always felt that we were in this together. So I

> I didn't want an unfair advantage, because I have always felt that we were in this together.

had a lot of guilt. Why me? Why was I given this opportunity? Someone, please help my friends.

But I continued. I continued the drug. I watched some of my fellow fighters surrender to the effects of the drug. It was making them worse, and that's something you don't need to happen when you're already fighting a losing battle. On the flip side, some were doing well. Some were feeling amazing things from the drug, me included. My body started to feel a tiny bit stronger, a smidge more energetic perhaps, and a few minor improvements in my symptoms. Subtle changes, but changes nonetheless. Before I knew it, I had been taking those crazy pink pills for eight months. I was at work and I noticed I hadn't coughed much that day. I was breathing easier even. As I sat there and started to think some more, I realized my most dreaded symptom hadn't been present in my life for the last four months. My monthly lung bleeds had been nonexistent for four months! How did I not notice that? Wow. I finally felt like this may be the moment I was hoping and dreaming for. And again, I wiped off that darn liquid that was dripping from my eye.

So here I am, exactly two years later. And you may be wondering how I am feeling now after more time has passed. In a nutshell, I still feel like I have CF. I still have good days and bad days that come with having a chronic illness. I still have the usual problems that people with CF have, and have to continue on my same treatment regimen. However, there are many things that have changed, and if I look back to where I was on August 14, 2015, I realize that things are quite different. Two years ago I was in a bad place. My health was starting to decline and I could feel it throughout my soul. I spent a lot of time thinking about what was happening to me, how I was going to handle the next disruption to my life, and I felt alone. And when I look back and remember how I was feeling back then, I realize and appreciate that I am in a much better place now. I have days where I lay in bed at night, hear the silence of my effortless breathing, and realize that I am feeling so good. Like, "Man, I felt great today!" I don't have symptomless days by any means, but I have days where I feel like I'm not as much of a sick person. Where I may have gone the day without thinking much about my CF. Which is a huge improvement from before. And because of that, I am thankful to say that Orkambi has simply changed my life.

Here's the kicker. You know what's the best news of all? My dear friends will soon have a medicine too. So many advancements and options are coming down the line in the near future, that we will all have a fair chance to live in this world. I can't wait for everyone to have moments like I do. I will no longer have to leave my fellow fighters behind on the trail. As soon as I can, I will be running back on the path, picking up all of the warriors along the way. Together we will walk easier, feel the warmth of the sun rays, see the amazing colors...and find that beautiful something at the end. ▲

Leah Sands is 36 and has CF. She lives in Michigan with her wife and two sons. She enjoys playing softball and volleyball, traveling and photography. You can reach her at leah.sands@gmail.com.

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http://tinyurl.com/y9ddhpcm

CF Study Shows Interactions Between CFTR Protein and Insulin May Contribute to Disease

Scientists have discovered that the defective CFTR protein leads to faulty insulin signaling, which, in turn, can cause diabetes and also contribute to cystic fibrosis complications. Insulin is a hormone that regulates metabolism, or the conversion of food to energy. Diabetes is common in cystic fibrosis patients. Scientists believe it stems in part from fibrotic scarring of the pancreas, the organ that produces insulin. However, lack of functional CFTR protein in the pancreas, with a resulting imbalance in chloride ions, may make it more difficult for insulin-secreting cells to get insulin through their membranes. In healthy people, glucose (blood sugar) helps trigger a cell's release of insulin, with help from ions inside the cell. But when the concentration of chloride ions is abnormal with a resulting loss of ion balance, the process gets disrupted, impairing insulin secretion. Meanwhile, research on the lungs has shown that CFTR protein is needed for the cells lining the airways to absorb glucose. Without a functional CFTR protein, insulin is unable to prompt glucose to enter the cells. The increased sugar content on the cell surfaces offers ideal conditions for bacteria to grow. Normally, insulin also increases what scientists call barrier immunity by strengthening connections among surface cells that prevent microbes from entering the body. This feature also requires the assistance of a functional CFTR protein.
http://tinyurl.com/ybxmh8vm

CF Study Indicates Chronic Achromobacter Infections Lead to Worse Patient Outcomes

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CFRI: Relationships and Presentations 2017

By Jeanie Hanley

Sitting comfortably at the table among the enormous slide screens and brightly decorated conference room, I was zapped out of my reverie as participants hurriedly entered and took a seat. The room came to life, animated with laughter, elbow bumps and conversation. This was a daily occurrence at the conference and I was excited to be a part of it. Every year has been more special than the last by meeting more people with and without CF. Relationships with them have also strengthened.

The Focus topic for this issue is on relationships and, although I will mostly summarize CFRI speaker presentations and the exceptional information disseminated, getting to know others occurred easily at the 2017 CFRI National Cystic Fibrosis Family Education Conference. It took place in Redwood City, CA, from July 28, 2017, to July 30, 2017. There were a multitude of outstanding speakers, a raffle, fun, dancing and celebrating – all while maintaining safe and strict infection control measures among all participants, especially those of us with CF.

Let's move on to a brief synopsis of some of the conference presentations. If they interest you, then watch them at www.cfri.org, where a complete list of videotaped presentations can be viewed.

Ray Pool, author of his book entitled *Lessons from a CF Cornerman*, gave a dynamic and entertaining talk on key ways to advocate for yourself and others. These lessons became evident during his journey of caring for his wife with CF - from her hospitalization and critical care to transplantation and recovery. In addition to the videotape recording, you may want to check out the Spring 2017 issue of *CF Roundtable* for the superb book review by Piper Beatty Welsh or just buy the book!

Kristin Riekert, Ph.D., from Johns Hopkins is an expert on treatment adherence issues. She provided insight on the barriers to our CF care. What gets in the way of fully following your treatment plan and being "adherent"? Her lively session hammers out some fascinating stats on how her research team measures adherence. She goes on to describe what happens to adherence when you're faced with declining health such as IV meds, lower lung function, more severe disease etc. and the ways you can overcome it.

JP Clancy, M.D., started Saturday morning with an update on CF research and provided a deeper understanding of the pesky CFTR protein that is mutated in all of us. Just as important is how the newer drugs available and on the horizon will enhance the function of CFTR at the DNA, RNA or protein level. He also added the benefits of newer agents such as anti-infectives, anti-inflammatory and mucolytics. There are many up and coming medications in the pipeline, so listening to his presentation and understanding how they'll help you is well worth it.

An awesome CF researcher, Lucas Hoffman, M.D., Ph.D., from Seattle Children's Hospital presented, "What to Expect When You're Expectorating" – a yummy good title to see just before lunch. Here you'll learn about sputum and biofilms, the importance of small colony variants, microbiomes in our lungs and the paths to discerning their significance and that of the bugs that love our CF lungs. The wonderful finishing touch was the discussion on the ways that new CFTR modulators such as ivacaftor (Kalydeco®) positively alter the bacterial environment in our lungs.

The outstanding Paul Quinton, Ph.D., from UCSD took on the enormous task of breaking down the last forty years of CF research breakthroughs. His relaxed style and humor

LAURA MENTCH, USACFA BASKET RAFFLE WINNER, KRISTIN SHELTON, AND JEANIE HANLEY AT THE CFRI CONFERENCE.

made this task quite interesting. You'll learn quite a bit about where we've been and where we're headed. We've come a long way, baby!

Stanford pulmonologist, John Mark, M.D., graced us with an exceptionally Zen presentation on "Mind Body Therapies in CF Care." He answered any question you could possibly have regarding alternative and integrative health from meditation and yoga to hypnosis, imagery, tai chi, music therapy and more. I didn't want this talk to end. But like all good things, it did, leaving us very optimistic that these therapies will positively affect our health and may be instituted by trained healthcare teams in the future.

Not summarized here are other excellent presentations such as "Adult Health Issues" by Cathy Chacon, an Adult CF Nurse Coordinator from National Jewish in Denver, covering a range of issues near and dear to our CF. Laura Mentch has contributed an overview for this autumn *CF Roundtable* issue of a very eye-opening and fascinating PT session by Karen von Berg. Our very own columnist, Isabel Stenzel Byrnes, filled us with "Belonging & Identity"- a deeply introspective session on growing up with CF.

The raucous CFRI Banquet and Dance Party Reception on Saturday evening included many delectable baskets that were raffled off, including our very own USACFA/*CF Roundtable* basket (see photo) filled with "Chocolate, Tea, Coffee & Fun" themed items that represented each of our directors' locales. I am happy to report that Kristin Shelton, RRT, at Stanford won our gift basket!

Having CF at the conference meant that you were treated very well and rarely had to lift a finger except to use your utensils. All this was done in an effort to reduce cross-infection. Dedicated hotel staff served your food, others at your table poured your water, salted your food or got you coffee. It was difficult to readjust once I returned home. Not really, but it was a very nice perk of having CF at the conference. I hope you can join the conference next year and experience this and the camaraderie, deepening your relationships in the beautiful CF community and gaining oodles of new knowledge. If you aren't able to attend, make sure to join the live streaming of the presentations, so you don't miss out. See you next year. ▲

Jeanie is 54 and is a physician who has CF. She is a Director of USACFA and is the President. Her contact information is on page 2.

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The risk of cystic fibrosis patients with chronic Achromobacter infections dying or having to have a lung transplant is twice as high as in patients who don't have the bacteria. Researchers grouped patients into three categories: (1) no history of infection, (2) intermittent infections or (3) chronic infections. They found that those with chronic Achromobacter infections were twice as likely to die or need a transplant as those with no history of infection. The average age of death or a transplant among those with a chronic Achromobacter infection was 21.5 years. It was 29.5 years for patients with intermittent Achromobacter infections, and 30.3 years for those with no history of infection. Patients with chronic Achromobacter infections also had poorer FEV1 percent scores, but their scores did not decrease further after they developed the chronic infection. While the study demonstrated that patients with chronic Achromobacter infections are at higher risk of dying or needing a transplant, it failed to answer the question of whether the infections occur in sicker patients or whether infections contribute to more severe lung disease.
http://tinyurl.com/yck62toz

As more adults are diagnosed with cystic fibrosis, radiologists look for patterns

Marked improvements have been made over the past few decades in managing cystic fibrosis, but as more adults are diagnosed with the disease radiologists can do more to monitor the wide spectrum of CF in adults, including nonclassic imaging findings. Although CF is usually diagnosed in children with progressive multisystem involvement, up to 7% of CF cases are currently diagnosed de novo in adults with subtle manifestations distinct from the typical features of classic CF. Recurrent pancreatitis, chronic sinusitis, and CBAVD are several of the ways in which CF is identified in adult patients with relatively rare mutations and with overall milder manifestations. It is important for radiologists to recognize the wide spectrum of CF to optimally monitor disease progression and response to therapeutic interventions in distinct adult patient populations.
http://tinyurl.com/yd2bg6a5

Good Nutrition May Slow Cystic Fibrosis Patients' Lung Disease Progression, Review Reports

Good nutrition may slow the progression of cystic fibrosis patients' lung disease. It's especially important that cystic fibrosis patients with an end-stage lung disease not limit their consumption of healthy food, even if they have diabetes. People with chronic lung diseases are at risk of developing pulmonary cachexia, a condition of muscle wasting and weakness. Pulmonary cachexia is the final result of malnutrition in patients with end-stage lung disease. In addition to speeding up the

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CFRI: CF Beyond The Lungs: The Role Of Physical Therapy

Presented by Karen von Berg, DPT, PT

Reviewed by Laura Mentch

On Saturday morning at the CFRI conference, Dr. Karen von Berg, Physical Therapist at Johns Hopkins CF Center, treated us to an informative and lively presentation highlighting the role of physical therapy (PT) in cystic fibrosis care. Her message about the importance of posture resounded with many in the audience. Throughout the conference, whenever her name or presentation was mentioned, many of us were provoked to sit up straight.

As a physical therapist, Dr. von Berg began caring for people with CF in the hospital providing support for airway clearance. Later she brought PT to patients in the CF Clinic, developing the outpatient physical therapy program at Johns Hopkins CF Center. Following her presentation, many of us wished we had PT in our clinic, too.

Physical therapy contributes to many aspects in the spectrum of cystic fibrosis care. A physical therapy assessment can include observation of physical health, including:

- Developmental milestones
- Breathing mechanics
- Posture
- Range of motion
- Strength
- Exercise assessment
- Pain
- Urinary incontinence
- Airway clearance

Working with children and adults with CF, physical therapists address physical body mechanics toward the goal of improving breathing capacity and lung function. Basic to the function of breathing are the core muscles of the back and abdomen, the diaphragm at the top of the core and the pelvic floor muscles at the bottom. When these muscles are working together, breathing is easier and more effective. The diaphragm rises and pelvic floor descends with inhalation; with exhalation the diaphragm lowers and the pelvic floor rises. When these muscles work together in this manner breathing is more effective.

How does physical therapy help with CF?

Exercise

If it seems that everyone is on the bandwagon of exercise for CF, physical therapists are leading the parade. A physical therapist will evaluate a person's capacity for exercise and recommend specific activities to support each individual's needs. When a person is ready and willing to make the first steps to exercise, a physical therapist can offer tools, support and encouragement.

Education

Our muscles, bones and organs are all connected and need support to work well together. Did you know that good posture encourages effective breathing and that stronger core muscles can reduce incontinence? Working with a physical therapist, we can learn about and receive support for stress urinary incontinence, healthy bowel movements, effective nebulizer treatments including huff coughing, and safe, helpful exercise. Dr. von Berg introduced many to the Squatty Potty® which encourages healthy bowel elimination with postural support. Look it up! We participated in a demonstration of breathing with slouching and upright sitting postures. Guess which posture makes breathing easier and encourages effective treatments and airway clearance? What is your typical position when doing nebs?

I still hear Karen von Berg's words, "Posture! Posture! Posture!"

CF care is expanding. The Cystic Fibrosis Foundation has recently provided funding to support physical therapists in CF clinics. Some centers include PT in clinic as well as during hospitalization. If you are interested in working with a physical therapist and don't have one in your clinic, check the American Physical Therapy website: www.apta.org to find a physical therapist in your area. It is important to work with someone who will listen to you and communicate with your CF center.

As a member of the CF team, the physical therapist understands the special considerations of our disease and the variety of presentations of CF and provides recommendations tailored for each individual, guiding us with exercise. Like our other CF providers, physical therapists know us well over time and can help us adjust our activity and therapy when our circumstances change. Health and quality of life for children and adults with CF can improve with physical therapy support.

Interested in hearing more of what Karen von Berg shared with us at the conference? You can view her presentation and others from the conference through the YouTube link in the lower right corner of the home page on the CFRI website, www.cfri.org

Have you thought about trying yoga? The CFF provides a Yoga for CF DVD. Contact resources@cff.org to ask for this resource. ▲

Laura is 64 and has CF. She is a Director of USACFA. Her contact information is on page 2.

Benefactors

BRONZE

Anonymous (In memory of Steven Riley and Douglas Riley)
Anonymous (In memory of Scott Cherry)
Anonymous (In memory of Scott Cherry)
Janet Bennett (In memory of Scott Cherry)
Jim Duke
Tina Frisbee
Terri Lyn Glickman
Heather Hamilton (In memory of Scott Cherry)
Barbara Harison
Nancy Heitter (In memory of Scott Cherry)
Tracy Johnson (In memory of Scott Cherry)
Sanjit Korde – Korde & Associates, P.C. (In memory of Scott Cherry)
Monica Leavitt
Robert McLaughlin (In honor of Kerri Hiller's 40th birthday!)
Joel and Jen Mehr (In memory of Scott Cherry)
Eric Messier
Benjamin Richards (In memory of Scott Cherry)
Kathy & Paul Russell (In memory of Helen Eisenman)
Zina Scarpulla (In honor of Andrea Eisenman for being a great friend and role model)
Dan Seilheimer
Davina Silvanio (In memory of Scott Cherry)
Barbara Stilke (In memory of Scott Cherry)
Laura Tillman (In memory of Helen Eisenman)

Silver

Richard Leurig (In memory of Scott Cherry)
Penny Daigrepont - Shapiro and Daigrepont, LLC (In memory of Scott Cherry)

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course of the disease, poor nutrition complicates medical care and increases the risks of poor outcomes for patients having a lung transplant. Proper nutrition can improve patients' functioning, ability to exercise and quality of life. But maintaining nutrition is difficult for CF patients, who are unable to absorb fats and other nutrients because of pancreatic enzyme insufficiency. Liver disease and diabetes can also complicate CF. These factors mean patients commonly expend an abnormally high amount of energy. Stress and failing to stick with treatment may worsen malnutrition in CF. All of these elements combined make it necessary for doctors to closely monitor CF patients' nutrient intake and energy. Many patients should be taking supplements formulated for their condition that contain the fat-soluble vitamins A, D, E, and K and nutrients such as calcium, sodium, iron, and zinc. Doctors should also ensure that pancreatic enzyme replacement therapy is adequate, by monitoring symptoms of malabsorption. Since many patients are used to the symptoms, they may not react when they worsen. This means doctors should encourage patients to talk about their toilet habits and stool to detect subtle changes. Additionally, doctors need to keep an eye on patients' levels of salt, calcium, iron zinc and selenium.
http://tinyurl.com/yac234d3

PATHOGENS

Incident Stenotrophomonas maltophilia infection and lung function decline in cystic fibrosis. Barsky EE, Williams KA, Priebe GP, Sawicki GS. Pediatr Pulmonol. 2017 Aug 16. Epub ahead of print

This study was done to determine whether incident detection of Stenotrophomonas maltophilia (SM) in patients with cystic fibrosis (CF) is associated with accelerated lung function decline and increased hospitalizations and to determine whether this effect is more pronounced in individuals with subsequent chronic infection. Acquisition of SM in CF was associated with an acceleration in lung function decline. Among those with chronic colonization, acquisition was also associated with increased hospitalization rates.
http://tinyurl.com/y8f6enhs

16S rRNA gene sequencing reveals site-specific signatures of the upper and lower airways of cystic fibrosis patients. Sarah K. Lucas, Robert Yang, Jordan M. Dunitz, Holly C. Boyer, Ryan C. Hunter. Journal of Cystic Fibrosis. Article in Press

Metastasis of upper airway microbiota may have significant implications in the development of chronic lung disease. Bacterial communities of matched sinus and lung mucus samples from cystic fibrosis (CF) subjects undergoing endoscopic surgery for treatment of chronic sinusitis were compared. Mucus from one maxillary sinus and expectorated sputum were collected. 16S rRNA gene sequencing was then performed on sample pairs to compare the structure and function of CF airway microbiota. Findings indicate that while the lung may be seeded by individual sinus pathogens, airway microenviron-

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A Smörgåsbord Of Living

By Mark Schroeder

My name is Mark Schroeder. I was born in Massillon, Ohio, into a family of six, including the three of us boys with CF. All my life, I thought that I would die young. It became a belief and a nagging, dark cloud over my head. I shouldn't have wasted all that time and energy worrying. At 67, I'm too old to die young.

But I'm still breathing away; taking in oxygen and thankful for the opportunity. I like breathing. Feels good. Sometimes I sit with my eyes closed and pay attention to that fact.

I have read some accounts from others with CF who attribute their good health to following the regimen suggested by their doctors, including taking all the prescribed medicine correctly, getting exercise and sleep et cetera. I agree with what they say and have done the same, religiously, for all but a short time of experimentation, without medications, during my early twenties. It didn't work out well, but that fact reinforced my faith in medicine, which put me back on track.

In addition, I always have experimented, mostly in a safe way, with alternative medicines, diet, vitamins and herbals, therapies and health-related activities. I've wasted money and time on some of it, but I don't regret that, in consideration of the fact that, overall, I believe it has paid off for me. If a doctor told me that a particular thing I was considering was definitely harmful, I gave it up. But if he or she was simply without knowledge of a particular approach to health, I continued researching until I made up my mind to try something or leave it alone. For my health, doctors have always been my sounding board, but in the final analysis, since my health is ultimately in my hands, I am the one to decide what to do or not to do.

MARK SCHROEDER

Here are a few things that I've tried and I think have helped me, in one way or another: acupuncture was a definite plus. The first time I did it, I lay on the table wondering if the needles were going to hurt. The doctor was a surgeon from Malaysia and he used acupuncture there for anesthesia. I could feel a little bit of sensation, but it was not really painful. In a few minutes, I relaxed. My breathing became easier than I could remember it ever having been before. My perpetually clogged sinuses cleared up. My breathing was a joy to experience. A few hours after the session, I began to feel my usual self again, with the clogged sinuses and breathing restrictions.

The doctor giving me the acupuncture taught me some Chi Gong which I practiced regularly until a major life change, not of my choosing, interfered with both the acupuncture treatments and the Chi Gong. But the experience of both was wonderful.

Over the years, I have tried acupuncture with four different acupuncturists. The last time was a series of more than 20 sessions with a Chinese woman who teaches acupuncture at a university in China. She spoke no English. I went to the sessions with my Chinese sister-in-law who did the job of translating as well as getting acupuncture for herself.

Like the first time I did acupuncture treatments, they left me feeling very different, almost as if I had no lung problems at all. I felt very calm and clear-thinking, as well. But, as with the first experience of acupuncture, the improvements didn't last. I often wonder what the results would be after having the treatments on a regular basis for a year or so, or even on a continuing basis without let-up. I was genuinely sad to see the doctor go back to China. We became friends in the few months I went to her for treatments. Language was not a big barrier to developing a friendship.

As with the first time, the acupuncture lasted for a while after each session and then the benefits gradually seemed to lessen. However, I think the thing that lasted, for me, is the belief that I have the ability to feel better and maybe without having to return each time to where I was before. This inspired hope and a dedication to searching out alternative treatments to supplement my regular health regimen.

My experience tells me that not all acupuncturists are alike. In between the two whom I have described above, I went to two other acupuncturists and I felt nothing from their treatments.

For the past ten years I have been practicing Reiki on myself. It is as easy to do as falling off a log! Ha! I have been very selfish about it, not sharing it with others much. I did it on my mom in the evenings and usually it put her to sleep. It is relaxing, medita-

> "I shouldn't have wasted all that time and energy worrying. At 67, I'm too old to die young."

tive, can help relieve pain, and maybe even improve health. Reiki is not something you "learn" so much as something that is given to you. It's a gift! You still have to pay for it, though. A Reiki Master gives you something called an attunement; you pay him or her the bucks, and there you go! You can perform Reiki!

After ten years of it, I still don't know any more about Reiki, from a rational perspective, than I did when I had my first treatment, which is what got me interested in it. Reiki is an experience, a very wonderful experience for me, and if I never learn to understand its origins or rationale, that's okay by me.

I have no regrets about my healthcare choices and no regrets in general. However, I think my life would have been smoother-going if I wasn't so hard on myself; if I wasn't judgmental of myself. Judgment leads to shame. To some extent, I have always felt inferior and at least part of this is due to shame about having cystic fibrosis. Is there shame in having a disease? I thought I was less valuable than "normal" people because of my CF. I thought that making stinky poop, four times a day; coughing up yucky phlegm; having skinny limbs, a barrel chest and a bloated belly, made me an outcast, an undesirable.

As I reread the above paragraph, it makes me laugh. I still make stink, spit up crappy stuff and look like a toothpick with a bubble. And I still feel twinges of shame in connection with what I look like. I still feel shame over what I sometimes say or do or over the beliefs I hold concerning other issues that come under the heading of "shameful for Mark," and sometimes I feel shame for feeling shameful. Shame feeds itself.

I've almost never heard people talk openly about their shame or read something of that kind. This must mean that I am one of the few people in the world who has experienced it. Hmmmm...

But I've noticed the shame in others and the destructive nature of having shame and the great lengths that people go to cover it up. Been there. Though there are those who would have us believe that shame has benefit, I believe the only benefit of my shame is to empower those who would use it to shame me into being inferior and subjective to them.

Shame is not what I thought I was going to write about. I don't like writing about it or even thinking about it, let alone telling you about my own. Shame is a taboo subject. If you don't believe it, then try talking about the subject to someone else and see how long it takes them to change the subject or even try to shame you for mentioning it. But avoiding the taboos makes them sacred, in a negative way; makes them powerful and unknowable and, consequently, out of our control.

Well, that's it. That is all I want to write. Ha! This writing started out pretty matter-of-fact and then delved into what is risky territory, for me. I need a rest. I need to see the bright side. Laugh a little bit. I'm going to walk through the muck and mire when that seems to be what I need to do, but as soon as I see some light, I'm going for it, man!! Balance is everything! ▲

Mark is 67 and has CF. He and his wife and daughter live in Salt Lake City, UT. He would like to hear from anyone who feels moved to correspond. You can contact him at: Mark_s83@yahoo.com

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ments harbor distinct bacterial communities that should be considered in selecting antimicrobial therapies.
http://tinyurl.com/y8qh7nn7

TREATMENTS

Clofazimine-Containing Regimen for the Treatment of Mycobacterium abscessus Lung Disease. Yang B1, Jhun BW, Moon SM, Lee H, Park HY, Jeon K, Kim DH1, Kim SY, Shin SJ, Daley CL4, Koh WJ. Antimicrob Agents Chemother. 2017 May 24;61(6)

Patients with lung disease caused by Mycobacterium abscessus (M. abscessus) typically have poor treatment outcomes. Although clofazimine (CFZ) has been increasingly used in the treatment of M. abscessus lung disease in clinical practice, there are no reported data on its effectiveness for this disease. This study sought to evaluate the clinical efficacy of a CFZ-containing regimen for the treatment of M. abscessus lung disease. Overall, there was an 81% treatment response rate based on symptoms and a 31% response rate based on radiographic findings. This study suggests that CFZ-containing regimens may improve treatment outcomes in patients with M. abscessus lung disease.
http://tinyurl.com/yb6k7csb

Outcomes associated with antibiotic regimens for treatment of Mycobacterium abscessus in cystic fibrosis patients. Alison DaCosta, Cameron L. Jordan, Olivia Giddings, Feng-Chang Lin, Peter Gilligan, Charles R. Esther Jr. Journal of Cystic Fibrosis. July 2017 Volume 16, Issue 4, Pages 483–487

Mycobacterium abscessus infection is associated with declining lung function in cystic fibrosis (CF), but there is little evidence on clinical efficacy to guide treatment. The authors

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Embracing Life With Cystic Fibrosis

By Colleen Veitengruber

When I was born in 1988, I was expected to live about 20-25 years. Today, I am 29 years old, have been married for seven years, have a three-year-old daughter and a son on the way. I graduated college with a bachelor's degree in elementary education in 2010 and have been teaching for six years, currently teaching third grade. If raising a family, working full time and being pregnant aren't enough "normal," I just added getting my master's degree on top of it all, something I never thought I would do.

Ever since I can remember, I have never let having a diagnosis of cystic fibrosis control the path of my life. I have always wanted to go to college, get married and have a family, and I've always pictured myself working while raising that family.

I am very lucky that my health has allowed me to follow my dreams, but it hasn't always been easy. While in high school, I had my lowest lung function because I wasn't educated about my disease and did not take my treatments seriously. When I went away to college, just over an hour away from home, I wanted to prove to my parents and myself that I could thrive and be "normal." It was at this time when two important things happened in my life. I met my now-husband, Tim, and I also switched to the adult CF clinic in my hospital. With the motivation of wanting a long relationship with Tim, and a big reality check from my new doctor, I learned that I had to start taking care of myself.

After Tim and I were together for a while, we started talking about our future together. I began to realize that it was up to me, and only me, to keep myself as healthy as possible to be around long enough for

COLLEEN VEITENGRUBER

him and our future. Here I was in college, pursuing a career and talking about starting a family with someone – things that people with CF rarely did, but I was determined to do it! The only way that was going to happen was if I stayed on top of my health.

Thankfully, my lung function remained pretty stable throughout college and, after graduating and getting married (only a few months apart), I entered the education field. For two years, I worked as a special education paraprofessional. Then, finally, in 2012, I got my first job offer to teach kindergarten! Tim and I had to move three hours south, but I couldn't wait to start my first year as a classroom teacher. My CF doctor wasn't thrilled with the age of my students (so many germs!), but I was so excited to get my foot in the door and start teaching.

My first year, I ended up losing a lot of weight. The stress of the job had me losing calories faster than I could consume and keep them on. Looking back, I was very skinny and underweight; I didn't realize then how low my weight really was. Thankfully, the next year, in August of 2013, I was able to begin Kalydeco, which has helped me tremendously by allowing my body to gain over 20 pounds and keep it on.

Also, during my first year teaching, Tim and I started talking about adding to our family. We quickly learned that we were going to have to get help through fertility treatments. I wanted to make sure that I was in the absolute best health I could be to carry a child, so I tried exercising a bit more and being absolutely compliant with all of my CF treatments. In December 2013, we found out that we were finally pregnant! Pregnancy agreed with me. The baby sat low most of the time and didn't squish my lungs. I was considered high-risk due to my CF, but I had no complications and felt great throughout my pregnancy. In August 2014, our daughter, Anna, entered the world on one of the most memorable days of our lives.

I was 25 years old, in my third year of teaching, learning how to breastfeed a newborn, pumping at

work to supply milk for her, trying to keep up my calorie intake as well as staying on top of my treatments to keep myself healthy. It was a lot to juggle, but it was everything I had ever wanted in life. I remember just being so happy about finally being at this stage in my life. I was doing exactly what I had dreamed of for so many years – it felt amazing. I was a wife. I was a teacher. I was a mommy!

Don't get me wrong, life was not easy, but I loved it. The first few months of Anna's life, I struggled to find time for my treatments because I just wanted to love on my baby that I had dreamed of for so long. But then that voice in the back of my mind would remind me that I have to keep myself healthy to live and be her mommy for as long as I possibly can. She and Tim were, and still are, my motivation to live. We chose to bring this human into the world and it is my responsibility to be around as long as my sick body will let me. I will battle my CF and fight to continue to be a wife and mother.

Over the next year or so, the decision of furthering my education or enlarging our family kept creeping into my head. I had been teaching for more than four years now and it was about this time that many of my peers were getting their master's degrees, but I had no desire. I wanted to grow my family and enjoy what I had. Plus, who wants more student loans? I decided to put furthering my education on the back burner, something I didn't think I would ever do. In all honesty, this was because I didn't think I would live long enough to reap the benefits of it.

In the education field, when teachers go back to school to get their master's and beyond, they get a pay increase. It's a nice little increase with each extra degree, but the benefits really pay off when you've been teaching for a longer period of time. As much as I say I don't let CF run my life, and I really try not to let it, it's a part of my life that I can't really ignore either. I am most likely not going to live to be 70-80 years old like others my age. Getting a master's degree, with more years of student loans to pay off, wasn't worth it to me if I wasn't going to be healthy enough to teach for 30-plus years to get the salary benefits. I had never really voiced this part of my thinking before and just told people I'd rather focus on family at this point – which was true.

> "If raising a family, working full time and being pregnant aren't enough "normal," I just added getting my master's degree on top of it all, something I never thought I would do."

Tim and I decided to continue to work on growing our family. In November 2016, we found out that I was pregnant with twins! It was exciting and terrifying all at once. Unfortunately, in February of 2017, about halfway through the pregnancy, we lost both of our boys. We were deeply saddened by this tragic event, but knew that we would continue to try to grow our family as soon as we were physically and emotionally ready.

After this happened, an excellent opportunity at work presented itself that would help me further my education, work financially with our family's needs, and allow me to still work and focus on my family. It was an opportunity that I couldn't turn down. I am participating in a two-year federally funded grant to learn about incorporating math and science with engineering into my classroom and instruction. Through this program, I am able to receive graduate college credits toward a master's degree. I never, ever thought I would go back to college to get my master's, but through this grant program, and the help of the Lauren Melissa Kelly Scholarship from the United States Adult CF Association (USACFA), I am able to pursue a Master's in Teaching and Learning, while teaching third grade, raising my three-year-old daughter, and being pregnant again, with "Little Brother."

I've almost completed my first semester of graduate school and I've lived to tell about it! Life has never been so busy, but I am so proud of myself for pushing through all of the extra work this semester. It's tough now, but I know that it will all be worth it when it's done. I am excited to be doing something that I never thought I would do – just another notch in the belt against CF. I cannot let the words "life expectancy" influence my life decisions. I need to live in the here and now, and I love what that looks like for me and my future. ▲

Colleen is 29 and has CF. She lives in Decatur, IL, with her husband, Tim, their daughter, Anna, and is currently pregnant with "Little Brother." She is a third grade teacher, but in her spare time, she likes to spend time with her friends and family, read, volunteer with Great Strides and relax when she can. She also likes to blog about her life with CF at: http://howilivelaughlovewithcf.blogspot.com/

Aging With Cystic Fibrosis

By Barbara M. Harison

Here I was thinking I was invincible! I am fortunate to receive and benefit from Kalydeco/ivacaftor and had not been hospitalized for two and a half years. Yet in August I went into the hospital with a pulmonary infection, triggered by a flu virus. I was treated with IV antibiotics in the hospital and then at home. Oxygen was also delivered to the house. This was new for me. I have recovered and am still working on getting my stamina back. Sure glad I made the trip to Alaska's Kenai Peninsula and Kodiak Island in June. I am happy to be back to my active lifestyle swimming laps, walking and golfing. All I could do with the PICC line in was take easy walks. Although this illness reminded me that I am not invincible, I believe I am healthier now at 71 years old than I was in my 50s.

After years of lung problems, I was finally diagnosed with CF in 2010 at 64 years of age. I lost my younger sister to CF 46 years ago when she was just 21 years old. I, too, struggled for many years with bronchitis and pneumonia and as I aged it got worse. In the 1990s I read about new treatments available for those living with CF and thought I might benefit from them. Unfortunately, the many pulmonologists I visited seemed to ignore me. They did not get it — even when I told them about my sister and her CF. They always asked me if I smoked. I never smoked in my life! They diagnosed me with bronchiectasis but said I was too old to have CF. Life went on and I assumed I was a carrier with symptoms of CF.

When I finally got a referral to Dr. Richard Belkin (Santa Barbara Pulmonary Associates), who specialized in adults with CF, I had all the genetic testing and other diagnostics done. In my case getting a diagnosis was a positive occurrence. I am fortunate now to have good care and all the new treatments available.

BARBARA M. HARISON

I truly believe swimming laps for over 40 years kept my lungs working. I knew swimming was a good cardio workout and sometimes made me cough. I still think it beats the Vest any day of the week! I am now retired after a busy and successful career in parks and recreation management and consulting. I recently got back into yoga and enjoy the mindfulness as much as the breathing.

My husband and I have lived in coastal Ventura, CA, for 30 years. With easy access to the beach, I take regular beach walks and connect with Salacia, the female divinity of the sea, worshipped as the goddess of salt water who presided over the depths of the ocean. Salacia was the wife and queen of Neptune, god of the sea and water. Regular beach walks breathing in the salty ocean air with vistas of the Channel Islands is hard to beat. It is therapeutic for life and aging. We often visit other CA beaches: Santa Barbara, Morro Bay and Monterey and walk the beaches gathering shells, sea glass and enjoying sunsets.

Recently I was introduced to a poem titled "I Want to Age Like Sea Glass" from "Slow Family Living" by Bernadette Noll. This is a great metaphor for how to live life at any age. It inspired me as I recovered from the recent lung infection and hospitalization. As soon as I could, I went to the beach to reconnect with Salacia and ocean air. I hope it inspires others living with CF when "you are caught between a rock and a hard place." Selections from the poem are shared here.

I want to age like sea glass so that
when people see the old person
I'll become, they'll embrace all
that I am.
They'll marvel at my exquisite
nature, hold me gently in their
hands and be awed by my well-
earned patina.
Neither flashy nor dull, just the
right luster.
And they'll wonder, if just for a
second, what it is exactly I am
made of and how I got to be in
this very here and now.
And we'll both feel lucky to realize,
once again, that we have landed
in that perfectly right place at that
profoundly right time.
I want to age like sea glass. I want to
enjoy the journey and let my
preciousness be, not in spite of the
impacts of life, but because of
them.

It is not always easy but I am enjoying the journey and I remember to breathe in positive – exhale negative. Namaste. ▲

Barbara is 71 and has CF. She and her husband, Rich, live in Ventura, CA. She is retired after a long career in public parks and recreation administration. When she is not swimming laps or golfing, she volunteers for community organizations and served on CFLF Board of Directors for six years.

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found that treatment of M. abscessus resulted in short term improvement in lung function that is inversely related to pre-treatment FEV1.
http://tinyurl.com/y9df7ooj

Recovery of lung function following a pulmonary exacerbation in patients with cystic fibrosis and the G551D-CFTR mutation treated with ivacaftor. Patrick A. Flume, Claire E. Wainwright, D. Elizabeth Tullis, Sally Rodriguez, Minoo Niknian, Mark Higgins, Jane C. Davies, Jeffrey S. Wagener. Journal of Cystic Fibrosis. Article in Press

Pulmonary exacerbations (PEx) are associated with acute loss of lung function that is often not recovered after treatment. Lung function recovery following PEx for ivacaftor- and placebo-treated subjects was studied. The investigators determined that ivacaftor treatment reduces the frequency of PEx but does not improve on the rate of complete lung function recovery after PEx when compared with placebo.
http://tinyurl.com/y8dhqbxk

Retrospective observational study of French patients with cystic fibrosis and a Gly551Asp-CFTR mutation after 1 and 2 years of treatment with ivacaftor in a real-world setting. Dominique Hubert, Clémence Dehillotte, Anne Munck, Valérie David, Jinmi Baek, Laurent Mely, Stéphane Dominique, Sophie Ramel, Isabelle Danner Boucher, Sylvaine Lefeuvre, Quitterie Reynaud, Virginie Colomb-Jung, Prissile Bakouboula, Lydie Lemonnier. Journal of Cystic Fibrosis. Article in Press

Ivacaftor has been shown to improve lung function and body weight in patients with CF and a gating mutation. Real-world evaluation is warranted to examine its safety and effectiveness over the long term. It was determined that the clinical benefits of ivacaftor reported in previous clinical trials were confirmed in a real-world setting two years post-initiation, also reducing treatment burden.
http://tinyurl.com/ybmvt3fg

Exploring probiotic use in a regional cystic fibrosis consortium. K.D. Gonzalez, J.B. Zuckerman, E.H. Sears, B.S. Prato, M. Guill, W. Craig, C. Milliard, E. Parker, T. Lever, M.M. Griffin, L.W. Leclair. Journal of Cystic Fibrosis. Article in Press

Studies in cystic fibrosis patients have reported reduced rates of pulmonary exacerbation and hospitalization with probiotic use. Additionally, regular intake of probiotics has been associated with improvements in gastrointestinal (GI) symptoms, levels of inflammatory markers, and quality of life. However, little has been published about clinical use of probiotics in CF, and a recent review highlighted the need to better understand the appropriate role of these agents in long-term disease management.
http://tinyurl.com/y9sonqyx

TRANSPLANT

Mechanical Ventilation and Extracorporeal Membrane Oxygenation as a Bridging Strategy to Lung Transplantation: Significant Gains in Survival. Hayanga AJ, Du AL, Joubert K,

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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

Cara Brahm
Lebanon, KY
41 on September 10, 2017

Donovan Couture
Milton, VT
49 on December 23, 2016

Kerri Hiller
Elmhurst, IL
40 on September 15, 2017

Transplant

Cara Brahm, 40
Lebanon, KY
Bilateral lungs
3 years on September 8, 2017

Andrea Eisenman, 52
New York, NY
Bilateral lungs
17 years on April 25, 2017

Stephanie Rath, 48
Brownsburg, IN
Bilateral lungs
4 years on August 31, 2017

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Marie T, Baird R, Pilewski J, Morrell M, D'Cunha J, Shigemura N. Am J Transplant. 2017 Jul 11. Epub ahead of print

Mechanical ventilation (MV) and extracorporeal membrane oxygenation (ECMO) are increasingly used to bridge patients to lung transplantation. The impact of using MV, with or without ECMO, prior to lung transplantation on survival after transplantation was investigated. There was no difference in overall survival between the MV and MV+ECMO groups. Recipients in the MV+ECMO group were more likely to require ECMO after lung transplantation. There were no differences in duration of postoperative MV, hospital stay, graft survival, or the incidence of acute rejection, renal failure, bleeding requiring re-operation, or airway complications. Thus, the combination of MV and ECMO was a viable bridging strategy to lung transplantation that led to acceptable patient outcomes.
http://tinyurl.com/y8xvrapa

Clinical outcome of cystic fibrosis patients colonized by Scedosporium species following lung transplantation: A single-center 15-year experience. Parize P, Boussaud V, Poinsignon V, Sitterlé E, Botterel F, Lefeuvre S, Guillemain R, Dannaoui E, Billaud EM. Transpl Infect Dis. 2017 Jun 15. Epub ahead of print

Scedosporium fungi are emerging pathogens responsible for severe infections in lung transplant recipients. These infections are associated with poor prognosis and some centers consider Scedosporium species colonization as a contraindication to lung transplantation (LT) even though no published evidence demonstrates that Scedosporium species colonization is associated with higher morbidity or mortality after LT. The authors felt that scedosporial colonization may not be a contraindication for LT in CF patients, as long as S. apiospermum complex is involved and a life-long azole prophylaxis prescribed.
http://tinyurl.com/y8sttxhg

Tracheal diverticula in advanced cystic fibrosis: Prevalence, features, and outcomes after lung transplantation. Siddhartha G. Kapnadak, Gregory A. Kicska, Kathleen J. Ramos, Desiree A. Marshall, Tamara Y. Carroll, Sudhakar N. Pipavath, Michael S. Mulligan, Christopher H. Goss, Moira L. Aitken. Journal of Cystic Fibrosis. Article in Press

Tracheal diverticula (TD) are rare anomalies that may harbor infected secretions, posing potential risk to patients with lung disease. In an end-stage cystic fibrosis (CF) cohort, the characteristics and associated post-lung transplant (LTx) outcomes of TD are described. The researchers found a high prevalence of TD in end-stage CF, where diverticula may represent a large-airway bacterial reservoir. TD were not associated with differences in post-LTx outcomes.
http://tinyurl.com/ya8oclu6

CFRD

The use of fructosamine in cystic fibrosis-related diabetes (CFRD) screening. Grace Y. Lam, Michelle Doll-Shankaruk, Jan Dayton, Karina Rodriguez-Capote, Trefor N. Higgins, Dylan Thomas, Kimberley Mulchey, Maeve P. Smith, Neil E. Brown, Winnie M. Leung, Mathew P. Estey. Journal of Cystic Fibrosis. Article in Press

The authors sought to determine whether serum fructosamine correlates with glycemic control and clinical outcomes in patients being screened for cystic fibrosis-related diabetes (CFRD). They deduced that FSF correlated with both OGTT results and FEV1, and reliably identified patients with abnormal oral glucose tolerance test results. This simple blood test shows potential as an effective tool in CFRD screening.
http://tinyurl.com/y7akhwpg

EXERCISE

A single bout of maximal exercise improves lung function in patients with cystic fibrosis. Matthew A. Tucker,

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Reva Crandall, Nichole Seigler, Paula Rodriguez-Miguelez, Kathleen T. McKie, Caralee Forseen, Jeffrey Thomas, Ryan A. Harris. Journal of CF. Article in Press

Responses to a single bout of exercise may provide critical information for maximizing improvements in pulmonary function following exercise training in cystic fibrosis (CF). The authors attempted to determine if acute maximal exercise improves pulmonary function in patients with CF. They found that a single bout of maximal exercise acutely improves pulmonary function in patients with CF and improvements may be related to peak work and peak pulmonary ventilation.
http://tinyurl.com/y9ubyvve

ETC.

Technological advances shed light on left ventricular cardiac disturbances in cystic fibrosis. Zahra N. Sayyid, Zachary M. Sellers. Journal of Cystic Fibrosis. July 2017Volume 16, Issue 4, Pages 454–464

Cystic fibrosis (CF) causes chronic pulmonary disease and can lead to cor pulmonale (abnormal enlargement of the right side of the heart) with right ventricular dysfunction. The presence of the cystic fibrosis transmembrane

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conductance regulator (CFTR) in cardiac myocardia has prompted debate regarding possible defective ion channel-induced cardiomyopathy. Clinical heart disease in CF is considered rare and is restricted to case reports. It has been unclear if this is due to the lack of physiological importance of CFTR in the heart, the relatively short lifespan of those with CF, or a technical inability to detect subclinical disease. Extensive echocardiographic investigations have yielded contradictory results, leading to the dogma that left ventricular defects in CF occur secondary to lung disease. In this review, the authors consider why studies examining heart function in CF have not provided clarity on this topic. They then focused on data from new echocardiographic and magnetic resonance imaging technology, which are providing greater insight into cardiac function in CF and demonstrating that, in addition to secondary effects from pulmonary disease, there may be an intrinsic primary defect in the CF heart. With advancing lifespans and activity levels, understanding the risk of cardiac disease is vital to minimizing morbidity in adults with CF.
http://tinyurl.com/ybkcas4q

Screening for ADHD in adults with cystic fibrosis: Prevalence, health-related quality of life and adherence. Anna M. Georgiopoulos, Deborah Friedman, Elizabeth A. Porter, Amy Krasner, Sheetal P. Kakarala, Breanna K. Glaeser, Siena C. Napoleon, Janet Wozniak. Journal of Cystic Fibrosis. Article in Press

International guidelines recommend depression and anxiety screening in individuals with cystic fibrosis (CF), but Attention-Deficit Hyperactivity Disorder (ADHD) remains understudied. Adults with CF were screened using the Adult ADHD Self-Report Scale-v1.1 Symptom Checklist (ASRS-v1.1), Cystic Fibrosis Questionnaire-Revised (CFQ-R) and a self-report measure of treatment adherence. The authors found that the highly specific screening tool ASRS-v1.1 can ascertain previously undetected ADHD symptoms in adults with CF. ADHD was substantially more prevalent than expected in this population. Elevated ASRS-v1.1 screens correlated with poorer Health-Related Quality of Life (HRQoL) in some domains, but not with BMI, FEV1% pred or self-reported CF treatment adherence.
http://tinyurl.com/yb66ycbx ▲

Laura Tillman is 68 and has CF. She is a former Director and President of USACFA. She and her husband, Lew, live in Northville, MI.