CF Roundtable
A NEWSLETTER FOR ADULTS WHO HAVE CYSTIC FIBROSIS SUMMER 2018

2018 North American Cystic Fibrosis Conference To Be Live Streamed

And/Or Archived For All To View (with some exceptions – see below)

By Amy Sylvis

For the first time EVER, the Cystic Fibrosis Foundation (CFF) has agreed to live stream and/or archive all sessions of the North American Cystic Fibrosis Conference (NACFC) this October 2018! The only restriction is that presenters must agree to participate – which hopefully most will.

Special thanks to Drucy Borowitz, M.D., and John Mercer for their championing of this incredible breakthrough! The CFF should be applauded for listening to the community and providing those with CF access to this treasure trove of information.

What: The North American Cystic Fibrosis Conference is put on annually by the Cystic Fibrosis Foundation. It is a 3-day conference jam-packed with the latest advances in cystic fibrosis care.

Who: Sadly, people with CF aren't allowed to attend NACFC due to CFF Infection Control Guidelines. Even if the guidelines didn't exist, some with CF may not have the health, the funds or the time off from work or school to attend.

Why: The best advocate for people with CF are the people themselves. Live streaming and archiving of all NACFC presentations (as long as presenters agree) empowers people with CF to be informed and proactive about optimizing their health in a collaborative effort with their care team.

The CF Foundation's efforts at co-production (shared decision-making between patients and providers, patient-centered care etc.) is a fantastic initiative that can increase personalized care. What better way for a person with CF to engage in co-production with care teams than by having access to the latest information on his or her own disease from NACFC?

People with cystic fibrosis don't fit neatly into a clinical box – in fact, the disease varies so widely in presentation,

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EDITOR'S NOTES

Summer is here. Hooray! I am ready for some warm weather and blue skies. I hope that your weather is pleasant, too.

We welcome **Beth Sufian** as the new Vice President. **Meranda Honaker**, **Lauren Jones Hunsaker** and **Brennan Reeves** have left the Board of Directors. We thank them for their service and wish them well.

I hope you have read the front page. **Amy Sylvis,** with the assistance of **John Mercer** and **Drucy Borowitz, M.D.,** have made a major accomplishment. Because of that we will be able to access presentations at the North American CF Conference. We thank the CFF for accepting us (adults who have CF) as an important part of CF care.

Our Focus topic is: What Is Most Important To You When A Disaster Strikes? **Jeanie Hanley** talks about earthquakes in California, where she lives. **Nicole Matthews** talks about blizzards in the part of New York near Buffalo. In "Speeding Past 50" I continue the theme with talk of wildfires and being ready to "go."

Julie Desch writes of mindfulness and CF in "Wellness." In "Spirit Medicine" **Isabel Stenzel Byrnes** talks of making comparisons. **Amy Sylvis** uses "Voices From The Roundtable" to discuss a diagnosis of CF-related diabetes and how it impacts our CF health. In "Transplant Talk" **Andrea Eisenman** tells of having another diagnosis added to all that she already has.

"Ask The Attorney" finds **Beth Sufian** discussing COBRA and how income can affect SSI and SSDI benefits. **Brad Johns** tells of a personal experience in "Clinical Trials." Surrogacy is the theme of "Family Matters" by **Molly Pam**.

John Mercer explains about making changes in career and job selections in "Financial Matters." Once again, **Laura Tillman** has done a great job of gathering "Information From The Internet."

Colleen Adamson was a Director of USACFA and served as Treasurer for a long time. We are happy that she is the subject of "In The Spotlight."

If you are looking for a speaker for a fundraiser or education day, please check out the info on page 5 about getting a speaker from USACFA.

As always, we thank everyone who has made a donation to USACFA. Every little bit helps. We appreciate the support. See the president's thank you on page 26.

I had to do a lot of thinking to write my column ("Speeding Past 50") in this issue. Much of it is stuff that I take for granted. To actually sit down and list all of it was a good exercise for me. I hope that each of you will do something similar for your disaster preparedness plans. It is extremely important that each of us, even those who don't have CF, has a good and workable plan for when a disaster strikes.

Stay healthy and happy,
Kathy

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

Information From The Internet...

Compiled by Laura Tillman

PRESS RELEASES

Cystic Fibrosis Treatment Options Detailed in Updated 2018 GeneFo Guide

GeneFo has put together an updated 2018 guide on treatment options for people with **cystic fibrosis (CF)** after recent developments again focused attention on the high price tag of some of these medications and the difficulties patients face in accessing them. The guide includes information on financial and clinical resources to help patients afford CF medications, such as assistance funds, clinical programs and free insurance counseling/tax reduction programs, as well as access programs that provide free treatments in return for patient feedback. GeneFo created the guide to keep patients and caregivers updated on the various opportunities available for help with funding CF medication, treatments and gear. http://tinyurl.com/ybaxs4ff

LAURA TILLMAN

Cystic Fibrosis-Related Pathogen Risk With Chronic Azithromycin Use

Compared with azithromycin nonusers, azithromycin users had a significantly lower risk for detection of new nontuberculous mycobacteria. In addition, there was a trend noted toward higher risks for new multidrug-resistant Pseudomonas aeruginosa (MDR-Pa) in azithromycin users compared with nonusers. The investigators found no differences between azithromycin users and nonusers for risk for acquisition of the other study pathogens: Pseudomonas aeruginosa (Pa), Stenotrophomonas maltophilia, Aspergillus species and Achromobacter xylosoxidans. Azithromycin users also had significantly higher rates of exacerbations compared with nonusers in the Aspergillus matched cohorts. The authors found that azithromycin users had lower risks for new acquisition of three key CF

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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**

Summer (current) 2018: What Is Most Important To You When A Disaster Strikes?

Autumn (November) 2018: Changes We Experience As We Age. (Submissions due September 15, 2018.) What changes have you noticed in your body and health as a result of aging? How do you approach your aging so as to handle it with ease?

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

Spring (May) 2019: Planning For Education And Careers. (Submissions due March 15, 2019.)

ASK THE ATTORNEY

Questions From Our Readers, Answers From Our Attorney

By Beth Sufian, J.D.

Readers of *CF Roundtable* asked questions related to insurance coverage and the ability to work and still receive Social Security benefits. No reference made in this column is to a specific situation. Nothing in this column is meant to be legal advice about a specific situation but is only meant to be information.

The CF Legal Information Hotline (CFLIH) can answer questions related to Social Security benefits, Medicare, Medicaid, health insurance, employment and education. If you have questions please e-mail CFLegal@sufianpassamano.com to schedule a time to speak with the CFLIH.

Question 1: Can a person who stops work due to his health continue on his employer's health insurance plan without opting for COBRA?

Answer: A small number of employers allow a person to stay on the employer's health insurance policy as a covered employee if the employee is receiving short-term disability benefits and/or long-term disability benefits.

There is no federal law that requires a continuation of health insurance benefits with the same employer contribution that was made when the employee was working at the company. For example, if the employer was paying $400 a month for the insurance policy and the employee was paying $200 a month, the employer does not have to continue paying $400 a month if the employee is on short-term or long-term disability. However, if the person is on FMLA leave, the employer does have to continue contributing to insurance premiums if before the employee went on leave the employer was contributing to the health insurance premium cost.

A limited number of employers do continue to pay a share of health insurance costs if an employee is on short-term disability or long-term disability offered by the company. Typically, the continued payment of health insurance premiums will happen only at large companies. But, occasionally, a smaller employer will continue to pay the health insurance premiums. Keep in mind this rarely happens. This means that a person with CF who is considering stopping work due to his or her health should make sure he or she has the funds to pay the full cost of the health insurance premiums under COBRA. It is best to at least take one month of COBRA coverage if a person thinks he or she will be able to secure other insurance coverage. Once the time period to enroll in COBRA expires, the person cannot then ask for COBRA coverage.

BETH SUFIAN

If the employer does not provide a continuation of the employer's help with health insurance premiums, then the person can usually extend coverage under COBRA (the employer who has fewer than 20 employees does not have to offer COBRA). The employee then pays the full cost of coverage. The employee receives 18 months of COBRA if he leaves his job for any reason. If the person becomes eligible for Social Security Disability benefits, then the person will get an additional 11 months of COBRA:

1. The person obtains a positive decision from Social Security within 18 months of stopping work due to disability; and

2. The employee provides a copy of the Social Security Notice of Award of benefits to their employer and insurance company within 60 days of receiving the SSA Notice of Award.

A person who receives Social Security Disability benefits will become eligible for Medicare after 29 full months of being unable to engage in substantial gainful employment, which SSA defines as being unable to work more than 20 hours a week and unable to make more than $1,180 a month from work activity (before taxes are taken out of the check).

Question 2: I sometimes receive small payments when I speak at CF-related events. Usually a CF-related drug company sponsors the speech and pays me an honorarium and pays for my expenses such as airline tickets, hotel and food costs. When the company pays for expenses I incur, is that money counted toward the work earnings limit set by Social

Security?

Answer: It depends.

Increasingly companies that sell CF-related medication or supplies are paying people with CF to speak at a variety of events. The drug company then pays the expenses the person incurs to travel to the event such as airline tickets, hotel costs, rental car and food costs. If reimbursement of these costs is characterized as payment for services rendered, then the money may be reported to the IRS as work earnings income.

If a person receives SSI or SSDI benefits, then the person should be concerned about expense reimbursement being reported to the IRS as work earnings. If the person is also paid an honorarium, then payment of expenses as work income could put the person over the allowable work income limit set by Social Security.

The person with CF should ask the company sponsoring the speech and providing payment to the person with CF how an expense reimbursement will be reported to the IRS. If it is possible for the drug company to pay for expenses directly, that will avoid the problem of SSA counting expense reimbursement as work earnings.

Question 3: How much can I make each month from work if I receive Social Security Disability Insurance benefits?

Answer: A person who receives Social Security Disability Insurance (SSDI) benefits has a monthly allowable amount for work earnings of $1,180 (before taxes are taken out of the check).

Social Security may allow a person a *total* of nine months in which the person can go over the $1,180 amount. Many people with CF think that SSA *must* give them nine months of trial work months. However, in the past year, Social Security offices around the country are saying there is not a mandate that a person be given a nine-month trial work period. Instead SSA says that their rules allow them to find a person is capable of substantial gainful activity (making more than $1,180 and working more than 20 hours a week) if the SSA representative determines there is evidence the person can engage in substantial gainful activity. Even if the person has not worked for nine months over the allowable amount, some SSA representatives are terminating benefits because the representative finds that the person is capable of working over the allowable monthly amount.

This interpretation by many SSA representatives of the application of the nine-month trial work is *very* important to people with CF who are working part time and receiving benefits.

A discussion of the trial work period on the SSA website is confusing. The SSA website makes it seem like a person uses up only one trial work month if he works making more than $1,180 in a month. However, every time a person works making more than $860 a month (before taxes are taken out of the work check) one trial work month is used. This confusion often leads to a person with CF going over the allowable amount because he thinks he has not used all of his nine-month trial work period. Once the person has used up his nine-month trial work period, the next month the person makes over the $1,180 the person loses benefits.

Typically, SSDI takes at least 12 months to see that a person has gone over the allowable amount for work earnings and so then the person has an overpayment for any month in which benefits were paid after the month the person went over the allowable work earnings amount.

For example, a person has an SSDI check of $1,000 a month. The person goes over the allowable work earnings amount in March 2018. SSA figures this out in March 2019. SSA issues an overpayment notice in April 2019 and the person owes SSA $12,000 for the 12 months after the person went over the allowable work earnings amount. ▲

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

USACFA Speakers Bureau

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

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

SPIRIT MEDICINE

The Spirit of Comparisons

By Isabel Stenzel Byrnes

Today I went to clinic and sat in the waiting room for a painful five hours. Looking around I saw people who were in wheelchairs, on oxygen, skinny, overweight, young, old and of all backgrounds. We all had lung transplants; we wore all kinds of masks. Some of us were just weeks out of the surgery and people like me were years out. My new nurse didn't expect me to look so athletic at 14 years post-transplant. I felt guilty for being so healthy among so many people struggling.

While I don't go to CF clinic anymore, this clinic made me contemplate living with cystic fibrosis relative to so many other CF patients out there. We all have such diverse experiences with CF. So today I'd like to write about comparisons. How can we look at our own situations compared to others in ways that are spiritually wholesome and healthy?

As someone who grew up with a twin with CF, I spent my life comparing my illness to Ana's. She had worse GI problems, I had worse lung problems. She had more ABPA and I had more hemoptysis. She needed a transplant before me. And she died of cancer and I'm still living. Yet, I consider it a gift from God to be born a twin. Having someone to share my disease with confirms the Chinese proverb, "Joy shared is joy doubled; sorrow shared is sorrow halved." And Ana helped me to gain perspective with comparisons.

Comparing ourselves to others can be dangerous. Neuroscience tells us our minds are wired to put people into categories. This helps us organize the world and people around us. Humans have adapted to be relational; how we see others helps us see ourselves. This habit has created all kinds of social problems around race, class and gender. People with CF are no exception to this tendency to put people into boxes and come up with explanations for those boxes. So, if we see a CF person on Facebook critically ill, waiting for a transplant, we might think, "Well, at least I'm not that bad off." We might become judgmental and think, "Hmm, maybe he didn't do his treatments. But I'm doing mine so I won't get that sick." This may be a bias we have been taught since a very early age. It feels empowering when we believe we have control to not get that sick. We see others with CF and may label them as "severe" – which helps us reassure ourselves that maybe we don't fit into that box; that we might be "mild" or "moderate."

Sometimes I hear about people with CF who are very healthy. I celebrate my peers who are running marathons and doing Ironmans. Their successes with lung disease and their ability to persevere the torture of running are extraordinary. Or, I meet people with CF who don't need enzymes or have given birth or don't do their treatments and still have good lung capacity. To be honest, my mind sometimes falls into the categorization habit. I think, "They must be mild. They must not have real CF." Those are judgments that I'm not proud to admit. And I also acknowledge someone reading this might be comparing my story to theirs. There are six-year-olds with CF who need a transplant; I was 32 when I got mine. And there are people who die within a year of their transplants, and I'm 14 years post. It isn't fair.

Lately, I heard about the new diagnosis, CFTR-Related Metabolic Syndrome (CRMS). This group of patients has borderline sweat test and genetic test results. They do not have CF, yet their lives are full of symptomatic uncertainty. When recently attending an event with parents of CF and CRMS children, I overheard

> *"The spiritual lesson of seeing people sicker than us is not guilt nor judgment, but rather to feel grateful for our health."*

ISABEL STENZEL BYRNES

comparisons between the parents. This is human.

With my CF friends, sometimes we talk about how many hospitalizations we've had or how much insulin we use or how much hemoptysis we've had. I love the term, "competitive suffering" or "competitive victimhood." It is natural in people who are struggling to view their struggle relative to others. This one-upping serves a purpose. It validates our efforts, becomes a badge of honor and even boosts our self-esteem for enduring as much or more than others. Yet it is arrogant and can diminish others' hardships. To need others' suffering to pale in comparison to ours is toxic.

And, of course, there are healthy people. I don't know anyone with CF who has not compared their lives to those of healthy people. Being athletic, the ease of finding a date, having babies, being able to work full time, earn a living...these are examples of what "they" have that we might never have (or have with great effort). We think healthy people have easy lives... but, newsflash, some still don't.

But where does this comparison come from? Envy and jealousy. Most spiritual traditions consider these two feelings as unhealthy or sinful. "You shall not covet" is the tenth commandment, after all. Jealousy is when we resent that others have what we don't have. And jealousy distracts us from gratitude for what we do have. It becomes a barrier for true love and compassion for others. It takes joy away. Envy, another form of jealousy, has desire in it; a yearning and attachment for something other than what we have. There are many negative consequences to the self when the emotions of envy and jealousy are fed.

Good spirit medicine means putting a raincheck on our comparisons. The spiritual path means re-directing jealousy to loving your neighbor who is better off, to trusting that your time for good things will come and that you will be rewarded for your patience. The spiritual lesson of seeing people sicker than us is not guilt nor judgment, but rather to feel grateful for our health. The spiritual lesson of seeing people healthier than us is to give us hope... maybe someday, post-transplant or after a cure, we'll be like that. And if that's not possible, we learn empathy and celebrate others' delight.

Another spiritual attitude to take on when comparisons arise is to ask the question, "What is this teaching me today?" Comparing ourselves to others can teach us humility. We can't have it all; we take turns to be strong, sick, thrive and struggle. To see someone else with CF running a marathon can inspire us to see what we are capable of. Most of all, the spiritually healthy thing to do is to live your own life. As a child of God and the universe, we are given one chance. I believe that path is perfect the way it is, with all its trials and tribulations. Everyone has their own story with CF. ▲

Isabel is 45 and has CF. She lives in Redwood City, CA, with her husband, Andrew.

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pathogens (nontuberculous mycobacteria, methicillin-resistant Staphylococcus aureus and Burkholderia cepacia complex), a trend toward higher rates of acquisition for one pathogen (MDR-Pa), and no difference for four other pathogens. The authors also noted that patients who regularly received azithromycin were sicker on average compared with nonusers.
http://tinyurl.com/yced9u5w

Gene Panel Predicts Course Of Cystic Fibrosis

Researchers at National Jewish Health have identified 10 immune-related genes whose activity during a respiratory infection predict the long-term prognosis for cystic fibrosis patients better than conventional measures. Five years after being evaluated, patients in the lowest-risk group were all alive and doing well, whereas 90 percent of patients in the highest risk cluster had been admitted to an intensive care unit, put on mechanical ventilation, referred for lung transplant, had a transplant or died. By looking at the underlying biology of the immune response, this panel allows subsets of CF patients to be identified who are likely to do better or worse in the long term. This information can help guide treatment plans in order to deliver the best outcomes for patients. FEV_1 and exacerbation history tell about a patient's disease history and existing lung damage. The gene panel tells more about a patient's biological response to infection, which provides valuable information about the future course of the disease.
http://tinyurl.com/ycqrga4k

CF Severity Linked To Variations In Patients' Airway Bacteria Mix

Researchers have linked variations in the mix of microorganisms in cystic fibrosis patients' airways to their disease outcomes. CF patients typically have particular strains of bacterial and fungus in their airways. The usual bacteria suspects include Pseudomonas, Achromobacter, Burkholderia, Haemophilus, Staphylococcus and Stenotrophomonas. Other bacteria and fungi also inhabit CF patients' airways, including anaerobic species that do not need oxygen to grow and spread. Not only do the microbial communities in

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FAMILY MATTERS

Surrogacy: A Primer

By Molly Pam

I have never had a desire to be pregnant. I'm weird, I know. Sometimes I think there is something biologically wrong with me that I haven't had this urge that seems so natural for everyone else. But just because I have never wanted to be pregnant does not mean I haven't wanted children. And the great news is there are many ways to have a family. As my husband and I are researching our options, I thought I would share some of our research about alternative ways to build a family, starting with surrogacy.

I first became fascinated by surrogacy in high school. A kid in my class was born via a surrogate and was very open about it. Even at that point in my life, I assumed I would never be pregnant, so my best friend offered to be my surrogate. Neither of us knew much of anything about surrogacy, but that set off my desire to learn more about it – I even wrote my senior thesis on it in college!

Why is surrogacy a good option for those of us with CF?

Surrogacy is a great way to have a child that is genetically related to you without having to carry the child. For many women with CF, carrying a child for nine months is risky. With surrogacy, you can do a tune up right before the baby comes to make sure you're in the best possible health. You can also split childcare duties much more easily since breast feeding is a moot point. The best arrangement I've heard from a woman with CF who did surrogacy was that her husband was in charge of all the night feedings from day one so that she could get a full night's rest every night and have the energy to care for their baby during the day.

What is surrogacy?

Surrogacy is an arrangement whereby a woman agrees to carry the child of another person(s) and relinquish parental rights after the birth. Here are some of the key terms used in surrogacy:

- **"Traditional" surrogacy**: Sperm is injected into the surrogate mother to impregnate her, using the surrogate's eggs. This is very rare and usually not legal due to the entanglement of the surrogate being related to the child.
- **Gestational surrogacy:** An embryo is created through in vitro fertilization (IVF) then implanted into the surrogate. The eggs and sperm can be from the intended parents, or from other donors, but NOT the surrogate or her partner. This is the most common type of surrogacy that is legal in most of the U.S.
- **Intended Parent**: This is the person or people who initiated the surrogacy process so that they can have a child.
- **In Vitro Fertilization, IVF:** A process by which eggs are fertilized by sperm in a laboratory setting. The fertilized eggs are then transferred to a woman's uterus for implantation. This can be used for surrogacy or for people who are having trouble getting pregnant.
- **Preimplantation Genetic Screening, PGS:** Genetic screening for embryos prior to implantation, most commonly done to screen for chromosomal abnormalities to reduce the chance of miscarriage. It can also be used to screen embryos for CF if your partner is a carrier. This is optional and can be controversial for some.
- **Birth Order:** The legal way to make sure the intended parents are the legal parents of the baby, usually done through a court order. The process for this varies from state to state and depends on whether surrogacy is legal there.

MOLLY PAM

Is surrogacy legal?

Laws regulating surrogacy vary widely from state to state, and even county to county in some places. The best resource I have found for finding out about surrogacy laws is: https://www.creativefamilyconnections.com/us-surrogacy-law-map/

How do I obtain a birth order?

Some states, like California and Connecticut, grant a legal right to parenthood through pre-birth court orders, and the intended parents are placed on the birth certificate from the get-go. The couples or their lawyer must go to the court and fill out the paperwork ahead of the birth. In other states, like New York or New Jersey, surrogacy is illegal, so the contracts are unenforceable. A couple must rely on the good will of the surrogate to go to court and relinquish her parental rights after which the intended parents must formally adopt the child, which can take many months to years. The best

option if you live in a state that bans surrogacy is to find a surrogate in a state that does recognize surrogacy contracts, let the baby's birth be registered in that state, then return home as the baby's legal parent(s).

Most states' laws are somewhere in between California and New York, and laws continue to evolve. Many states have only legal precedent with no relevant laws on the books. New York is currently exploring legislation to make surrogacy legal, and this spring New Jersey's legislature passed a law legalizing gestational surrogacy. It is now on the governor's desk so by the time this article is published surrogacy could be legal there!

What is a surrogacy contract? Do I need one?

Even when you live in a state that allows surrogacy, you will need legal help. Both you and your surrogate must have your own lawyer to represent your interests in drawing up the contract. The contract will spell out:

- Financial agreements surrounding compensation for medical and other related expenses.
- Agreement about healthcare providers and birth plans.
- Confirmation that the surrogate has at least one child of her own.
- Acknowledgement that all parties understand the process and medical risks it entails. This can be done through separate psychological evaluations for the intended parents and surrogate, and is often required by agencies.
- Agreement for the intended parents and surrogate to obtain the birth orders.
- Relinquishment of parental rights by the gestational surrogate and her partner.
- And, optionally, social media clauses preventing both parties from posting about the pregnancy on social media unless mutually agreed upon, or any other provisions you and your surrogate discuss.

How do I find a surrogate? Should I ask a friend?

There are surrogacy agencies in many states that will help you find a surrogate and guide you through the relevant laws and paperwork. Generally, a gestational surrogate needs to have a child before she can become a surrogate. Surrogates through an agency tend to be more expensive because the agency and usually the surrogate charges fees. Sometimes women who seek to be a surrogate contact fertility clinics that then facilitate the process without the need of an agency.

If a friend or relative offers to be a surrogate, this eliminates the agency middle man, but fertility clinics will still require the surrogate to have her own lawyer and have a completed contract before they will proceed with the embryo transfer. This ensures the surrogate's intent to transfer legal custody of the child to the intended parents, and also lays out the financial arrangement.

What is my doctor's role in the process?

As with everything in CF, you want your doctor to be on board with your decision. Becoming a parent through any means will take a toll on your health, so the best place to start talking about your plans is with your partner, then your doctor. If you undergo egg harvesting, there are a lot of intense hormone regimens that may need to be coordinated with your CF medications. You may want to have your partner tested for the CF gene before you start so that can decide if you want to screen the embryos for CF before implantation. It is not unusual to need many rounds of IVF, which can take a toll on your body – and mind.

Before the baby arrives, you can do a tune up so that you are as healthy and strong as possible. Your doctor and social worker can help counsel you about parenting techniques to maximize your own health once the baby is consuming a lot of your time and energy. Your CF team is there to support you, and the more you tell them the more they are able to help you.

This is all great, but how much does it cost?

A lot. There is no getting around the fact that surrogacy is expensive, but there are ways to help minimize your costs. Typical costs include paying for IVF (potentially many rounds), medical care for your surrogate, sometimes compensation for medical leave for your surrogate, and legal fees.

The first thing to check is what your insurance covers. Many insurance companies cover at least some of the costs of the IVF, embryo creation and storage. You might also find the surrogate's own insurance will cover many of the costs of her pregnancy, with you just footing the bill for co-pays and deductibles. There are also secondary insurance plans you can buy to help cover some of these costs. It's important to ask these questions before you get too far into the process if money is a concern. It is possible that your insurance company will cover more than you thought. At the end of the day, my best advice for this and all financial planning is: plan for the worst and hope for the best. Having a baby is expensive for everyone, but having a baby through surrogacy adds a number of costs if you want to ensure the process is legal. The Internet is full of horror stories, so it's best to be prepared!

I hope I've given you a better understanding of surrogacy and what questions to ask when you are considering it. If you want to talk with someone who has been a surrogate, the CF Foundation Peer Mentorship program can match you with someone who has done it to tell you about their experience. ▲

Molly is 30 and has CF. She lives in NYC with her husband, Adam. She loves to tend her rooftop garden and cook from her bounty. You can follow her on Instagram @ chefmollypam or e-mail her at mpam@usacfa.org.

SPEEDING PAST 50

Am I Ready?

By Kathy Russell

This May has been incredible. We have had such lovely weather that I almost feel guilty for having it so good. (Notice that I said "almost," because I really don't feel guilty. We deserve it as much as anyone else.) The only problem with having such a warm May is that we have had almost no rain. We need rain to keep everything green. After the wildfires that we had last year, we hope that this year will be easier on the firefighters. From my point of view, I'd like to have fewer fires so that I can go out into the great outdoors and still be able to breathe. Last year I spent many weeks staying inside because of smoke from a wildfire that still has some hotspots. Which takes us right to our Focus topic for this issue – What Is Most Important To You When A Disaster Strikes?

None of us likes to think of being caught by any disaster, but many of us will be caught by one disaster or another at some time. Right now many people in Hawaii and Guatemala are dealing with active volcanoes. People in other places are having to put up with fires, earthquakes, landslides, floods, storms of many types and other problems of which I am not aware. Some of these will cause people to have to leave their homes in a hurry. Evacuations are difficult enough for most people, but they can be really hard for people who have a chronic health condition such as CF. It is truly important for us to have a plan in mind.

We need to have a plan for what we absolutely must have with us and how to get it quickly. It is important to keep a list of what medicines we take and what the doses are and when we take them. Actually, we all should have such a list that we keep with us at all times. I know that it makes my doctor visits much easier to be able to hand such a list to the person who is getting all the info at the beginning of a visit.

It is important to have our medicines available to access easily so that we can be ready to go in only a short time. We shouldn't have to spend limited minutes looking for this med or that one. If some of our meds need special handling, such as Pulmozyme, we must make plans for how to keep it at the correct temperature. I always have chemical ice packs frozen in our freezer that we can use to keep Pulmozyme cool in an insulated cooler. By putting the air-filled packing around the meds, I don't have to worry about the meds getting too cold.

Next there are the pieces of equipment that we use. There are several that come to mind. I use a Waterpik for irrigating my sinuses. I also have a compressor to power my inhalations. There are nebulizers to go with that. Many of us also have some kind of vest and its power source. Another somewhat cumbersome piece of equipment is my portable oxygen concentrator (POC) and its extra batteries and chargers.

> *"Evacuations are difficult enough for most people, but they can be really hard for people who have a chronic health condition such as CF."*

KATHY RUSSELL

I feel that it is important to talk about potential disasters and how we will handle them. We need to put into our brains that *when* there is a disaster (notice that I didn't say *if*, it is important to think *when*), I will take...and make lists of what those things are. Be sure you know where each item is kept and how to get to it. Have plans for how you will keep temperature-sensitive meds at their correct temperatures.

I know that I can run my POC and my nebulizer compressors on the power of my car. I have those connectors and cords in my car. In an emergency, I don't have to hunt for them. That should help me get through at least the first few hours or days of any disaster that might strike here. Fortunately, we live high enough that we don't have to be too concerned about flooding. (If we got flooded, it would mean that water had risen 400-500 feet above nor-

mal. Highly unlikely, I think.)

Our largest concern is fire. We live in a wooded area and have many tall trees around our house. Whenever there have been fires in our area, we have paid very close attention to which way the wind was blowing and have been ready to get out if we needed to. So far, we have been lucky. No fire has gotten nearer to us than about two-and-a-half miles. That's far enough away to be manageable and as long as the wind is away from us I am able to breathe okay.

My husband and I always have kept "go bags" ready. A go bag is a small pack such as a DOP kit or a shaving kit. In these we each keep items such as a toothbrush, toothpaste, mouthwash, shampoo, deodorant, powder, bandage strips (like Band-Aids), dental floss, razor, hair gunk, hair brush and comb, small mirror, bathing soap, little sewing kit and so on. These items always are handy to grab and go. We know where they are and that they always are ready for us.

I have a down-filled comforter that I keep in my favorite chair. I use it to keep warm, since I always seem to be a little chilly. I know that I definitely want to have that with me. It is exactly the right weight and size to keep me happy.

Another very important item is my pair of ear plugs. They protect my ears from the loud sounds of my various pieces of equipment. Since I am old, I know that my hearing isn't what it used to be. I want to be sure to protect all that I have left. My eyeglasses fit into that same category. Must protect those old eyes. These items don't take up a lot of room, but they do provide important protection.

Along with all of this medical equipment, we have food and clothing ready to go. We have one of the prepackaged containers of MREs and keep another similar-sized container of utensils and equipment for cooking ready to grab at a moment's notice.

We have bottled water on hand in the house as well as in the car. This is a necessity all of the time. (Our capital city just went through a "don't use the water" advisory, because of an algae bloom in the reservoir. Many people didn't have water on hand and had to rush out to buy some.) It is better to be prepared.

So, let's see, we have lists of meds and equipment, plans on how to gather and use them, go bags with personal items, food and water (if necessary) and clothes. Just making lists and thinking about where everything is and how you will access it can help to get you prepared.

Of course, I hope you never have to use your disaster plan, but I feel that you will be able to handle it much better if you are well prepared. Remember to think "when a disaster happens." I wish you safe harbor.

Stay healthy and happy. ▲

Kathy is 74 and has CF. She is Managing Editor of CF Roundtable. *Her contact information is on page 2.*

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CF patients' airways vary by type of microorganism, but also in the relative abundance of each species. Researchers decided to see if the prevalence and relative abundance of typical CF pathogens and anaerobic microorganisms play a role in the severity of patients' disease and their lung function. The team classified the stage of patients' disease on the basis of their lung function scores. Researchers classified disease aggressiveness — mild, moderate or severe — on the basis of change in FEV_1 relative to age. They discovered a link between variations in the prevalence of the six typical CF pathogens, plus nine anaerobic species and changes in a patient's disease stage and lung function. One finding was an increase in the relative abundance of anaerobic microorganisms when patients were experiencing pulmonary exacerbations before antibiotic treatment and a decrease during treatment and patients' post-treatment recovery phase.

http://tinyurl.com/yankwxuw

Lung Function In Cystic Fibrosis Improved Via Autogenic Drainage, Study Says

Autogenic drainage physiotherapy can effectively improve the underlying lung mechanical impairment of cystic fibrosis (CF), a study shows. Autogenic drainage is a commonly used, self-administered airway clearance technique based on repetitive exercises of controlled inhalation and exhalation to move mucus buildup from the small to the large airways. In the study, researchers evaluated the respiratory changes induced by a single session of autogenic drainage. Participants' lung function was evaluated before and after the treatment session based on the respiratory system resistance and reactance measured using the forced oscillations technique (FOT). This technique consists of applying repetitive oscillatory forced signals to the subject's airways during normal respiration opening and measuring the pressure, flow and volume of the lungs. Results showed that all participants had moderate-to-severe obstructive respiratory disorder, characterized by reduced forced expiratory volume in 1 second (FEV_1) and FEV_1/forced vital capacity (FVC) ratio — all specific measures of lung function.

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FOCUS TOPIC

WHAT IS MOST IMPORTANT TO YOU WHEN A DISASTER STRIKES?

Shake, Rattle and Roll

By Jeanie Hanley

I was blissfully asleep early one Los Angeles Monday morning in January 1994 after a busy weekend, when I was suddenly and literally jolted awake by the bed shaking. I felt as if I was in a boat, over choppy seas. There was rolling and shaking. The house beams sounded stressed and the windows rattled while the earth rumbled. As I jumped out of bed, my husband and I took one look at each other and hightailed it to the rooms of our two young children – our four-year-old son, Kevin, and our eight month-old daughter, Maria. We knew immediately that it was the Southern California "big one" – the earthquake that had been expected for years, later named the Northridge earthquake.

We had already lived through an even bigger "big one" in San Francisco of 1989, when my son was two months old. We had learned during this catastrophic quake to never stand underneath doorways after seeing the destruction around homes and doorways that fell to the ground in the aftermath around our San Francisco neighborhood. This was contrary to what we were taught during earthquake drills. Luckily our apartment building held and we just had to deal with lots of broken glass and broken furniture.

Back to Los Angeles in 1994, we knew what to do and huddled under our wooden tabletop until the earth stopped convulsing. We were very fortunate to have electricity and water afterwards as many homes didn't. What we realized later was we were woefully unprepared had there been any structural damage to our home, or if water and/or power had been lost. What would we have done?

Without my nebulizers, Vest or clean water and power, I may have had to battle L.A. traffic to get any kind of help. Even then, UCLA Medical Center, where I worked, had closed due to structural damage, serving only the most emergent of needs in an emergency facility. The local hospitals were overwhelmed with patients. Where would I get care if all I needed at that point were preventive meds and electricity, e.g., power for nebulization of my daily medications? I knew that I would have to be as self-sufficient as possible should another disaster occur.

Since then, I'd like to say that I am totally prepared. But I'm not certain that's possible. I can say I'm more prepared than ever. And if I can remember to keep my wits about me as the emergency unfolds, all the better. My best plan is my travel list that outlines all the items I need with the most emergent items listed first. Because I never know if I'll have enough time to pack everything, I can only hope that the most critical items at the top of the list can be grabbed in a pinch – such as baggies filled with emergency medicines of antibiotics, steroids, inhalers and nebulized medicines. I would also need to pack my portable nebulizer, refrigerated medicines, supplies, ice packs and cooler etc.

JEANIE HANLEY

Envisioning an emergency drill for practice, I know I'd have to make sure that everyone in the house is safe first and foremost. Then if the house is structurally sound for the moment, I can start gathering critical items from my list, which is stowed in different areas of my house, on my phone and computer for easy access anywhere, anytime. If we lost power and water, we could rely on our stored bottled water, food and solar lights for a while. If we have to leave our house due to structural damage, flood or fire, our backup plan is to use our car's power (and hope that we had a full tank of gas) and drive to any one of our relatives living throughout California. My car has the portable nebulizer already inside, but I'd also have to include as many things on my list as time would allow.

I revise and fine-tune my list almost every time I pack for a trip to keep it current. I usually start packing a day early, a luxury that isn't available in an emergency. The fastest packing I've ever done was when I had to pack quickly for a surprise weekend away last year. I packed in an impressive two hours only to find out halfway into our drive that I had jumped in the car barefoot and hadn't packed any shoes. Still – not bad. Shoes can be bought (and were). This "drill," so to speak, was done under happy excitement not

> My best plan is my travel list that outlines all the items I need with the most emergent items listed first.

duress and panic. So it wasn't surprising to have successfully rounded up the essentials for my health - all medicines (including extra in case we stayed longer), machines (Vest, portable oxygen concentrator, two nebulizers), clothes, toiletries, water and snacks for the trip.

Two hours to pack may or may not be available during the next calamity. While Mother Nature has endowed Southern California with many perks, She has also afflicted it with fires, floods, droughts (intense traffic that's not to be blamed on Her) and the frequent tremors, all of which could create more adversity in trying to flee a disaster-laden area.

As I write this, there is more that I know I can do but may also be impractical or very costly. I am fortunate to have neighbors who have other essentials. One has a generator; another has a large water storage tank. There's also the house at the end of the block that's at the highest elevation and least likely to flood. As a parting thought, it pays to know your neighbors and how you can help each other, especially when you have CF. ▲

Jeanie is 56 and is a physician who has CF. She is a Director of USACFA and is the President. She lives in Los Angeles with her husband, John, and youngest daughter, Jessica. She is also her Neighborhood Watch Captain.

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They also all experienced increased bronchial resistance and decreased reactance. After the physiotherapy session, patients who performed autogenic drainage showed small but significant improvements in FEV_1 and FVC measures compared with CF patients who did not. Similar positive effects were also reported in inspiratory resistance and reactance.
http://tinyurl.com/ycaqn5dq

Drug For Cystic Fibrosis Patients Associated With Higher Hospitalization

Long-term use of a class of drugs that suppress stomach acid is associated with an increased risk of hospitalization for cystic fibrosis patients. The study looked at a class of acid-suppressive drugs called proton pump inhibitors, which are used to treat reflux, heartburn, ulcers and other conditions. The study found patients using PPIs long term were more often hospitalized for complications of the disease than patients not on the drugs. This research underlines the need for physicians to more closely evaluate whether a cystic fibrosis patient can benefit from the acid-suppressant drugs or if other alternatives might be available.
http://tinyurl.com/yb283h9y

Oral Antibiotics Associated With Increased Risk For Kidney Stones

Use of five classes of antibiotics is associated with increased risk for kidney stone disease (nephrolithiasis). After multivariable adjustment, use of sulfas, fluoroquinolones, cephalosporins, nitrofurantoin/methenamine and broad-spectrum penicillins was associated with higher risk for nephrolithiasis, compared with nonuse. The authors speculate that antibiotic-induced alteration of the gut microbiome could change macronutrient metabolism, thus leading to kidney stones.
http://tinyurl.com/ycz8bz88

CF Patients, Especially Post-transplant, At High Risk Of Gastrointestinal Cancers, Study Finds

Patients with cystic fibrosis (CF) are at higher-than-average risk of developing gastrointestinal cancers, especially those who underwent a lung transplant. Transplant patients with

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it's almost amazing that we all have the same diagnosis. Some people with CF are diagnosed soon after birth due to failure to thrive; some are diagnosed at menopause due to symptoms worsening; some are diagnosed in their 20s or 30s due to infertility; and still others aren't diagnosed until their 60s after suffering from symptoms for decades. Some with CF have liver disease; some have sinus issues; some are pancreatic sufficient; some have CF-related diabetes; some culture aspergillus and some don't. CF really impacts each and every patient differently. CF care teams can't possibly attend all sessions at NACFC; so in 2018 - where information can be life-saving power for those with CF - it stands to reason that our community should have access to any and all presentations at NACFC (as long as presenters agree).

When: October 18-20, 2018

Where: Streaming and archiving websites to be announced. We encourage everyone to take full advantage of any and all live-streamed and/or archived content that the CF Foundation provides at NACFC– we must make sure that we show that the demand from people with CF is there for this essential information. ▲

Be Prepared

By Nicole Matthews

> "The most important thing that he [my husband] brings to not only my survival but everyone in our houses, his calmness."

Living in Orchard Park, NY (just outside of Buffalo, NY). I worry about something happening, a massive blizzard, something at Niagara Falls or anything else that could happen. But, I feel like I am prepared.

It also helps that my husband is extremely amazing at being prepared for everything (thank goodness, I have enough on my plate). I have all my medications in one Tupperware bin, easy to access and ready to go in case we need to grab it ASAP. I also have my refrigerated medications all in the same area in the fridge. I feel that we as people with cystic fibrosis do seem a little more prepared than "the norm."

I know at my work I always have food at my desk in case I need it. I have frozen lunches in the fridge in case I forget. I see others who don't have anything and end up asking me for food. Luckily, I am always prepared (I think).

I would personally write a list of everything you may need or even create a bag. I have a bag of clothing in my car in case I ever get stranded in a blizzard at work or on the road. (The bag includes everything from socks, underwear, hoodies, hats, gloves, to you name it, it's probably in there.) I even make sure I have enough food in my car for at least a couple days; meal bars, protein shakes and nuts are great go to items.

But, I have to say my survival in an emergency will fall heavily onto my husband, Michael. He is an extreme preparedness man. We have MREs to last at least a month in our house, and they're gluten free for me!! He has cases of water all over in case we need them. We have flashlights that do not require batteries, but are crank. And the most important thing that he brings to not only my survival but everyone in our houses, his calmness. He is always prepared, he even had a pocket knife in his pocket at our wedding (which helped for random strings on the outfits). I fully trust that he will guide us through any and all disasters that happen.

NICOLE AND MICHAEL KOWAL ON THEIR WEDDING DAY, POCKET KNIFE AND ALL.

When "SnowVember" happened in Buffalo a couple years ago, we were unable to leave our home for more than four days. I had enough medications to last me a month (I get the highest amount of prescription possible). We also stuff our pantry during sales of canned goods and anything we can live on for a bit. We keep a case of water on every floor and building of our house, rotating them out after a year.

We also are very blessed to live in Buffalo, the city of good neighbors. Throughout the snowstorm, we had help from our neighbors and we helped them. We also enjoyed a couple of drinks after snow-blowing. I believe that in a disaster you need to remain calm, you need to have faith not only in yourself but humanity, sometimes horrible things can bring us together. We get dumped on with snow and yet we help one another shovel out, plow and snow-blow.

But, I knew we were fine with food and water because we are prepared. It doesn't take long to become prepared, it just takes a little planning. And, a little mental note: don't panic, just let whatever happens, happen. Things can get crazy, but if you have yourself ready for anything, it will be okay. ▲

Nicole is 30 and has CF. She and her husband, Michael, live in Orchard Park, NY.

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Disaster Ahead: Cystic Fibrosis-Related Diabetes (CFRD)

The Complication That's Impacting Your Lungs More Than You Realize

By Amy Sylvis

Cystic fibrosis-related diabetes (CFRD) is the CF complication that I feared my whole life. I love sugary foods and I hate needles, so CFRD was definitely something I always wanted to avoid no matter what! I felt such a sense of ease every year when I passed my oral glucose tolerance test (OGTT), meaning I didn't have CFRD. To further reassure me, my A1C was around 6.5-7, which according to my CF team, also confirmed I was in the clear. Life was good!

From 2010 to 2012, when I was ages 29-31, I started to feel so exhausted that I could barely get off the couch after work and on weekends. My FEV1 was high, but I expected that as I got older, CF would simply just take its toll and make me more fatigued. Over the years, I found myself sacrificing social time with friends, sacrificing time for hobbies and even sometimes sacrificing time for self-care more and more often – all so I could sleep more. I knew I had to work to pay for my rent, food and medical bills, so I powered through my 40 hour/week job and slept when I could.

With the little spare time I did have in between work and sleeping, I did research online: I read scholarly articles about CF, I chatted with other patients with CF who had been through the various progressions of the disease and, of course, I constantly questioned my CF team for solutions. Thyroid problems? Hormone imbalance? Change in sputum cultures? Elevated white blood cell count? Elevated CRP? IgE? Nope. All was well!

AMY SYLVIS

Enter a new nurse to my CF team who suggested that I try to test my blood sugars on my own with a meter. Not my favorite idea because of my fear of needles and blood, but I was so desperate for answers after two years of such fatigue, I would try anything. What would you know? I was getting blood sugars in the 180s, 190s and low 200s mg/dL two hours after eating.... sometimes. For reference, anything above 120 mg/dL is considered to be elevated blood glucose (hyperglycemia). But the elevated sugars didn't happen all the time – even when I would eat the exact same meal (or dessert).

Off to the endocrinologist where I learned that I had caught CFRD quite "early" – early enough to where my pancreas was working in fits and starts – meaning my precious pancreas would sometimes release enough insulin to cover what I was eating, but other times it simply just didn't. I was put on long-acting insulin (basal insulin) and almost overnight I felt like I gained my life back; it's no exaggeration that even lowering my sugars slightly reduced fatigue dramatically. To say I was ecstatic would be a complete understatement.

I plugged along for the following months, tracked my blood sugars several times a day with a meter, recorded what I ate, counted those fun carbs and sugars that I LOVE and what activity I did during the day and when. It was quite a bit of work, but it was so incredibly worth the effort because my new energy level made me feel like a new woman! Eventually my tracking led me to initiate fast-acting insulin depending on what I ate, and I learned the ins and outs of how my body reacted to exercise, heat, different foods and time of day.

All this is to say that being diagnosed with CFRD at age 31 was quite the adventure – but I have learned over the years my situation wasn't that atypical. Sadly, the standard diagnosis techniques for CFRD may be missing some patients. Here's why that matters:

According to retrospective studies including one by Rolon, MA, et al., "the years preceding therapy for CFRD, reductions in FEV1, forced vital capacity (FVC), and body mass index (BMI) are observed."[1] Meaning that before a person with CF initiates therapy, FEV1 can decline – the best predictor we have of health in a person with CF. In fact, as early as 1988, it was reported that less than 25% of CF patients with diabetes reached the age of 30 compared with about 60% of those with-

[1]Rolon MA, Benali K, Munck A, Navarro J, Clement A, Tubiana-Rufi N, et al. Cystic fibrosis-related diabetes mellitus: clinical impact of prediabetes and effects of insulin therapy. Acta Paediatr (2001) 90(8):860–7. doi:10.1080/08035250152509555

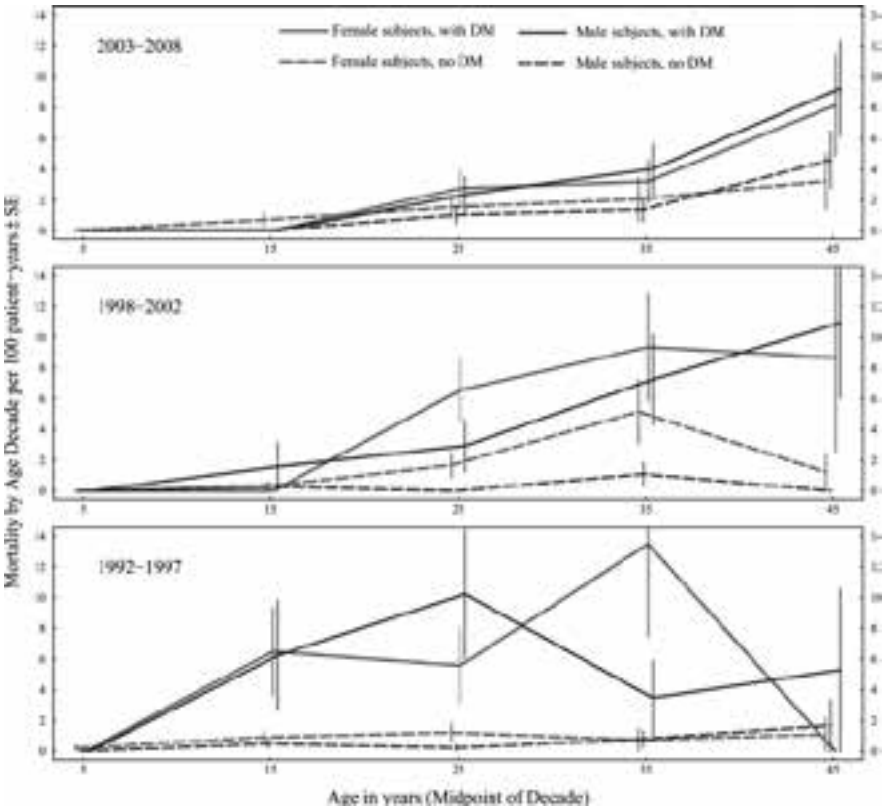

Figure 1 Moran et al. Diabetes Care. 2009 Sep;3(9): 1626-1631

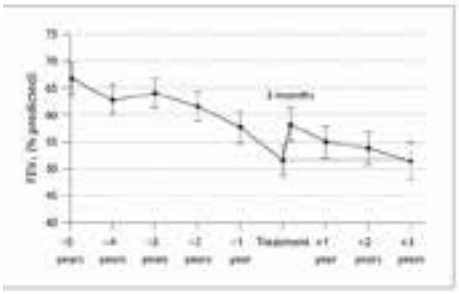

Figure 2 Mohan, Kamlesh et al.

FEV¹ decreases 3 years prior to diagnosis. Mean (SE) changes in FEV¹ 5 years before and 3 years after the onset of insulin

out diabetes.[2] Dr. Moran et al. said that earlier diagnosis and treatment of CFRD decreased mortality over 20 years. Let that sink in – CFRD may really have an impact on lung health – but unlike me, a person with CF may not feel any symptoms of CFRD, particularly in the early stages.

What's more concerning is that current guidelines suggest utilizing diagnostic criteria that may miss the presence of CFRD, let alone pre-diabetes. The Cystic Fibrosis Foundation's CFRD Guidelines from 2010 recommend an annual OGTT for people over the age of 10 who have CF – and a diagnosis of diabetes occurs when blood glucose two hours after initiation of the test is ≥ 200 mg/dL (11.1 mmol/L)[3]. An OGTT involves the patient drinking a high sugar drink - and glucose and insulin levels are measured typically at one and two hours post consumption. According to María Clemente León et al., an OGTT may very well miss hyperglycemic episodes and a Continuous Glucose Monitor (a small device that is worn on the body that measures blood sugar levels every few minutes) more frequently detects these episodes.[4] This is so fascinating for me to read because I definitely fell into the category of an OGTT missing my elevated blood sugar episodes – but that didn't mean they weren't happening. I wish I had known this was possible in the years prior to my diagnosis! Let's summarize: not only can CFRD cause lung function decline, but diagnosis can be missed in the CF population because of tools that are recommended in diagnosis.

The literature seems to be clear, as verbalized by Mainguy, C., et al. in 2016: "The conventional OGTT, however, has been shown to have weak capacity to diagnose [CFRD] in CF individuals as the inherent variability of the test and the variability observed in individual CF patients over time means it does not accurately reflect glucose handling."[5]

What's more, the threshold for diagnosis may need to be updated based on some new research since the 2010 guidelines were published by the CFF. The CFF says a patient has CFRD if his or her two-hour OGTT blood glucose is above 200 mg/dL. In a study on CFRD by Brennan, AL., et al., showed that not only did glucose appear in CF patients' airways when blood glucose reached above 140 mg/dL, but this glucose in the lungs fueled bacteria growth.[6] Perhaps diagnostic guidelines should be reconsidered so that we're not fueling bacterial growing in patients' airways?

Where did 200 mg/dL for CFRD diagnosis even come from? "The cut-offs

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[2]https://www.ncbi.nlm.nih.gov/pmc/articles/PMC2992212/
[3]https://www.cff.org/Care/Clinical-Care-Guidelines/Other-CF-Related-Conditions-Clinical-Care-Guidelines/Cystic-Fibrosis-related-Diabetes-Clinical-Care-Guidelines/
[4]https://www.sciencedirect.com/science/article/pii/S2530016417302380?via%3Dihub#bib0150
[5]Mainguy, C, Bellon, G, Delaup, V, Ginoux, T, Kassai-Koupai, B, Mazur, S et al. Sensitivity and specificity of different methods for cystic fibrosis-related diabetes screening: is the oral glucose tolerance test still the standard? J Pediatr Endocrinol Metab (2017) 30(1):27–35. doi:10.1515/jpem-2016-0184
[6]Brennan, AL, Gyi, KM, Wood, DM, Johnson, J, Holliman, R, Baines, DL et al. Airway glucose concentrations and effect on growth of respiratory pathogens in cystic fibrosis. J Cyst Fibros (2007) 6(2):101–9. doi:10.1016/j.jcf.2006.03.009 https://www.cysticfibrosisjournal.com/article/S1569-1993(06)00093-2/fulltext

SYLVIS continued from page 17

defining CFRD were taken from Pima Native Americans with Type 2 Diabetes, in which they were used to forecast microvascular complications (e.g., retinopathy) and may not be appropriate to use in CF,"[7] states Hameed, S., et al. So really, the diagnostic criteria for CFRD doesn't have much to do with CF patients at all – it was set by the American Diabetes Association to reduce microvascular complications (damage to kidneys, eyes, heart, nerve disease), not lung function decline. I would strongly argue that we need to aim criteria that more closely aligns with what matters in CF – lung issues that lead to early death.

Lastly, those with CF and CFRD tend to have inhibited first phase insulin release[8], therefore, there can be a 30-minute or 60-minute spike in blood glucose after eating. This further demonstrates that a two-hour glucose measure with OGTT may be completely missing hyperglycemic episodes – and therefore missing a diagnosis. In fact, I can recall getting low blood sugar episodes in my teenage years (12+ years before my diagnosis) as a result of blunted first phase insulin release after a carb-heavy meal. I was getting an overly eager second phase insulin response, which caused two- to three-hour post-meal blood sugar lows – turns out this is a common early sign of decreased insulin in a person with CF called reactive hypoglycemia.

"It may be advisable to begin insulin therapy early for people with or developing CFRD, as glucose tolerance declines, especially as CFRD is the result of progressive cell dysfunction," states Kayani Kayani et al. [9]

Given the unquestionable impact hyperglycemia has on cystic fibrosis patients, their lungs and prognosis, it is my hope that the Cystic Fibrosis Foundation will take into consideration the bountiful new research that has been published since 2010. At best, these current CFF 2010 CFRD Guidelines are preventing CFRD cases from being appropriately diagnosed and, at worst, the guidelines may be contributing to worsening lung function by many in our community - something we all simply cannot ignore. More large-scale, multi-center research needs to be funded by the CFF to bring CFRD guidelines to reflect the latest research. Our lungs simply don't have the luxury of waiting any longer. Looking forward, we are eagerly awaiting results of studies to see if using insulin in patients before they have CFRD will increase survival. Stay tuned for those results hopefully soon!

I would like to thank Attain Health and Dr. Colleen Wood for opening my eyes to the latest research in CFRD. Without them, I would not have the slightest inkling about this space. In fact, you can access a webinar on CFRD here: https://youtu.be/U8mpHtUWako ▲

Amy is 36 and has CF. She is a Director of USACFA. Her contact information is on page 2.

[7]Hameed, S, Jaffé, A, Verge, CF. Advances in the detection and management of cystic fibrosis related diabetes. Curr Opin Pediatr (2015) 27:525–33. doi:10.1097/MOP.0000000000000251

[8]Lanng, S, Thorsteinsson, B, Røder, ME, Orskov, C, Holst, JJ, Nerup, J et al. Pancreas and gut hormone responses to oral glucose and intravenous glucagon in cystic fibrosis patients with normal, impaired, and diabetic glucose tolerance. Acta Endocrinol (1993) 128(3):207–14. doi:10.1530/acta.0.1280207

[9]https://www.frontiersin.org/articles/10.3389/fendo.2018.00020/full#B18

TILLMAN continued from page 13

CF were found in this retrospective study to have a five-times greater risk of gastrointestinal cancer than those who have not had a transplant, emphasizing a need for careful screening for small intestine and colon cancers particularly, but also for biliary tract and pancreatic cancers. Compared to the public-at-large, all CF patients had a 20 times higher risk of cancer of the small intestine, and 10 times higher for colon cancer.
http://tinyurl.com/y8k8gbkw

Toothpaste Ingredient May Bust Up Cystic Fibrosis Biofilms

A common antibacterial substance in toothpaste may combat life-threatening diseases such as cystic fibrosis when combined with an FDA-approved drug. Researchers have found that when triclosan, a substance that reduces or prevents bacteria from growing, combines with an antibiotic called tobramycin, it kills the cells that protect Pseudomonas aeruginosa by up to 99.9 percent.
http://tinyurl.com/y7dud4sk

BUT

Common Toothpaste Ingredient May Promote Colon Cancer

Researchers claim that there is an "urgent need" to test triclosan. After preliminary studies, scientists conclude that it might cause colonic inflammation and promote colon cancer. Although generally considered safe, the chemical has become worryingly universal. Because of its potential role in antimicrobial resistance and endocrine disruption, and its theoretical effects on the immune system, it has been deemed a "minant of emerging concern" by the United States Geological Survey. A study tested whether triclosan might have any negative effects on gut health. In order to investigate, they tested the impact of small doses of triclosan on a range of mouse models. The scientists gave each mouse model a brief, low-level exposure to triclosan. In all mouse models used, triclosan prompted inflammation of the colon, worsened symptoms of colitis (inflammation of the lining of the colon) and promoted colitis-associ-

Continued on page 27

PHOTO BY STEPHEN BOYER

Fight

Was the last thing that I ever said to her.
Oh my God I was terribly mistaken.
When she said that she wanted to go home,
I thought she meant Heaven.
It didn't occur to me
That she wanted to go to our home,
Find a career, get married, raise a family
And live happily ever after.
The things that every twenty-seven-year-old girl dreams of.

Fight
Was the last thing that I ever said to her.
She fought longer than three, three-minute rounds
She fought twenty-four hours a day
Three hundred sixty-five days a year
Her prize meant more to her than a title
A ranking
Or a medal
Her prize was one extra day of her life.

Fight
Was the last thing that I ever said to her.
Then she closed her eyes and she died.

-R. Rohde, 2006

FROM OUR FAMILY PHOTO ALBUM...

MOLLY PAM AND HER HUSBAND, ADAM, ENJOYING HVAR, CROATIA, IN JUNE 2018.

AARON CUNNINGHAM CELEBRATING HIS 40TH BIRTHDAY IN 2017 WITH HIS NEPHEW CONNOR.

ANDREA EISENMAN WITH HER DOG, ERNIE, AT THE OCEAN IN HAMPTON BAYS, NY.

DAN MCAULIFFE (BROTHER), KATHY MCAULIFFE (MOM), DAN MCAULIFFE (DAD), COLLEEN ADAMSON, AND SCOTT ADAMSON CELEBRATE HER PARENTS' 50TH WEDDING ANNIVERSARY, AT TEUGEGA COUNTRY CLUB IN ROME, NY, ON MAY 13, 2017.

JOEL GUTIERREZ AND AMY SYLVIS.

PHOTO BY ANNA DELORES PHOTOGRAPHY

JEANIE HANLEY AND HER DAUGHTER, JESSICA HANLEY, CELEBRATING JEANIE'S 56TH BIRTHDAY IN MANHATTAN BEACH, 2018

SEARCHING FOR THE CURE

One Patient's Perspective

By Brad Johns

Growing up, I never gave much heed to clinical trials. Now, they are one of the most important things in my life. Clinical trials mean that you get to participate in the cutting edge of research, try out medicines way before the average person and help improve the lives of every person with CF in the world.

Seven years ago, I began my search to participate in clinical trials because of a friend. She and her family had been invited to watch a CF podcast about a new drug that was supposedly going to change the way that CF would be treated. High expectations were put on this drug because it had seen very good results in its clinical trial stages of development. This drug would later be known as Kalydeco.

After seeing the podcast, she immediately e-mailed her friends and family about the news. Six months prior to this, I had met with her daughter and son-in-law concerning their child. He had recently been diagnosed with CF and they wanted to speak with someone with whom they could talk about their situation very candidly. I met with them and honestly expected to never hear from them again.

When I got the e-mail, I'll be honest and say that I didn't really want to spend the time reading it. However, as I began to read it, I was filled with excitement. I was so excited, in fact, that I wanted to scream and shout. I was in my college library at the time, and containing myself so that I did not disturb others, was very difficult.

I immediately began to research this drug and where I could find information about the trials that were taking place. It was at that time that I learned that there was a second drug, later it would be called Orkambi, that could possibly work for my genetic mutation. Oh, my, could this be real? Could this really work for me? Could my life be extended because of some new pill? It was hoped that this new drug would increase CFTR function in a person with CF. My mutation basically causes there to be very little, if any, CFTR function, so any increases in it would go a long way.

I was excited. I could not believe what I was reading. This new medicine could possibly extend my life and there were hospitals around the country wanting to test it out on CF patients. After looking at the centers that were involved with this study, I realized that the closest one to me was about four hours away. I didn't know a soul over there. Why would they choose me? They do not have a clue who I am and have no reason to invite me to participate.

Even though the odds were not in my favor, I decided to write the head investigator an e-mail. Yes, I would write her an e-mail and plead my case to be involved in their research. Carefully, I crafted the e-mail so it would not sound like I was begging to be part of their newest endeavor.

BRAD JOHNS

I sent the e-mail that day. That is a day that I will never forget. It was the e-mail that changed my life forever. Patiently, I waited for a response. Finally, after about two weeks, it came.

The e-mail went something like this:

"Dear Mr. Johns, we are currently in the development of the Phase 2 trial for VX-770/809. We are not ready to begin the trial that you are speaking of. However, we would like to begin getting information from you and your doctor so that we could have everything in order when we do. Please send us a copy of your medical record so that we may begin the process of assessing your health to see if you are a good candidate for this drug study."

I couldn't believe what I was reading. They were going to allow me the opportunity to try this new drug. Six months later, I was sitting in their unit screening for my first Vertex drug trial.

I screen-failed for that trial. I was not allowed to participate. I was CRUSHED. However, another opportunity appeared with a new drug, VX-661/770 (Symdeko) seven months later. I was one of the first CF patients in the world to try it. It wasn't even in its current form yet. It was a much smaller dose as to make sure that it would not do harm to a person with CF.

As time has gone on, my passion to help with these drug trials has grown. My life has been greatly affected over the last six or seven years because of the opportunities that I have been given through these trials. Since my first drug trial, my lung function has

improved. That never would have happened if I had not gotten involved in clinical trial research.

When you participate in clinical trials, you help not only yourself, but you also help the 70,000 CF patients worldwide. Clinical trials are very important to increasing the outlook for all who have CF. As a CF patient, if you have the opportunity, I would encourage you to participate in drug trials. They are the KEY to improving the life and health of every CF patient in the world. ▲

Brad is 46 and has CF. He lives in Mendenhall, MS. He enjoys being active in the CF community. You may contact him by e-mail at: Brad.johns@rcsd.ms

Pay It Forward

BRONZE

Michelle Belt (in honor of Laura Davis)
The Benevity Community Impact Fund
Frances Birkner
Pete Caliendo (in honor of Laura Davis)
Tina Carlsen Cederquist (in honor of Laura Davis)
Charles & Maureen Cokelet (in honor of Amanda Ahlert)
Robert & Nancy Coleman
William Coon
William Coon, Jr. (in memory of Diane Farley Coon)
Kevin Corr
Janie Davis (in honor of all those with CF)
Laura Davis (in honor of Laura Davis)
Pauline DiNello
Ed Fleischman (in honor of his 76th birthday)
Anne & Chris Gautier (in memory of Stephanie Rath)
Kristen Gleason (in honor of Laura Davis)
Elizabeth Hissing (in memory of Maryann Bean)
Lauren Hunsaker (in memory of Stephanie Rath)
Dr. Neil Isdaner (in honor of Ali R. Isdaner)
John & Joanne Jacoby
Brooklyn Kupsak (in honor of Laura Davis)
Ruth Ann Layton (in memory of Joseph & David Donaldson)
Nicole Levas (in honor of Laura Davis)
LindQuist, Corp.
Tia Lucchesi (in honor of Laura Davis)
Shannon Lynn (in honor of Laura Davis)
Zachary Marotte (in memory of Joseph & David Donaldson)
Sydna Marshall (in memory of Stephanie Rath)
Stone McGowan (in honor of Laura Davis)
Emily Meyer (in honor of Lise-Courtney D'Amico)
Nick Mitcheff (in honor of Laura Davis)
Loretta Mulatz (in honor of Debra Radler)
Heidi Neargarth (in honor of Laura Davis)
Sheila Rathbone (in honor of Lynn)
Jeannine Ricci (in memory of Stephanie Rath)
Nancy Rieker
Salesforce.com
Michael Schnitzer
Gretchen Scott-Moore (in honor of Vicki Thompson)
Nancy Schueneman (in honor of Laura Davis)
Karen Scott
Kenneth Seil (in honor of Laura Davis)
Max & Iris Shaw
Jane Simi (in honor of Laura Davis)
Beverly Sufian (in honor of Beth, Sandy & Aviva Sufian)
Kristin Castricone Tepovich (in honor of Laura Davis)
Susan Tickell (in honor of the Desch family)
Jon & Joyanne Van Bloom (in honor of Sydna Marshall)
Katie Lathrop Venvertloh (in memory of Joseph & David Donaldson)
Laura Watts (in honor of my 20th transplant anniversary)
Dr. James Yankaskas

SILVER

Janice Friedeborn
Michael Gallagher
Dr. Jeanie Hanley (in memory of Stephanie Rath)
Gary Lazur (in honor of Delayne Santos)

GOLD

Norman Young

PLATINUM

Genentech

SUSTAINING PARTNERS

Kroger Specialty Pharmacy
Two Hawks Foundation

PEARL SUSTAINING PARTNERS

Boomer Esiason Foundation
Cystic Fibrosis Foundation

IN THE SPOTLIGHT
With Colleen Adamson

By Jeanie Hanley and Andrea Eisenman

Colleen just celebrated her 20th anniversary of lung transplant and eighth for kidney transplant. She is delightful, humorous and engaging and has dealt with so much with grace and determination. There was only one exception – a difficult time when she couldn't have salt in her diet! We're sure you'd all agree there. Other than that, her experiences will inspire and educate you and probably reduce anxieties you may have about the transplant process. With her challenging health issues, she has had an exciting career, long and happy marriage and has felt the love and support from family and friends. Although I've (Jeanie) known Colleen for many years, as our former Treasurer, I didn't know very much about her life and transplant experience. I'm honored that she so openly shared her life with us. Read on to meet our latest star. Spotlight, please!

Stats – 49 years old, living in Alexandria, VA

When were you diagnosed with CF? Who was instrumental in your diagnosis?

I was 13 months old when I was diagnosed with CF. My mom recognized all the signs of the disease by reading the Dr. Spock book. She had to insist on me being tested, because the doctors didn't think I had it since there wasn't a history of it in our family (there was a history of lung issues on both sides of my family, but it wasn't called CF back then).

How long have you known your husband and how long have you been married?

I have known my husband, Scott, for 25 years. We were married on June 28, 1997, and have been married for 21 years.

What CF issues did you have before transplant?

COLLEEN AND SCOTT ADAMSON AT THE HONDA CLASSIC GOLF TOURNAMENT IN PALM BEACH GARDENS, FL, IN FEBRUARY 2018.

Lung and sinus infections, pancreatic insufficiency, gall bladder issues and I was very skinny.

What job did you have at the time?

I was a cost analyst for the Navy, estimating the costs of building, operating and maintaining ships, aircraft, missiles and information technology systems.

When and where did you get your lung transplant?

I received my lung transplant at Fairfax Hospital in Falls Church, VA, on July 3, 1998. My mom called it "Colleen's Independence Day."

What was your health like before the transplant process started? Why did you get listed for transplant?

My health started going downhill the year before I got married. The stress of buying a house, traveling a couple of times to England for work and planning a wedding all at the same time took a toll on my health. In December 1997, I was at Johns Hopkins Hospital for a tune-up, which started out fairly normal except I had a G-tube for feedings since I was losing weight. About a week into the tune-up, I suddenly went into respiratory failure. I don't remember anything about that and I don't remember the month of January 1998. My husband of six months was my next of kin, so even though my parents were there my husband had to make the decision to stop treatment or have the doctors put me on a ventilator. He decided to have them intubate me. I was in the hospital for two months on the ventilator. My CF doctor completely gave up on me; he came to see me once and never came back. The doctors told my family I only had six months to live and I would never get off the ventilator (I took that as a challenge!). I was sent to a rehabilitation facility near my house at the end of February 1998, and by the end of May 1998, I was miraculously off the ventilator and back home. However, I was on six liters of oxygen and still very frail. I could not go up a flight of stairs without sitting and resting. I ended up with a lung infection. I was doing nebulizer and vest treatments about eight hours a day and slept a lot. I had a high level of anxiety. I couldn't be alone at all, and I had to take anxiety drugs when doing the vest and taking a shower because they made me claustrophobic.

How difficult was it to be on the ventilator for so long and how did you get off of it?

It was very scary to be on the ventilator. First of all, I had no idea what a ventilator was and that I was even on one. I didn't know that I couldn't speak or eat while on the ventilator, which made it so scary because sometimes my nurse call button would fall off my bed and I couldn't reach it. I was anxious all the time. I was afraid to even move. The only thing that helped was my mom being there every day, all day and my husband coming in the evenings. They were my voice. It was also terrifying to get off the ventilator. I had been depending on that to breathe for me for so long that it was excruciating to be off it at all, even for just one hour, which is how they start to wean you off it. I couldn't wait to be put back on it. My dad noticed this and had a heart-to-heart talk with me. He told me that the ventilator was not my friend and that I had to get off it to have any chance of getting out of that rehab facility. I really needed to hear that, because after that talk, I did change how I thought about the ventilator and realized it really was a bad thing to be on. Luckily, I was weaned off it and was able to go home at the end of May 1998.

How long did you have to wait to start the pre-transplant process and get listed?

When I was at Johns Hopkins, they would not even consider me for a lung transplant because their wait list (which was based on time on the list) was one year long and they thought I would only last six months. One of the doctors recommended my mom call Fairfax Hospital because they had a lung transplant program that was fairly new and didn't have as long a wait list. I did end up getting listed there and got my lung transplant within 12 hours of being listed. In our opinion this was an absolute miracle because my time was literally almost up. It just so happened that there were seven people ahead of me on the wait list, but the lungs that came in were B blood type and I was the only one on the list with B blood type! I was extremely lucky to be at the right place at the right time.

How did you feel right after the transplant?

I remember watching Wimbledon (I love watching tennis) and I remember the nurses getting me up and walking the day after the surgery. I don't remember a lot of pain, I do remember being able to breathe easily for the first time in a very long time, which was wonderful. I felt so great that about a week after the transplant I got out of bed and immediately fell — having been bedridden for so long on the ventilator prior to my transplant, my legs weren't strong enough for me to stand! When you can finally really breathe, you think you can do anything.

When did you go back to work and what was that like?

I went back to work about six months after the transplant. I was in the hospital recovering for three weeks, which was a relatively long time compared to other lung transplant recipients. I was very frail when I had the transplant and it took me a long time to gain enough strength to go home and care for myself. I also went through pulmonary rehabilitation once I was strong enough to do that. Going back to work was wonderful; getting back to normal and seeing my friends/coworkers made everything I went through worthwhile.

Do you have any limitations now?

I still try to stay away from crowds when possible. I can't do any gardening because of potential fungus in the dirt. I don't drink alcohol because of how it can affect my medication and my liver. I try to stay out of the sun because I've had a lot of skin cancers, and I don't have any children and try to not be around children because of germs.

What led up to your kidney transplant?

My kidneys failed in 2005 due to one of the immunosuppression drugs I was taking. They failed quickly and I had to go on dialysis for about six months. The dialysis dietitian told me I had to go on a no-salt diet; that was the first time I outright cried! We CFers need our salt! I didn't even cry when I was told my kidneys were failing!

Dialysis was really hard. It made me very nauseated and tired, and I lost 30 pounds in one month. My best friend, Kelly (we met in college and were roommates for two years), was adamant about donating a kidney to me because she felt she "had to save my life." I tried to talk her out of it but she was insistent. She ended up being a good match and my kidney transplant was on March 7, 2006, a day before my birthday. What a great friend and what a birthday present!

How did your husband deal with your CF and transplants?

Scott is so wonderful because he is very even-keeled and goes with the flow. He was the only one who could read my lips when I was on the ventilator and it calms me down to just look at him when I am stressed out. Also, he lived more with my mom than with me the first year we were married – talk about a test for a new husband!

Did your coworkers know about your health issues?

Yes, I've always been very open about my health issues because I think people can learn from each other in this area. Plus, I think it's fair to explain why I coughed so much (before the lung transplant). After my kidney transplant, one woman who I barely knew at work told me she became a donor on her driver's license because of my story. I was so touched by that.

When did you go on disability and why?

I just retired on medical disability at the end of April 2018. This was not

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an easy decision for me to make. It was very hard to accept that I was not as good as I once was with my memory and being able to answer questions as quickly as I used to. As a cost analyst, I was expected to remember numbers and associated technical information on the spot, and with my increasing memory issues and not being quick to answer questions, it was getting harder and harder and, quite frankly, very sad for me, because being a cost estimator was a big part of my identity and a big part of my social circle.

Do you volunteer for any organizations?

I volunteer for the Washington Regional Transplant Community (one of the 58 organ procurement organizations nationwide), raising organ donation awareness by speaking to people at health fairs and various community events. I am also a mentor for the Fairfax Hospital lung transplant program pre-transplant patients, talking to them about what to expect post-transplant and relating my experiences.

How will you spend your time now that you're retired?

I plan to do more volunteer work for WRTC and write a book about my life, with a focus on my transplant experiences. Also, I want to focus more on my health by exercising more, including walking my dog, Penny.

Who inspires you?

I know how lucky I am to have had support from a lot of people to get me as far as I have gotten. First of all, my family, especially my mom. She has been such a fighter for me all these years, and I have learned determination and how to be my own advocate from her. My dad and brother have always been there for me and keep me smiling and sane. Scott is my rock and lets me know, sometimes without even saying a word, that everything is going to be okay. My friends and other family members are super supportive to me and my family. And I can't leave my miniature Schnauzer, Penny, out! She makes me laugh every day and loves to go on walks. She is now 11 years old and still spunky and spoiled; I wouldn't have it any other way!

What advice would you give to those who are considering a transplant?

Know that you are trading one disease for other ones, but I think it is worth it. The number of medications and potential side effects can be overwhelming at first, but it becomes second nature pretty quickly, especially I think for people with CF who are used to taking medications. Also, vigilance is key because any change in your body (temperature, spots on skin, mood, blood pressure, breathing etc.) could mean something could be going on, and catching anything out of the ordinary early is crucial. ▲

Jeanie Hanley is 56 and is a physician who has CF. She is a Director of USACFA and is the President. Her contact information is on page 2. Andrea is 53 and has CF. She is a Director of USACFA and is the Webmaster and Executive Editor of CF Roundtable. *Her contact information is on page 2. If you would like to be interviewed for "In The Spotlight," please contact either Andrea or Jeanie.*

Paying It Forward - Thank You!

On behalf of *CF Roundtable* and the directors of USACFA, I bestow a BIG THANK YOU to all of you, our generous donors, who once again have shown amazing support for our CF community. Because you paid it forward during the CF Awareness Month of May, we can continue to provide *CF Roundtable* for free to you, your friends and family.

Our board is made up of all volunteers who have cystic fibrosis. As a result, 100% (yes, 100%!) of your contributions are used in the production of *CF Roundtable* and our programs such as providing college scholarships and our Speakers Bureau. Our Speakers Bureau is a special panel of our directors, all of whom have CF and are freely available to speak at your next CF event.

Please continue to share your milestones (birthday, anniversary etc.) and your experiences with CF, either in the form of an article or simply e-mail us at cfroundtable@usacfa.org. We are so grateful and overwhelmed by your kindness and support. We would love to hear from you.

— Jeanie Hanley, President of USACFA

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

Amanda Ahlert
Hazlet, NJ
39 on June 6, 2018

Ed Fleischman
Plainview, NY
76 on December 24, 2017

Debra L. Radler
Roselle, IL
56 on May 31, 2018

Kathy Russell
Gresham, OR
74 on April 17, 2018

Wedding

Kathy & Paul Russell
Gresham, OR
53 years on March 27, 2018

Transplant

Amanda Ahlert, 39
Hazlet, NJ
Bilateral lungs
19 years on June 16, 2018

Paul Albert, 58
Catasauqua, PA
Bilateral lungs
25 years on February 10, 2018

Andrea Eisenman, 53
New York, NY
Bilateral lungs
18 years on April 25, 2018

Laura Watts, 66
West Grove, PA
Bilateral lungs
20 years on May 3, 2018

TILLMAN continued from page 18

ated tumor growth. Next, the scientists wanted to understand exactly why triclosan seemed to have this pro-tumor, pro-inflammation effect. They found that triclosan altered the composition of the gut microbiome significantly.
http://tinyurl.com/ybedf3vs

Antioxidant-enriched Vitamin Reduces Respiratory Illnesses In Patients With Cystic Fibrosis

The study looked at the effects of a "cocktail" of multiple antioxidants on inflammation and health outcomes in patients with CF. CF patients ordinarily do not adequately absorb important dietary antioxidants including carotenoids such as beta-carotene, tocopherols (vitamin E), coenzyme Q10 (CoQ10) and selenium that help to neutralize inflammation in the body. To address this issue, the antioxidants used in the study were delivered in a capsule specifically designed for individuals with difficulties absorbing fats and proteins. Antioxidant supplementation was safe and well-tolerated. Supplemental antioxidants increased antioxidant concentrations in the bloodstream in treated subjects and temporarily reduced inflammation in the blood at four weeks but not 16 weeks. Antioxidant treatment appeared to both prolong the time to the first respiratory illness requiring antibiotics and reduce the frequency of respiratory illnesses. Half as many of the patients taking the supplemental antioxidants experienced a pulmonary exacerbation (or respiratory illness) requiring antibiotics compared to the group taking the control multivitamin without added antioxidants at 16 weeks. In addition, the antioxidant treated group experienced a lower frequency of respiratory illnesses compared to the control group.
http://tinyurl.com/ydfx48qh
AND
http://tinyurl.com/y85n8h33

Cystic Fibrosis Severity Linked To Immune Overreaction To Fungus, Study Reports

Disease severity in cystic fibrosis (CF) may be associated with an overreaction of the immune system to the fungus Aspergillus fumigatus, particularly due to a type of white blood cell called a phagocyte — which ingests and kills invading organisms. Researchers found that phagocytes from CF patients release higher amounts of harmful reactive oxygen species in response to Aspergillus fumigatus. Up to 58% of CF patients are colonized with this fungus, and an estimated 47.7% of adult patients are affected by either allergic reactions or infection caused by the fungus. Persistent infections with A. fumigatus are also known to be adversely correlated with lung function and hospitalization. The data suggest that a hyperresponsive state in CF phagocytes plays a crucial role in the hyperinflam-

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FINANCIAL SOLUTIONS

The Way We Work, Earn And Save Is Drastically Changing

By John Mercer

The world of work and the way we earn money is changing more drastically than most people can even imagine, and it would be a disaster to avoid thinking about that when planning for your future — both financially and professionally. To coincide with these drastic transformations, the way we think about and look at cystic fibrosis and the role that it plays in our lives is changing more now than ever in the course of human history. The future is starting to look more and more different.

The development of drugs is beginning to provide an unheralded level of optimism that some have never been able to imagine. Think about this: You've imagined looking at your life one way and thinking you knew or had an idea of what was inevitably coming and unavoidable. Then you planned your future accordingly for that. Now, due to amazing discoveries in the pharmaceutical industry, people with CF and their families are beginning to envision a future they may never have imagined before. The power of that is more substantial than anyone truly understands and very few people are talking about that. The course of one's history or how one previously imagined it needs to change. Along with that, the working world is going through the most notable disruptions since the 1970s. All of it is for the better; it would be a disaster to not plan accordingly.

The workforce of today is dramatically different from that of five years ago, let alone 10 or 20. Can you imagine what it is going to look like in 2025? All of these changes can be related to social, technological, environmental and behavioral issues. Generally, most of it is for the better. The majority of mundane, boring and simple task-based jobs will be phased out of the workforce, and how people earn money and work is having to adapt to all of it in order to keep up with the times. You need to look ahead at what that looks like in order to be prepared for your future, both financially and professionally. It's such a fascinating time in our lives and we should all have a high degree of excitement for the possibilities ahead. These are some significant trends that will hopefully alter the way you look at your work, earning potential and professional life.

> *"People with CF and their families are beginning to envision a future they may never have imagined before."*

JOHN MERCER

Stop Thinking "Jobs"

While the U.S. job market is relatively weak, there are some underlying themes that pose new problems for individuals and their employers. Statistically, the length of time a person remains at a job is staggeringly low, and the time it takes before a person begins a new job search after starting a new job is mind boggling. People born after 1968 average 12 jobs throughout their career, and as millennials begin to take over the workforce that average number continues to rise to an estimate of 15 jobs over a lifetime for those born after 1980. Even more concerning is the length of time it takes a person to begin a new job search after starting a new job. Current estimates state that a new employee begins a job search within 17 months in their new role. Also staggeringly high is the rate of voluntary unemployment, or not having a job by choice. Estimates state that around 20 percent of those who are unemployed are so because they choose to be, as they can't acquire employment that will make them happy. The way we think about a "job" is dead; it's an antiquated mentality.

The mindset of looking for and thinking about a "job" is unhelpful because it encourages you to think about and envision a monolithic and rigidly fixed, uniform, pre-decided lump of time that often is viewed as something unbecoming of who you are

or who you want to be. A job often is described as a "chore" that one just has to do, regardless of how it impacts who one is. In organizational behavior, a task or goal mindset is far superior to that of the "job" mentality because the greater degree of freedom, interest, creativity and intellectual autonomy one has in a potential role increases the immunity from becoming subject to the multitude of psychological pitfalls associated with having a "job." When you think about how you spend your time earning money, you must think at great lengths about what will challenge you intellectually, the creativity and freedom you may have, the interest you actually possess in a particular role and (most importantly) how something fits into your life both now and in the future. If you are more selective about how you pursue roles that earn you money based on the above factors, you tend to earn significantly more money both in the long and short term. The "job" is a problem, not your ability to earn money, to make a great living and to be happy. People who describe themselves as happy at a job tend to make over 30 percent more money in their lifetimes than those who don't.

The Upside Of Changing Jobs

The newly minted term "job-hopping" was coined as millennials have become substantial within the workforce. Job-hopping is the act of *trying out* a variety of roles, workplaces, duties, cultures and countless new skills. While job-hopping, or testing out jobs first, may possess a negative connotation among older generations, it actually has an immense upside. Younger generations are drastically more obsessed with finding happiness and fulfillment in their work than previous generations. Rather than predicting what will make you happy and making a commitment to that, testing out different jobs, roles, skills and professions has an indescribable value. It's the theory of trial-and-error — a methodology characterized by repeated, often varied attempts in various contexts, which are continued until individualized success is achieved. This can be done in any number of ways besides applying to a job and then quitting. People are always willing to talk about their jobs and what they do to aspiring professionals; you just have to ask. You can also watch the thousands of videos online about what certain jobs may entail. Or, don't be afraid to e-mail someone for whom you may want to work, asking if you can spend a week with them. You'd be amazed how quickly they say yes.

Find Positions That Are Suited To Meet Your Needs

Research shows that in order for one to prepare for the future of work, to be happy and to be financially secure it is imperative that you possess a job that is of interest to you. If you aspire to achieve financial security, you need to earn money in a way that is of interest and meaning to you. Also, it has to fit in with the greater vision that you have over the foreseeable future. When people report happiness and interest in their work, they make substantially more money over a lifetime...not to mention the huge health benefits associated with that! It is never okay to settle on something that doesn't help you achieve the goals you have in life or the level of health you desire.

The most significant reasons people change jobs are: higher pay, finding a less stressful job, escaping negativity at work, changing career focus, work/life balance, more interesting work, abilities and skills that don't fit, and lack of recognition and respect for the work they do. If you avoid finding roles with these negative aspects and pursue something you more strongly desire, the money will follow. Also, improved mental, emotional and physical health are associated with finding roles that have significant meaning and interest to you.

Next Steps Financially

Now that you've found something that generates income, the next step is to learn how to save your money. Guess what? The financial world is also going through drastic changes that can save you a great deal of money. Since technology has drastically changed the landscape of the world, the outdated financial models have changed as well. Active money management or a traditional professional who helps manage your money is rapidly fading away to an online presence and a number of communities that actively share knowledge, tips and tricks. Most of it is free! Whereas your traditional savings and retirement options skim more money than they should off the top to advisors, taxes, fees etc., online companies are offering savings options that are customizable and cost significantly less. Combine that with your ability to acquire knowledge and information, and you're all set. The rest is up to you!

I challenge you to do your homework and find companies and solutions that will do just that and are totally transparent about what they do, what they'll return and what they'll charge you. Or, find a fiduciary professional who will also do the same and compare all of your options against one another. Keep asking until people don't want to hear from you; then ask some more! Our lives are ripe with greater change than most can ever expect; it's a great time to be alive. Keep your standards high and let's go change the world. ▲

John is 34 and has CF. He is an investment planner and senior consultant in Pasadena, CA. He is a house party enthusiast and organizational strategist. You may contact him at: jmercer@usacfa.org.

WELLNESS

Living Mindfully With CF

By Julie Desch

I probably should just change this to the Mindfulness Column, because that's all I seem to write about these days. But when I think about topics, this is all I come up with, so I'm either too old to think of new ideas, or because I'm old, mindfulness is the thing that matters most to me now. Or both.

As many of you may know, I have been teaching a class entitled "Mindfulness-Based Stress Reduction (MBSR)" to people with CF and their caregivers/loved ones over the last few years. This class follows a structured protocol—the class that I teach is the same one you might find in a community hospital or local community center. It is eight weeks in duration, each class is two hours long, with specific "homework assignments" between classes and a half-day "retreat" on a weekend day between classes six and seven. The main modification that I have made, which makes this class unique, is that it is done online instead of in person.

I think the class has been successful and I look forward to teaching it again. But my personal venture into mindfulness has led me to take a mindfulness meditation teacher training course with two well-known teachers in the Western mindfulness movement, Tara Brach and Jack Kornfield. As a result of this training, my practice has deepened quite a bit, and with that, I am seeing more and more clearly how having a more mindful presence to my life has eased the struggle of living with CF.

Nothing has changed with the CF, of course. I still get sick out of the blue and end up on IVs. More and more antibiotics are unavailable to me due to either bacterial resistance or my hearty immune system saying, "Nope." I still have to start my fitness program back up from ground zero after exacerbations. I still read about friends who die of this crappy disease. I still miss my sister and brother. I still get angry, and sad, and frustrated.

What I see changing (slowly) is my relationship to the illness, to the emotions that come up almost daily, to the worried thoughts that carry me away into a fantasy future or that come up with stories about each bodily pain that arises.

To me, living with CF is a little like running a marathon. Maybe not in terms of the length of the race, but in the mental and physical stamina that it takes to finish. It's hard. Sometimes it feels like things are going along smoothly, and other times you hit the wall. Somewhere around mile 15, the body starts to complain not just because your lungs aren't "normal," but because you're well past halfway through and, dang it, you're now "middle-aged."

JULIE DESCH, MD

Whoever designed the course apparently decided it was a good idea to make the runners go uphill for the last 10 miles. Evil. Now on top of crappy lungs, your back hurts, your knees hurt, you suddenly can't see without glasses, your list of medical "providers" requires two pages and you forgot what you are writing about...

Oh yeah—the race. So when you decide to run a marathon, you train for it, right? That is, if you want to do it well, you set up a schedule, maybe even get a coach, modify your diet and put in lots and lots of miles "practicing" for the big day.

The analogy breaks down a bit here, because obviously we can't train for our life. We're in it. But I've started to think of "the rest of my life" as the marathon. I want to run it well, and to me that means being as present for it as possible...to not waste any more time on autopilot, being tossed around by my emotions, believing every thought that crosses my mind, regretting the past or worrying (much) about the future. This is a tall order, of course, and I fail almost every day.

But guess what I also do every day? I train. I'm in training to live mindfully for what remains of my life with CF. The training occurs not just during meditation, although that's where it starts. In sitting quietly, a remarkable amount of insight about how the mind works becomes available. You get to see your thought patterns and how they interact with emotions and even with the physical body. You ideally see these interactions from a non-judgmental and kind perspective. We call it "practice" because to live this way continually is impossible to do (or it seems that way to me). But just as with practicing a musical instru-

ment or practicing an intricate dance move, you do get better over time. Then as you understand how to observe the mind during practice, it gets easier to do it out in the world, and you become less controlled by an unruly mind. You have more choice in how to respond to difficulties in life. You have more power.

And now, drumroll please, you can join me in the training for the low, low price of.... oh yeah, it's free.

As one of the requirements for the mindfulness teacher training course, I designed a new course called "Living Mindfully With Cystic Fibrosis." I've essentially taken many of the concepts from the MBSR course, but hopefully tailored them to be more specific to CF, both for adults with the disease and for those who love them. As with my MBSR course, this new class is offered free of charge through the generosity of CFRI and grants from Vertex, Genentech and Gilead.

As a brief overview, the course is six weeks in duration. Each online class is 90 minutes and, instead of a six-hour "retreat," we will just spend one weekend morning in silent meditation. The schedule is as follows:

Class One (July 17): Introduction to Mindfulness
Class Two (July 24): Mindfulness of Body and Breath
Class Three (July 31): Mindfulness of Thinking
Class Four (August 7): Mindfulness of Emotion
Mini-Retreat (Saturday, August 11): Morning Meditation
Class Five (August 14): Working with Uncertainty
Class Six (August 21): Self-Compassion and Resilience

While there is definite overlap with MBSR, even if you have taken that class, I'd encourage you to check this out. I put much more emphasis on building resilience, self-compassion and how to live with the uncertainty that we all face. As I read somewhere, everyone knows and understands at some level that they are going to someday die, they just don't know how or when. But we understand this more acutely. We can learn to live with this uncertainty with more equanimity, and we can build the skills of resilience and compassion. Of course, many people turn to religion to fill this role, and there is absolutely nothing wrong with that! But this course is not about believing any doctrine. This is a course where you learn what is true about your own mind by direct experience. It can fit comfortably into any religious practice, or none at all.

The class will be offered one more time this year. If interested, go to CFRI.org and you'll see it listed as one of the psychosocial support classes offered. I hope to see you online! ▲

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

Stephanie Devine Rath
January 26, 1969 – April 7, 2018

Stephanie Rath, "Steph," was a hero to the CF Community, bringing her optimism and love of life to all. She was an inspiration, just like her love of flying pigs, her personal mascot and motivational symbol for overcoming the impossible. Even though her health was constantly a challenge requiring more of her time and attention, she forged ahead never to miss an adventure.

Steph had overcome many CF health challenges including rectal cancer, lung transplant and kidney failure. Being on dialysis didn't stop her from going on her annual music cruise with friends in January. Upon returning, she suffered serious health setbacks and despite having returned home from an ICU hospitalization and feeling like recovery was imminent, she passed away two days later.

In 2012 Stephanie was elected to the USACFA board of directors. We were so incredibly fortunate to have her as our Treasurer since 2014. She was a skilled accountant who was a joy to work with, gave her time tirelessly and made USACFA better in so many ways. She also was USACFA's Chair of Fundraising. She served on the USACFA board until her death.

During her 49 years, she touched many lives in many ways – being the mover and shaker for a yearly music cruise with friends, CFF fundraisers in Indy, volunteering at the local nursing home and many more. Her love of music was included in all she did.

She treated everyone with kindness and respect. Steph truly lived in the moment and was a beautiful reminder for all of us to embrace life with gusto. She is survived by her husband, Randy Rath, her mother, Robin Devine, and her two Boston terriers, Rocky and Rosie. We will sorely miss her.

TRANSPLANT TALK

I Have What Now?!

By Andrea Eisenman

As I sit quietly in my individual "pod" in the infusion suite at Columbia Presbyterian, I reflect on my recent diagnosis. I am between wanting to sleep due to the Benadryl and jumping out of my skin from the steroid push. My infusion of Rituxan comes shortly after these medications and will last around three to four hours.

Have you ever been at that point that you are incredulous about a new diagnosis? Have you ever been angry about it? Scared? Have you ever thought: I have WHAT now? And why? Normally, I answer "no" to all of the above. Just having started treatment for Post-Transplant Lympho-Proliferative Disease (PTLD), I went through a range of emotions from disbelieving, scared, angry to finally accepting it. But it took me many months and a lot of processing.

I feel I am constantly coping and adapting to a new normal. Whether it is a decline in my PFTs, a pulled muscle or osteo-arthritis that limits my ability to exercise, skin cancer or my having constant sinus infections, I try to work around it when possible to live my life. I am always pushing to find options to my various situations. I am constantly working around my sinus pain or just keeping up with my doctor appointments to stay on top of it all. But having this cancer, PTLD, really threw me. One of the hardest parts was waiting for a correct diagnosis. The testing and having a surgical biopsy were more traumatic and painful than I expected.

The last year brought many health issues directly following the death of my mother—melanoma, two rounds of IV antibiotic treatments within eight weeks for sinus infections, a blood clot from the PICC line that involved a hospital stay and then these mysterious lumps that were showing up in a few parts of my body (the lymph nodes protruding leading to PTLD). Even though my mom had not cared for me physically, as she did when I was waiting for a transplant, she had been my emotional support.

That period of extreme loss and depression really was a challenge. I missed my mom horribly. I cried almost every day. I wanted to die. And I am embarrassed to write this, but that was how I felt for a few weeks. Then I realized, my mother would not want this, she did everything in her power to keep me alive and feeling well. After that the proverbial doo-doo hit the fan. It started with the sinus infection that lasted most of the summer. And then the rest followed.

My mom also taught me how to cope and problem-solve. Being proactive this past year was one of the things that helped me stay focused and feel like I was trying to solve the problem(s). I kept lists of when to see doctors, when to stop taking Coumadin for the surgeries, when to order my meds etc. I tried to stay orderly even though at times my mind was in disarray from the loss and diagnosis. Also, staying with a regimented exercise routine, even if it was only 30 minutes a day and was a walk in the park with my dogs, I did it. These things usually helped me clear my head to be less anxious.

ANDREA EISENMAN WITH WILLIE, HER RECENTLY INHERITED DOG.

There was fear of the looming "what if" people face with cancer. There is no cure, only remission; what if it comes back? I had to acknowledge my fear personally and with others to process it and then move forward.

I didn't have the time to feel sorry for myself and even if I did, it really would not have served me. Victimhood can be stifling in its own way. Mainly, I was angry. And maybe that anger propelled me to start to try to just take care of things as best as I could. During my IVs, I got the melanoma removed. Then after that, eventually, I dealt with the PTLD diagnosis to start the treatment of infusions that would hopefully get me into remission. I tried to stay positive. I distracted myself when possible with tennis and learning pickleball, which are both more social than just biking or swimming can be. Initially it was hard to be around others, but since it was usually fun to be in a group, my hectic health life melted away for the duration of play.

For the depression, I knew I needed help to work through my sadness after my mom died. I met with the transplant center's psychiatrist who gave me an antidepressant that I had been on before. This helped me sleep and function. Just speaking to the psychiatrist to "unload" was a start. I then found a great CSW whom I now see

weekly. She initially helped get over my guilt and loss over my mom's death. I am more at peace with her death, but I will never be over it entirely. Talking to someone who "gets it" really makes a difference. She also helped me process my anger with the surrounding cancer, too. Of course, I spoke to those who knew me but I didn't want to be a broken record and that was how I felt. I was stuck on my sad song.

I considered a bereavement group but realized I would be exposed to people with colds and possibly flu. Having to explain why I cannot do group hugs or be close to others during these sessions of crying and letting go would get tiresome. Wearing a mask might be too off-putting for something like that. I just felt, as I moved through this journey, seeing someone one-on-one was my best recourse.

Mom frequently told me, things could be worse. There always are people who are worse off than you. During all of this upheaval, I tried to remain grateful and see the bright side of things. I was grateful for many things: having my mom around for so many more years than I expected, being alive and celebrating 18 years post-transplant in relatively decent health, even the PTLD I was diagnosed with was the more benign kind. And I recently learned that I was in remission. I was not taking those things for granted.

Learning all I have in difficult situations (having CF, receiving a lung transplant, spinal fusion for scoliosis, late onset diabetes etc.) has taught me to discard misgivings and regrets and try to focus on what is happening now to actively move forward. Moving through a minefield is easier said than done. Going through life with many challenges helped me learn to communicate with my physicians: speaking up and advocating for myself. It helps me with tracking my health insurance—taking notes and whom I spoke to when and on what day. And, I really saw how it served me in taking care of my mother's will. The lawyer's office did a lot of it, but I still had to do the footwork: calling and going many places, mailing things to close her accounts and making copies of everything, keeping track of who I sent what etc. Even though it was tedious, being organized and staying focused got the job done. There were days I could not do it or I reached a roadblock. Usually, I could just put it all away and go back to it the next day...something I could not do in a crisis with CF.

For years, I lived in fear of how the loss of my mom would create a huge hole in my life. But now, almost a year later, I am more settled. I still cry about her not being alive. But I somehow came through this whole experience of losing her and going through a gauntlet of health challenges a bit stronger. And I feel that my coping and adapting skills that she instilled in me helped the most. ▲

Andrea is 52 and has CF. She is a Director of USACFA and is Webmaster and Executive Editor of CF Roundtable. *Her contact information is on page 2.*

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matory response upon exposure to A. fumigatus.
http://tinyurl.com/ybgwl4la

Natural Sniper Kills Hospital Bacterium

Bacteria produce proteins to take out specific competitors. One of these proteins can kill the hospital bacterium Pseudomonas aeruginosa. Microbial geneticists have unraveled how this protein launches its attack and ensures that the bacteria die very quickly. In the long term, these proteins hold potential for new antibiotic cocktails. One type of these proteins — LIpA bacteriocins — is highly effective in eliminating the hospital bacterium Pseudomonas aeruginosa. The LIpA protein has a specific target in the outer wall of the bacterial cells. That target is a protein as well: the BamA protein, which is involved in maintaining the bacterial cell wall. Without the BamA protein, bacteria cannot survive. LlpA binds to that BamA protein and, by doing so, shuts it down. Unlike standard antibiotics, LIpA proteins don't even need to get inside the bacteria; they recognize their target and then sabotage it from the outside.
http://tinyurl.com/y86xdbgy

Guidelines Needed For Listing CF Patients With Nontuberculous Mycobacteria Infections For Lung Transplants, Survey Finds

Clinical centers around the world differ in the standards they use when considering cystic fibrosis (CF) patients with nontuberculous mycobacteria (NTM) infections for lung transplants, and few have a clear written policy. Researchers found that most centers surveyed do not consider NTM infections in cystic fibrosis as an absolute contraindication for transplants. However, NTM infection in cystic fibrosis patients has been regarded by some clinicians as an absolute contraindication for lung transplant due to poor post-transplant outcomes. In particular, the presence of M. abscessus complex before a transplant has been linked with severe complications and poor outcomes after the procedure. But some

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single-center case series have indicated that patients with an NTM infection who undergo a lung transplant have similar outcomes to those without an NTM infection. Because different approaches can be taken, researchers in this study investigated current worldwide practices concerning the listing of cystic fibrosis patients with NTM for lung transplants. Results showed that only 29% of centers have a clear written policy regarding NTM. In addition, 16 centers, or 76%, require molecular identification of specific NTM species. Only four centers considered infection with M. abscessus complex a contraindication for listing, but 76% regard it as a relative contraindication, which means additional caution should be used if other conditions or contraindications are present. Results also revealed that 86% of centers require treatment before the transplant. Only 61% of the clinical centers surveyed reported having policies in place regarding the segregation of patients depending on NTM status before a transplant, and just 48% reported having such a policy for patients after the procedure. In order to better inform decision-making, the team emphasized that there is an urgent need for further high-quality clinical research to comprehensively investigate the outcomes and international experience of patients with NTM after lung transplantation.

http://tinyurl.com/ycjeh5qm

CF Therapy Maker JHL Plans Phase 1 Trial Of Its Biosimilar To Pulmozyme

JHL Biotech plans to start a Phase 1 clinical trial of its cystic fibrosis (CF) therapy JHL1922, a biosimilar to Pulmozyme (dornase alfa) that is designed to improve lung function. Pulmozyme (marketed by Genentech) was one of the first approved treatments for CF shown to prevent mucus accumulation. It is an engineered enzyme, called recombinant human deoxyribonuclease I or rhDNase I, which selectively breaks down DNA molecules in mucus. This helps thin the thick secretions. The result is better airway flow, improved ability to expel mucus and better ability to prevent bacterial infections. JHL Biotech expects JHL1922 to be a less expensive but equally effective alternative to Pulmozyme. JHL1922 was designed to achieve biochemical and safety profiles, as well as therapeutic activity, similar to those reported for Pulmozyme. Experts estimate that the average cystic fibrosis patient has to spend between $12,000 and $40,000 a year just on Pulmozyme. This underscores patients' need for more affordable therapies.

http://tinyurl.com/y8v5fdoj

CF Therapy RPL554 Improves Lung Function, Phase 2a Trial Shows

Verona Pharma's nebulized RPL554 can significantly improve lung function in patients with cystic fibrosis, according to data from a Phase 2a clinical trial. The latest results revealed that single administration of both high and low doses of the investigative drug significantly increased patients' average forced expiratory volume in one second ($FEV_{1)}$. This significant positive effect was sustained for at least eight hours. In addition, RPL554 displayed favorable stability and distribution profiles, and was well tolerated by the patients. RPL554 is an inhaled dual inhibitor of two enzymes, phosphodiesterase 3 and 4, which Verona designed to act as an anti-inflammatory as well as bronchodilator.

http://tinyurl.com/ya5r79n2

Proteostasis Stock Soars On Cystic Fibrosis Breakthrough Status

Regulators handed out breakthrough status to the company's cystic fibrosis therapy. The drug, called PTI-428, is meant to boost levels of the CFTR protein, which is lacking in cystic fibrosis patients. The therapy is an add-on treatment for patients with the F508del mutation in the CFTR gene who are already taking an approved CFTR modulator or as part of Proteostasis's triple combo regimen that includes PTI-808, a potentiator, and PTI-801, a corrector. PTI-428 can potentially be added to current and future standards of care.

http://tinyurl.com/y7wc7yea

AND

http://tinyurl.com/y6uqk9tr

Potential CFTR Corrector PTI-801, Plus Orkambi, Seen To Treat CF Patients In Phase 1 Trial

Treatment for 14 days with Proteostasis Therapeutics' PTI-801 in cystic fibrosis (CF) patients also being treated with Orkambi (lumacaftor/ivacaftor) led to statistically significant improvements in sweat chloride, body mass index and rescued blood glucose levels in a subgroup of patients with diabetes, Phase 1 trial data show. PTI-801 is a third-generation CFTR corrector. The trial's primary objectives are to assess the therapy's safety, tolerability and pharmacokinetics. Parameters like sweat chloride and body mass index (BMI) and changes in percent predicted FEV_1 ($ppFEV_1$, a measure of lung function) will also be analyzed. Patients with CF-related diabetes (CFRD) were also analyzed for changes in blood glucose levels. Results in the patients showed that PTI-801 was generally well-tolerated, with only mild or moderate adverse effects. The most common was pulmonary exacerbations. Patients randomized to the therapy's highest dose, 400 mg, showed significant improvements in both sweat chloride and BMI. At a dose of 200 mg, improvements were found to be significant only for the sweat chloride. Participants treated with PTI-801 also showed improvements in $ppFEV_1$ across all treatment doses, although the difference didn't reach statistical significance. In hyperglycemic CFRD patients, all three doses of PTI-801 were effective in normalizing the

glucose levels.
http://tinyurl.com/yd6pg9sz
AND
http://tinyurl.com/y75pm5ro

Laurent Pharmaceuticals Reaches Agreement With U.S. Cystic Fibrosis Foundation To Increase Development Award To $5M

Laurent Pharmaceuticals, Inc., announced that it has reached an agreement to receive an additional $2 million commitment from US-based Cystic Fibrosis Foundation to further support the upcoming Phase 2 clinical study aimed at demonstrating the safety and efficacy of the company's lead compound LAU-7b in adult patients with cystic fibrosis (CF). LAU-7b is a once-a-day oral pro-resolving therapy with potential to treat chronic pulmonary inflammation that leads to irreversible lung damage in patients with CF, regardless of their CFTR genotype. As opposed to typical anti-inflammatory drugs that inhibit the activation of the inflammatory response, LAU-7b uses the body's own ability to timely terminate inflammation without interfering with the natural defense mechanisms. LAU-7b was tested in adults with CF in a dose-ascending Phase 1b study, showing good safety and pharmacokinetic profiles, coupled with promising positive effects on specific markers of inflammation. The goal of the upcoming Phase 2 trial is to evaluate LAU-7b's effect on the preservation of lung function in patients with CF by reducing persistent, unresolved inflammation in the lung and stimulating its return to homeostasis.
http://tinyurl.com/y83uvudw

UC Researcher Granted Patent For Potential Pulmonary Infection Treatment

A researcher in the University of Cincinnati (UC) College of Medicine has been granted a U.S. patent for a potential treatment for a pulmonary infection in patients with cystic fibrosis (CF). The treatment, known as AB569, is a potential treatment for many antibiotic-resistant organisms, including Pseudomonas aeruginosa (P. aeruginosa). Earlier work on CF found that P. aeruginosa was susceptible to destruction by slightly acidified sodium nitrite. The researcher also discovered a synergistic effect by adding disodium ethylenediaminetetraacetic acid to acidified sodium nitrite, which led to the development of AB569. AB569 is to be administered to patients as a nebulized (inhaled) solution or powder.
http://tinyurl.com/y8p6xb4t
AND
http://tinyurl.com/y9y96aqm
AND
http://tinyurl.com/y8zycbna
AND
http://tinyurl.com/y9r4e8nu

USFDA Approves Proteostasis's Triple Combination Program For CF

Proteostasis Therapeutics' proprietary triple combination includes a novel cystic fibrosis transmembrane conductance regulator (CFTR) amplifier, third generation corrector and potentiator, known as PTI-428, PTI-801 and PTI-808, respectively. The FDA's Fast Track program is designed to facilitate the development and expedite the review of new drugs that are intended to treat serious or life-threatening conditions and that demonstrate the potential to address unmet medical needs.
http://tinyurl.com/ydz42c9p

Improved Treatment Targeting Root Cause Of Cystic Fibrosis May Be On Horizon

A clinical trial led by a pharmaceutical company has shown that a combination of two medications provides significant clinical benefit in a subgroup of patients with cystic fibrosis (CF). Using knowledge of the different CFTR mutations, scientists have developed candidate medicines that may help restore sufficient CFTR function to partially alleviate CF symptoms. The current study tested a combination of ivacaftor and a promising new (not yet FDA-approved) CFTR-stabilizing drug, tezacaftor, in people who have one copy of Phe508del and one copy of a residual-function CFTR mutation. The researchers found that the amount of air that participants were able to exhale per second — a standard measure of lung function in people with CF — rose by an average 4.7 percentage points with ivacaftor treatment alone, and 6.8 percentage points with the combined ivacaftor-tezacaftor treatment, compared to a placebo.
http://tinyurl.com/y8846h44

CF Study Identifies Possible Shortcoming Of Orkambi's Dual-component Approach

The two components of Orkambi act separately to trigger a liver enzyme that decreases the therapy's ability to fight cystic fibrosis, a study reported. One concern the researchers found is that the therapy's combination strategy may be limited due to antagonistic drug-drug interactions. The team has been looking at how ivacaftor and lumacaftor work in the body, as well as the workings of a CFTR regulator called tezacaftor (VX-661). They found that ivacaftor is broken down into two main components in the liver — an active metabolite called hydroxymethyl-ivacaftor (M1) and an inactive one called ivacaftor-carboxylate (M6). The enzymes CYP1A2 and CYP3A4 play the main role in breaking down ivacaftor. The key finding was that ivacaftor-M6 and lumacaftor trigger the CYP3A4 liver enzyme. This leads to patients who receive Orkambi having significantly less ivacaftor in their system, reducing the treatment's effectiveness against CF.
http://tinyurl.com/y9o49lrl

ProQR Announces Presentations At

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ECFS On Eluforsen For F508del Cystic Fibrosis And At CFF Research Conference

Eluforsen, formerly known as QR-010, is a first-in-class RNA-based oligonucleotide designed to address the underlying cause of the disease by targeting the mRNA in CF patients that have the F508del mutation. Eluforsen is a single agent designed to bind to the defective CFTR mRNA and to restore CFTR function. Eluforsen is designed to be self-administered via an optimized eFlow® Nebulizer.
http://tinyurl.com/ycv2r2er

AmpliPhi Biosciences Announces Presentation Of AB-PA01 Bacteriophage Therapy Case Study At 41st European Cystic Fibrosis Conference

The presentation describes the case of a 26-year-old patient with cystic fibrosis (CF) listed for a double lung transplant, who developed multiple episodes of multi-drug resistant (MDR) P. aeruginosa pneumonia and had multiple CF exacerbations. Prior to treatment with AB-PA01, the patient received multiple courses of antibiotics, including colistin, but due to renal failure, colistin administration was discontinued. The U.S. FDA granted an emergency IND to administer AmpliPhi's AB-PA01 as an adjunctive treatment to systemic antibiotics. AB-PA01 was administered via intravenous route every six hours for eight weeks. Treatment with AB-PA01 was well tolerated and the patient's infection resolved. No recurrence of pneumonia or CF exacerbation was reported during the two-month follow-up period after the completion of treatment with AB-PA01. The patient's renal failure resolved. In this case study, bacteriophage therapy has demonstrated to hold much promise for patients with cystic fibrosis suffering from multidrug-resistant pulmonary bacterial infections.
http://tinyurl.com/yahwfxgp

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

Results of a Phase 2 trial showed that treatment with Spyryx's investigative drug SPX-101 can improve lung function in patients with cystic fibrosis, regardless of their background genetic mutation causing the disease. SPX-101 is a small protein fragment that was developed to target epithelial sodium channels (ENaC) in the lungs and prevent them from taking up sodium. The drug reduces sodium absorption, allowing fluids to be retained on the airway surface, making mucus clearance an easier task. The effectiveness and safety of SPX-101 is currently being evaluated in the HOPE-1 trial (NCT03229252). During the trial, SPX-101 was found to be safe and well-tolerated. The most common adverse events reported were increased sputum production and cough.
http://tinyurl.com/ycbj3zmu

FYI

Prevalence Of Severe Fatigue Among Adults With Cystic Fibrosis: A Single Center Study. Merel M. Nap-van der Vlist, Marcella Burghard, H.J. Hulzebos, Wytze R. Doeleman, Harry G.M. Heijerman, Cornelis K. van der Ent, Sanne L. Nijhof. Journal of Cystic Fibrosis. May 2018Volume 17, Issue 3, Pages 368–374

With life expectancy increasing among patients with cystic fibrosis (CF), the prevalence of complications such as fatigue is also expected to increase. The aim of this study was to investigate the prevalence of severe fatigue among adults with CF and to identify factors associated with fatigue. Fatigue is a clinically important and highly prevalent issue among adults with CF and is associated with a significant reduction in health-related quality of life and participation in society. In addition, fatigue is associated more strongly with the patient's perception of symptoms and well-being than with clinically measured parameters.
http://tinyurl.com/y95nb42d

A Prospective Analysis Of Unplanned Patient-initiated Contacts In An Adult Cystic Fibrosis Centre. Espérie Burnet, Dominique Hubert, Isabelle Honoré, Reem Kanaan, Rosewilta Panzo, Pierre-Régis Burgel. Journal of Cystic Fibrosis. Article in press.

Timely response should be provided when patients contact the cystic fibrosis (CF) center in between scheduled visits. Little data exist on unplanned patient-initiated contacts in CF adults. Answering unplanned patient-initiated contacts represented a significant workload for CF caregivers. Increased disease severity was associated with high contact frequency.

In Memory

Marcia Klucas Moes, 40
Watertown, SD
Died on February 6, 2018

Stephanie Devine Rath, 49
Brownsburg, IN
Died on April 7, 2018

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:
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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. March 2018 .Volume 17, Issue 2, Pages 204–212

The authors compare bacterial communities of matched sinus and lung mucus samples from cystic fibrosis (CF) subjects undergoing endoscopic surgery for treatment of chronic sinusitis. The findings indicate that while the lung may be seeded by individual sinus pathogens, airway microenvironments harbor distinct bacterial communities that should be considered in selecting antimicrobial therapies.
http://tinyurl.com/y6vtqafe

The Effect Of Enteral Tube Feeding In Cystic Fibrosis: A Registry-Based Study. Denis Libeert, Dimitri Declercq, Simeon Wanyama, Muriel Thomas, Sabine Van daele, Frans De Baets, Stephanie Van Biervliet. Journal of Cystic Fibrosis. March 2018. Volume 17, Issue 2, Pages 264–270

Long-term effect of enteral tube feeding (ETF) in cystic fibrosis (CF) remains equivocal.

ETF introduction improved BMI z-score and stabilized FEV_1, was associated with fewer hospitalizations and IVAB treatments. Higher mortality and transplantation in the ETF cases, leading to drop-outs, made determination of the effect size difficult.
http://tinyurl.com/ybkupb5t

Attention Deficit Hyperactivity Disorder Symptoms In Patients With Cystic Fibrosis. Malena Cohen-Cymberknoh, Tzlil Tanny, Oded Breuer, Hannah Blau, Huda Mussaffi, Diana Kadosh, Silvia Gartner, Alma Salinas, Lea Bentur, Vered Nir, Michal Gur, Joel Reiter, David Shoseyov, Eitan Kerem, Itai Berger. Journal of Cystic Fibrosis. March 2018Volume 17, Issue 2, Pages 281–285

Cystic fibrosis (CF) is a chronic life-threatening disease. In patients who suffer from chronic disease, Attention Deficit Hyperactivity Disorder (ADHD) is associated with functional impairment that can affect adherence to treatment and consequently influence prognosis. The occurrence of ADHD symptoms in patients with CF is substantially higher than in the general population and should be recognized as a co-morbidity of CF. As ADHD can impair adherence to therapy, further research is needed to investigate the effect of ADHD therapy on adherence.
http://tinyurl.com/y7em42tl

Risk Of Gastrointestinal Cancers In Patients With Cystic Fibrosis: A Systematic Review And Meta-analysis. Akihiro Yamada, MD, Yuga Komaki, MD, Fukiko Komaki, MD, Dejan Micic, MD, Samantha Zullow, MD, Atsushi Sakuraba, MD. The Lancet Oncology. Published: 26 April 2018

The management and life expectancy of patients with cystic fibrosis have improved substantially in the past three decades, which has resulted in an increased number of these patients being diagnosed with malignancies. The aim of this study was to assess the risk of gastrointestinal cancers in patients with cystic fibrosis. Results suggests that patients with cystic fibrosis had a significantly increased risk of gastrointestinal cancer compared with the general population, including small bowel, colon, biliary tract and pancreatic cancers. These findings highlight the need to develop individualized screening strategies for site-specific gastrointestinal cancers in patients with cystic fibrosis.
http://tinyurl.com/ya7zbk7a

TREATMENT

Lumacaftor/Ivacaftor In Patients With Cystic Fibrosis And Advanced Lung Disease Homozygous For F508del-CFTR. Jennifer L. Taylor-Cousar, Manu Jain, Tara Lynn Barto, Tarik Haddad, Jeffrey Atkinson, Simon Tian, Rui Tang, Gautham Marigowda, David Waltz, Joseph Pilewski on behalf of the VX14-809-106 Investigator Group. Journal of Cystic Fibrosis. March 2018. Volume 17, Issue 2, Pages 228–235

The objective of this investigation was to evaluate the safety, tolerability and efficacy of lumacaftor/ivacaftor in

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Paul Albert
Catasauqua, PA

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patients with cystic fibrosis (CF) with severe lung disease. Compared with patients with higher lung function, respiratory events were more common in patients with ppFEV$_1$ < 40; aside from these events, the lumacaftor/ivacaftor safety profile was consistent with previous studies. Results suggest that patients with ppFEV$_1$ < 40 may benefit from treatment initiation at a lower dose with augmented monitoring before increasing to the full dose.
http://tinyurl.com/ya54xx2e

Data From The US And UK Cystic Fibrosis Registries Support Disease Modification By CFTR Modulation With Ivacaftor. Leona Bessonova, Nataliya Volkova, Mark Higgins, Leif Bengtsson, Simon Tian, Christopher Simard, Michael W Konstan, Gregory S Sawicki, Ase Sewall, Stephen Nyangoma, Alexander Elbert, Bruce C Marshall, Diana Bilton. Thorax. May 23, 2018

Ivacaftor is the first cystic fibrosis transmembrane conductance regulator (CFTR) modulator demonstrating clinical benefit in patients with cystic fibrosis (CF). As ivacaftor is intended for chronic, lifelong use, understanding long-term effects is important for patients and healthcare providers. This ongoing, observational, post-approval safety study evaluates clinical outcomes and disease progression in ivacaftor treated patients using data from the US and the UK CF registries following commercial availability. Analyses revealed favorable results for clinically important outcomes among ivacaftor treated patients, adding to the growing body of literature supporting disease modification by CFTR modulation with ivacaftor.
http://tinyurl.com/yb94dyfu

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. March 2018. Volume 17, Issue 2, Pages e20–e21

Studies in cystic fibrosis (CF) patients have reported reduced rates of pulmonary exacerbation and hospitalization with probiotic use. Furthermore, 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/y8yt8xlx

Colistin Resistance In Pseudomonas aeruginosa And Achromobacter spp. Cultured From Danish Cystic Fibrosis Patients Is Not Related To Plasmid-mediated Expression Of mcr-1. Maya G. Pedersen, Hanne V. Olesen, Søren Jensen-Fangel, Niels Nørskov-Lauritsen, Mikala Wang. Journal of Cystic Fibrosis. March 2018. Volume 17, Issue 2, Pages e22–e23

The emergence and global spread of a new plasmid-mediated resistance mechanism to colistin, the mcr-1 gene, can have significant implications for the treatment of pulmonary infections in cystic fibrosis (CF) patients. The polymyxins, colistin (polymyxin E) and polymyxin B, are antimicrobial peptides with broad-spectrum activity against Gram negative bacteria, including common CF pathogens as Pseudomonas aeruginosa and Achromobacter species and are frequently administered as inhaled antimicrobial therapy to CF patients. A transfer of colistin resistance to multidrug-resistant CF pathogens would limit treatment options.
http://tinyurl.com/y775pg9u

Treatment Of Chronic Rhinosinusitis With Dornase Alfa In Patients With Cystic Fibrosis: A Systematic Review. Gopi B. Shah MD, MPH Linde De Keyzer MD Joy A. Russell PhD, MLS Ashleigh Halderman MD.

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International Forum of Allergy & Rhinology Volume 0, Issue 0. 11 January 2018

A major component of sputum in cystic fibrosis (CF) patients is polymerized DNA, a byproduct of degraded neutrophils. Dornase alfa (dornase) selectively cleaves extracellular DNA and reduces the viscosity of sputum. It improves mucociliary clearance and pulmonary function. The benefit of dornase on CF-associated sinusitis is less clear. Therefore, the objective of this study was to systematically review the use of dornase on chronic rhinosinusitis (CRS) in CF patients. Findings indicate that topical intranasal dornase appears to improve sinonasal symptoms in CF patients to a greater degree than saline alone.
http://tinyurl.com/y7umqqpr

Treatment Compliance In Cystic Fibrosis Patients With Chronic Pseudomonas aeruginosa Infection Treated With Tobramycin Inhalation Powder: The FREE Study. Francesco Blasi, Vincenzo Carnovale, Giuseppe Cimino, Vincenzina Lucidi, Donatello Salvatore, Barbara Messore, Marta Bartezaghi, Elisa Muscianisi, Pasquale Alberto Porpiglia. Respiratory Medicine. May 2018. Volume 138, Pages 88–94

A high treatment burden with neb-

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ulized therapies in cystic fibrosis (CF) patients is the major limitation for treatment compliance; moreover, studies on treatment compliance with inhaled antibiotics are limited. This study assessed compliance to TOBI® Podhaler™ (TIP) treatment in CF patients with chronic Pseudomonas aeruginosa (Pa) infections in a real-world setting. The authors concluded that TIP was convenient to use and led to improved treatment adherence in CF patients with chronic Pa-infection.
http://tinyurl.com/ycgwtddx

CFRD

Prevalence Of Hypoglycemia During Oral Glucose Tolerance Testing In Adults With Cystic Fibrosis And Risk Of Developing Cystic Fibrosis-related Diabetes. Lisa A. Mannik, Kristy A. Chang, Pascalyn Q.K. Annoh, Jenna Sykes, Julie Gilmour, Ronalee Robert, Anne L. Stephenson. Journal of Cystic Fibrosis. Article in press

Hypoglycemia in cystic fibrosis (CF) patients during the oral glucose tolerance test (OGTT) has been reported. Few studies have examined whether hypoglycemia during the OGTT increases the risk of developing CF-related diabetes (CFRD). Objectives of this study were to describe the characteristics of CF patients with hypoglycemia during the OGTT and to determine the incidence and time to development of CFRD in those with hypoglycemia. Results show that hypoglycemia following OGTT is common in CF; however, the 10-year risk of developing CFRD in these patients was low. Males and those who were heterozygous deltaF508 were at higher risk for hypoglycemia.
http://tinyurl.com/yccqp4nd ▲

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