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

Expanded Access Programs — A Lifeline For Some

By Jeannine Ricci

In November 2014, I was able to access off-label Kalydeco and have enjoyed much success with it. The immense gratitude I felt when I realized Kalydeco was working could only be matched by my urgent need to help others in my similar situation, those who weren't included in clinical trials but who had the potential to benefit greatly from Kalydeco. My biggest worry was that this drug, which had a well-established safety profile, would not be available to someone who was in dire need of it, despite evidence of its potential efficacy for those with residual function mutations. This was unacceptable to me. Lives were at stake. I could not find any indication that there was a compassionate use program offered by Vertex, so over the next two years, I focused my efforts on increasing awareness of off-label prescribing. Little did I know that there was a compassionate use program available (also called expanded access program) for some residual function mutations, but its existence wasn't well known among either patients or CF physicians. This past fall, when I discovered this, I questioned how such a vital program could be so elusive.

As I researched, it became evident that many others with serious and life-threatening illnesses shared my frustration with decreased awareness of expanded access programs. Certain provisions have been made in the past to increase awareness. For instance, in 2007 it became law that when a drug company is registering a clinical trial on clinicaltrials.gov, they are required to specify whether there is an expanded access program (EAP) for the investigational drug available for those who do not qualify for enrollment and how to obtain information about such access. However, even with this rule in place, EAPs remain elusive, leaving some patients feeling as if their rights to be an active, informed participant in their healthcare are being disregarded.

In response to rising concerns of patients and advocacy groups regarding this lack of transparency, legislative action has recently been taken. On December 13, 2016, President Obama signed the 21st Century CURES act into law. This law supports many healthcare and research initiatives, among them expanded access to experimental drugs. A pharmaceutical company of an investigational drug must now make public and readily available its policies on evaluating requests for expanded access, such as posting such policies on a publicly available Internet website. The company must include a hyperlink or other reference to the clinical trial record containing information about the EAP. It is important to note that this legislation does not

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EDITOR'S NOTES

Hoorah! Hooray! Spring has arrived! Can you tell that I am happy that winter finally is over? I believe that this was a fairly difficult winter for most of us and we feel fortunate to have survived another one. Wahoo!

We have some sad news to share, former USACFA Director and President, **Paul Feld**, died December 17, 2016. You may read his obituary on page 39 to get more information. He was such an important part of our USACFA family and he is sorely missed.

Now for some happier USACFA news. The Board of Directors has grown by three. You may read about **Lauren Jones Hunsaker** on page 12, **Brennen Reeves** on page 12 and **Jeannine Ricci** on page 13. We welcome all three of them and are happy that they have joined the group.

If you already have read the front page, you know that one of the new Directors, Jeannine Ricci, has written about expanded access programs for some of our expensive medications. **Beth Sufian** in "Ask The Attorney" addresses the expected change from the Affordable Care Act to the American Health Care Act and how we can try to get the politicians in Washington to protect the portions that make such a difference in our lives. She also answers a couple of questions regarding Medicare and Medicaid.

Once again, **Laura Tillman** has compiled a great amount of "Information From The Internet" with press releases and news of research. **Piper Beatty Welsh** reviews *Lessons from a CF Cornerman: 38 Lessons I Learned During My Wife's Illness and Lung Transplant*, a new book by Raymond L. Poole.

In "Voices From The Roundtable" **Anna Payne** writes of why it is so important for us to use our voices and to speak up for ourselves and our community. **Devin Wakefield** writes of the CFRI retreat and invites all of us to attend. **Ella Balasa** encourages us to keep a positive attitude as our CF progresses.

Our Focus topic is Traveling With CF. **Jeanie Hanley** discusses using oxygen on airplanes. **Sarah Albright** tells of her special masks and other things she uses to make travel safer. **Andrea Eisenman** discusses keeping lists to help remember all that we need to make a trip enjoyable and safe. **Nicole Kowal** relates her travels with a lot of baggage. **Isabel Stenzel Byrnes** tells about travels in our minds in "Spirit Medicine." In "Speeding Past 50" I reminisce about trips I made when I was young and how we can make travel easier.

Emily Kramer Golinkoff is "In The Spotlight," where she answers questions about herself and CF. **Aimee Lecointre** uses "Active For Life" to talk of exercise and travel. In "Parenting," **Dana Giacci** shares ways to keep kids engaged when you are sick.

Be sure to check out the new USACFA scholarship winners, **Diana Wasserman** and **Grace Knight**, on page 11. Also, page 44 and page 45 have information about the CFRI Conference and Retreat.

Stay healthy and happy.

Kathy

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Information From The Internet...

Compiled by Laura Tillman

PRESS RELEASES

CF Patients' Kidneys Not Harmed by Once-daily Tobramycin Treatment, Study Says

Once-a-day treatment with the antibiotic tobramycin does not harm the kidneys of patients with advanced cystic fibrosis (CF), a new study has found. Physicians often prescribe tobramycin and other aminoglycosides to control P. aeruginosa, but when taken three times a day – as was often the case in the past – they reportedly caused acute and chronic kidney failure. Today, a once-daily treatment is more common, and seems to be just as effective and less toxic to the kidneys. Previous reports regarding the use of aminoglycosides three times a day in CF patients showed links between treatment and kidney injury, but P. aeruginosa infection itself may also cause kidney failure, the researchers said. A more recent study showed no such links. This study is the first to analyze kidney function in CF patients exclusively treated with tobramycin once a day. Its findings indicate that even frequent treatment with tobramycin does not necessarily lead to kidney injury in CF patients.
http://tinyurl.com/zpg99fj

Many Cystic Fibrosis Patients Have C. Difficile Infections Without Symptoms, Study Says

The bacterial species Clostridium difficile is often found in the gut of cystic fibrosis (CF) patients, but rarely makes them ill. In those instances, atypical symptoms can delay diagnosis and proper treatment, and may put patients' lives in jeopardy. CF patients have asymptomatic C. difficile in the gut more often than healthy people or those hospitalized for other reasons. It is not clear why the bacteria make some patients ill, but the researchers said several factors could be at play. People with CF have more bacterial growth in the gut, so it's possible other species counteract the effects of the Clostridium bacteria. It is also possible that CF patients' more acidic guts are a less-than-optimal environment for the disease to develop.
http://tinyurl.com/znnebac

Pregnancy in CF Patients Raises Risk of Diabetes and Premature Delivery, Large Study Reports

Pregnant women with cystic fibrosis (CF) are more likely to develop diabetes and deliver prematurely by cesare-

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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**

Spring (current) 2017: Traveling With CF.

Summer (August) 2017: Problem-Solving With CF. (Submissions due June 15, 2017.) How do you go about solving a problem related to having CF? Whom do you ask or with whom do you discuss it? What helps you make decisions? Do you feel you are a good problem-solver because of your CF?

Autumn (November) 2017: Dating And Relationships With CF. (Submissions due September 15, 2017.) Do you have any good advice for us about dealing with others when it comes to your CF? When do you tell someone that you have CF?

Winter (February) 2018: Becoming A Parent With CF. (Submissions due December 15, 2017.)

ASK THE ATTORNEY

Medical Records And Increase In Continuing Disability Reviews

By Beth Sufian, J.D.

In the past three months *CF Roundtable* readers have asked for an assessment of Congress's attempts to repeal the Affordable Care Act. Most people in the CF community are very concerned about the United States Congress and its attempts to repeal the Affordable Care Act. These are scary times. However, we cannot put our heads under our pillows and say it is just too upsetting to be engaged.

We must all stay informed about efforts by our government to deprive our community of the health insurance we need to live longer and healthier lives. Every person with CF has benefitted from at least one section of the ACA, even if they do not realize it.

Current proposals in Congress keep a few sections of the ACA but drastically change or delete other sections. Even if only certain parts of the law are repealed, many people with CF will suffer. For example, if the subsidies that thousands of people with CF receive to help pay monthly insurance premiums are no longer available, then thousands of people with CF will not be able to afford health insurance coverage.

If Medicaid is no longer available to adults who have low household incomes, then thousands with CF will lose Medicaid coverage. Currently, under the ACA Medicaid expansion, 32 states offer Medicaid to adults who meet low income criteria. Many years ago the U.S. Supreme Court held that the ACA's mandate of Medicaid expansion to low income adults could only be voluntary and that states could not be forced to expand Medicaid. After this decision, 18 states did not expand Medicaid to low income adults.

In those 18 states adults who have low income cannot access Medicaid unless they are also receiving SSI benefits. Prior to the enactment of the ACA, in most states the only way adults over the age of 18 could obtain Medicaid coverage was if the adult also received SSI benefits. In addition, women who were pregnant and had low income could be eligible for Medicaid but only during their pregnancy and for a short time afterward.

Children under the age of 18 who live in low income households are eligible for Medicaid. The ACA did not affect Medicaid benefits provided to children living in a low income household. Repeal of Medicaid expansion will put the CF community back where we were before passage of the ACA in the 32 states that did expand Medicaid to low income adults.

I am writing this column on March 15, 2017. It is unclear if the American Health Care Act will be approved by the House of Representatives. If the House passes the bill it is unclear if the Senate will vote to approve the law.

BETH SUFIAN

People with CF should continue to let their representatives know the importance of access to Medicaid, including premium subsidies, no caps on coverage, no pre-existing condition clauses, limiting the ability of a dependent child to stay on a parent's policy until age 26 and the right to have an independent review of an insurance company's denial of coverage for medical treatment. In addition, the ACA mandates all health insurance policies provide coverage for 45 medical services. These are just some of the sections of the Affordable Care Act that have helped people with CF. The ACA also has sections that help Medicare recipients.

The CF Foundation has a section of its website that helps you send e-mails to Congress. Go to: act.cff.org for more information. Many legislative experts say that calling your Congressman's office is also effective. Often it is hard or impossible to reach your representative's office in Washington, DC. However, it is usually easy to reach their local office, often called a satellite office. It is important to express your concern about efforts to make it harder for people with CF to access health insurance. Go to: https://www.usa.gov/elected-officials to access a list of phone numbers for your Congressional representatives.

In just the past few days, members of Congress have proposed changes to the proposed American Health Care Act. These proposed changes are based on advocates who told Congress the bill needs to provide more help to older Americans. Congress is listening to those who speak out. Your voice can

make a difference.

A few people tell me they have good insurance through their employer and so they are not worried if the ACA is repealed. Many sections of the ACA apply to ALL health insurance policies. For example, the ACA mandated an independent review of an insurance company's denial of coverage for medical treatment or services. The ability to have an independent review has helped many with CF who have had coverage for transplants, medications or IV treatment denied. This section of the ACA applies to all health insurance policies.

In addition, the most vulnerable people in the CF community are those who live in poverty and rely on Medicaid as their sole source of insurance coverage. Those of us who are lucky enough to have good jobs or helpful parents need to speak up for their needs. We are a strong community of caring individuals and our fellow brothers and sisters who have CF need us to raise our voices on their behalf.

During such uncertain times we can find hope in many places. First, we can all find comfort in knowing that there are scientists in laboratories across the country and around the world who are working very hard to bring us new and better medications to treat CF. I am very grateful for their efforts.

Second, we can find comfort knowing there are many organizations working to help us. I am grateful for Dr. Preston Campbell who is leading the CF Foundation at such an important time. He cares not just for the fortunate members of our community. He also cares about those with CF who have very little and face great hardship. Dr. Campbell's efforts to do what is right for every person with CF are an inspiration to many.

> *"We must all stay informed about efforts by our government to deprive our community of the health insurance we need to live longer and healthier lives."*

Sue Landgraf, who leads the CFRI, is also making sure that various needs of the CF community are met. For example, CFRI offers funds to help people access counseling services and offers an excellent course on mindfulness led by the amazing Dr. Julie Desch, an adult who has CF who gives back to her community every day. CFRI also funds research and support groups for those living with CF or who have a family member with CF.

Boomer Esiason and the Boomer Esiason Foundation address a number of unmet needs in the community, by providing transplant grants to those in need, offering educational scholarships and raising money for research. There are also so many other smaller groups that provide help to people with CF.

None of us has a crystal ball to predict what will happen in Congress but we should find some comfort in knowing that there are many who are working to help the CF community. Our energy and efforts are needed to continue to help each other. Writing an article for *CF Roundtable* or becoming involved with the organization is a good way to impact the lives of others in a positive way.

Two other questions were posed by readers.

1. I have Medicare and I need to buy a Medicare supplement policy to pay for the Part B 20 percent co-pays. However, every time I apply for a policy I am turned down. Do they have to sell me a Medicare supplement policy?

Answer: Approximately half of all states do not require an insurance company to sell a Medicare supplement policy to a person who has Medicare because he is also eligible for Social Security Disability Insurance (SSDI) benefits. In states where there is no requirement that an insurance company sell such a policy to a person who receives Medicare because he receives SSDI benefits, it is difficult (and in many states impossible) to find an insurance company who will sell such a policy. Medicare Part B is the section of Medicare that provides coverage for outpatient services, such as physician office visits, blood work, X-rays etc. Medicare Part B also provides coverage for a limited number of prescription medications.

Medicare Part B pays 80 percent of the allowable charges and the Medicare recipient must pay the other 20 percent. If a person has a Medicare Supplement policy, that policy will typically pay the 20 percent portion that Medicare does not pay. There is no cap on how much a person can pay out of pocket in terms of the 20 percent cost share under Medicare Part B.

Most CF medications that are covered under Medicare Part B will have patient assistance programs (PAP) that may provide assistance paying the 20 percent co-pay for the medication. However, most PAPs have income eligibility guidelines. Those individuals whose household income is over the allowable amount will not be eligible for help from the PAP. Each PAP has its own household income eligibility guidelines. There are a few drug companies that will NOT provide co-pay assistance to a person on Medicare.

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SPIRIT MEDICINE

Mental Travels

By Isabel Stenzel Byrnes

The theme of this issue is traveling when you have CF. I could share many stories about global traveling. But I'm confident this issue will have plenty of stories from other jet-setters. I am so grateful that people with CF are living healthier and longer lives, allowing them to do "normal" things like travel. However, I'd like to ponder the kind of travel that people with CF can do when they don't have the money, time, health or opportunity to venture out into the world to exotic far off places.

What does traveling do for us? It gives us a change of scenery from our ordinary lives. We get to witness the vast natural beauty, diverse cultures, social bonds, riches and poverty, and joy and suffering that exists in the world. It opens up our perspectives, as we notice what we have easier and what we have harder. It changes our minds: to a "beginner's mind" as they say in Buddhism, where the newness of everything around us can inspire a sense of wonder, excitement, curiosity, friendliness and awe.

When we want to travel but can't, for whatever reason, it is natural to feel disappointment. So often, our minds dwell on what "can't" be done, rather than what "can" be done. It's important to ask what we can do to experience the closest thing to real travel. Maybe, because of my immune system, I can never go to East Africa, but I can certainly go to the zoo or to our local attraction, "Safari West," to see the large herd animals of the Serengeti. Maybe I can't go to the Amazon, but I can watch "Survivor: The Amazon" and live vicariously through the people in the show! In fact, there are many ways to enjoy travel without travel: films, video games and virtual reality, tripping out on drugs, reading and so on. But in this issue of "Spirit Medicine," I'd like to ponder the spiritual kind of travel: mental travels.

Our minds are our most valuable plane ticket. Our minds provide endless imagination, fantasy, myth and metaphor, taking us out of this reality on a moment by moment basis. Children are our best teachers! Even when we are asleep, our minds provide us a steady stream of dream imagery, so that when we awake, we wonder where we have just been! We are the only animals that can think about our own thinking. Our minds can view our lives as a great travel itinerary. We can ask big picture questions like, "What's ending so I can begin?" or What's at the end of this journey?" or "Where am I going?"

ISABEL STENZEL BYRNES

Living with CF can present us with physical discomfort, loneliness, uncertainty and numerous disappointments. CF may be a terrible disease, but at least we are cognitively intact. Our minds are our greatest resources to cope with these losses. An active imagination gives us respite from the mundane suffering of CF and of life in general. But what if escapism or avoidance is unhealthy? Since I'm a grief counselor, I've studied the "dual process model" of grief, developed by Stroebe and Schut. They found that healthy grieving means engaging in a dynamic process of oscillating between a focus on the loss and a focus on "restoration" or life. In other words, a griever will oscillate between confronting the loss and avoiding the loss. So, I'd extrapolate and say that it's okay, and in fact healthy, to let our minds oscillate between reality and our fantasies, as long as we don't get stuck in one place.

I'd like to share one of my favorite stories about the power of the mind to overcome suffering. In "Man's Search for Meaning," Viktor Frankl described a miserable scene: he and other men were forced to walk in darkness, over wet and rough roads, to another concentration camp, with Nazi guards shouting at them and hitting them with their rifles, for miles. He writes, "Hiding his mouth behind his

upturned collar, the man marching next to me whispered suddenly: 'If our wives could see us now! I do hope they are better off in their camps and don't know what is happening to us.'

"That brought thoughts of my own wife to mind. And as we stumbled on for miles, slipping on icy spots, supporting each other time and again, dragging one another up and onward, nothing was said, but we both knew: each of us was thinking of his wife. Occasionally I looked at the sky, where the stars were fading and the pink light of the morning was beginning to spread behind a dark bank of clouds. But my mind clung to my wife's image, imagining it with an uncanny acuteness. I heard her answering me, saw her smile, her frank and encouraging look. Real or not, her look was then more luminous than the sun which was beginning to rise.

"A thought transfixed me: for the first time in my life I saw the truth – that love is the ultimate and the highest goal to which Man can aspire ... I understood how a man who has nothing left in this world still may know bliss, be it only for a brief moment, in the contemplation of his beloved. In a position of utter desolation, when Man cannot express himself in positive action, when his only achievement may consist in enduring his sufferings in the right way – an honorable way – in such a position Man can, through loving contemplation of the image he carries of his beloved, achieve fulfillment."

Wow. You have just traveled with Viktor to this miserable scene of his life during the Holocaust, but you have just also just seen how his contemplation of his wife brought him love, joy, comfort and affection in this very dark moment. If he could do this, we surely can with our struggles with CF.

Sometimes, the more we struggle, the more active our minds can become. When I was critically ill, my mind was vibrant and I wrote poem after poem. I'd like to share "Mental Travels," written three weeks before my transplant:

In my hospital room I am staring at the ceiling at night.
Two weeks here is enough and I yearn for the outside world.
Lonely but surrounded by people,
Free of responsibility but imprisoned,
In my fear I seek refuge.

My mind is my solace and I travel far far away from this place.
In the dark I hear the loud bubbling of my oxygen humidifier
And I embark on a captivating deep sea journey
Where I am scuba diving, looking for mermaids and bottom dwelling creatures
Colorful fish tickle my feet as I glide past them going deeper and deeper
The pressure of the deep water squeezes my chest
So I breathe quickly and heavily while tethered to my gear
And I gasp for air as I swim to the surface.

I open my eyes and am surrounded by metal and hard, artificial surfaces
There are no organic smells nor any life around
I hear the buzzing of machines and pipes in the walls
And I realize I am in a space ship
Traveling the galaxy for thousands of miles from my homeland Earth
The black box above my head is blank
"Houston, can you read me?"
I mutter with no response.
I am floating, disconnected from everything.
My bed is my command station with lights that blink and buttons that change my position
Finally, I push the red one and hear from the Mother Ship, "Can I help you?"
And through the closed blinds at the window I see a bright light
I think it's a star I must be approaching
The stars are brilliant from this vantage point
Then I remember it's just another passenger in her spaceship across the wing with her lights on.

I open my eyes to a bright light overhead, the kind I saw once when my body almost left this world.
Its brilliance invited me to a safe place of comfort and peace
And I follow it with wonder.
Soon I am surrounded by this light
And I see Jesus and he comforts me and we smile at each other.
He reaches for me and lays his gentle hands on my chest
I feel tremendous warmth and healing
He says it will be okay.
I blink and I see him and it's Charles, my wonderful respiratory therapist
And he's laughing and I'm laughing and he's laying his hands on me
I feel his healing.

My reality and fantasy are blended.
In my fantasy there is fear and loneliness.
In my reality there is goodness and refuge.

What a trip! I hope you, too, can embark on your own mental travels when this helps you deal with the stressors of life and CF. Be your own pilot! At the end of my life, I may not remember the details of my trips to Costa Rica, Japan, Peru, Germany or Italy, but I will feel complete knowing that I used my mind to the best of its ability to help me live the best life possible – no matter the circumstances. ▲

Isabel is 45 and has CF. She lives in Redwood City, CA, with her husband, Andrew. She is 13 years post-lung transplant.

SPEEDING PAST 50

Travel Creates Great Memories

By Kathy Russell

I was looking back at an issue of *CF Roundtable* from 20 years ago in which I had written about traveling with CF. The first couple of lines caught my eye. I said, "Traveling is one thing I really enjoy." That was so true, then. Not so much, now. I went on to say that I had "flown in excess of 500,000 miles on pleasure trips. Most of those miles proved to be a lot of fun." I remember many of those trips and they were a lot of fun. I flew another 100,000 miles before I had to cut down on travel.

I used to love to travel. When I was young and didn't have a lot of medical equipment to haul around, travel was so easy. My husband worked for a major airline and we had excellent flight benefits. We took advantage of them. We live on the West Coast and his folks lived in NYC, so we made a lot of cross-country trips. Those flights were anywhere from four-and-a-half hours to as much as seven hours in duration. I had no trouble dealing with such long flights. I didn't tire as easily as I do now and could get by on much less sleep than I now need.

We could (and did) make quick round-trips to San Francisco, frequently. I remember one time when we didn't have a lot of ready cash that wasn't spoken for in our budget. We really wanted to do something fun, but couldn't think of anything that we could afford that appealed to us. We hit on taking a flight to San Francisco for lunch. In those days, airline food was quite good and we usually were able to go first class. We came home on a dinner flight, after an afternoon of window shopping. The dinner was excellent, too. So we had a wonderful day with two good meals and all for only the price of an employee's pass fee.

Of course, this was long before the present type of security measures had been started so it was easy to run out to the airport and just hop on a plane. We didn't have to get to the terminal two hours before flight time. There was more than one time where we walked into the terminal only a few minutes before a trip was to leave and had no trouble making the flight. That would be very hard to accomplish today.

Now, even if the trip were free, I couldn't handle such trips. Having to wait in long security lines is very exhausting, even if I use a wheelchair to get to the gate. I get too tired so quickly. It just wouldn't be worth the energy expense.

> *"I always carry my patient profile with me, so that is not something that I have to gather before travel."*

When I want to go anyplace, and I mean anyplace at all, I have to get my portable oxygen concentrator (POC) and all the stuff to go with it. Fortunately, my POC sits in a wheeled bag that has room for extra tubing, extra batteries and my water bottle. Since I keep the tubing and batteries in the bag, I don't have to gather anything but my water bottle for it to be ready to go. Thankfully, it is approved by the FAA for use on airlines, so I can take it on airplanes. I have made only a few trips by plane since I started using continuous O_2, but on the trips I have taken things went pretty easily.

KATHY RUSSELL

Some airplanes have electrical outlets where it is possible to plug in a POC. (The battery must be removed before plugging it into the airplane.) This helps with keeping power for long periods of time, which can be a real lifesaver when there are delays or flights take longer than planned.

I won't go into all of the sanitizing and cleaning that might need to be done before sitting in an airplane seat. I know that others have written about their routines when flying. No need to duplicate information.

I have traveled by automobile, ship and train, as well as plane. They all

necessitate some kinds of preparation. My POC has adaptors for use in autos, as well as having the plug for use with electricity. That allows me to keep my batteries charged, which gives me a little more freedom to move about.

There are some basic rules to remember when traveling. The first one is to have a patient profile. I always carry my patient profile with me, so that is not something that I have to gather before travel. My profile says who I am, what medical conditions I have, what medicines I take and their dosages, allergies (especially to medicines), surgeries I have had, doctors' names and contact information, my preferred pharmacy and whom to contact in case of an emergency. You may add anything else you think would make it easier for someone who doesn't know you to treat you. Your doctor may be willing to help you create such a profile.

Make sure that you have enough of each medication to last the duration of your trip. Add a little extra in case you have to stay longer or get delayed getting home. If you are getting close to having a prescription run out, check with your physician to get the necessary prescriptions or refills. You don't want to run out of meds while you are traveling. That is a real downer.

I copied some of the following from my old article. The information still is relevant and helpful. All medicines must be in properly labeled containers. That means that prescription drugs are in containers with a prescription label attached and non-prescription drugs, such as aspirin, are in their original containers. Since many of us get medicines in large supplies, we may have to ask the pharmacist for a smaller, labeled container for travel. Many of us are in the habit of carrying pill boxes or small bottles for our daily doses. Although this may be okay most of the time, it is not a good idea when we are crossing international borders or even some state borders. Many states have adopted "zero-tolerance" policies. This means that if you are found in possession of *any* medicines that are not in properly labeled containers, you are subject to arrest. You may be able to explain to a judge that you have prescriptions for all of your medicines, but it may take a long time and in the interim you've interrupted a trip unnecessarily. The time it takes to get everything into proper containers is time well spent.

Always keep your medicines and essential equipment with you while on public transportation. For instance, keep your medicines in a small bag or backpack. Don't "check" your medicines or equipment as baggage. If they get lost, you are without your proper treatments until they are returned to you. If they get damaged, you may be "out of luck." It is important that you be in control of these essential items at all times.

Make sure all of your equipment is in good working order. You don't want to have to spend time getting equipment repaired while traveling and, even worse, you may be unable to get necessary repairs everywhere you go. If you have battery-operated pieces, make sure that the batteries are good. If you are going somewhere that the electrical plugs are different from the U.S., be sure to get the proper adaptors before you go.

Be aware of the public hygiene conditions of your destination. This may not be a problem if you are visiting relatives in the next town. However, if you are going somewhere that isn't quite up to 21st century with their plumbing facilities, you may want to be prepared. When we went to China, shortly after they had opened their borders to Americans, I was grateful that I always carry tissues in my pocket. I also had little packaged wet wipes. Since they didn't have toilet paper or soap and water for hand washing, those things came in handy.

Remember to request smoke-free accommodations and any other special needs in advance. Most hotels have rooms where smoking is prohibited. Many hotels have entire floors that are smoke-free. You can request a smoke-free room when you book your reservation. This applies to rental cars, too. Be sure to ask for a no-smoking-allowed car when you make your reservation and again when you pick up the car. All commercial airline flights are smoke free. Amtrak trains are all no smoking. Fortunately, many states have gone smoke free in public buildings. It really makes a difference.

If you need a refrigerator for medicines, be sure to request one when making your reservations. You want a refrigerator that is free, if possible. Be careful about opening the little bar refrigerators that are in some hotel rooms. Some are made to charge you just for opening the door. Check with your hotel about the type of refrigerator they have available and make sure they understand that you need refrigeration for meds. Some hotels do not charge you when it is needed for meds.

Masks, hand sanitizer and sanitizing sprays are all good things to keep with you. You never know when you will want any or all of these items. If you are able to keep your surroundings a little cleaner, you may be able to avoid catching some rotten bacteria.

All of this can sound overwhelming at first, but I guarantee that a little pre-planning can make one's travel much more enjoyable. I hope you are able to travel to many places and to have a lot of great times. ▲

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

RICCI continued from page 1

require a company to provide the experimental drug on an expanded access basis.

If you have severe disease and think you may qualify for the EAP for certain residual function mutations, discuss it with your physician to see if he/she would consider you a suitable candidate (program described below). One concern regarding expanded access programs is that they can run the risk of eliminating potential participants in clinical trials. This can be especially problematic for clinical trials for rare diseases like CF, where patient recruitment is already limited. Whenever possible, enrolling in a clinical trial is preferable because this can generate data that may lead to the approval of drugs and to broader access. Data collection from EAPs is unlikely to result in efficacy information that the FDA would consider when reviewing the drug for approval. However, the FDA doesn't exclude the possibility that data from EAPs could be used.

In the expanded access program offered by Vertex for those with certain residual function mutations, the criteria to be considered for the program are very clear. If you meet these criteria, then your low lung function or unstable disease would most likely prevent you from participating in a CF clinical trial, based on Vertex's and other sponsors' usual requirements for inclusion. Therefore, it seems unlikely that this program would significantly jeopardize clinical trial enrollment.

> "EAPs remain elusive, leaving some patients feeling as if their rights to be an active, informed participant in their healthcare are being disregarded."

Here are the details of the expanded access program offered by Vertex:

Program designed for individuals two years or older with specific residual function genotypes and severe lung disease meeting the following criteria:

- One of 23 residual function mutations: 2789+5G-> A, 3849+10kbC-> T, 3272-26A- > G, 711+3A- > G, E56K, P67L, R74W, D110E, D110H, R117C, L206W, R347H, R352Q, A455E, D579G, E831X, S945L, S977F, F1052V, R1070W, F1074L, D1152H and D1270N.
- FEV_1 of less than 40 percent predicted OR decline of 20 percent predicted in a six-month period and sustained for one month OR being evaluated for a lung transplant.

Even if you meet the above criteria, this does not guarantee that Vertex would provide your physician with a letter of authorization (LOA). If an LOA is obtained, the next steps would be for your physician to fill out an application for Individual Expanded Access and submit it to the FDA and to submit the protocol to their facility's Institutional Review Board (IRB). The FDA authorizes over 99 percent of expanded access requests it receives. Obtaining IRB approval, however, can sometimes prove to be more challenging due to time and cost constraints. Different rules exist for emergent requests of individual expanded access so that the process can be expedited.

Expanded access programs offer hope to those patients with serious or life-threatening diseases who lack other therapeutic options. It is encouraging to see that legislative action has been taken to help increase transparency of these critical programs. For those with CF, awareness of an EAP could potentially lead to access to a treatment that could prevent unnecessary suffering and loss of lung function. Ultimately, awareness of these programs can make the difference between life and death. ▲

Jeannine is 45 and has CF. She is a Director of USACFA. Her contact information is on page 2.

Cystic Fibrosis Mothers

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

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

If you would like to join our highly active and friendly community, please message Emma or Alison on one of these links:

https://www.facebook.com/emma.harris.16
https://www.facebook.com/alison.w.smith.7

USACFA Scholarship Recipients Announced

This scholarship is in honor of the late Lauren Melissa Kelly.

The United States Adult CF Association (USACFA) is proud to announce the recipients of our second **Lauren Melissa Kelly Scholarship**. In our evaluation, we look for students who demonstrate tremendous academic achievement, community involvement and a powerful understanding of how having CF matched with these achievements places them in a unique situation to gain leadership roles within the community. This round, we opened our scholarship to all pursuing any sort of degree. From associate's degrees to Ph.D.s, we at USACFA believe that any degree is a strong foundation for advocacy and involvement in CF.

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

We are pleased to announce **Diana Wasserman** and **Grace Knight** as the recipients of this year's Lauren Melissa Kelly Scholarship**.** They each will be awarded $2,500.

Diana Wasserman is a young lady who graduated high school in Wilmington, North Carolina. She is a motivated leader and volunteer in her community. Annually, she participates and fundraises for the Cystic Fibrosis Foundation Great Strides walk, and is an ambassador in chapter events. She has won many awards for her scholastic accomplishments, including having the highest AP calculus score and is the recipient of other CF scholarships. She attends the University of North Carolina at Chapel Hill and will be a sophomore in the autumn of 2017.

DIANA WASSERMAN

Grace Knight is an accomplished AP student, athlete and musician from Tyler, Texas. Not only does she excel academically, she has been on the swim, cross country and track teams her entire high school career. She also has volunteered countless hours to help raise awareness and funds for cystic fibrosis research by speaking at galas and tournaments in her community. Grace demonstrates leadership and drive in every aspect of her life. She attends the University of Pennsylvania and will be a sophomore in the autumn of 2017.

GRACE KNIGHT

Both Diana and Grace demonstrated the leadership, intelligence and drive of Lauren Melissa Kelly. We at USACFA look forward to seeing them further develop their leadership and advocacy in the cystic fibrosis community. ▲

Meet A New Director

Hello! My name is **Lauren Jones Hunsaker** and I'm 30 and have cystic fibrosis. I also have cystic fibrosis-related diabetes, chronic kidney disease and am partially blind in my left eye as a result of diabetes. I grew up in Houston, Texas, and was diagnosed with CF when I was six months old.

My family was heavily involved in the CF Foundation when I was young and I served as poster child for the Gulf Coast Chapter of the CF Foundation for three years. Prior to its closure following implementation of cross-infection precautions, I attended CF camp where I met other patients with my disease. For a time, nearly all of my friends had CF and those friendships were among the most impactful relationships I've ever had. As a practical result of cross-infection safeguards, CF can be an isolating disease and our community has to find unique ways to stay involved and connected.

I'm immensely proud to serve as a director for USACFA. *CF Roundtable* provides a vital outlet through which the CF community can engage and share experiences with each other concerning all challenges of the disease – medical, social, professional and financial.

LAUREN JONES HUNSAKER

I received a B.A. from the University of Pennsylvania and a J.D. from the University of Texas School of Law. For the past five years, I've practiced law in Texas. My first two years as a lawyer were spent at the law firm of Sufian & Passamano, LLP, where I provided advice to and represented people with CF in the areas of disability and health insurance benefits, employment and education. Beginning in 2013, I worked for a real estate firm and specialized in commercial real estate transactions. I recently retired from law, due to health, and now work remotely as a copywriter for a marketing company. I married my law school sweetheart and we live in Houston with our two dogs – a King Charles Cavalier named Shelby and a Lhasa Apso named Max. ▲

Meet A New Director

Hi folks, my name is **Brennen Reeves** and I'm 25 years old. I was born and raised on Hilton Head Island, SC, alongside my brother and my mom and dad. Just eight weeks after being born, I was diagnosed with cystic fibrosis and 19 years later, in 2011, I received a bilateral lung transplant at Duke University.

After overcoming my medical obstacles, I received a scholarship to attend the College of Charleston, where I majored in Theatre Performance. Since my graduation in 2014, I have spent much of my time performing my one-man show, titled: "BREATHE. A TRUE STORY," which I wrote and produced. The show, named one of the top five shows of Charleston's 2015 Piccolo Spoleto Festival, tells the story of my experiences with cystic fibrosis and recalls my life-saving surgery and my outlook moving forward.

BRENNEN REEVES

I continue to perform in Charleston, regionally, and at various venues around the country in addition to working as a comedian, writer and public speaker. This past year, I was honored by being named among Charleston's Best and Brightest, which is a designation awarded to young professionals in the low-country who excel in leadership, charity and career achievements. I'm hoping to continue my studies through a creative writing graduate program this coming autumn. I currently reside in Charleston, SC. Here are links to my pages and sites: brennenreeves.com; facebook.com/reevesbrennen/ ▲

Meet A New Director

Hello. My name is **Jeannine Ricci**. I'm 45 years old and was diagnosed with CF at the age of seven. My exposure to the medical field during my childhood inspired me to pursue a career in nursing. I landed my dream job and worked on the pediatric oncology unit at The Children's Hospital of Philadelphia for seven years. The lessons I learned while helping these courageous children and their families battle cancer were invaluable and I carry them with me throughout my life. With the birth of my first daughter, I decided to stop working and focus all of my energy on motherhood and staying as healthy as possible. I'm savoring every moment with my two girls, ages 13 and 15. I did eventually return to the field of nursing, but on a part-time basis, and currently work per diem as a school nurse.

I've been involved with the Cystic Fibrosis Foundation (CFF) for about 20 years as a Great Strides team leader. Our team has raised over $100,000 through the years. I'm currently working with the CFF by sharing my experiences and ideas in an effort to raise awareness of clinical trials. I'm a strong supporter of the concept of participatory medicine and believe that patients should be viewed as equal partners in their healthcare.

JEANNINE RICCI

Education, connection and communication are key components to helping patients feel empowered to advocate for themselves, and *CF Roundtable* is the perfect tool toward this goal. I remember receiving my first *CF Roundtable* newsletter about 15 years ago. I read it cover to cover and then over again, not realizing until that moment just how much I craved connection with others with CF. I've learned so much from *CF Roundtable* over the years and am excited to now be able to give back and share some of my knowledge and experiences.

On a more personal note, I live in New Jersey with my husband and two daughters. I love being outdoors enjoying nature, whether it is hiking in a national park or walking my dog, Buddy, at our neighborhood park. I enjoy traveling, photography and spending time with friends and family. I look forward to being a part of this wonderful organization. ▲

TILLMAN continued from page 3

an than other women. They are also more likely to have babies with birth anomalies, although not necessarily life-altering ones. Due to their chronic lung disease, women are also at higher risk of serious complications during pregnancy and delivery. Specific genetic mutations associated with CF also appear to increase the risk for diabetes in pregnancy. Compared to pregnant women without CF, those with CF were found to have a higher risk of hypertension, gestational diabetes, and to deliver before 37 weeks of gestation. Importantly, researchers found that their babies' survival rate was normal, and these infants were not at increased risk for significant neonatal morbidity or mortality when adjusted for gestational age. Still, premature babies of CF mothers had significantly increased rates of jaundice. The premature delivery may be explained by the mother's poorer pulmonary function or malnutrition. The health status of the mother may also contribute to a tendency toward reduced fetal growth, a condition known as intrauterine growth restriction (IUGR). This study is one of the few large-scale studies of pregnancy and CF, and it may be of aid in counseling patients who decide to have children.
http://tinyurl.com/jefskkr

CF Patients Using Kalydeco Show Better Lung Function But Bacteria Lingers, Study Says

Kalydeco (ivacaftor) is known to increase the activity of the cystic fibrosis transmembrane conductance regulator (CFTR) protein in cystic fibrosis (CF) patients. A new study says the drug also improves lung function and reduces inflammation, but does not eradicate Pseudomonas bacteria infection.
http://tinyurl.com/hra3gfn

VX-659/tezacaftor/ivacaftor for Cystic Fibrosis

VX-659 is the third next-generation corrector being developed by Vertex Pharmaceuticals. Preclinical results of the triple combination of VX-659 with tezacaftor (VX-661) and ivacaftor

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(Kalydeco) have already shown greater effectiveness and potency in restoring the cystic fibrosis transmembrane (CFTR) function. Both VX-659 and tezacaftor (VX-661) are CFTR protein correctors, and ivacaftor (Kalydeco) is a potentiator of this protein. While the mechanism of action of the first two is to move the defective CFTR protein to the correct place on the cell's surface, ivacaftor aids in the opening of this protein. In this way, the transport of chloride and sodium (salt) across cell membranes is enhanced and thin mucus, which lubricates and protects internal organs and body systems, is produced.
http://tinyurl.com/gmgu9b7

AbbVie, Galapagos's Kalydeco challenger clears Phase 2

Galapagos has posted Phase 2 data on the cystic fibrosis drug it is developing with AbbVie. The data suggest the potentiator - GLPG1837 - can play a role in challenging Vertex's Kalydeco and its follow-up programs. The ALBATROSS Phase 2a study is a multicenter, randomized, double-blind, placebo-controlled, parallel group study to evaluate two doses of orally administered GLPG2222 in adult subjects with a diagnosis of CF harboring one F508del CFTR mutation and one gating mutation. The primary objective of ALBATROSS is to evaluate safety and tolerability of GLPG2222 in patients. Secondary objectives will include the assessment of FEV_1, changes of sweat chloride, and CFQ-R.
http://tinyurl.com/z3uz5ja

PTI-428 for Cystic Fibrosis

PTI-428 is a cystic fibrosis transmembrane conductance regulator (CFTR) protein modulator, also known as an "amplifier." The oral treatment, which is being developed by Proteostasis Therapeutics, demonstrated positive effects on CFTR protein activity in preclinical tests when used in combination with existing treatments. Unlike other approved and investigational therapies that address specific CF gene mutations, such as F508del, PTI-428 is being developed to treat multiple CF mutations. PTI-428 modulates the CFTR protein. The drug acts as an amplifier, meaning it increases the amount of an immature form of the CFTR protein, providing substrate for other CFTR modulating agents, such as potentiators and correctors, to act upon.
http://tinyurl.com/hknuk4t

Spyryx Biosciences Announces Successful Completion of Phase 1 Clinical Trial for SPX-101 and Planned Phase 2 Initiation in Cystic Fibrosis During 2017

Spyryx Biosciences, Inc., a clinical stage biopharmaceutical company developing innovative therapeutics to address severe lung diseases, today announced the successful completion of a Phase 1 study for its lead compound, SPX-101, in healthy volunteers. SPX-101 is an inhaled SPLUNC1-derived peptide with a novel biological mechanism for regulating epithelial ion channels in the airway. The drug is designed to restore a natural pathway in the lung that regulates airway hydration and promotes mucocilliary clearance, which is dysfunctional in cystic fibrosis (CF). The mechanism of action of SPX-101 is independent of the genetic mutations that cause CF and has the potential to provide disease-modifying therapy to all CF patients. Summary of Key Findings:

- SPX-101 was safe and well-tolerated in all dose cohorts of the SAD and MAD arms.
- SPX-101 had no adverse effect on lung function.
- SPX-101 demonstrated very low systemic exposure and rapid clearance, with no effect on electrolyte levels or indication of hyperkalemia.

http://tinyurl.com/h2yqthj

Cystic Fibrosis Patients Dosed in Phase 2 Study of Lynovex to Treat Flares

The first patients have been dosed in the Phase 2 clinical trial evaluating NovaBiotics' Lynovex (NM001, cysteamine) as an adjunct therapy for infectious exacerbations in cystic fibrosis (CF). CARE-CF (NCT03000348, NBTCS-02) is a randomized, double-blind, parallel group, and placebo-controlled study designed to investigate the optimal dose regimen, effectiveness and safety of oral Lynovex in adult patients being treated for exacerbations in CF-associated lung disease. The objective is to assess Lynovex's benefits as an adjunct therapy to standard of care (SoC), and six dose regimens will be tested. Lynovex, an orphan drug candidate, works through a triple mode of action:

- Mucolytic — Disrupting the mucus in CF patients' airways
- Antibacterial — Addressing current (such as Pseudomonas aeruginosa) and emerging pathogens

In Memory

Paul Feld, 59
St. Peters, MO
Died on December 17, 2016

Paul Drury, 50
Plantsville, CT
December 19, 2016

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

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

• Antibiofilm — Addressing mucus buildup within the airways
http://tinyurl.com/hqo8g5f

VX-371 (Formerly P-1037) for Mucociliary Clearance

VX-371 (formerly P-1037) is an inhaled epithelial sodium channel (ENaC) inhibitor being developed by Vertex Pharmaceuticals (in collaboration with Parion Sciences) that blocks the sodium channel in airway cells. By blocking this channel, VX-371 may prolong the duration and maintenance of fluid in the airways after hypertonic saline use. Additional fluid in the lungs aids in making mucus thinner and easier to clear.
http://tinyurl.com/z383g2t

Concert Pharmaceuticals Initiates Phase 2 Clinical Trial Evaluating CTP-656 for the Treatment of Cystic Fibrosis

Concert Pharmaceuticals, Inc., announced the initiation of a U.S.-based Phase 2 clinical trial evaluating CTP-656 (deuterated ivacaftor), a next generation CFTR potentiator being developed for the treatment of cystic fibrosis. CTP-656 was created based on Concert's application of deuterium chemistry to modify ivacaftor (Kalydeco®). In contrast to Kalydeco, Phase 1 clinical results from CTP-656 support dosing it once-daily with food, without regard to the fat content of the food, potentially enabling CTP-656 to support improved adherence and provide real world benefits for patients with cystic fibrosis.
http://tinyurl.com/gnmupde

Protalix BioTherapeutics Announces Last Patient Enrolled in the AIR DNase™ Phase II Clinical Trial for Cystic Fibrosis

Protalix BioTherapeutics, Inc., announced that the last patient has been enrolled in the company's Phase II clinical trial of AIR DNase™ (PRX-110) for the treatment of cystic fibrosis (CF). AIR DNase is a plant cell-derived recombinant form of human deoxyribonuclease I (DNase I) that the company has designed, through chemical modification, to be resistant to inhibition by actin. Given actin is a potent inhibitor of DNase I activity, the company's AIR DNase has the potential to enhance the enzyme's efficacy significantly in CF patients when compared to the currently approved DNase treatment (Pulmozyme®). AIR DNase is administered through Philips Respironics' I-neb AAD Inhaler System. The I-neb AAD is a small, lightweight, virtually silent device that is fully portable and has a unique vibrating mesh technology that allows for faster administration than conventional jet or ultrasonic nebulizers.
http://tinyurl.com/zkbbnq2

Resunab (formerly JBT-101) for Cystic Fibrosis Lung Inflammation

In normal lungs, alveolar macrophages (immune system cells present in the lungs) promote inflammatory processes to defend against pathogens and infection. In CF patients, however, this inflammatory response is overactive and persistent, causing long-term and irreversible damage to the lungs. Resunab is a synthetic oral drug that activates a group of proteins called CB2 receptors. These receptors are expressed on activated immune cells and, when activated, trigger molecular pathways that resolve inflammation and halt

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Information For People Who Travel On Airlines

In December of 2011, the Transportation Safety Administration (TSA) introduced a toll-free helpline that answers questions for fliers with disabilities and medical conditions. Disabled travelers may call ahead about screening policies, procedures and what to expect at the security checkpoint. The purpose of the new helpline is to inform passengers with disabilities about certain policies before they fly so they may properly prepare for travel. The helpline may be helpful for people with cystic fibrosis who must travel with durable medical equipment, portable breathing machines, needles, liquid solutions (inhalant medications, insulin etc.) and other medical equipment.

Travelers are encouraged to call at least 72 hours prior to a flight. People with CF and other medical conditions may call the "TSA Cares" toll-free number at: 1-855-787-2227. The helpline is available Monday through Friday, 9 a.m.-9 p.m. (Eastern Time) and is closed on all federal holidays.

Individuals also may find information on traveling with special medical needs on the TSA Website at: http://www.tsa.gov/travelers/airtravel/disabilityandmedicalneeds/tsa_cares.shtm.

FOCUS TOPIC

TRAVELING WITH CF

The List Is All-Important

By Andrea Eisenman

> "The best thing I did was to create my packing list in MS Word and print it out a few days before my trip."

Sometimes I wonder what life might be like if I didn't have CF and have to use so many pieces of apparatus to keep me healthy. One thought is: What would I spend all my free time on that I spend on cleaning and maintaining my equipment, counting my pills, doing nasal lavage and breathing treatments, using and changing an insulin pump site, exercising and cooking most of my food? Oh yeah, I might be able to work full-time and have a completely different outlook on life. But really, I wonder what it is like to travel as a person without CF and transplant. I see what my husband packs for a four- or five-day trip compared to what I have to pack and envy is the main emotion I feel. But then again, due to him not having so much to bring, I can sneak some of my gear into his checked suitcase.

Traveling can be a joy but, for me, forgetting something such as extra pump cartridges for my insulin pump, surplus medication etc. can ruin a trip. Once, for some reason I forgot my weekly pill pack organizer at home (about 25 daily pills) but brought only my cyclosporine. On that trip, I also forgot my inhalation machine and nebulizer but brought my hypertonic saline. Forgetting an important item while traveling happens less often, due to extensive list making and checking the all-important list multiple times.

I think what helped is that I go to my mom's for a few days a week periodically and need to bring most of my daily meds in the weekly pill pack plus extra medications I take for a migraine or supplements to help me sleep. I have started to leave things at my mom's and

ANDREA EISENMAN

at home but then am not sure entirely what is where. Still, going back and forth and using my double-check system has helped alleviate forgetting major things like my daily pills or enzymes. And my husband asks me sometimes if I have all my important things. This helps me stop and think about what I did or didn't pack. I also go through my bag to check it again: completely worth it.

Something to note: as I get older, go through menopause and remain on my transplant meds, my memory has worsened. I write everything down. I make daily lists for reminders of tasks, even if it is just ordering more medications because I am planning a trip and may need extra while I am away. The best thing I did was to create my packing list in MS Word and print it out a few days before my trip. I then start gathering items I will need to bring and eventually sort them into either checked or carry-on baggage. I also leave stuff on my list that I might not need on that trip, if not traveling with my dogs (they get medication too) or traveling in warm weather versus cold, or even if I am going to be swimming or playing tennis while away. These items vary greatly, but I am able to leave all of those options on my list and either ignore or put slashes through if they are not currently needed.

Packing, though, can be a challenge. I know that in my carry-on bag, I have to pack my weekly pill pack plus an extra few days' worth, cyclosporine, enzymes, sleep meds, extra salt tabs, electrolytes tablets, Nystatin, (if on them) antibiotics, my inhalation machine (including tubing and nebulizer and hypertonic saline), insulin pump supplies, syringes – just in case the pump fails, extra batteries for pump and glucose meter. For my nasal lavage, I bring my nasal steamer, glacial acetic acid (less than 1 ounce), Kosher salt in a Ziploc (it does get some funny looks), my Neti pot and a two-cup Pyrex measuring cup. It must be packed so that it all remains intact. Then add in my computer and my iPad, plus their chargers, and some snacks and that is about

all I can put on board with me. I pack the mask I wear in-flight and buy at least a liter of water in the airport to stay hydrated. When I finally get to my seat in the airplane, I have to take several minutes to get really organized so I am not trying to find things I know I will want during the flight. It gets cumbersome carrying it all on.

A challenge has been the addition of a new machine, my CPAP. For this device, I need to make sure I buy distilled water when I land for the water chamber and vinegar plus mild detergent liquid to clean its parts daily. Getting these items in a remote part of Puerto Rico was challenging. Normally, these are items I can find in a local drug store. The CPAP machine is supposed to fly in the cabin with me, but I cannot get it in my carry-on. I may have to invest in a bigger bag. Actually, this usually gets put in my husband's checked bag as mine is too full with my leg brace and tennis racquet (if I play tennis), my clothes and, because I sweat a lot, many changes of socks, shirts, underwear and exercise wear. I make sure I keep up my daily exercise regimen, if possible. And then maybe something nice to wear? I try.

Every time I fly, I do try to pare it down, but I would rather bring too much than not enough and then have regrets. But if I follow my list and check it over several days, crossing things off as they go into the travel bags. I usually get where I am going well-equipped. Sometimes, I have forgotten my hairbrush because it was not on the list and usually the last thing I would pack. It is now on my list. I have learned a lot from leaving things behind. ▲

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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In Memory of Dr. Lisa Marino
twohawksfoundation.org

DIAMOND SUSTAINING PARTNERS

IN MEMORY OF
LAUREN
MELISSA KELLY

Getting There Is Half The Fun, And Half The Battle

By Sarah Albright

"Is your cough contagious?" "Can I please move my seat?" "What do you have? You should get it checked out." Those are just some of the countless comments I have heard on a plane while traveling. I know the CF cough is not pretty sounding, nor is the spitting, sniffling or clearing our throats; but I can't leave my CF behind while I step out into the world for a vacation. Therefore, I deal with symptoms and annoyances of the disease while navigating through airports, train stations, bus stations and cars. Not everyone knows about CF, so I have learned to respond to the questions and glares by educating others about the disease. Traveling with this illness can be complex and exhausting. However, when packed appropriately, taking a trip can be a breeze.

As a 25-year-old, I have caught the traveling bug, and I love to pack up and see the world. But CF doesn't make that easy. However, my travel experiences have taught me how to make the process easier. For starters, get yourself a good bag or case to help keep all your medications together in one area. When they're all together you know where to find something quickly. I have a teal and gold toiletries bag that perfectly fits all my pills, inhalers, spacers and equipment. If you bring it in your carry-on, make sure to put all the liquid medications in the approved-size baggies.

I also bring along my Vogmask. This is a mask that has a filter for breathing in all the recycled air on planes. I know it looks peculiar, but the Vogmask makes appealing designs that can match your personality with colors and patterns. I personally don't like wearing it in public besides in a hospital setting, but I'm protecting myself from those on the plane who might actually be contagious.

My purse is an arsenal of goodies for the plane, train or car. I keep a lot of hand sanitizer in my pockets and purse. You are touching dirty surfaces that thousands of people also have touched. It's good to stop the spread of germs and clean your hands for snacks and meals as well. I make sure to pack enough snacks and light meals for the trip. Flights, bus rides and trains don't really feed you anymore, so to keep my calories up I bring along bags of chips, cookies, gummies, cheese and crackers, and some fruit. This is usually enough to get me through the travel period. Next I have inhalers and a spacer in my purse for when I'm half way through the flight feeling tight in the chest. Just a few quick puffs, and I'm feeling better already. Between the mask and the inhaler, passengers usually get the hint that my cough is chronic and not contagious.

SARAH ALBRIGHT

Next, I have a portable nebulizer machine that is my go-to for traveling. It is small, can be battery-operated or plugged in and even has a car adapter. It's lightweight and easy to use so it doesn't take up a lot of space in my luggage, and the case can even hold the tubing and some tubes of hypertonic saline. I went to a music festival in Delaware for five days in the middle of the woods, with nothing but a tent, clothes, food and a mini burner. I had this machine, and the car adapter, so every night I snuck into the car and did my breathing treatments. Small advances in technology like this help to not stop the fun and allow you to keep enjoying the things you want to do.

One thing I found difficult in the past was trying to haul The Vest® along with me on my travels. I recently got the AffloVest, which is completely portable and I love it! There is no heavy machine, and no tubing; it's just the vest! It is battery-operated so that makes it a little heavier, but I'll take it over all the heavy machinery. It has a remote to control the frequency, time and the type of thumping. You can choose from vibration, percussion and drainage, or do all three consecutively. The battery is removable to charge when needed. It also comes with a bag with wheels to help carry it around the airport.

Another large issue I have come into contact with while traveling is getting through security smoothly and without embarrassing myself. I make sure to have a list of medications I take and the dosage, as well as the packaging to all my medications, especially for Pulmozyme because it's liquid and can cause problems going through security. I don't know how many times I have

been stopped and asked about the clear liquid, stating, "No, it's not a bomb."

When you finally get to your destination, there might be a sense of relief, but don't breathe yet (not that I can anyway). Depending on where you are going, the elevation might be higher so take that into consideration. I always carry my pulse oximeter in my purse to measure my oxygen saturation. I went to Palm Springs in the autumn and took a tram up 8,000 feet through the clouds. It was a breathtaking view (literally), but I de-saturated quickly to about 75 percent, so I stayed up there for only a few minutes. I know that I try to do it all while on vacation, but it's important to know your limits and stay safe.

If traveling abroad, a lot of the buildings and monuments in Europe are extremely old. Therefore, not many towns and cities have elevators or escalators. We hope that everyone is accommodating for the disabled, but it's not a law everywhere. When I went to Greece, our hotel was built into a volcano and, as beautiful as it was, I couldn't breathe walking up and down the stairs. Luckily, the staff was extremely accommodating and moved my room so I didn't have to take the stairs. Also, when I visited Mexico, there were no elevators, just stairs, so I had to walk up three flights of winding stairs. My friend, who was traveling with me at the time, carried me up them many times. But in case you don't have someone to carry you around, be sure to look at the hotel layouts and the places you will be visiting. It's important to know what you will be getting yourself into and if you should make accommodations.

If you're traveling to a different country, make sure to remember your adapters. Not every country uses the same plugs that we do, so you could have all your breathing equipment and have no way to use it. Before I go to a country, I make sure to search online to check the type of outlet they use. You can purchase an international adapter kit online, in an electronics store or even from SkyMall. Sometimes hotels provide adapters to their foreign travelers as well.

Traveling abroad can be exciting; there are many new sights, sounds, foods and an overall different culture. But sometimes the culture isn't good for the lungs. In many European counties almost everyone smokes. I remember feeling so uncomfortable in a restaurant trying to eat while the people next to me were blowing smoke into my food. It is not illegal to smoke in public, restaurants or bars in Europe. Take this into consideration while dining out. Try to get a table by the window or even sit outside. Almost all of the cafés have outdoor seating.

A lot of the medications and treatment regimens are uncommon and unknown to others, so it's helpful just to have as much information as you can about the paraphernalia you will be bringing along while traveling. Be prepared to answer questions, give information and deal with ignorance. Some people have never even heard of cystic fibrosis, so be patient and understanding. I have educated many while traveling over the years, and I hope that CF awareness is spread all over the world, where I travel. ▲

Sarah is 25 and has CF. She lives in Manhattan Beach, CA. She is currently waiting for a bilateral lung transplant. In the interim, she is an EEG technician at a local hospital, studying for the GRE and planning to apply to graduate school in the autumn for health psychology. She is also writing a book about living with a chronic illness. You may e-mail her at: sealbright291@gmail.com.

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fibrosis. Preclinical and early clinical studies involving Resunab showed a favorable safety, tolerability and pharmacokinetic profile. In preclinical models of inflammation and fibrosis, it stopped tissue scarring, essentially "turning off" chronic inflammation and fibrotic processes.
http://tinyurl.com/j3mjpym

LAU-7b for Lung Inflammation in Cystic Fibrosis

LAU-7b is an oral form of the retinoid fenretinide being developed by Laurent Pharmaceuticals. Retinoids are a group of compounds related to vitamin A. Fenretinide may help reduce the inflammatory response in the lungs of people with cystic fibrosis (CF). Administered as a once-a-day, solid dose of fenretinide, LAU-7b is designed to correct the defective metabolism and modulate chronic inflammation due to the genetic defect that causes CF. Arachidonic acid (AA) and docosahexanoic acid (DHA) are two essential fatty acids that play a key role in maintaining an effective immune-inflammatory response. The genetic defect that causes CF leads to exaggerated AA-mediated inflammation and low DHA-mediated resolution, causing lung infection and local tissue damage. LAU-7b works by correcting the defective metabolism of AA and DHA, and controlling chronic inflammation. Resolution of inflammation is a new approach that uses the body's own ability to modulate inflam-

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Up In The Air: Do You Need Supplemental Oxygen?

By Jeanie Hanley, M.D.

The first time it hit me that I needed supplemental oxygen was during a six-hour flight. Several hours into the flight, I woke up feeling faint with chest tightness and shortness of breath. Forcing myself to breathe slowly and deeply helped, so I continued this way for the remainder of the flight and, in retrospect, was lucky I didn't pass out. Respiratory symptoms ensued for the next few days – hemoptysis (coughing up blood), productive cough, wheezing – which led to hospitalization for the next two weeks. I attributed the need for oxygen in flight to a flare-up of my CF lungs and downplayed the role of high altitude.

Pondering several of my previous flights over the years, a clear pattern emerged: feeling well prior to the flight, increased pulmonary symptoms during the flight and CF flare-up after the flight. So when it came time to schedule another flight, months later, I knew I needed to be certain if the high altitude cabin pressure in flight was the culprit for this pattern.

According to the American Thoracic Society guidelines, it is difficult to predict who will need oxygen in flight using only oxygen (O_2) levels and FEV_1 (forced expiratory volume in 1 second, a measurement taken during spirometry), especially when you have significant respiratory disease such as CF. Because FEV_1 and O_2 levels alone can't predict who will need oxygen, all of us should be evaluated prior to flying. Even so, these measurements taken at rest can be helpful. If O_2 levels at rest are above 95 percent and FEV_1 is over 50 percent, then the odds are supplemental oxygen is not essential. Below 95 percent O_2 and/or under 50 percent FEV_1 suggests strongly that oxygen is justified. In my case, I was borderline at 60 percent FEV_1 and 95 percent O_2 saturation, but with my history of in-flight respiratory symptoms, the odds of requiring oxygen surged.

To fully answer the question, my CF pulmonologist recommended that I undergo HAST testing, or a hypoxic high altitude simulation test. During this test, a low concentration of oxygen is inhaled while monitoring your blood O_2 level by pulse-oximeter. If the O_2 level drops below 85 percent, oxygen will be necessary during flights.

On the day of the HAST test, my FEV_1 was in my usual 60 percent range and I was feeling well. I was hooked up to oxygen via mask. Normally we all breathe in 21 percent oxygen (at sea level), but the oxygen given during HAST was 15 percent which simulates what you breathe in an airplane cabin at 8,000 feet, over a mile and a half high. During the test, I read or closed my eyes so as to replicate my most frequent activities on planes. Lo and behold, the oxygen saturation started diving from an initial 95 percent into the low 80s. I breathed deeply to improve the O_2 level, but the moment I relaxed, the O_2 level would descend again. The verdict was clear — that I would need oxygen for my next trip.

JEANIE HANLEY, MD

I began to investigate the options available for oxygen. My kids were away at colleges in the Big Apple, and one was graduating soon, so the trek from California to NYC was fast approaching. I learned that none of the airlines I planned to use provided oxygen to passengers unless the situation was emergent. That left portable oxygen concentrators (POC) as my best option. They are relatively lightweight, battery-operated and there were many to choose from on the list of FAA-approved POCs.

If you need oxygen only when you fly, then insurance coverage may be challenging. Medicare Part B (under Durable Medical Equipment) covers 80 percent POC costs if: oxygen levels are significantly low; overall health may improve with oxygen therapy; and/or arterial blood gas levels are low. These are the same criteria used for those who need oxygen for normal daily activities of living, which do not take flying or HAST test results into account. So if the only criteria are a positive HAST, then you are on your own for covering costs of oxygen.

Since my insurance did not cover it, the next decision was whether to buy or rent a POC. I initially opted to buy because a payment plan was

> "Because FEV_1 and O_2 levels alone can't predict who will need oxygen, all of us should be evaluated prior to flying."

offered. POCs are very expensive, but spreading the cost over a year seemed prudent at the time. It was very convenient not to deal with renting, which I thought would be a hassle and too costly over the long run. The other benefit of the POC was for car trips at higher altitude locales and for impromptu trips by air without spending time renting. The downside was the POC lasted only three years and the warranty was finished. Once my first POC went belly up, I have rented from local medical suppliers and found that, except for the first rental, subsequent POC rentals were relatively smooth and not very time-consuming. Given I haven't traveled as much as in the past, it has been less costly than buying a new POC.

Another hurdle is obtaining the proper documentation for bringing the POC on board. Each airline requires its own unique set of documents, which once filled out, can be used for 12 months. Check online that your POC is FAA-approved for your airline: www.faa.gov/about/initiatives/cabin_safety/portable_oxygen/. Notify the airline way ahead of time that a POC will be used during flight. Some air carriers will have their own medical liaison who can work directly with you. Print out the airline's forms for POC use, which a physician will need to complete. Make a copy of the documents for your purse or backpack and luggage. E-mail or fax a copy to the airline/liaison.

I carry my POC in a small suitcase that fits under the seat in front of me. The case also has plenty of room for the nasal cannula tubing, extra batteries, antibacterial wipes, tissues, charcoal-filtered facemasks, water, book, computer and chocolate.

Arrive at the airport earlier than usual because the ticket agent will need time to check the POC to ensure that it is FAA-approved, that you have a physician note and documents and enough POC batteries to ensure that there is enough to last one-and-a-half times the required flight time. Most airlines make you sit in the window seat. If you have a different seat, then they may change it. The silver lining is I happen to enjoy the window seat. Also, anyone with a POC gets to pre-board without paying extra. More importantly, pre-boarding allows more time to sanitize the area around your seat and anything within arm's reach, turn off the air outlets above your seat and in your aisle, don a facemask, set up the POC, get comfortable, take a big breath and relax. ▲

Jeanie is 54 and is a physician who has CF. She lives in Los Angeles with her best flying companion, her husband, John. They have three adult children. Jeanie also has three siblings with CF. She is a Director of USACFA and is the President. Her contact information is on page 2.

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matory responses, with the goal of addressing inflammation without inducing immunosuppression.
http://tinyurl.com/z8pr7ec

GS-5745 for CF Lung Inflammation in Cystic Fibrosis

GS-5745 is a monoclonal antibody that may help reduce inflammation in the lungs, leading to improved lung function in people with cystic fibrosis (CF). GS-5745 inhibits matrix metalloproteinase 9 (MMP-9) — an extracellular enzyme involved in matrix remodeling, tumor growth and metastasis — and has antineoplastic potential. Once administered, GS-5745 binds to MMP-9 and inhibits its enzymatic activity. A Phase 2 study to evaluate the effect of GS-5745 on FEV_1 (forced expiration in one second, a measure of lung health) in adults with cystic fibrosis (NCT02759562) is recruiting participants. The study will evaluate GS-5745's effect on pre-bronchodilator FEV_1 in adults with CF after eight weeks of treatment.
http://tinyurl.com/gl289bc

Pitt study links cystic fibrosis lung inflammation to opportunistic bacteria

This study details that the bacteria, P. aeruginosa, thrives in inflamed lungs, secreting an enzyme called Cif that sabotages the body's ability to make a key molecule called a pro-resolving lipid mediator, which helps halt inflammation. Patients with higher Cif levels in their lung secretions experienced less signaling to stop inflammation and increased levels of IL-8, a marker for inflammation. Increased Cif levels were also linked to reduced lung function.
http://tinyurl.com/zbceo9f

Improving the Lives of Patients with Respiratory Diseases

Pulmatrix is developing an inhaled

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Traveling With Baggage, And Lots Of It

By Nicole Kowal

Recently my husband and I went on our honeymoon, to Alaska! It was amazingly beautiful, breath-taking and absolutely outstanding! I couldn't have asked for a better honeymoon and trip with the man of my dreams, although I could have asked for a lot less baggage. We did a cruise up there, but traveling to the cruise ship and then from the ship was a bit of a struggle.

I don't know about you, but when I fly I wear a Vogmask. Whenever I use public transportation I wear one, and I bring many different styles to go with different outfits. As we left Buffalo, NY, I grabbed my luggage, which included not only all my clothes for two weeks, but my lunchbox with neb meds, my The Vest® in its very convenient carrying case, enough medications just in case we get delayed anywhere, and of course regular stuff like makeup, hair things and I surely can't forget slippers! I looked ridiculous loading up the truck at 5 a.m. and my dad making fun of all my "crap" - lol. Now here is where it gets even more fun, going through security.

Have you ever taken your vest through security at the airport? If not, be sure to be prepared to be pulled off to the side and having it wiped down, and being questioned on what it is, what it is used for etc. I was surprised that they didn't question any of my medications, but I am sure they see a lot of medications daily there. Now, boarding the plane while wearing my mask was actually quite nice, because I was able to get on there with all the wheelchair access personnel. All I did was ask at the desk at the gate if that was possible, due to trouble breathing, all my carry-on (I carry on my The Vest® and place it above me) and to make sure I am not taking up an emergency section. It does amaze me how people seem to not want to sit near a person wearing a mask, which makes it sort of nice for my husband and me. (Bring on more room!)

We landed in Seattle and that was all fine, even getting in and out of the Ubers and rentals we took was all great. Throughout all the different transportation, I never lost sight of my The Vest®, nebs and medications. I could always buy clothing, but man oh man do I need my medications.

The cruise ship was amazing with where I set up my treatment in our room, and having the mini fridge to hold my nebs, and the fact of simply having room. It was great! I am very open when people ask what I am carting around or why I wear a mask and any other questions thrown my way. I truly believe that helps make traveling and life that much better. I believe that telling people I have CF spreads awareness and opens their minds to what else is out there.

MICHAEL AND NICOLE KOWAL AT CLINIC.

I make it sound like the whole honeymoon was easy with traveling, but I can tell you this much, when we got off the ship, it downright sucked. We decided that we could walk from the ship terminal to the train terminal, eight blocks, with our luggage, through Vancouver. And crazy enough, it was uphill a lot of the way. I had my The Vest® and carry-on, and my poor husband carried everything else. (Big kudos to him!) That was hard. Then came the waiting for our train back to the States. The five-hour train ride turned into a sort of sick and twisted laughing/giggling fest between my husband and me. Our assigned seats were behind a gentleman who was not very nice and very afraid of germs and bodily functions. I wore my mask, just like I always do on public transportation. Every single time I coughed, he swore at me, he freaked out and he would give me the finger or some other vulgar gesture. But, instead of my husband and me letting it ruin our honeymoon, we laughed and still to this day joke about it.

I believe that having an open mind, being open about CF and the luggage we carry every day has an impact on others. It shows that we, as "sick" as we can be, know how to live and enjoy life. We are not afraid to travel to different places just because we have a few extra

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THROUGH THE LOOKING GLASS

PHOTO BY DAVID MYERS

Road Couch

I refer to my bike as my “road couch”
It’s what I do to unwind.

It frees me and works like therapy
to help me clear my mind.

-J. Lindic, 1998

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

FROM OUR FAMILY PHOTO ALBUM...

ANNA PAYNE BETWEEN TWO BANANAS, MAKING NEW FRIENDS, WHILE WAITING TO SEE BERNIE SANDERS SPEAK AT THE LIACOURAS CENTER AT TEMPLE UNIVERSITY, APRIL 2016.

FAMILY KRAMER GOLINKOFF: MOM LIZA, BROTHER COBY, EMILY AND SISTER ANNIE, AFTER COBY AND ANNIE RAN ON #TEAMEE IN THE PHILADELPHIA HALF MARATHON!

PHOTO BY CHEYENNE GIL PHOTOGRAPHY

MADDIE, LAUREN, JEANNINE AND MICHAEL RICCI IN LONDON, ENGLAND, AUGUST 2016.

NICOLE AND MICHAEL KOWAL, JR., VISIT ALASKA.

ELLA BALASA AT DUKE GARDENS, NC, WHEN SHE WAS AN INPATIENT BEFORE SURGERY.

LAUREN JONES HUNSAKER ON VACATION AT THE HOTEL DEL CORONADO IN CORONADO, CALIFORNIA.

BOOK REVIEW

Lessons From A CF Cornerman:

38 Lessons I Learned During My Wife's Illness And Lung Transplant

By Raymond L. Poole

Reviewed by Piper Beatty Welsh

As a married, two-time bilateral-lung transplant recipient with cystic fibrosis, I'll admit that I sat down to read Raymond L. Poole's *Lessons from a CF Cornerman* with some degree of trepidation. Living with CF and transplant can often seem overwhelming enough on its own – did I really want to dive headfirst into another couple's experience with "my" disease?

It took about ten pages of reading for me to answer that question with a resounding "Yes!" Poole's book follows him and his wife Rebecca, as they meet, fall in love, learn to negotiate life with her CF as a couple, then face new challenges together when she experiences a sudden decline and listing for bilateral-lung transplant. Rebecca's story – and Poole's expert narration of it from his perspective as husband, caretaker and, well, "cornerman" – is at once both completely unique and heart achingly relatable.

For a reader outside the CF community, this is an inspirational read about love for better or for worse (and sometimes through even worse than that!) in sickness and in health. For those of us within it, the book has added dimension, reflecting back on us the raw truths of CF through a candid and sometimes humorous lens. Through this book, we are treated to unique insights about life with CF from "the other side" of the marital aisle. Poole takes us from the halls of the college where he and his wife met, down the roads of their early relationship and marriage, into the grim world of the ICU, through the operating room and back out again. All with an unshakeable narrative voice that somehow manages to be both brutal and comforting. Scattered along the way are the many invaluable lessons (38 of them, to be exact) that he took home with him. And although some of these insights made me squirm (note to readers: please do not try to pull a PICC line at home in your bathroom to go on a business trip!), every single one of them was meaningful, personal and, above all, honest. Because at the end of the day, that is Cornerman's greatest lesson to all of us: that honesty, love and a willingness to adapt to new circumstances are our best weapons not just in the fight against CF but in everything we do.

Perhaps the best review I can give of this book, however, is not in my words but in my actions. So absorbed was I in this read that I barely put it down from start to finish, despite the tears, smiles and laughter I experienced along the way. And this afternoon I gave the book to my husband – my very own transplant and "CF cornerman" – for him to read. I can't wait to hear what lessons he learns, too. ▲

Piper is 35 and has CF. She is a former director of USACFA.

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ACTIVE FOR LIFE

Exercise and Travel

By Aimee Lecointre

When I travel, whether for leisure or work, I still make every effort to stay active. For me, exercise is just as important as everything else required to take care of and maintain my health. It's just as important as my enzymes, breathing treatments and other medications. As I've shared before, exercise has played a critical role in my overall health.

You may not be able to keep up your usual or regular exercise regimen, but that's no excuse to just ditch exercise or activity altogether. In fact, it's incredibly easy to stay active while traveling, no matter the reason for your travel; you just have to make it a priority along with all your other CF treatments.

Here are some tips to stay active and exercise while you're out of town.

1. If staying in a hotel, utilize the fitness room. Nearly every hotel has one and when they do it's free to use. Not all of them are created equal and you may have to do something you normally don't but don't let that hold you back. Tip: bring your own disinfectant wipes if you are worried about germs.

2. Bodyweight and plyometric exercises require zero equipment! Squats, lunges, jump switch lunges, tuck jumps, curtsy lunges, squat jumps, jumping jacks, high knees, burpees, mountain climbers, dips using a chair, push-ups etc. Create your own sequence and change it up daily.

3. Bring your own equipment. Jump ropes and exercise bands take up very little room in your suitcase. My go-to when traveling these days is my EmPack. It's a backpack that comes with water bladders that can be emptied for travel then converted into an exercise pack. Once you get where you are going you can fill the bladders up with water (each is 15 pounds) and place them in the backpack. Now you have a weighted bag that can be used for pretty much anything – squats, deadlifts, chest press etc. Or just use it to add some weight while running stairs or hiking.

4. Walk. Weather permitting, and depending where you may be, get out and walk! Or if you're a runner, go for a run. One of my favorite trips was to Seattle. I went with one of my best friends and we walked everywhere. It kept us active, saved money on cabs/Ubers/public transit and we got to see so much more of the city. We found restaurants to try we wouldn't have otherwise found and got to see some amazing views of Puget Sound.

5. Yoga. No mat required even. I often travel with my yoga mat, but it's not necessary. You can follow along with an online video (www.yogawithadrienne.com is my favorite free yoga resource) or just flow as you go.

AIMEE LECOINTRE

6. Check out the local cross-fit boxes, yoga and Pilates studios or local gyms. Let them know you are visiting and ask if they have a one-week deal or a pass for just a few classes. Most places are willing to give you a deal if you just ask!

7. Some of the larger gym chains have locations nationwide and even offer membership levels that allow you access to every location while traveling.

8. Go on an adventure. Depending on where you are and why you're traveling, there may be plenty to do that allows you to have fun, make memories and get exercise in without even feeling like you're exercising. Kayaking, SUP (stand-up paddleboard), surf lessons, hiking, jet skiing, skiing etc. Endless possibilities!

It used to be that when I traveled, near or far, I would take a break from exercise. This "break" always turned out to last much longer than my trip, and it was always a struggle for me to get back on track. I have found that not only is it easier for me to keep up my much-needed exercise regimen when I exercise on vacation, I also find it to be a great way to start the day. It gives me energy and I end up feeling much better throughout my trip and upon returning home.

I hope that when you next travel you find these tips useful! ▲

Aimee is 31 and has CF. She lives in Utah with her husband, two pugs and a cat. She is a registered yoga teacher and nutritional therapy practitioner. She can usually be found hiking in the mountains, spending time with family or cooking up something delicious in the kitchen. You may contact her at: alecointre@usacfa.org.

PARENTING

Parenting When Sick

By Dana Giacci Rogers

I love seeing those commercials for cough and cold medicine where the terribly sick parent with the stuffy nose and scratchy throat hobbles into the toddler's bedroom and tells him or her that they are taking the day off because they are just too sick that day to keep up. Then the children, usually causing some half disaster or standing confused looking them right in the eye, are speechless. Because even those kids know, there are no days off in parenting. Usually at about that time they deliver the punchline of some great cough medicine to get their day back and conquer their cold symptoms as the commercial cuts out.

Parenting with CF, some days, feels exactly like those commercials. Unfortunately, there is no magic cough syrup that will work to eliminate all our symptoms and make the world right again in a matter of hours. So how do we do it? After a few years of trial and error, as well as helpful tips from loving grandparents who enjoy watching their grandkids, I have developed a list of ideas to stash away for those sick days. These are just some ideas that will hopefully help you get the brainstorm started to find creations of your own tailored to your family's lifestyle.

1. A special tote or shoe box with toys that are played with only during sick days. Put these toys away on a shelf and only get them out when you need downtime. It will make them feel special over and over again and capture attention for possibly hours depending on what is in it. Coloring book or crayons that are new, puzzles or games, perhaps even a couple of special books that are fun or some action figures. Notice again that all the toys in the box are also easy lying down activities so you can participate. The only limit to what is in the box is how big it is. Put these toys away when you are feeling better. If they are played with every day they won't feel like special toys anymore and therefore won't capture the same amount of attention.

2. Let your kids play doctor. There are endless doctor sets on Amazon and at places like Target and Walmart. This idea will work depending on the age of your child. But in playing doctor they can bring you your meds, wash treatment gear, start you an extra breathing treatment, put you down for a nap, give shots, check your vitals, even prescribe and bring you extra oxygen.

3. Nail polish, was always a winner with my oldest daughter. She would paint my toes over and over and, sure, it wasn't a salon-caliber pedicure, but I got to rest and even do a treatment while she had a great morning all the same.

4. Something that has recently been working with my three-year-old is timing her as she runs a pre-determined course around the house. All of which is timed from my watch while I am taking whatever downtime I need. She does it over and over, trying to beat her previous time. By the end of it, she is tired and I have rested.

DANA GIACCI ROGERS

5. Another downtime game that my youngest and I tend to do together is reading one book at a time and then drawing in her drawing book certain pages out of the book or parts of the story that we liked. This has entertained her for hours before. And if we play this game while lying down in her teepee fort it lasts even longer. In the end, there is story time then drawing time and all of it happens while I am lying down.

6. Take the cushions off the couch and put them on the floor. Any configuration you want. We do a line and it becomes gymnastics tumble track, but you could also spread them out and play "lava" where they can't step anywhere but on the cushions. The possibilities here are endless and you can be sitting and watching as the audience. Every good game needs an audience.

7. Don't forget the classic board game is always a winner. Conversation and learning ensue when playing games like Candy Land, Chutes and Ladders, Connect Four, and even a game of war with cards. With my three-year-old I pull out the cards from ace to five to play war. Then she has to count how many symbols of the suit are on each card and decide which is higher to find out who won. Great counting and number recognition. Could always go up to ace to ten for a bit older children. Games like these are great. It is one-on-one time, but it is also something you can do while doing a treatment or rest-

ing on the couch. They assist in learning colors and numbers as well as taking turns and winning or losing.

8. PLAY-DOH! Goodness I love Play-Doh. It can last up to an hour at times. It is simple, creative and quiet play. Great for a down day.

9. Parade. We love playing dress-up and what does every parade need? You guessed it, an audience. If you are really wanting it to last longer, you can toss a piece of candy (yes, granted it is the opposite direction from usual) for each new outfit.

10. Finally, the best down day activity in the world. Take a bath together. Good downtime activity and all while soaking in the tub. Add some Epsom salt and ease your achy joints while you are at it.

Keep up the good work CF parents. Even when you are feeling sick, it is my experience that you are most likely still doing a terrific job. ▲

As Always,

Dana

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

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antifungal product, named PUR1900. The product, which has received Orphan Drug designation by the FDA, combines an antifungal drug with the company's unique dry powder iSPERSE™ technology, enabling patients to inhale the drug deep into the lungs where it can fight the infection. Because Pulmatrix's iSPERSE™ technology has the potential to deliver drugs much more efficiently to the lungs than exiting approaches, its products offer the promise of high effectiveness with fewer side effects than current inhaled drugs.
http://tinyurl.com/hctlxwo

Cystic Fibrosis Study Links Patients' Risk of Fungal Infections to Bacteria

Certain bacteria may increase the risk of fungal colonization in cystic fibrosis (CF) patients. CF makes a patient susceptible to chronic airway inflammation and infections, resulting in lung tissue damage. Scientists have long known that bacteria such as Pseudomonas aeruginosa and Staphylococcus aureus play roles in airway infections, but the role of filamentous fungi (in infections) is poorly understood. An increasing number of CF lung infections are caused by the Scedosporium apiospermum fungi species or Lomentospora prolificans (Sac-Lp). It ranks second among filamentous fungi colonizing CF airways, after Aspergillus fumigatus. Sac-Lp fungi are opportunistic pathogens that can cause infections such as pneumonia and sinusitis. Researchers found an association between Sac-Lp colonization and an increased incidence of allergic bronchopulmonary aspergillosis (ABPA). They also found an association between Sac-Lp and an increase in the mucoid bacterial strain of Pseudomonas aeruginosa. Another finding was that Sac-Lp co-colonization was reduced in patients with H. influenzae and Candida species infections.
http://tinyurl.com/glykpaz

AeroVanc (Vancomycin Inhalation Powder) for Lung Infections in Cystic Fibrosis

Vancomycin inhalation powder (trade name, AeroVanc) is the first dry-powder inhaled version of the antibiotic vancomycin being developed for the treatment of methicillin-resistant Staphylococcus aureus (MRSA) lung infection in people with cystic fibrosis (CF). Vancomycin inhalation powder is a dry-powder form of vancomycin, self-administered using a capsule-based device. Vancomycin is the treatment of choice for MRSA-related bronchopneumonia. However, this antibiotic has systemic side effects and requires intravenous (IV) administration. Because AeroVanc delivers vancomycin directly to a lung infection, it is expected to improve the drug's clinical efficacy and lessen the adverse effects caused by broad-spectrum antibiotics.
http://tinyurl.com/jd4otvn

Fosfomycin/Tobramycin Inhalation Solution for Cystic Fibrosis (CF) Lung Infections

Fosfomycin/tobramycin inhalation solution (FTI) is a broad spectrum antibiotic for the treatment of CF lung infections that consists of fosfomycin, an antibiotic with activity against both gram-positive and gram-negative bacteria, and tobramycin, an aminoglycoside that has potent gram-negative activity. FTI has shown improved effectiveness against lung infections caused by bacteria compared to fosfomycin or tobramycin alone. FTI rapidly kills a variety of gram-positive and gram-negative bacteria commonly present in people with CF experiencing lung infections, and demonstrates effectiveness in killing bacteria relative to fosfomycin and tobramycin alone. FTI also shows a low frequency of resistance relative to fosfomycin and tobramycin alone.
http://tinyurl.com/z8l9mmb

CFF Grant Will Support Early Studies of Oral Antibiotic, MAT2501, to Treat NTM Infections in Cystic Fibrosis

Matinas BioPharma and Colorado State University (CSU) will jointly con-

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IN THE SPOTLIGHT

With Emily Kramer Golinkoff

By Andrea Eisenman and Jeanie Hanley

Emily is quite determined and, talking with her, I [Andrea] could barely keep up. I think she is just so passionate about finding new research for CF and wants to help others so badly, she needs to do things quickly so she can fit everything in. She feels time is of the essence. Mainly her focus is on what is important to her, leaving no stone unturned in finding a cure for CF. That and her friends and family are clearly what keep her going. She and her family tirelessly fundraise for a foundation they started five years ago, called Emily's Entourage. It began as an effort to accelerate research and drug development for those with CF who have nonsense mutations and may not respond to the new mutation-dependent medications like Kalydeco and Orkambi. She travels all over the world speaking on advocacy and stresses the importance of storytelling in social media. She has received recognition from the White House as a "Champion of Change" for precision medicine. She is a doer.

Please welcome our newest star, **Emily Kramer Golinkoff**. Spotlight, please!

Age: 34

Diagnosed: At six weeks of age. I had failure to thrive and pneumonia.

How many siblings? I have three siblings. I am the oldest and the only person with CF in my family.

Do you feel your siblings feel guilty about your diagnosis? No, not guilty, I don't think. CF has become the family's shared disease. We are a close-knit family. We all struggle with CF and our life-quest to cure it. My family is emotionally connected and we all have scary thoughts about my future health status, so they feel fear more than guilt. It is the progressive aspect of CF that is scariest. If I could stay frozen in my current condition, late-stage CF, it would not feel so threatening. One of my sisters has special needs and that fostered acceptance of all of our medical issues. My family and I have become masters at creative solutions for adapting to and solving problems. Growing up, we all worked within limitations and contributed to a framework to moving forward and having fun, rather than feeling overwhelming guilt or sadness. We make the best of the life and what we are given and that is a lot!

EMILY KRAMER GOLINKOFF

PHOTO BY CHEYENNE GIL PHOTOGRAPHY

How important is family to you? Family is incredibly important. In just emotional and practical ways, I could not do what I do without my family's support. CF is a shared illness. I feel intensely grateful for my family, their unconditional love and support, and the values they taught me from a very young age.

What are your mutations? Double W1282X (a nonsense mutation).

What does knowing mean to you? It is only because I know about my mutations that I could dig into research. It was a motivating factor for me. There was not enough research or drug development efforts for nonsense mutations and that was precisely what propelled me forward. If I wanted any hope, my family and I had to take the research into our own hands to start Emily's Entourage.

I was tested as a kid for my CF mutations. My parents had participated in clinical research and had me tested at that point. I had known my mutations when Kalydeco was being developed. And knew I would most likely not benefit with my nonsense mutations. Reading more and digging to see what was happening with nonsense mutations was a big factor in why I got involved. I had not planned to start a foundation, but no one else was looking into nonsense mutations and it seemed like the only way to get things done. More research needs to be done as nonsense mutations are the cause of approximately 12 percent of all genetic diseases.

What propelled you to start Emily's Entourage? My family and I had no intention of starting a foundation, it just happened. We had started to fundraise for a one-time campaign and through initial efforts saw such a high return, how eager people were to support the cause, that we decided to take science into our own hands. We were amazed by the compassion that exists, so many people wanting to help. People who donated realized that there was something they could do by donating funds.

With all the big institutions in place driving medical advances and research, it can be easy to take a laissez-faire approach. But I realized you do not have to be the best or smartest person in the world to realize gaps. It

just takes one motivated voice or movement to propel ideas into action. We had to be that voice.

Can you discuss donating your cells? From the beginning, Emily's Entourage (EE) realized that the most impactful thing we could do was bring the community together to advance research and drug development. One major barrier was that research scientists didn't have cells for nonsense mutations. So I have been donating nasal epithelial cells and induced pluripotent cells for years to researchers and labs around the world. It is hard to come by these cells since I have two copies of the same rare mutation. The idea is that when they have these cells they can test drug samples on them and make research easier and faster. EE also supplies grants to researchers at universities. Grants are given, but they are imbedded in the process and EE remains involved with the advisory board. We stay focused on the key questions to answer and the key researchers to work with. It is a partnership between researchers, industry, clinicians, patients and families.

Science is methodical and slow, which is not ideal for those with a progressive, fatal disease like CF. EE brings this urgency to the table because we are a family and patient organization: the biggest threat is time.

How many hours do you spend taking care of yourself daily? About three to four hours a day. I use the Vest with inhalations (inhale albuterol plus hypertonic saline with Vest, Pulmozyme once a day and take pills). I rotate inhaled antibiotics – Cayston, Vancomycin and take oral doxycycline. Then I sterilize nebulizers, after using them. I am compulsive about my regimen but consider it an investment in my health and future.

What is your motivation to keep going? My family and friends keep me going. On top of that, I have big eyes, lots of dreams and plans, and I'm very ambitious. I feel there is much to do and don't want it to be hindered by CF. And, not just for me, but for others with CF, too. I want to see the members of the CF community reach their own potential and I don't want CF to stand in their way. The Entourage has been a major motivator too. The kindness and passion that EE has sparked in the community has been so awe-inspiring. I have had a chance to experience firsthand the kindness and love of people who give, without strings attached. It is just remarkable to me and that solidarity around a shared mission – to cure CF – is the most empowering feeling in the world.

Your biggest fear? Being outside the cusp of breakthroughs. Working so hard for a breakthrough and it not coming on time. But this is a motivator too to keep going and keep moving as fast as we can. I find peace in feeling like I am doing everything possible.

What are you most hopeful for? My hope is that they will find a huge breakthrough for everyone with CF, and then we will take all the lessons we've learned from EE and apply them to a new rare disease.

Do you see a cure for CF in your lifetime? I am fighting my hardest to get one. Cure would be great but control is good, too.

Last thoughts? Sometimes it takes someone whose life is on the line to make others step up, add rigor and raise the bar for everyone else. My life and others' are dangling in the balance and change and progress are up to us.

To find out more about Emily's Entourage, here are some helpful links. Website: www.EmilysEntourage.org.

Video: https://youtu.be/1rWh_CwxSS4. Facebook: https://www.facebook.com/EmilysEntourage.

E-Newsletter: http://bit.ly/EEInsider. ▲

Andrea Eisenman is 52 and has CF. She is a Director of USACFA and is the Webmaster and Executive Editor of CF Roundtable. Her contact information is on page 2. Jeanie Hanley is 53 and is a physician who has CF. She is a Director of USACFA and is the President. Her contact information is on page 2. If you would like to be interviewed for "In The Spotlight," please contact either Andrea or Jeanie.

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duct preclinical studies assessing the efficacy of MAT2501, a potential oral antibiotic, in treating non-tuberculous mycobacterium infection (NTM) in a cystic fibrosis (CF) lung model. NTM are universal organisms responsible for opportunistic infections with a broad spectrum of virulence. These infections are difficult to treat, and progressively more resistant to most available antibiotics. MAT2501 is an encochleated formulation of the broad spectrum aminoglycoside antibiotic agent amikacin. But amikacin is now given by intravenous injection, and its use is associated with major side effects, including toxicity to the kidneys and ototoxicity (permanent loss of hearing) with long-term use. The efficacy of MAT2501 against several mycobacterium species has been established. MAT2501 is an orally administered version of amikacin (encochleated amikacin) based on a lipid-crystal, nanoparticle delivery technology for more targeted treatment, improving its safety and tolerability to allow for mul-

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2. If I get married and my spouse's income results in my losing SSI benefits because his income puts us over the household income amount for SSI, will I lose my Medicaid? If I lose my Medicaid is there another way for me to qualify for Medicaid?

ANSWER: Marriage will result in the new spouse's income and assets being considered by Social Security for purposes of determining eligibility for SSI benefits and for purposes of Medicaid eligibility. If the household income and assets are over the allowable amount, then the person loses SSI and Medicaid. In some states a person may be eligible for Medicaid even if the person does not have SSI, but the person will still have to have a household income that is below the allowable Medicaid income and asset criteria.

Many people with CF tell Medicaid representatives that they should be allowed to keep Medicaid benefits because otherwise they will not be able to access health insurance coverage. Sadly, this is not a consideration under Medicaid rules.

Social Security considers a person to be married if the person "holds themselves out as married." If a person does not obtain a marriage license, Social Security can still find the person is married under the Social Security definition of marriage.

If you have other questions, feel free to contact the CF Legal Information Hotline at: CFLegal@sufianpassamano.com. It is best to e-mail in order to obtain a specific time to talk. Watch the *CF Roundtable* blog for answers to other questions. Nothing in the above column is meant to be legal advice but is meant only as information. ▲

Beth is 51 and has CF. She is an attorney who specializes in disability law and is a Director of USACFA. Her contact information is on page 2. You may contact her with your legal questions about CF-related issues.

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tiple dosing.
http://tinyurl.com/z6jj953

Nitric Oxide (Inhaled) for Lung Infections in Cystic Fibrosis

Nitric oxide (NO, brand name Thiolanox) is a naturally occurring gas that, when inhaled, may reduce lung infections, leading to improved lung function in people with cystic fibrosis (CF). The gas possesses various biochemical characteristics, such as efficient broad spectrum anti-infective properties, as well as antimicrobial activity against gram-positive and gram-negative bacteria, yeast, fungi and viruses both in vitro and in vivo. Thiolanox is an investigational drug product of Novoteris and is not approved for sale in the U.S. Nitric oxide is one of the major signaling molecules in biological systems. It has been shown that it can promote the growth and activity of immune cells at low concentrations, while at higher concentrations it establishes connections with DNA, RNA, proteins, or lipids that contain iron or sulfur to inhibit or kill target pathogens. It also has lipophilic and hydrophilic properties that allow it to cross cell membranes and dissolve readily in water. Nitric oxide is also a mucolytic and vasodilator. It is a muscle relaxant with an effective short-acting bronchodilator. This effect could contribute to the removal of secretions and maintain airway stillness. Administration of NO to the airways before or during lung infections may reduce viral load and, therefore, bacterial susceptibility. It can thin secretions, restore the mucociliary system and increase removal of secretions. During an infection, NO may improve oxygenation as well as local blood flow to the respiratory tract due to its vasodilatory effect. This increased blood flow could increase an influx of nutrients and white blood cells while also increasing local temperature, all of which are useful for the resolution of an infection. Based on laboratory tests, in vivo models and healthy human data, NO is administered in 30-minute sessions, three to five times a day for multiple days at a dose of 160 ppm.
http://tinyurl.com/zyrhglb

Compound in Soil Bacteria May Combat CF Pseudomonas Infections

Researchers may have come one step closer to more effective treatments against Pseudomonas aeruginosa growing in layers known as harmful biofilms. A compound found in soil bacteria prevented the bacteria from forming biofilms, which is a serious health threat to patients with cystic fibrosis. Pseudomonas owes a large part of its persistence in people with CF due to its tendency to form biofilms — layers of bacteria embedded in mucus. When growing in this form, the bacteria are effectively protected from antibiotics, making it difficult to eliminate with current treatment approaches. Researchers studied a compound secreted by the bacteria, called pyocyanin. The substance, which is known for its bright blue color, has long been used to diagnose infections with the bacteria. However, researchers recently discovered that it is valuable for more than diagnostic purposes. The compound helps the bacteria grow into biofilms, making it a possible target for treatment of Pseudomonas infections. The research team then isolated another type of bacteria present in soil. Analyzing

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> "It does amaze me how people seem to not want to sit near a person wearing a mask, which makes it sort of nice for my husband and me. (Bring on more room!)"

bags. Sure, packing for the trip stinks – making sure you have enough room for all treatments, medications and, in my case, I bring my own gluten free snacks just in case my sugar drops too much and I need something fast.

That was just one story of us traveling. We have been to a few places and it never gets easier. We simply just become smarter with packing. To gain room in our luggage, we don't pack as many clothing options but bring those little Tide packets that allow you to wash your clothes in the sink. (We did that twice on the honeymoon.) Being smarter with the way we pack has us being okay with anywhere we want to travel. In the years we've been together (almost six), we have traveled to Boston; Siesta Key / Sarasota; Seattle; Callaway Gardens, GA; Nashville, and, of course, our Alaska trip. We make it fun and try not to let my baggage be too heavy for our trips.

Now, packing for camping is a whole different story. Tent supplies, clothing, the dog, food and my lovely CF baggage. But that attitude I talked about earlier helps a lot. I create my own area for my medications and treatment to plug in, and I make sure I try to keep that outlook with everyday life. I believe that life is in the eye of the beholder. No reason to make mountains out of mole hills, right? So here is to carrying our baggage with strong arms, and an even stronger outlook. ▲

Nicole is 29 and has CF. She and her husband, Michael, live in Buffalo, NY. You may contact her at: nkowal@westherr.com.

factors released from this bacterium, they found one substance capable of changing the chemical structure of the pyocyanin. The newly discovered compound, which the scientists called pyocyanin demethylase, or PodA, prevented Pseudomonas from forming biofilms when grown in lab dishes. However, researchers are years away from translating the findings into treatments for human disease.
http://tinyurl.com/z4vfftv

Stripping away an infection's armor

A protein that could help fight serious infections has been identified. The research focused on biofilm, which allows bacteria to stick to materials and form a protective layer. Research focused on the bacterium Pseudomonas aeruginosa. The bacteria works by attaching itself to lung tissue and producing a dense and slimy alginate biofilm, where bacteria can embed themselves and stay protected from the body's immune system. The researchers have found a way to strip the protective layer away so the bacteria can be attacked. The breakthrough comes with the identification of a protein that may be able to stop the biofilm latching on to tissues and causing an infection, but also may help the body stop the bacteria spreading to other parts of the body and causing a chronic infection. The protein influences the chemical composition of the bacteria.
http://tinyurl.com/jottm3h

CF Treatment Using Bone Marrow Cell Transplants May Improve Lung Function, Study Says

Bone marrow cell transplant can improve overall lung function and may have therapeutic benefits in treating cystic fibrosis (CF). Previous research showed that cell therapy could restore production of functional CFTR protein. However, this approach has been limited due to low levels of engraftment, or incorporation of the cells. A team of researchers found that — using a mouse model of cystic fibrosis — an optimized delivery of normal bone marrow cells contributes to the expression of CFTR proteins in the epithelial cells of the airways and restores fatty acids, which are altered in cystic fibrosis. The delivery of bone marrow cells also delayed lung infection with the pathogenic bacteria Pseudomonas aeruginosa and increased survival of the mice. The researchers concluded that bone marrow cells confer a dual benefit: an early non-specific beneficial effect against infections — possibly by recruiting macrophages and other immune cells that fight off infections — and a late beneficial effect linked to the expression of functional CFTR protein. Some potential problems associated with the clinical applications of cell replacement therapy have, however, been noted. Therefore, the researchers urge further study before translating their results to people.
http://tinyurl.com/gujau8u

AzurRx BioPharma Announces First Three Patients Included in Phase IIa Study with MS1819-SD for Exocrine Pancreatic Insufficiency

AzurRx BioPharma, Inc., announced

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Mailbox

I am writing to ask you to remove my name from the snail mail *CF Roundtable* newsletter that you send out periodically. I love your newsletter and follow it regularly, now online and on Facebook. Thank you so much for the work you do to compile such interesting and helpful topics. I will continue to read your publication online, as it has been very helpful to read stories from others with CF and to see how others handle the daily struggles of living with this disease.

Thanks again,
Jonathan Liston
Syracuse, UT

I am a 58-year-old CF adult (diagnosed at 3 years old) and have seen too many losses to this breath taking disease.

Lynn Pancoast
Allentown, PA

[This is] for all the heart and love you put into bringing your best to CFR. I appreciate your efforts. It's come a long way since Lisa McDonough started her newsletter, and our first board took it forward after her. She would be most proud! Website looks phenomenal.
Blessings be,

Melinda Anderson
Lighthouse Point, FL

Paul [Feld] received a lung transplant from the sudden death of our son. We are happy we got to meet Paul and that he was able to enjoy 12 more years of life. He was a very nice person and will be a pleasant addition to Heaven.

Terry, Carole & Scott Sproul
Knoxville, TN

Thank you for a great publication!

Mike Darrar
Post Falls, ID

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that the first three patients in the Phase IIa clinical trial with MS1819-SD for patients with exocrine pancreatic insufficiency (EPI) caused by chronic pancreatitis (CP) have been enrolled. The primary objective of this Phase IIa study is to investigate the safety of escalating doses of MS1819-SD in patients with CP. The secondary objective is to investigate the efficacy dose response of MS1819-SD in these patients by analysis of the coefficient of fat absorption (CFA) and its change from baseline. Safety will be assessed at the end of each treatment period with particular attention paid to immunoallergic effects, digestive symptoms and clinical laboratory tests. MS1819-SD, an oral non-systemic biologic capsule, is a recombinant enzyme that is derived from the yarrowica lipolytica lipase, and is entirely plant-based/vegan.
http://tinyurl.com/zyqnnos

Aberdeen drug firm launches full-scale trial of cystic fibrosis drug

NovaBiotics is currently testing its treatment for cystic fibrosis (CF), Lynovex. The randomized, placebo-controlled double blind study follows promising preliminary trials which found the oral capsule version of the drug was safe and well tolerated by patients – and showed some improvement for those with CF. The latest trial, the CARE-CF-1 clinical study, is designed to test how well the drug capsule treats "infectious exacerbations" of CF. The private equity-backed company is also working on an inhaled form of Lynovex, which it expects to start testing in proof-of-concept clinical trials this year.
http://tinyurl.com/zqa6d42

SINUS

Manuka honey sinus irrigation for the treatment of chronic rhinosinusitis: a randomized controlled trial. Lee VS, Humphreys IM, Purcell PL, Davis GE. Int Forum Allergy Rhinol. 2016 Dec 9

Manuka honey (MH) has been shown in vitro to be effective against biofilm-producing bacteria. This study assessed the effectiveness of MH for patients with active chronic rhinosinusitis (CRS) and prior sinus surgery. In patients with active CRS and prior sinus surgery, both MH and saline improved outcomes, but there was no statistically significant difference between these groups. However, in the subset that did not receive oral antibiotics/steroids, culture negativity was statistically better on MH, suggesting that MH alone may be effective for acute exacerbations of CRS.
http://tinyurl.com/h9w2l6x

Determinants and outcomes of upfront surgery versus medical therapy for chronic rhinosinusitis in cystic fibrosis. Ayoub N, Thamboo A, Habib AR,

MILESTONES

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

ANNIVERSARIES

Birthday

Janie Davies
Arlington, TX
70 on January 16, 2017

Ed Fleischman
Plainview, NY
75 on December 31, 2016

Zach Hays
Vernon, CT
28 on November 26, 2016

Stephanie Rath
Brownsburg, IN
48 on January 26, 2017

Delayne Santos
Gulfport, FL
50 on February14, 2017

Wedding

Delayne & Gerry Santos
Gulfport, FL
20 years on September 19, 2017

Transplant

Paul Albert, 57
Catasauqua, PA
Bilateral lungs
24 years on February 10, 2017

Jill Doran, 42
Durham, NC
Bilateral lungs
14 years on January 3, 2017

Darvin Mazariegos, 23
Los Angeles, CA
Bilateral lungs
One year on January 4, 2017

NEW BEGINNINGS

Engaged

Anna Payne and **Kyle Ferretti**
Philadelphia, PA
On December 9, 2016

Nayak JV, Hwang PH. Int Forum Allergy Rhinol. 2017 Feb 20

The indications for surgical management of chronic rhinosinusitis (CRS) in patients with cystic fibrosis (CF) are poorly defined. In this study outcomes of medical versus surgical treatment were compared and trends associated with the transition from medical to surgical therapy in CF patients were examined. The researchers concluded that surgery effectively reduces CRS-related symptoms in CF patients but may not improve pulmonary function. In patients who first pursue medical therapy, symptomatic decline may prompt eventual conversion to surgery. Patients who delay surgery may achieve similar outcomes as those who pursue surgery upfront.
http://tinyurl.com/z37ghxq

FYI

Clostridium difficile carriage in adult cystic fibrosis (CF); implications for patients with CF and the potential for transmission of nosocomial infection. Burke DG, Harrison MJ, Fleming C, McCarthy M, Shortt C, Sulaiman I, Murphy DM, Eustace JA, Shanahan F, Hill C, Stanton C, Rea MC, Ross RP, Plant BJ. J Cyst Fibros. 2016 Nov 29

Clostridium difficile is an anaerobic gram-positive, spore-forming, toxin-producing bacillus transmitted among humans through the fecal-oral route. Despite increasing carriage rates and the presence of C. difficile toxin in stool, patients with CF rarely appear to develop typical manifestations of C. difficile infection (CDI). In this study, the carriage, toxin production, ribotype distribution and antibiotic susceptibility of C. difficile was examined in a cohort of 60 adult patients with CF who were pre-lung transplant. C. difficile was detected in 50 percent of patients with CF by culturing for the bacteria. All strains were susceptible to vancomycin, metronidazole, fusidic acid and rifampicin. The high prevalence of hypervirulent, toxigenic strains of C. difficile in asymptomatic patients with CF highlights the potential role of asymptomatic patients with CF in nosocomial transmission of C. difficile.
http://tinyurl.com/jcvmxmd

Airway inflammation in mild cystic fibrosis. Jonas Eckrich, Ulrich M. Zissler1, Friederike Serve, Patricia Leutz, Christina Smaczny, Sabina Schmitt-Grohé, Daniela Fussbroich, Ralf Schubert, Stefan Zielen, Olaf Eickmeier. Journal of Cystic Fibrosis.

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Raise Your Voice

By Anna Payne

ANNA PAYNE, ON MSNBC, WATCHING BERNIE SANDERS SPEAK AT THE DNC.

This July I had the privilege and honor to be able to go to the Democratic National Convention (DNC) as an elected Bernie Sanders delegate for the Eighth Congressional District in Pennsylvania. It was an experience I will never forget, and it has propelled me into the next political chapter of my life.

I wanted to share a little bit about not only the experience of being a delegate, but also what we all need to do now, regardless of our political party affiliation, to make sure our legislators make the right decisions for the CF community.

The first day at the convention I was totally fired up and excited to meet new people and check out all the parties. There are so many parties; I had so many invites in my e-mail I couldn't keep track. The parties are where the free food is; I was all about free food. I made it to two parties the whole time and they occurred on the first night there. I'll be honest, I don't know how people keep up with the schedule and party until the wee hours in the a.m. I was beat. By the time I got back to my room from just the convention it was always 11 p.m. or later. We had to be up as early as 6 a.m. the next day to get breakfast and our daily credentials to get onto the convention floor. We had breakfast and then a meeting and a few hours in-between to get ready to go to the convention floor.

The earlier you got to the Wells Fargo Center the better your chances were to make sure you got a seat. Seats filled up fast especially on Monday, Wednesday and Thursday. Those days had more popular speakers. My candidate spoke on the first night. Ironically I was late that day. I was lucky enough that a nice woman gave up her seat so I could sit close enough to see. Since I am only 4 feet 10 inches, a good seat was very important to me. I won't lie, it was very hard to fit in all my treatments and get proper rest. I did my best to do

Genetic Mutation Information Resource

Collaborators from several institutions around the world and the U.S. Cystic Fibrosis Foundation are excited to announce that a new resource – CFTR2 – is now available for public use! This is the result of an international research collaboration to provide information about specific cystic fibrosis gene mutations to patients, their families, researchers, health professionals and members of the general public. We hope that you will find the information useful.

The website is available at www.cftr2.org. Once you have reviewed the website, please take a few minutes to complete the user-satisfaction survey located in the blue box "How can you help us improve the website?" in the "Quick Links" section of the left margin. Your responses will help us improve the website. Please feel free to contact cftr2@jhmi.edu with any comments, questions or suggestions, but please note that we are unable to answer any questions about the medical care of individual patients, since we are the research team that helped develop the website and not a clinical care team.

two treatments a day. It wasn't easy, especially when I wouldn't get to sleep until 2 a.m. and had to be up at 6 a.m. the next morning.

I feel like the first day I ran on excitement and then the second day I was exhausted. I had to recharge my batteries and take a nap, and try and get to bed before 2 a.m. By the last day, I was definitely ready to go home, or at least sleep for a full day. While by the end I was exhausted, and the rigorous schedule did take a toll on me, and it probably took me about two weeks to recover, I wouldn't trade the experience for the world. I got to meet and speak with so many people from across the country about not only issues they cared about but also what motivated them to get involved. It was truly inspiring. I made friends across my home state and I got to talk to elected officials about CF and healthcare and why it's so important for our community. I learned a lot as well; I was able to attend some classes about advocacy, issues and even one about running for office.

This experience has propelled me to my next adventure of running for local office in my township. I'm so excited, the election isn't until November 2017, but I have already started to get things in place for my campaign. I'm running for Township Auditor. I don't have a primary opponent which gives me extra time to prepare, so that is great (especially since I'm working full-time). I hope that if I win, I can learn more about the township's inner workings and hopefully this will help set me up for bigger and better things in the future.

As I mentioned I have met so many people over the last year, some of whom are currently elected officials. I will be going to Harrisburg in the next week for PA advocacy day and I'm super excited. Since I have gotten more involved politically, I have been able to make connections with my local state legislators. So for me it will be like sitting down with friends, which provides an advantage especially when talking about CF. I won't be just a face. I will be a familiar face of someone they know personally. I hope that will help when trying to get them to either join the caucus or support our various causes.

I know right now it is more important than ever to stand up for our community, make sure that our voices are heard and our needs are met. It doesn't matter what political party you are affiliated with, our representatives locally and nationally need to hear from all of us. Maybe we don't agree on every issue, but we need to ensure that when new laws are drawn and decisions are made, the chronically ill are not left behind. We can't just assume that we won't be forgotten or that someone else is going to rise up and be the voice. We must be our own advocates just like we are at our clinic appointments. This is no different; the setting is different and the scenery, but the idea is the same.

People with CF are living longer. Adults with CF are experiencing new problems, such as applying for social security, disability coverage or receiving health insurance, and how this impacts us daily, not only health-wise but financially as well. These are all reasons we have to make our voices heard. The more we make others aware of our situation the better off we are not only as individuals but also as a community. I will keep speaking out, and I hope you will too. ▲

Anna is 30 and has CF. She lives in Levittown, PA, with her mother, step-father and dog, Geoffrey. She is recently engaged and purchasing a condo with her fiancé. Some of her goals this year include hoping to raise $30,000 for the CFF and getting elected as her township's Auditor. She currently works full-time at a local credit union. In her free time, she loves to watch Law and Order SVU, *and* WWE *programing and relax with her dog and fiancé.*

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Pay It Forward

BRONZE

Melinda Anderson
Anonymous (In memory of Paul Feld)
George Bahorich
Janet & Ray Bernacki (In memory of Paul Feld)
Cyndi Bondy (In memory of Paul Feld)
Eva Bradin (In memory of Paul Feld)
Brenda Bunge (In memory of Paul Feld)
Isabel Stenzel Byrnes (In memory of Ana Stenzel)
Mary Carmody (In memory of Paul Feld)
Kristine Casteel (In memory of Paul Feld)
Lisa Cissell (In memory of Paul Feld)
Judy Dale (In memory of Paul Feld)
Mike Darrar
Janie Davies (In honor of my 70th birthday and 6-year cancer survivor)
Ruth Dunafon
Laura Ewald (In memory of Paul Feld)
Nina Ferrell
Ed Fleischman (In honor of my 75th birthday)
Rusty Fry (In memory of Paul Feld)
Valje Fulton (In memory of Paul Feld)
Doreen Gagnon (In memory of Joe Kowalski)
Holly Godsey (In memory of Paul Feld)
Judy & Ken Greenburg (In memory of Robert & Leigh Anne Hoehn)
Ruth Haddox (In memory of Paul Feld)
Mark Hale (In memory of Jennifer Hale)
Dennis & Val Hallebusch (In memory of Paul Feld)
Joan Hamlin (In memory of Paul Feld)
William & Karen Hams (In memory of Paul Feld)
John & Jeanie Hanley (In memory of Paul Feld & David Sproul)
Richard Harris (In memory of Kathleen Harris)
Cindy Hays (In memory of Zach Hays)
Michael & Paula Henry (In memory of Paul Feld)
Jean Howe (In memory of Paul Feld)
Harriett Hubbard (In memory of Paul Feld)
Gertrude Huntington
John & Joanne Jacoby
Barbara Jenkins (In honor of Steve Jenkins)
Theodore Kowalski & Philip Devine (In memory of Joseph Kowalski)
Margaret Krahman (In memory of Paul Feld)
Gay Lazur (In honor of daughter, Delayne F. Santos, and celebration of wedding anniversary of Delayne & Gerry Santos)
Alana Lehr (In memory of Paul Feld)
Diana Lewis (In memory of Paul Feld)
James & Debra Lohe (In memory of Paul Feld)
Randy Oppold (In memory of Paul Feld)
Lynn Pancoast (In memory of friends, brother and Larry Culp)
Emily & Matthew Piette (In memory of Paul Feld)
Tami Prentis (In memory of Paul Feld)
Ray & Janet Pullen (In memory of Paul Feld)
Sidney Rabinowitz (In memory of Ilana Davida Schwartz)
Denise Rubin (In memory of Paul Feld)
Sandra Ruhmann (In memory of Paul Feld)
Paige Russell
Nancy Saunders (In memory of Paul Feld)
Rodney Scott (In honor of Nick)
Bettye Seibels (In memory of Paul Feld)
Janice Siegel
Gaylord & Katherine Smith (In memory of Paul Feld)
Kristal Sproul (In memory of Paul Feld)
Terry, Carole & Scott Sproul (In memory of Paul Feld)
Jennifer Staashelm
Laura Tillman (In memory of Paul Feld)
Nancy Tinkham
Team Transplant St. Louis (In memory of Paul Feld)
St. Louis Urological Surgeons (In memory of Paul Feld)
Jill Walker (In honor of Jonathan Miller)
Mary Weiss (In memory of Paul Feld)
Terri Williamson (In memory of Paul Feld)
D. Lynn & Daniel Witthaus (In memory of Paul Feld)
Lester & Serene Witthaus (In memory of Paul Feld)
Steve & Erin Witthaus (In memory of Paul Feld)
Adria Wood (In memory of Paul Feld)
Michael & Angela Wood (In memory of Paul Feld)
William & Bernidine Wood (In memory of Paul Feld)

SILVER

Jill Doran (In celebration of bilateral lung transplant 14th anniversary)
Norman Young, Jr.

GOLD

Beth Sufian (In honor of Kathleen & Mike Beatty)
Beth Sufian (In memory of Will Cooper)

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A Former Director Of USACFA
Paul Feld
May 9, 1957, to December 17, 2016

By Andrea Eisenman

Former director of U.S. Adult CF Assn. (USACFA) and a frequent contributor to *CF Roundtable*, Paul Feld died from complications of stage-four kidney failure. Paul started as a director for USACFA in 2002 and departed in 2015. He was President, Vice President and chair of the fundraising committee during his tenure. Dedicated to raising awareness regarding CF and for treatments that could help people with this disease, he was well suited for being part of this organization and made a big impact on helping it grow. He wrote articles about receiving a bilateral lung transplant, being a proud father and then becoming a grandfather. He shared the challenges he faced, with the many skin cancers and the Mohs procedures he had.

He received his bilateral lung transplant in 2004 at Barnes-Jewish Hospital. Since then he was able to compete in three U.S. Transplant Games. He was thrilled to win medals in his donor's honor. But for Paul, it was more about being at the Games to promote organ and tissue donation. At his first Games, he met his donor's family and ran with his donor's wife in the 5K race to celebrate his donor's life and the "gift" he received.

As long as I have known Paul, he always had a positive perspective on things. It sometimes helped me reassess my own view to see him handle his health issues with aplomb. I admired him and it was easy to do as he was a very generous person with his time and gave you his full attention. We shared our trials and tribulations as it related to living with a transplant. I usually felt better after our e-mail correspondence. He was a very giving person.

He did impressive things. Once, he invited a friend to stay at his house with him and his wife while waiting for a lung transplant at Barnes-Jewish. It made it easier on the person's wait to be close to the hospital when the call came. She stayed for weeks. This act was much appreciated. It removed the stress of travel added to whether the call would come in time. He wanted others to experience the gift of life as he did, and he showed this desire with many acts of kindness.

Paul worked in IT for 24 years at BJC Healthcare and it was there that he met his wife, Kristi. He made a big impact at work as well, as many people who donated to his memory were from his job. Paul was proud he was able to work full-time and never let his CF get in the way of things he wanted to do. He was determined to live an independent life and did so until he had to have help. His family –wife, daughter and sister– then gladly stepped in and provided as much support as he needed. He attributed his surviving so long in stage-four kidney disease to his family who were always there for him.

He always loved the CFRI summer retreat, in Northern California, where he connected with friends, old and new. Even when he didn't feel so well because of his kidney disease, he went to the retreat. It rejuvenated him. And, maybe he didn't know this, but I bet as one of the oldest attendees, he gave others with CF, who were younger, hope.

Paul is greatly missed in the CF community and beyond. I thank him for all he did to make lives better for everyone he knew. ▲

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Because I Live And Breathe ...

By Ella Balasa

I live my life by this motto. Because I live and breathe... fill in the blank. Anything can be written at the end of this sentence. This means anything is a possibility. I've not always viewed life in this way, but through maturity, experiences, gaining self-confidence and finding my identity, I have come to believe in, and to live, a fulfilled life. Fulfillment to me means to achieve a dream, to pursue a passion, to strive to be happy every day and find joy in what you do. To say you did your best and you made every moment count. That is living a fulfilled life. Everyone's best and everyone's dreams are vastly different. The importance is finding the fulfillment of the dreams you want, not what you think you should want or what society has deemed worth achieving.

I believe having those dreams and feeling fulfillment comes from motivation. Motivation to do and to be better for whatever parameters you set for yourself. My motivation for life comes in the most innate form, which is the will to live — to live the fullest life I can, in the time I am given to live it.

Having cystic fibrosis has shaped me to want to live to the fullest each and every day without letting time go by that I can't get back. My motivation to do this has grown with each passing year and especially, most recently, as I've felt the effects of my disease more than ever before. In a way, it has scared me, but from that fear, I feel I've grown to have an even greater will to go on and to have an appreciation for the life I have. I've come to realize there is a lot I want to do and to achieve, and though my time may be more limited statistically than the average person's, it certainly won't deter me from striving for even more.

Over the past year, I have struggled with keeping lung infections at bay and having very resistant Pseudomonas bacteria has required me to be on constant antibiotics, either oral or intravenous. During these episodes of infection onset, I have required using oxygen more and have now started using it during sleep. As of most recently, about three months ago, I developed a pneumothorax (collapsed lung) which persisted and finally after about six weeks in the hospital on and off waiting for it to heal, I had surgery to fix the collapse. This will hopefully eliminate this issue completely.

Prior to this episode, I was working part-time, participating and organizing after-work social gatherings, as well as exercising on a treadmill five days a week and practicing yoga at a studio. I want to continue to be able to participate and do things on my terms, and not be at the mercy of the functionality of my own body. I am a young woman, in the prime of my life. I should be able to make spur of the moment plans with friends and stay out a bit longer than I probably should without having to worry about the potential health consequences.

ELLA BALASA

I have come to realize, however, especially after this recent illness, it requires more and more effort to keep up and, no, I can never let my guard down. I feel weaker and require more rest. Spending close to two months in a hospital deconditioned my body in a way I never felt before. At the time of discharge, one flight of stairs was unthinkable. Even now, it's an excruciatingly slow climb. That's both literal and metaphorical.

I have always been one to adhere to my treatment regimen, and now I know I will have to work even harder and for longer to try to regain the strength I had, but I accept it because I have to. I have to because I want to live and breathe as normal a life as I can. And you know what? It's incredibly frustrating at times.

I'm not going to write this without an underlying hint of discontentment. It sucks knowing that I have to work extra hard, each and every day, just to maintain my health; and maybe – just maybe – after several months – I'll see a fraction of improvement – *if* I don't catch a cold or get ill in the meantime.

I've thought about the fact that my disease is progressing and will only get more difficult from here. I have the drive, the motivation and potential. It isn't primarily my fears, my insecurities or my doubts that hold me back, it is my physical body. This frustration isn't constant, it ebbs and flows, and as I build some strength from this health setback, I know it will become a less significant thought. I also have started redirecting my focus and deciding to write more personal stories from which I gain fulfillment as well. If, in the long term, I won't be able to continue my

> "To say you did your best and you made every moment count. That is living a fulfilled life."

job and doing as many activities outside my home because of failing health, then I will continue to achieve fulfillment in other ways, through writing and interacting more within the cystic fibrosis community.

The message I want to get across is that although I have limitations physically, and it's hard to accept a progression of this disease, I won't let it hold me back from being happy, feeling accomplished and staying motivated. I won't let it also limit me mentally. It's futile to waste energy on being sullen and wishing for it to be different, because this is the life I have been given. Therefore, I shall make the most of it, while I can. I won't dwell on the possibility that I won't ever make a full recovery from this setback. Instead, I try to be hopeful that I will.

I feel as though many people get caught up in insignificant details about life. People dwell on past mistakes, wishing for changes to occur without really acting and lacking the motivation to make a change and, because of this, they lack a feeling of happiness in their present lives. Most are inhibited by fears, which are our mental limitations: fear of judgment, fear of failure, fear of the unknown, fear of hurt. These are all rational fears, but they don't help us achieve our dreams or our happiness, rather they hinder us. The only person stopping you is yourself. Precious time is wasted dwelling on the past, regretting or wishing for future ideas and moments that seem impossible to achieve. Instead, try to focus on the present and what can be done now to live a life of happiness. I believe happiness determines the quality of our experiences. Having a deep-rooted sense of happiness about yourself and who you are determines the quality of life, where we can "smell the roses," being truly active in every moment.

Most of the population doesn't face their own mortality in a way individuals with a chronic or terminal illness do. Therefore, can their will be as strong? Not having the knowledge that one day a specific disease will be the ending in a way is a less burdened life; concurrently, a person will take the moments, days, weeks and years for granted, just floating through life, without the drive, without motivation, without finding purpose. People believe they still have their whole lives to one day "figure it all out." I certainly believe there are many individuals without significant obstacles in their lives who also live life to the fullest, but I think there may be a higher proportion of those who face extreme adversity and rise above with greater will. I encourage not just CF patients, but everyone, to find motivation which leads to fulfillment in every day. Find happiness in yourself and in others around you. Don't be fearful to love to the fullest, to be bold, to speak your mind, and to let others experience everything you are; to bear your soul. This way, you won't have any of those life regrets, for you lived the best life you could.

I fight every day for one more breath, to accomplish one more thing. Living, that's my motivation. I hope one day I will breathe deep once again with fully functioning lungs, by receiving a double lung transplant, but for now, I am coping, accepting, and being fulfilled every day ... because I live and breathe. ▲

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

YOU CANNOT FAIL

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

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

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

Newcomers To The CF Summer Retreat

By Devin Wakefield

Hi everyone! After my article last year about CFRI's (Cystic Fibrosis Research Incorporated's) CF Summer Retreat, we recruited some newcomers and I wanted to share their experiences and perspectives with *CF Roundtable* readers. The following was culled from interviews with three first-timers. Also, to offer a broader perspective, I talked with a long-time retreat attendee who has come for decades.

As a brief refresher, CFRI's CF Summer Retreat is held annually in Menlo Park, CA, for CF adults, family and friends. We absolutely encourage friends and family of CF patients to come, as everyone benefits from their presence. The retreat provides a relaxed and safe atmosphere to learn more about CF on several levels: practical day-to-day, biological and psychosocial. We do this through support groups, speakers, fun events and simply hanging out. To make retreat as safe as possible, attendees follow CFRI's infection control policies. You can find out more about that at the bottom of the article.

I interviewed several new attendees, including Colleen, who came all the way from Pennsylvania, invited by a friend. She said, "I wasn't working at the time and had moved back in with my parents and learned there were scholarships." Ricky, from Maine, shared, "I didn't know that the retreat existed until I read *CF Roundtable*. Applied soon after and it happened fast." Before, "I had inquired about support groups for CF people, but it's never really encouraged by anybody. The only way to do that was online through what I found on a website. That's not the same thing." Jeanie had been to the CFRI CF Conference (where we met) and, after some encouragement, came to the retreat! She says that before coming, "I wasn't sure what to expect, since the last time I went to a retreat was over 30 years ago in high school, where we all bunked in one large room. I knew that, at the very least, it would be much better than that." Finally, I interviewed a long-time retreat attendee, Alex, to help give more perspective. He first attended camp – as it was called in the 1980s – when he was eight to ten years old. The first time Alex attended, he recalls "immediately feeling a connection," and he "got hooked."

Let's start with the atmosphere of the retreat. Colleen says she "was pleasantly surprised it wasn't convention-style presentation. The activities were more communication and patient-centric focused than medical." Ricky agreed, saying, "I didn't think it would be that down to earth; it turned out to be a fun thing... Who knew it wasn't going to be a clinical experience, but a human, fun experience?" Jeanie, too, "did not expect it to be so fun. I didn't expect to bond with anyone for the short time I knew I'd be there." I think with cystic fibrosis, we patients tend to think CF-related events will focus entirely on the medical aspect of the disease (as many do!), ignoring the everyday and social aspects. The retreat works very hard to include a broad range of topics. As Alex put it, "all three parts of cystic fibrosis – mental, physical and spiritual – get addressed. Whether intentional or not, the organizers always include this." Jeanie added how she loved "the sharing! Especially the sharing and openness of so many." It can be hard to open up, but once one person starts to share, everyone gets into it. Colleen added, "everything was optional. If you didn't want to do arts and crafts, for example, you didn't have to."

DEVIN WAKEFIELD

Come for the people! Ricky "loved the camaraderie and that there's a group of people with CF in common. I thought it crazy that I was 52 and hadn't met anyone else with CF." Colleen said, "Everyone was very welcoming. I expected to be [my friend's] little buddy, but everyone was, 'Hi and welcome! You're one of us!' That was cool." Ricky "felt like the bar was raised, and the amount of things people with CF could do increased" for him, and "helped me get out and do more!" Alex said he "loves the people, loves being able to go to a place and just unwind and let go of the stresses of everyday life." He also loves "meeting new people and connecting with old friends."

Everyone learned a lot. Colleen said she "learned a better way to deal with survivor guilt. I also learned more about life experience with CF. Everyone is very individualized; I'm pleasantly surprised. Everyone is so different. I

COLLEEN LEWIS

learned from [another attendee] about asking for new hours at her job, because I did the same thing. I was never not learning, even if it was just how someone goes about fixing some problem we all face." Ricky had new realizations. "More than anything, my personal bar got raised a lot. I learned there are a lot of other people with CF stepping it up every day and that's invaluable; no excuse to not be doing that. I learned what was possible for myself and others. I was also really surprised how many people regularly traveled outside the country. One went to London with an oxygen tank and I was impressed." Alex added that even after all the years he has come, he still learns from others and "shows up with an open mind to learn as much as I can." Over the years, Alex learned about "issues with getting older – how do you take care of yourself and work, live a life that's still fulfilling? Maybe you can't work or stay in school, but there are things you can do to keep your mental health going so that depression and other things don't set in. There's physical, spiritual and mental health; no matter what your beliefs, background and upbringing, I think we all agree that these are equally involved in our overall health. These are things I've learned about at retreat."

As for the organization for the event, Ricky was "impressed by the volunteers and the event as a whole. Walking in, I felt like it was a promising experience from the get-go. I was surprised at how well organized it was." Alex said, "Every year is a great program. I'm surprised every year that there's so much thought and planning to get worthwhile speakers and guests who can share about new treatments." Alex also highlighted "the yoga that happens," as did Ricky ("I loved the yoga") and Colleen ("I got to practice yoga and walk around the beautiful grounds"). Jeanie "loved that in just two and a half short days, the memory of this retreat still makes me smile. The retreat itself was entertaining, fun and informative. I felt very grateful to be a part of our exceptional CF community."

We're glad to have you too, Jeanie! I hope you enjoyed reading everyone's own words about retreat as much as I enjoyed discussing it with them. Please, if you have any questions, contact me at devin.wakefield@gmail.com or contact CFRI through one of the following options:
E-mail: cfri@cfri.org
Phone: (650) 665-7559
Web: http://www.cfri.org/summer_retreat.shtml

RICKY ALBANO, WHO HAS CF, WITH HIS SON, NICHOLAS ALBANO.

ALEX JENKINS AND HIS DOG, ROSIE.

CFRI's Infection Control Policies: http://www.cfri.org/pdf/CFRI_Infection_Control_Policy.pdf

CFRI's CF Summer Retreat is generously sponsored by AbbVie and Gilead Sciences. ▲

Devin is 25 and has CF. He lives in Seattle, WA, and works at Microsoft as a software programmer. He enjoys the bracing rain and cold of the Pacific Northwest. That's because he doesn't sweat as much (meaning he doesn't have to lick himself to get his salt back!) and he can wear his snazzy leather jacket and gloves as his long hair billows iconically in the wind.

JEANIE HANLEY AND HER GRAND-BABY POOCH, LADY FRANCESCA.

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Cathy Chacon, RN, National Jewish Health: Addressing Multi-Faceted Health Issues Beyond CF
John Clancy, MD, Cincinnati Children's Hospital: The CF Pipeline
Lucas Hoffman, MD, PhD, University of Washington: New Strategies to Treat CF Pathogens
John Mark, MD, Lucile Packard Children's Hospital, Stanford: Adjunctive Therapies and CF
Ray Poole, MBA, Fort Thomas, KY: Perspectives from a CF Cornerman
Carol Power, RT, CF Center at Stanford: Respiratory Therapy – Options New and Old
Paul Quinton, PhD, UC San Diego: CFRI-Funded Research: 40 Years of Progress
Kristin Riekert, PhD, Johns Hopkins School of Medicine: Adherence to CF Therapies
Karen Von Berg, DPT, Johns Hopkins Hospital: The Role of Physical Therapy in CF

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For the safety of participants, Retreat attendees must be at least 18 years old, must complete a sputum culture after June 15, 2017, and must submit CFRI's Medical Release Form and Cross-Infection Liability Release Form to CFRI's office by July 27, 2017. Please download forms at www.cfri.org.

cfri Cystic Fibrosis Research Inc.

TILLMAN continued from page 35

January 2017 Volume 16, Issue 1, Pages 107–115

Airway infection and inflammation play major roles in the progression of cystic fibrosis (CF) lung disease. In patients with mild disease, airway inflammation is a clinically relevant and often under-diagnosed feature. Lung function, sputum cell counts and cytokine profiles in CF with mild disease might be different in patients with and without involvement of small airway disease (SAD). This study demonstrated that patients with CF with mild disease defined by lung function might be further endotyped according to their involvement of SAD. In patients with CF and SAD, airway neutrophilic inflammation is more pronounced and is in part distinct from that seen in patients without SAD.
http://tinyurl.com/jnj6ytm

End-stage cystic fibrosis lung disease is characterized by a diverse inflammatory pattern: an immunohistochemical analysis. Lammertyn EJ, Vandermeulen E, Bellon H, Everaerts S, Verleden SE, Van Den Eynde K, Bracke KR, Brusselle GG, Goeminne PC, Verbeken EK, Vanaudenaerde BM, Dupont LJ. Respir Res. 2017 Jan 10;18(1):10

Cystic fibrosis (CF) lung disease is characterized by vigorous airway inflammation eventually resulting in severe lung damage. This study aimed to describe the diversity of the inflammatory pattern in end-stage CF lungs by evaluating and quantifying which components of the innate and adaptive immunity are involved, and by assessing whether this is gender-specific. The data demonstrate a diverse inflammatory response in the CF lung, reflected by an increase of both myeloid and lymphoid immune cells. Inflammation in the CF lung appeared to be gender-specific in this study population, as the significant increase of eosinophils, mast cells and CD4 T cells was especially notable in the female subpopulation.
http://tinyurl.com/gp3loge

The changing prevalence of pulmonary infection in adults with cystic fibrosis: A longitudinal analysis. Kay

Continued on page 46

A. Ramsay, Harpreet Sandhu, James B. Geake, Emma Ballard, Peter O'Rourke, Claire E. Wainwright, David W. Reid, Timothy J. Kidd, Scott C. Bell. Journal of Cystic Fibrosis. January 2017. Volume 16, Issue 1, Pages 70–77

Increased patient longevity and aggressive antibiotic treatment are thought to impact on the microbial composition of the airways of adults with cystic fibrosis (CF). In this study, the authors attempted to determine if a temporal change in the airway microbiology of adults with CF has occurred over time. They found that a decrease in the prevalence of Pseudomonas aeruginosa, Staphylococcus aureus, Burkholderia cepacia complex and Aspergillus spp. occurred. Significantly, an incremental improvement in lung function was reported for transitioning patients with current P. aeruginosa infections.
http://tinyurl.com/gttvmj5

Reduced survival in adult cystic fibrosis despite attenuated lung function decline. Claire Keating, Armeen D. Poor, Xinhua Liu, Codruta Chiuzan, Daniel Backenroth, Yuan Zhang, Emily DiMango. Journal of Cystic Fibrosis. January 2017 Volume 16, Issue 1, Pages 78–84

There is limited data on disease progression and survival in adult diagnosis cystic fibrosis (CF). This study evaluates change of lung function over time and rates of death/lung transplant in adult diagnosis CF. The authors determined that lung function declines at a slower rate in adult diagnosis CF. However, particularly in those with low lung function at diagnosis, rates of death or transplant in adult diagnosis CF after 10 and 15 years is not negligible.
http://tinyurl.com/j4z4tev

Impaired rib bone mass and quality in end-stage cystic fibrosis patients. Mailhot G, Dion N, Farlay D, Rizzo S, Bureau NJ, Jomphe V, Sankhe S, Boivin G, Lands LC, Ferraro P, Ste-Marie LG. Bone. 2017 Feb 22;98:9-17

Advancements in research and clinical care have considerably extended the life expectancy of cystic fibrosis (CF) patients. However, with this extended survival come comorbidities. One of the leading co-morbidities is CF-related bone disease (CFBD), which progresses with disease severity and places patients at high risk for fractures, particularly of the ribs and vertebrae. The combination of reduced bone mass, altered microarchitecture, imbalanced bone remodeling (maintained bone resorption but decreased formation), increased microdamage and a small increase of the degree of mineralization, may lead to decreased bone strength, which, when coupled with chronic coughing and chest physical therapy may provide an explanation for the increased incidence of rib fractures previously reported in this population.
http://tinyurl.com/hht4au4

BACTERIA

Longitudinal study of Stenotrophomonas maltophilia antibody levels and outcomes in cystic fibrosis patients. Jillian Wettlaufer, Michelle Klingel, Yvonne Yau, Sanja Stanojevic, Elizabeth Tullis, Felix Ratjen, Valerie Waters. Journal of Cystic Fibrosis. January 2017. Volume 16, Issue 1, Pages 58–63

Previous studies have shown an association between higher Stenotrophomonas maltophilia antibody levels and decreased lung function in patients with cystic fibrosis (CF). The purpose of this study was to assess the serologic response to S. maltophilia over time and to determine whether changes in antibody levels could predict clinical outcomes. It was determined that S. maltophilia antibody levels may be helpful to identify individuals at risk of exacerbation who may benefit from earlier antimicrobial treatment.
http://tinyurl.com/hrrw5ud

Change in Pseudomonas aeruginosa prevalence in cystic fibrosis adults over time. Crull MR, Ramos KJ, Caldwell E, Mayer-Hamblett N, Aitken ML, Goss CH. BMC Pulm Med. 2016 Dec 7;16(1):176.

Little is known about risk factors for chronic and mucoid Pseudomonas aeruginosa (Pa) infection in cystic fibrosis (CF) adults, and whether the prevalence is changing. The odds ratio of mucoid Pa infection was significantly less in individuals with better baseline lung function and those diagnosed after the age of 25. The prevalence of chronic and mucoid Pa is decreasing. Larger studies are needed to confirm these regional trends and their significance.
http://tinyurl.com/z4zg49z

TREATMENT

The cumulative effects of intravenous antibiotic treatments on hearing in patients with cystic fibrosis. Angela C. Garinis, Campbell P. Cross, Priya Srikanth, Kelly Carroll, M. Patrick Feeney, Douglas H. Keefe, Lisa L. Hunter, Daniel B. Putterman, David M. Cohen, Jeffrey A. Gold, Peter S. Steyger. Journal of Cystic Fibrosis. Article in press

Aminoglycosides (AGs) and glycopeptides are antibiotics essential for treating life-threatening respiratory infections in patients with cystic fibrosis (CF). The goal of this study was to examine the effects of cumulative intravenous (IV)-AG (amikacin and/or tobramycin) and/or glycopeptide (vancomycin) dosing on hearing status in patients with CF. Participants in the hearing loss group were significantly older than those in the normal-hearing group. After adjusting for gender and age at the time of hearing test, participants in the two highest-quartile exposure groups were almost five times more likely to have permanent sensorineural hearing loss than those in the two lowest-quartile exposure groups. There was a small group of CF patients who had normal hearing despite high exposure to IV-antibiotics.
http://tinyurl.com/hwe82jj

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Implementing Standardized Palliative Care Education for Children and Adults with Cystic Fibrosis. Claire Hailey, BS; Mary Prieur, PhD; Sarah Helms, PhD; Howard Schmidt, MD; Scott Carney, MD; Elisabeth Dellon, MD MPH. Journal of Pain and Symptom Management. February 2017. Volume 53, Issue 2, Page 427

People with cystic fibrosis (CF) can benefit from early introduction to palliative care (PC) while living with a life-limiting, progressive illness. However, currently there is no standardized approach to inform people with CF about PC. The authors' objectives were to test the feasibility of incorporating standardized PC education into routine CF care and to assess changes in knowledge and perceptions of PC after receiving education. Results of a 30-minute PC education session suggest that this standardized training period effectively increased patient and caregiver knowledge and established positive perceptions about PC. These findings indicate that PC education is well-received by adolescents and adults with CF and caregivers and it can be provided efficiently during outpatient CF care.
http://tinyurl.com/j82ejn2

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TILLMAN continued from page 47

Phage therapy is highly effective against chronic lung infections with Pseudomonas aeruginosa. Waters EM, Neill DR, Kaman B, Sahota JS, Clokie MR, Winstanley C, Kadioglu A. Thorax. 2017 Mar 6

With an increase in cases of multi-drug-resistant Pseudomonas aeruginosa, alternative and adjunct treatments are needed, leading to renewed interest in bacteriophage therapy. There have been few clinically relevant studies of phage therapy against chronic lung infections. Using a novel murine model that uses a natural respiratory inhalation route of infection, the authors show that phage therapy is an effective treatment against chronic P. aeruginosa lung infections. They also show efficacy against P. aeruginosa in a biofilm-associated cystic fibrosis lung-like environment. These studies demonstrate the potential for phage therapy in the treatment of established and recalcitrant chronic respiratory tract infections.
http://tinyurl.com/hohtbhs

Effect of probiotics on respiratory, gastrointestinal and nutritional outcomes in patients with cystic fibrosis: A systematic review. Jacqueline L. Anderson. Journal of Cystic Fibrosis. March 2017 Volume 16, Issue 2, Pages 186–197

An increasing body of research investigating the use of probiotics to improve health outcomes in patients with cystic fibrosis (CF) prompted the need to systematically assess and summarize the relevant literature. The findings suggest that probiotics may improve respiratory and gastrointestinal outcomes in a stable CF clinic population with no reported evidence of harm. There is inadequate evidence at this time to recommend a specific species, strain or dose of probiotic as likely to be of significant benefit.
http://tinyurl.com/hbnnkqn

LUNG TRANSPLANT

Cancer risk among lung transplant recipients with cystic fibrosis. Aliza K. Fink, Elizabeth L. Yanik, Bruce C. Marshall, Michael Wilschanski, Charles F. Lynch, April A. Austin, Glenn Copeland, Mahboobeh Safaeian, Eric A. Engels. Journal of Cystic Fibrosis. January 2017. Volume 16, Issue 1, Pages 91–97

Previous studies demonstrated increased digestive tract cancers among individuals with cystic fibrosis (CF), particularly among lung transplant recipients. The researchers describe cancer incidence among CF and non-CF lung recipients. They determined that CF recipients have increased risk for colorectal cancer, suggesting a need for enhanced screening.
http://tinyurl.com/hrew5j5 ▲

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