

## Listen Up! The Importance Of Monitoring Hearing While Receiving Aminoglycoside Treatments

By Angela Garinis, Ph.D.,  
CCC-A, Research Audiologist

As an audiologist, my primary goal is to improve the quality of life of patients with hearing loss. One way of categorizing hearing loss is whether this sensory loss is congenital or acquired. Congenital hearing loss is present at birth (or shortly thereafter) with a genetic etiology and is ultimately unavoidable. For these cases, the audiologist focuses on providing therapeutic options (e.g., acoustic amplification or electric stimulation) and coping strategies to minimize the impact of hearing loss. In contrast, acquired hearing loss typically occurs after birth and is often due to an external event, such as ear infections, head trauma, disease, exposure to loud sounds and/or inner ear toxicity from certain medications.



ANGELA GARINIS

As a researcher, I have a strong interest in preventing acquired hearing loss, particularly those caused by specific, prescribed medications, i.e., ototoxicity. Ototoxicity occurs when a drug or chemical damages inner ear

sensory cells or the auditory nerve. Drug-induced hearing loss initially affects the higher frequencies (pitches), and may progress to lower frequencies important for speech and language with continued dosing. This is of particular concern in younger pediatric patients who are still developing their communications skills. Patients with cystic fibrosis (CF) are at risk for acquiring hearing loss due to the obligate intravenous administration of ototoxic aminoglycoside antibiotics that are critical for treating life-threatening bacterial respiratory infections. To date, there are no established clinical protocols to determine which patients are at high risk for ototoxicity. Currently, physicians primarily rely on patients' reports to identify risk. An objective measure to determine who is susceptible to ototoxicity based on

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## EDITOR'S NOTES

**A**t last, summer is in full swing here in the great Northwest. We have been blessed with wonderful weather. The daytime highs have been from 70 to 90 degrees, with most days in the upper 70s. That is just about perfect in my opinion. Hallelujah!

We have many interesting articles in this issue. I hope you have read the front page article on ototoxicity written by audiologist researcher **Angela Garinis**. This is one of the problems that can affect many of us, and we appreciate her sharing this information with us.

In "Ask The Attorney," **Beth Sufian** answers questions from our readers about SDI, COBRA and Medicaid. In the "Conversation Corner," **Bill Coon** writes about hospitalization insurance, specifically AFLAC, and how it can be a great help. In "Voices From The Roundtable," we have an introduction to the CF Reproduction and Sexual Health Collaborative (CFReSHC) written by **Sandy Sufian**, **Laura Mentch** and **Emily Godfrey**. They explain what it is and how you can become a part of it. **Meranda Honaker** gives a one-year update of her participation in a clinical trial in "Searching For The Cure."

Our Focus topic is Problem-Solving With CF. **Erin Evans** talks about solving the puzzle of living with CF, and **Jennifer Kyle** talks about keeping the fun in life with CF. **Julie Desch** continues the theme in "Wellness" with accepting helpers and learning gratitude. "Spirit Medicine" finds **Isabel Stenzel Byrnes** discussing spiritual problem solving through prayer. In "Parenting," **Dana Giacci** talks of labor and delivery with CF. I add my two cents worth about various problems in "Speeding Past 50."

"In The Spotlight" features **Suzanne Joyce** in an interview by **Andrea Eisenman**. **Aimee Lecointre** interviews **Zack & Farrel DeBaltzo** about nutrition and exercise in "Active For Life." **Lauren Hunsaker** offers us a look at CF through haiku in the "Poetry Corner." As always, **Laura Tillman** has compiled a lot of interesting "Information From The Internet."

Meet the new directors on pages 10-11. We are happy to welcome **Lise Courtney D'Amico** and **Sydna Marshall** to USACFA.

Be sure to check out page 34 for information on how to nominate someone for either the USACFA Founders Award or the Jacoby Angel Award. Also, look at page 18, where **Reid D'Amico** gives us information about the new USACFA Speakers Bureau.

Please look at page 3 in the "Looking Ahead" box. Our suggestions for future Focus topics are there. Perhaps you would like to write about one of them. Please consider it. This is your newsletter and we want to hear from you. We are happy to assist you, if you are concerned that you are not a polished writer. Give it a try. You may find that you are better than you think.

Stay healthy and happy,  
Kathy

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# Information From The Internet...

Compiled by Laura Tillman

## PRESS RELEASES

### CF Therapy Anabasum Reduces Acute Pulmonary Exacerbations Up to 75%, Clinical Trial Shows

Corbus Pharmaceuticals recently reported that its Phase 2 clinical trial examining multiple doses of anabasum (formerly known as JBT-101 or Resunab) compared to placebo in patients with cystic fibrosis (CF) achieved the trial's primary endpoint demonstrating an acceptable safety and tolerability profile at all doses, with no serious or severe adverse events. Secondary outcomes of the trial included evaluation of changes in lung function and bacterial composition of sputum.

The company reported that patients

treated with anabasum showed dose-dependent reductions in the number of acute pulmonary exacerbations. Compared to placebo-treated patients, those treated with the highest anabasum dose (20 mg orally, twice daily) had a 75 percent reduction in the annualized rate of pulmonary exacerbations. Treatment with anabasum also led to a reduction in patients' inflammation-associated sputum cells. Patients' lung function, as measured by forced expiratory volume in one second (FEV<sub>1</sub>), remained stable. According to Corbus these results demonstrate that anabasum treatment reduces pulmonary exacerbations in CF patients. They also provide clinical evidence of the drug's biological activity in

treating innate immune responses in the lungs. Anabasum is a synthetic, oral endocannabinoid-mimetic drug for inflammation, thought to increase the production of anti-inflammatory mediators while reducing the production of those that increase inflammation. Reducing inflammation helps to prevent permanent tissue damage in the lungs of people with CF. Anabasum has been designated an orphan drug for treating CF in the United States and Europe. <http://tinyurl.com/y7u74g6p>

### Laurent Pharmaceuticals Receives FDA and Health Canada Clearance to Initiate APPLAUD Phase 2 Clinical Study of LAU-7b for the Treatment of Cystic Fibrosis

Laurent Pharmaceuticals Inc. announced the clearance of its Investigational New Drug Application ("IND") with the U.S. Food and Drug Administration and the Clinical Trial Application ("CTA") with Health Canada to initiate APPLAUD (a dou-

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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](mailto:cfroundtable@usacfa.org)

**Summer (current) 2017: Problem-Solving With 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.) How is your CF affecting your decisions about becoming a parent? Whether you already have children or are wanting to start a family, tell us about your journey to parenthood.

**Spring (May) 2018: Maintaining Mental Health With CF.** (Submissions due March 15, 2018.)



# ASK THE ATTORNEY

## Questions From Readers, Answers From The Attorneys

By Beth Sufian, J.D.

A person, family member or member of the CF Care Center team who has questions about the legal rights of a person with CF can contact the CF Legal Information Hotline directly by e-mail at [CFLegal@sufianpassamano.com](mailto:CFLegal@sufianpassamano.com). E-mailing is best. A specific time to talk to an attorney can be scheduled. If you do not have access to e-mail, you can call 1-800-622-0385.

CF Roundtable readers had these questions in the past three months.

**1. Question: I am trying to decide if I should stop work and apply for Social Security Disability benefits. I also paid for a private long term disability benefit through my employer. What are things I should consider when making this decision?**

Answer: There are many things a person should consider when deciding to stop work. The following is not an exhaustive list but will give readers things to consider when consulting with their CF Care Center team. Some CF Care Centers mistakenly think that a person must have a very low FEV<sub>1</sub> in order to be eligible for Social Security benefits or private long term disability benefits. This is UNTRUE. There are a number of medical criteria that Social Security takes into consideration including an increase in hospital stays or IVs and an increase in the number of hours it takes to perform medical treatments. Private long term disability policies typically have a more general requirement that the employee show she cannot work full time due to her medical condition.

It is important to educate the CF Center and discuss the reasons a per-

son thinks stopping work will help his or her health. While some people with CF want to work until their FEV<sub>1</sub> is dangerously low, some people with CF do not think that is the best path for them.

People with CF who want to stop work in order to preserve lung function should not be discouraged by their CF Care Center. Some adults with CF have decided that they want to preserve lung function so that when new medications to treat CF become available the person will be in the best shape to benefit from the medication. While not everyone may decide that is the right path, those who feel they would benefit from more time to take care of themselves should be supported and encouraged by their CF Care Centers.

Additional considerations are:

-Does the person feel she does not have enough time to do all of her prescribed medical treatments each day?

-Does the person with CF need more time to take care of himself to maintain his health?

-Has the person had more hospital stays for IV treatment in the prior year?

-Has the person experienced a decline in pulmonary function in the past year?

-Does the person's financial situation allow her to stop work?

-Will the person have access to insurance coverage once she stops work?

**2. Question: I have stopped work so I can take better care of myself. Once I start receiving Social Security Disability Insurance benefits I will become eligible for Medicare, 29 full months after my date of disability. I do not know if I should continue the health insurance I receive from my employer under COBRA. COBRA will allow me to continue my employer-sponsored insurance for a cost of \$500 a month.**

Answer: This column is being written on June 20, 2017. The Senate has just released its proposed healthcare bill and indicated a vote will be held before July 1, 2017.

There is concern that some insurance companies that offer health insurance on the Affordable Care Act (ACA) healthcare exchanges may stop offering plans if Congress passes a new law that takes away protections the ACA put in place in 2010. A few insurance companies have already announced they will not sell plans in a few states in 2018. Insurers have said that the uncertainty surrounding Congress's plans to make significant changes to the Affordable Care Act premium subsidies and cost-sharing reduction payments is a major factor contributing to instability in the



BETH SUFIAN



health insurance marketplaces.

The subsidies reduce deductibles, copays and coinsurance payments for some low-income people who purchase health coverage on the health insurance exchanges. In the past few months, the current administration has publicly stated it is considering **discontinuing the payments**, to gain leverage in its efforts to repeal the Affordable Care Act.

“Even if you live in a state or a market that still has multiple carriers participating in it, if the Trump administration decides to pull the plug on cost-sharing reduction payments, all bets are off,” says **Sabrina Corlette**, a research professor at Georgetown University’s Center on Health Insurance Reforms.

A person with CF who purchases a policy on an ACA Healthcare Exchange faces uncertainty regarding whether that policy will be offered in 2018. Of course if that is the only insurance option, then it is better to purchase a policy than to have no insurance coverage.

However, electing to COBRA an employer-provided health insurance policy that has provided good coverage while the person with CF was employed may provide more certainty than purchasing a policy on a Healthcare Exchange.

A person who works for an employer with 20 or more employees can elect COBRA for 18 months, if he leaves his job or is terminated. If the person applies for Social Security benefits and is approved within 18 months of stopping work, then the person may receive an additional 11 months of COBRA coverage. After 29 months of being eligible for Social Security Disability Insurance benefits, the person will become eligible for Medicare.

A person who elects COBRA, has to pay the full premium. Often employers contribute to the health insurance

*Those who feel they would benefit from more time to take care of themselves should be supported and encouraged by their CF Care Centers.*

premiums of employees while the employees are employed. When an employee stops work and elects COBRA, the person must pay the full premium. A small number of employers may keep paying for health insurance if the person has left work due to his health, but there is no legal requirement that the employer do so.

The Healthcare Exchange open-enrollment period begins on November 1, 2017. At that point a person could check and see what health insurance policies are offered for purchase. If a person enrolls in a plan by December 15, 2017, the coverage does not take effect until January 1, 2018. If there are policies that provide good coverage for a reasonable price, the person then could decide to enroll in a Healthcare Exchange Plan and when the plan goes into effect, COBRA coverage can be stopped. People should not drop COBRA coverage unless they are positive they will have other coverage that will start on the day the COBRA coverage is stopped.

Keep in mind once a person stops the COBRA coverage the person cannot re-enroll in the COBRA policy if he finds the new coverage he has purchased is not as good. Nothing in this answer is meant to direct any person to stop COBRA benefits or to elect COBRA coverage. The answer to this and all questions is meant to be information only. Each person must determine what insurance coverage is best for his or her situation.

**3. Question: Can a person with CF enroll in Medicaid if he is NOT**

**receiving SSI benefits?**

Answer: If the person lives in one of the 31 states or the District of Columbia that have **expanded Medicaid** coverage to adults with incomes up to 138 per-

cent of the federal poverty level (approximately \$16,000 for one person), then the person can check to see if she would qualify for Medicaid. The person must meet certain low income and low asset criteria in order to be eligible for Medicaid.

The Affordable Care Act made the provision of Medicaid to low income adults mandatory but the United States Supreme Court said that there could be no mandate that each state provide Medicaid to low income citizens. Medicaid expansion then became optional. While 31 states did expand Medicaid coverage to low income adults, 19 states did not expand Medicaid. In states that did not expand Medicaid the only way for an adult to obtain Medicaid is if the person is eligible for SSI benefits. A person who is pregnant and has low income and low assets can obtain Medicaid but only for the period during which the person is pregnant.

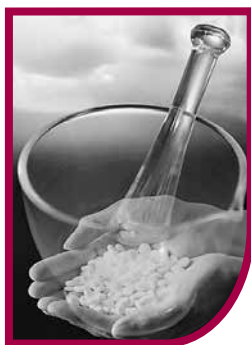
**4. Question: Can a person work full time while her application for Social Security Disability Insurance or SSI benefits is pending at Social Security?**

Answer: No. If a person works full time at any point during the application process, then the person is NOT eligible for Social Security Disability benefits.

If a person is working full time when he applies for Social Security Disability or SSI benefits, then the SSA application will be DENIED.

Under the SSA rules a person can

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# SPIRIT MEDICINE

## The Spirit Of Prayer

By Isabel Stenzel Byrnes

**T**oday's issue of *CF Roundtable* is about problem solving. Every moment of our day, people with CF are challenged to come up with solutions for their physical, social, emotional, financial and spiritual challenges. This disease makes us experts at problem solving! Today in "Spirit Medicine," however, I'd like to ponder spiritual problem solving...in the form of prayer.

When we have problems, it's natural to ask ourselves what we can do about them. We can tackle the problem, harness support persons, talk to experts—in essence, try our best to control the parameters around a problem to reduce its impact. What if we try to control everything we can, but the problem persists? What if the problem doesn't resolve in the time frame we want it to? What if there are uncontrollable elements of a problem? What do we do then?

Spiritually minded people from all religious backgrounds will say they pray. Prayer is focused attention to something or someone greater than themselves. They relinquish the problem to a higher power and ask God (or Buddha, gods, the Universe etc.) for guidance. Ninety percent of Americans say they pray—for their health or their love life or their final exams. Most prayers are a form of spiritual request for oneself or others. As my friend says, "I talk and God listens."

As someone with CF, I've prayed all my life for health. I've asked God to help me get through an important event, like high school graduation, my

wedding or my lung transplant. I've prayed for a cure for CF since I was nine years old. I especially prayed when I was scared, like when I had massive lung bleeds, a blockage, severe discomfort or other health crises. Since my transplant, my prayers have focused on gratitude to my donor family, for my ease of breathing, for my many adven-

tures since my transplant.

Recently, I had the privilege to go to Bethel Church in Redding, California, to attend their Healing Rooms with a few friends. There were hundreds of people gathered—some amputees, people in wheelchairs, as well as many with invisible conditions—who had faith that this kind of prayer would make them well.

There were people attending from all over the world. It was quite emotionally powerful to witness the faith and need for healing. I went with an open mind and curiosity, really to give thanks for what I've already received.

I was deeply touched by the concentration of prayer energy that was directed to each individual. We were ushered into a room where adults and children approached us and asked permission to pray for us. They put their hands on our shoulders and spent a few minutes praying for health. We were ushered to another room where each individual received more focused prayer by three church volunteers who asked us what our health prayer needs were. At the end of this gathering, our group was brought into another room and a facilitator asked who in the room experienced immediate healing. A few people volunteered to share that the pain in their leg disappeared, that their tinnitus of ten years disappeared, their vision cleared and so on. Unfortunately, the amputee did not grow a limb and people in wheelchairs did not get up and walk. Later that afternoon, my blood sugar was 300, so my diabetes was not cured.

*“Prayer allows us to surrender and relinquish our problems and this itself can offer a more peaceful and calm inner state.”*



ISABEL STENZEL BYRNES

Some of my friends are skeptical about this type of healing experience, saying that these sorts of healing prayers take advantage of vulnerable sick people. Thankfully, the experience was free of charge, which increased my trust in the process. To me, there is no harm to give and receive prayers, as long as it is all done unconditionally and without judgment. What struck me was the attention that the people who prayed gave to those who received the prayers. They firmly believed they had the power to channel God's energy to heal. Their directed compassion, love, care, devotion, good intentions and earnestness were palpable. And I appreciated that at the end of the session, we heard a sermon about being healthy in thoughts, words, relationships, diet, exercise and rest. Health is attitude and much of it is a choice.

Did I believe my CF, sinusitis, diabetes, back pain, osteopenia, skin cancer, myopia or arrhythmia would disappear after this session? No. I'm enough of a scientist to know that's not possible. (Some may say I lack faith.) Did I walk away feeling uplifted, grateful, hopeful, moved and graced? Yes. And that is a type of healing.

So, after this experience, I still ask, does prayer work? Everyone has an answer to that question. We all know some prayers come true, and some do not. As someone who counsels grieving people, often faith in God is rocked after the death of a loved one. People prayed for a cure or more time, but the loved one died anyway. Some people

turn away from God when prayers don't come true. I think many of us know that prayer can be helpful exactly in these times, to pray for strength, guidance, support and insight to understand how to navigate loss.

Our scientific culture has tried to understand prayer. In the late 1990s, there were a few studies that tried to see if intercessory prayer influenced recovery in people with heart surgery. At this time, all of those studies have been refuted because of flawed design. A 2006 randomized clinical trial by Harvard Medical School's Dr. Herbert Benson showed there was no difference in surgery recovery whether people were prayed for or not.

But maybe prayer can't be measured. As Michael Shermer in *Pseudoscience and Deception* writes, "scientific prayer makes God into a celestial lab rat." Maybe that's the point of faith. To believe in something regardless of proof or observable evidence.

I believe prayer serves many different purposes other than the desired outcome. Prayer is a direct path to Spirit. Prayer offers a one-on-one communication with a higher power. It is private, honest discourse. For some, it's praising God, or it's confessional, or gratitude, or supplication. Prayer allows us to surrender and relinquish our problems and this itself can offer a more peaceful and calm inner state. Prayer offers openness to mysterious experiences. It offers spiritual growth and insight through reflection and discernment. Prayer gives a sense of agen-

cy—we are exerting control over what might be uncontrollable. Finally, prayer disciplines the mind—it's a practice, sort of like yoga, for the mind. In fact, MRI studies by Andrew Newburg show that the frontal lobe, which is used for focus and concentration, lights up and the parietal lobes, used to process sensory information, darken during both prayer and meditation. The feeling of "oneness" with God or the Universe changes the brain.

People continue to pray day after day, decade after decade, century after century. Prayers come in as diverse forms as the people doing the praying. If prayer persists over time as a practice, then surely people must feel it is effective. In our little lives with CF, we have so much to "do" to take care of ourselves. Maybe adding prayer to our routines can reduce the burden of some of our treatments. In the CF community, we still have so many things to pray for. I pray for a "cure" for CF, I pray for a treatment for chronic rejection. I pray for reasonable drug prices. I pray for organ donors and their families. I pray for access to good health insurance. I pray my friends live long and happy lives. Some of these prayers are coming true. I feel like still being here is an answered prayer. Some prayers will not come true. Hopefully God hears them anyway. Prayer allows me to live in optimism, faith and hope. What does prayer do for you? ▲

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



## Mailbox

Thanks for the great articles.

Alice Todd  
Woodway, TX



## SPEEDING PAST 50

# Once It Is Fixed, It Isn't A Problem

By Kathy Russell

**W**ell, here we are with some summer weather, at last. Oh, how we have waited for this drier, warmer weather. I know that everyone thinks that it rains all the time in Oregon, but usually that isn't the case. However, it was "and then some" this year. I believe that I really may have rust on my skin and webs between my toes. It wasn't that we had so many deluges — although we did have a few of those — it was just that it kept on raining. It rained day-after-day for weeks. Egads! Enough, already! Now we are back to our days of sunshine — interrupted by a day or two of rain here and there — much easier to take.

I always feel better when the sun is shining. It makes me feel as if I have some energy, which I don't feel normally. With our somewhat dreary winters, I get something like seasonal affective disorder (SAD) and feel absolutely listless most of the time. I tend to stay home a lot during the cooler, wet weather. I just don't want to be around people who may have upper respiratory infections (URIs). Those pesky URIs can be so dangerous to me and to anyone who has compromised lungs. Staying home so much causes me to feel as if I never see the sun. We discovered that there are lamps that have bright light that mimics daylight. Some are especially for SAD and are quite expensive, but we found some that were easily affordable. By using those less expensive ones, I am able to avoid some of that listless feeling. This was a fairly simple fix to handle the problem.

Which leads right in to the Focus

topic of this issue of *CF Roundtable*, "Problem-Solving With CF." I am sure that we all face some problems because of our CF. Some of them are rather minor, but others may be tougher to deal with. I am very fortunate because my CF is what some docs have called "mild." I do not necessarily agree with that assessment. When I am ill, I am just as ill as anyone else who has CF. Since I have survived for more than 70 years, those disbelievers figure that my CF isn't very serious. This, sometimes,

has proven to be a problem for me.

When I have had to hire a new physician, I have had to go through the long, involved process of explaining how I can have CF and still be alive. (My current pulmonologist is the exception to this. He understood immediately, and did not doubt the accuracy of my diagnosis.) Some didn't make it past the first interview. (Yes, I interview a physician before hiring one.) Others *seemed* to understand, but they didn't *really* get it. I feel that one reason

I have been able to survive is that I have had some excellent docs working with me to keep me as healthy as possible. It is imperative that my docs understand that I will be a large part of my clinical care and that I will be

included in all decisions regarding my health. Once we get that clear, there is no problem.

As with all of us, I spend hours a day doing treatments and meds. When someone wants to meet with us early in the day, I may decline simply because I don't want to have to get up early enough to do my treatments before going to meet them. I am an old, retired person and I choose to sleep in until (as one of my friends used to say) the "crack of 10." My husband and I worked evening shift for many years and that schedule is what is comfortable for me. I like to do things in the late evening, when it is quieter and I am less apt to be bothered by phone calls. We don't turn out the lights until about 1:30 a.m. I get my eight hours of sleep and wake up refreshed and happy. I always hated being awakened by an alarm. It just seemed so harsh. Awakening naturally has been ever so

*In an effort to protect my ability to hear, I try to be mindful of things that could damage my hearing.*



KATHY RUSSELL



much more pleasant.

So now I'll go back to the problem of meeting people early. If they want to meet for breakfast, I simply suggest that we meet for lunch instead. That way I can sleep in a little later, get my meds done and still meet them at a reasonable time. By the way, I make all of my doctor appointments in the afternoon, if at all possible. I really, really don't like to have to awaken early.

I just remembered one of the things that was a real problem for me, when I was young. My feet sweat! I don't mean a little sheen of perspiration. I mean they pour out buckets of salty sweat. When I was working, I had to wear white, tie shoes. My sweat would eat the leather so fast that I had to buy new shoes every six weeks or so. That got a little pricey. Fortunately, I could deduct that cost from my income taxes. Now when I sweat out a pair of shoes, it is just a pair for the trash. My solution to the problem of wet feet was to wear slip-on clogs from Scandinavia. I had some that had punched leather uppers. The soles were made of cork and they were adequately supportive. Those lasted me a little longer and were very comfortable, as well as stylish.

Since my feet still tend to be wet all the time, I wear cotton socks and open sandals most of the time. I have custom orthotics that fit into my sandals and they give me good support. It would be nice to be able to wear fashionable shoes, but I think it is more important to be comfortable than stylish.

Chronic sinusitis is another problem that I have dealt with most of my life. Before I had my first sinus surgery, the pain was almost indescribable. It really limited my enjoyment of life. After that first surgery, I realized how negatively my life had been affected by my bad sinuses. It was wonderful to be able to move my head without pain and to be able to breathe through my nose. Of course, the polyps returned

and I had to have more surgeries. After my last surgery, I asked my ENT doc about irrigating my sinuses. We discussed it and came up with a plan that includes irrigating with a sterile saline solution and with another solution that includes sterile glacial acetic acid. I have been doing those irrigations for the past 21 years and have had no further recurrences of my sinus problems. Wahoo!

Another problem that goes right along with sinus troubles is ear woes. Like most of us, I have taken a lot of antibiotics in my life. It seems to me that I was on antibiotics almost continuously from the time I was a toddler until I quit working outside of my home, when I was in my 30s. Since I was home most of the time and was being exposed only to the germs of my own family, I suggested to the doctors that I might do all right without the constant dosing of antibiotics. I did have to be on them periodically, but I stayed off them for the most part. Even with the periodic respites from antibiotics, I still have issues with my ears and my hearing.

As I age, I expect to become more hard of hearing, we all do. However, the ringing in my ears (tinnitus), the difficulty with understanding what is being said and balance problems may all be attributable to antibiotic use. In an effort to protect my ability to hear, I try to be mindful of things that could damage my hearing. I had the audiologist at my ENT doctor's office mold ear plugs for me to wear when I use my nebulizers or any other loud equipment. My insurance did not cover the cost of ear plugs, but it was worth it to me to pay the price. As I recall, it wasn't that expensive anyway and my hearing is worth protecting. They are small and have a convenient carrying case. I make sure to stick them in my purse when we travel. They are especially good to wear on airplanes. They

cut out the majority of the noise and I still am able to hear conversation.

Insurance coverage can be another big problem for us. I am very fortunate to have excellent health insurance through my husband's retirement benefits. It isn't a cheap policy (between Medicare and our supplemental policy, we pay about \$6,000 per year), but it is a good policy. Every year, when it is time for open enrollment, we study all the information we are sent and figure what our costs are likely to be. Then we decide which insurance we will take. We stick with our current insurance because it has the best coverage for my medicines and durable equipment. On our income, we wouldn't be able to afford all of that without good insurance.

Even though it is a good policy, we sometimes hit a snag. Maybe they don't want me to take a specific medicine or they don't think I need some other med or treatment. If I am covered for whatever they are balking at, we appeal. That means there will be hours spent on the phone and paperwork to be completed. Fortunately, I am able to stay out of most of those negotiations, because my husband, Paul, serves as my advocate. He is my knight in shining armor when it comes to battling with oxygen providers, pharmacies, appointment desks and insurance companies. He spends many hours on the phone, talking with the various people who are making life more difficult for me (us) and keeps at it until he gets satisfaction. He does this not just to get Medicare and our health insurance to pay for what is covered under our policies, he also works with trying to get the prescriptions that my physicians order and getting my oxygen and the equipment for it when I need it.

Mentioning my oxygen reminds of one more problem. The cannula that carries the oxygen to my nose makes dents in my face while I'm sleeping.

Continued on page 18

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# New Director Of USACFA

**H**ey ya'll! My name is Sydna Marshall. I'm 36 years old and was diagnosed with CF at 11 when my adenoids were removed due to continual sinus and allergy problems. I am born and bred in Austin, Texas. I obtained my bachelor's degree in biology from The University of Texas at Austin, in December 2001. For the last 20 years, I worked in various capacities, including Marketing Assistant, for a boutique civil litigation law firm in downtown Austin. I recently retired for health reasons. In addition to CF, I'm also managing Graves disease, osteopenia and chronic sinusitis. I should be crowned the queen of CF sinus complications as I've had five-plus sinus surgeries, including a frontal obliteration.

I discovered yoga over a decade ago when I first read an article that Hatha yoga practiced two to three times weekly can increase breathing capacity by 70 percent. I haven't always had the chance to be consistent with my asana practice due to hospitalizations and surgeries. However, the breath work, self-acceptance and self-awareness inherent in any yoga practice have been life-saving tools

in handling the many CF challenges with resilience and grace.

I've met many people with CF, CF caregivers, doctors and more in my jour-



**SYDNA MARSHALL**

ney with CF. I endeavor to find the silver lining with each obstacle, with the hopes that younger people with CF will discover that CF doesn't define you as a person. Each of us is dealt our own deck

of cards, some easier than others to manage. The key is recognizing that most of life is how we respond to the deck of cards we're given.

I met and married my best friend and love of my life, Adam, while working at the law office. We have one loyal, feisty fur-baby boxer-mix named Husker (this Texan bleeds Nebraska red). Music is a huge part of our lives together, and we love the variety of live music around Austin and the surrounding hill country. On any given day you can find us spending time with family and taking in as much as possible. I've yet to meet a coffee shop or bookstore that I didn't adore from the moment I entered. I love the twinkly lights at Christmas as they elucidate not only the joy of the season, but the stillness of the moment. I love capturing the divine magic in that moment on camera. When I'm not staring at the lights around me, reading, practicing yoga on my mat, listening to music or spreading CF awareness, you can find me in the kitchen. Whole Foods is one of my happy places. Food, music and a good book will always nourish my soul.

E-mail: [smarshall@usacfa.org](mailto:smarshall@usacfa.org) ▲

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**TILLMAN** continued from page 3

ble-blind, randomized, placebo-controlled, Phase 2 study of the efficacy and safety of LAU-7b [fenretinide] in the treatment of cystic fibrosis in adults). This Phase 2 trial aims to evaluate LAU-7b's effect on the preservation of lung function in patients with cystic fibrosis by reducing persistent unresolved inflammation in the lung and stimulating its return to homeostasis. LAU-7b is a once-a-day oral pro-resolving therapy with potential to treat chronic pulmonary inflammation that leads to irreversible lung damage in patients with CF, regardless of their CFTR genotype. As opposed to tradi-

tional anti-inflammatory approaches that focus on the inhibition of the inflammatory response, LAU-7b works by using the body's own ability to resolve inflammation, which is a more natural way to modulate inflammatory response without directly interfering with defense mechanisms.

<http://tinyurl.com/ydxvw7o>

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## Clinical Trial Planned for Cystic Fibrosis Bacteria Treatment AIR001

The University of Pittsburgh has agreed to conduct a Phase 1 and 2 clinical trial of Aries Pharmaceuticals AIR001 for treating *Pseudomonas aeru-*

*ginosa* bacterial infections in patients with cystic fibrosis (CF). AIR001 is a sodium nitrate solution for intermittent inhaling. The formulation has been shown to benefit patients by dilating their blood vessels, reducing inflammation and slowing the growth of undesirable cells. Research has indicated that nitrite inhibited *P. aeruginosa* growth in primary cystic fibrosis airway cells at concentrations compatible with AIR001. This finding suggested that AIR001 could be a potential novel therapy for chronic *P. aeruginosa* infection in conditions such as cystic fibrosis. The main objective of the Phase 1/2

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# New Director Of USACFA

**H**il! My name is Lise Courtney D'Amico. I am excited to be a member of the USACFA board. I love reading *CF Roundtable* because I always gain so much knowledge from its pages. It is truly an honor to be able now to contribute to the publication myself.

I am 23 years old and I was diagnosed with cystic fibrosis at the age of two. I have an older brother and a two-year-old nephew, neither of whom has CF.

I grew up in the Washington, DC, metro area, which is where I currently live. After graduating from high school, I attended Boston College (BC), where I studied math and economics. I graduated from BC in May 2016, with a

degree in both math and economics. Now I work as an economic consultant specializing in antitrust and competition cases.



**LISE COURTNEY D'AMICO**

I have been blessed with fairly stable health throughout my life. I strongly believe in exercise as a form of airway clearance. When I am not at work, you can usually find me at the gym trying to clear my lungs. I also enjoy trying new restaurants, cooking and attending Orioles and Capitals games. In addition, I am involved with the CF Foundation and host several fundraisers a year. This year I am even training for the CF Foundation's Xtreme Hike in September.

I am so excited to be part of the USACFA board and I cannot wait to get to know the CF community even better.

E-mail: [ldamico@usacfa.org](mailto:ldamico@usacfa.org) ▲

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trial is to assess AIR001's safety. Researchers will explore its effect on lung function, exhaled airway nitric oxide and bacterial density.  
<http://tinyurl.com/ybnjbqne>

## Clinical trials pave way to new cystic fibrosis drug

In Phase 3 trials, Vertex's new Tezacaftor/Ivacaftor combination drug did not have any of the adverse pulmonary effects associated with another Vertex drug, Orkambi (Lumacaftor/Ivacaftor). Researchers think this new "corrector" combination will be better tolerated in patients in the long run because of the less frequent pulmonary side effects. Patients in the drug-treated group showed greater lung function improvement compared to the placebo-treated group. Additionally, patients in the drug-treated group did not report

more serious side effects than the placebo-treated patients. The exact mechanism of how Tezacaftor works is not known yet.  
<http://tinyurl.com/ycj2phz2>

## U.S. triples indications for Kalydeco without needing new clinical data

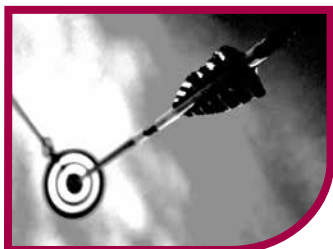
Vertex Pharmaceutical's Kalydeco was approved to treat 33 rare gene mutations – up from 10 – without new additional data being generated.  
<http://tinyurl.com/ycf5mh3l>

## Vertex Acquiring Cystic Fibrosis Treatment CTP-656 from Concert Pharmaceuticals

Vertex Pharmaceuticals is acquiring CTP-656, a potentiator treatment for the protein that is defective in cystic fibrosis, from Concert Pharmaceuticals. CTP-656, a deuterium-modified version

of the therapy ivacaftor, is a next-generation cystic fibrosis transmembrane conductance regulator (CFTR) potentiator. Potentiators are treatments that intensify the effects of other treatments. CTP-656 has received orphan drug designation in the United States, Europe, Canada and Australia. It has the potential to play a key role in once-a-day combination regimens of CFTR modulators as a treatment for cystic fibrosis. The deal also gives Vertex rights to all of Concert's other cystic fibrosis research and preclinical programs. Concert developed CTP-656 by using deuterium chemistry to modify Kalydeco (ivacaftor). CTP-656 is also a potential stand-alone therapy for CF associated with gating mutations of the CFTR gene. Results from a Phase 1 trial (NCT02599792) assessing the tolerabil-

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# FOCUS TOPIC

## PROBLEM-SOLVING WITH CF

# Putting The Pieces Together

By Erin Evans

Having CF can often feel like trying to solve a puzzle. A puzzle with many different, complex, colorful, weirdly shaped pieces. Sometimes there are pieces missing and you have to look everywhere to find them, otherwise you will have holes in your puzzle. CF is such a mystery, even now, after so many years of understanding the disease there is still no cure, which means there are still things we don't know about it. Even individually learning about your own CF and what works for you takes years of practice. What works for one person with CF may not work for another. Even siblings or twins with the same gene mutation may have very different successes with certain medications, therapies and overall lung function.

I have had almost 34 years of solving my own puzzle with CF and have found what works for me and what doesn't. It hasn't been an easy puzzle to put together though, and there was no one who could tell me exactly what would work and what wouldn't, since everyone's CF puzzle is so different. It is an ongoing process that takes your own willingness to really *want* to figure out. It wasn't until I was in my 20s and had my first hospital admission that the desire to really *want* to take on that role and responsibility set in.

The thing is, I have never been very good at puzzles. There is a patience that is required that I don't naturally possess. Having CF has made my idea of time slightly different from someone who doesn't have CF, or another chronic illness. There is a clock that's always ticking that I'm constantly in a race with. This is why sitting down,

“Taking a step back allows you to see things you may have missed or neglected and the things you've tried that aren't working.”



ERIN EVANS

sorting out all the pieces, slowly building the border and then finally filling in the middle, piece by piece, has always been challenging for me. I want to see the finished picture without the hours and hours of searching. But just like CF, you can't get there until you've put in the hard work.

It's ironic that the last few puzzles I've done have been while I've been in the hospital. Although I don't necessarily like going into the hospital, it does allow me to slow down and really take a step back to examine what I'm doing and how I can improve on my own CF care. I try to use my time in

the hospital not as a negative experience or a sign that things are getting worse but as a sign that it's time to slow down, take a deep breath, reflect and reevaluate.

Taking a step back allows you to see things you may have missed or neglected and the things you've tried that aren't working. Just like a puzzle you've been working for hours on, sometimes you forget what the end goal is and need to step back to remember and see what you're working toward. Having a clear idea of what you are working toward will drive your ambition to actually get there.

Over the years I have tried a number of things thinking they may be the missing piece to my CF puzzle. Some have helped and some haven't. Because of my gene mutation, the new CF drugs that now are on the market aren't an option for me. Instead, I have to keep trying things out, to see if they work for me. Living and succeeding with CF requires time and effort and an ability to accept that understanding your CF isn't going to be handed to you. Rather, it's a constant work in progress that you have to keep shuffling the pieces around in order to create the picture you want to see. ▲

*Erin Evans is 33 years old and has CF. She lives in Vermont with her amazing husband and dog. She works for the Cystic Fibrosis Lifestyle Foundation as the Program Coordinator. She can be contacted at [erin@CFLF.org](mailto:erin@CFLF.org)*



work part time while an application is pending and while the person receives benefits.

While the SSA website says part-time work is work less than 20 hours a week, SSA can find that working 20 hours a week is the equivalent of full-time work. In the past year SSA sometimes found that people with CF who are working at physically demanding jobs for 20 hours a week are able to work full time, given the strenuous job the person is performing.

Disability advocates recommend that if a person must work part time while their application for SSA benefits processes, it seems safer to work no more than 15 hours a week and work no more than four hours a day.

In addition, if a person works part time he cannot make more than \$1,170 a month (before taxes are taken out of the work check).

On October 6, 2016, SSA changed the medical eligibility criteria for SSA benefits for people with CF. Since that time, SSA is going through applications for benefits with a fine tooth comb. Each day it gets harder to be approved for SSA benefits. It is best to have the strongest application and to make sure that part-time work activity does not support a finding that the person could actually work full time.

Working part time increases the chance an application for SSA benefits will be denied. However, there are many people with CF who must work part time while an application for benefits pends at SSA in order to have money for food and rent. A person who works while an application for benefits is pending should make sure he does not work more than the allowable hours or for more than the allowable amount of money. ▲

*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.*

## SUSTAINING PARTNERS

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## PEARL SUSTAINING PARTNERS



## DIAMOND SUSTAINING PARTNERS





# Keeping The Fun In CF

By Jennifer Kyle

I am 52 years old living with cystic fibrosis (CF) that was diagnosed very late at the age of 37. Some would consider me new to CF having dealt with the medication schedule for only 15 years now. One area that continues to perplex me with this disease is that while we, as patients, clearly understand our medication schedule and how important it is to stick to, our friends and family members have difficulty understanding how it interferes with everyday life. I notice this most when I am on vacation or have company visiting, we are out having a great time and the day is quickly passing by us. When I announce that I need to go home to do my meds and have to stop the day short, I nonetheless hear the same reply, "Really? You have to go?" "Can't you stay just a little longer?" I know the people close to us want to be understanding about CF, but this is when I am faced with the problem that they really don't get how much it affects us. The answer to their question is, "Yes, I really have to go." I can't help sometimes feeling a touch of anger at this situation, but I am learning that this is life with a chronic illness. So off I go to do my meds knowing it's the right decision but disappointed that I am missing the fun.

Of course the other solution is to bring my meds with me, but this is not

always easy. That means preparing ahead of time, making sure there is a refrigerator or somewhere the meds can be kept cool and a place to plug in the nebulizer where the noise won't bother people. I once attended a weekend wedding and the hotel was so rustic that the only place to plug in my neb was in



JENNIFER KYLE

the bathroom and it was so small that I could not fit a chair in there. So the choice was to sit on the toilet or go down to the lobby to do my meds. There I was each morning seated on

the plush sofa in the lobby, as guests of the bride and groom said their hellos, while a plume of smoke surrounded my head. Next time I stay at a "rustic" hotel I am calling ahead to see just how rustic it is and then making my reservations at the Hyatt down the road.

I am very fortunate to have a few people in my life who do understand how CF affects everyday life. One of my closest friends never hesitates to take afternoon naps with me when we get together, and while spending a vacation in Disney World she would give me the time I needed in the mornings and evenings to get my treatments done without me feeling like I was missing anything. So the more we talk openly to our loved ones about this disease we will find people who can adapt to the daily rigors just like the patient. ▲

*Jennifer is 52 and has CF. She has two dogs, a golden retriever, Oscar, and a shepherd mix, Sadie. She dog sits to keep herself busy and volunteers as the Government Relations Chair for the Middlesex County Retired Educators Association of the New Jersey Education Association. She is recently divorced and her 81-year-old mother lives with her in Somerset, NJ. She retired eight years ago from teaching physical education, dance and health, a career she held for 16 amazing years.*



## Encourage Family and Friends to Sign Donor Cards

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patient factors (e.g., age, genetics, frequency of ototoxic treatments) would be highly valuable for CF patients.

Our research team at Oregon Health & Science University (OHSU) and the National Center for Rehabilitative Auditory Research (NCRAR) at the VA Portland Health Care System is dedicated to improving protocols for clinical monitoring of ototoxicity, as well as developing a screen for identifying “high risk” patients. We recently published an original research article in the *Journal of Cystic Fibrosis* entitled, “The Cumulative Effects of Intravenous Antibiotic Treatments on Hearing in Patients with Cystic Fibrosis (CF).” The study examined how lifelong exposure to intravenous aminoglycosides was related to the development of permanent ototoxic hearing loss. Our study group included 81 CF patients, ages 15 - 63 years, recruited from the OHSU adult and pediatric CF centers. All participants received a standard hearing test and were categorized into one of four groups based on their lifetime dosage of intravenous aminoglycoside antibiotics. We used two analytical strategies to determine if dosing was related to ototoxic hearing loss. The first strategy counted the total individual doses a patient received over his or her lifetime. The second strategy also incorporated the number of doses given per day.

Our results for both strategies revealed that increased lifetime intravenous dosing with aminoglycoside antibiotics was associated with a greater likelihood of drug-induced hearing loss in participants with CF. We also found that older participants tended to have a greater risk of hearing loss compared to younger participants. This was not surprising given that most CF patients receive their first course of treatment as a child once diagnosed with a severe respiratory infection caused by bacteria known as *Pseudomonas aeruginosa*. The frequency of these clinical treatments tends to increase with age and disease severity.

An unexpected but important finding was that some participants have normal hearing despite high cumulative ami-

noglycoside dosing over their lifetime. This suggests that some CF patients are “resilient” or have a higher threshold for developing drug-induced hearing loss. Understanding the factors (e.g., genetic, kidney function) that separate these individuals from other patients at high risk for ototoxicity is critical to implement an effective and cost-efficient ototoxicity monitoring program. Data from this study will lead to improved protocols for physicians to identify patients with CF who are at higher risk of drug-induced hearing loss. This will allow both the patient and physician to discuss possible modifications to their treatment regimen, particularly if alternative approaches are available. Persons with CF are living longer, increasing the importance of their quality of life, as hearing loss negatively impacts successful integration in family, academic, career and social activities leading to reduced quality of life and isolation. It is important to identify individuals at a higher risk for ototoxicity, so the physicians can start monitoring their hearing more closely to prevent hearing loss or the progression of an existing loss, when possible.

Patients with CF who receive intravenous aminoglycoside treatments should pay close attention to any potential symptoms of ototoxicity and share these with their doctor(s). These symptoms include ringing in the ears (tinnitus), difficulties listening in noise, balance issues, as well as hearing loss. Aminoglycosides are prescribed due to their potency and clinical effectiveness to treat life-threatening bacterial infections. Thus, patients should continue to use these treatments as directed by their care provider until an equally effective alternative medication can be identified or when safer dosing strategies are developed. Once a risk for ototoxicity is suspected, the patients should be referred to their local audiology clinics for routine monitoring of their hearing, particularly before, during and after intravenous aminoglycoside treatments. The audiologist can also inform the patients regarding rehabilitation (e.g., hearing aids, cochlear implants) and listening

strategies that reduce the impact of hearing loss on their daily life.

I hope our work will inform CF patients and their physicians about (i) the risks of hearing-related issues associated with routine intravenous aminoglycoside treatments, (ii) the effects of even mild hearing loss on quality of life and (iii) the resources available for their patients to seek information about hearing health-care (i.e., audiological services). I recognize that physicians use aminoglycosides to save lives, and this is ultimately essential. More importantly, I hope this work encourages both patients and physicians to discuss symptoms of ototoxicity before treatment begins so patients with CF can seek care early or be informed to report symptoms to their doctors as they occur.

I want to acknowledge the health-care workers who have dedicated their lives to extending and improving the lives of patients with CF. I have the pleasure of working with the providers at Oregon Health & Science University Adult and Pediatric CF Centers, and I have the utmost respect for the quality of care they provide to their patients. I hope this research continues to improve the quality of life of patients with CF (and others) as biomedical researchers continue to find a cure for CF.

*Disclaimer: The content of this article does not represent the views of the National Institutes of Health or the Department of Veterans Affairs. ▲*

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*Dr. Angela Garinis is a senior research associate and audiologist at Oregon Health & Science University with a joint appointment at the VA Portland Health Care System in the VA RR & D, National Center for Rehabilitative Auditory Research (NCRAR). Her primary research interests involve improving non-invasive techniques for the diagnoses of middle-ear and/or cochlear dysfunction. She also has a strong interest in the diagnoses and monitoring of ototoxic hearing loss in patients with cystic fibrosis receiving aminoglycoside treatments. Dr. Garinis may be contacted via e-mail at: garinis@ohsu.edu.*



# Women With CF Are Helping Guide Reproductive And Sexual Health Research

*By Sandy Sufian and Laura Mentch with Emily Godfrey*

**D**o you remember hearing about the Australian surfers who felt better by the ocean in the salt air? Their observation, shared with their doctors, led to nebulized hypertonic saline for CF, an effective and low cost treatment.

People with CF can make a difference. We are engaged patients who participate in our care, often problem solving with our doctors. And, we ask a lot of questions.

The story of this project begins with questions about reproductive and sexual health asked by Sandy Sufian, a woman with CF. “Why am I so sick around the time of my period?” Other women with CF ask the same question. The relationship is not well understood by our healthcare providers who often are unable to address this concern. The good news is that CF researchers have begun to study the influences of the female hormones estrogen and progesterone on CF health.

## **Our project begins with a friendship and leads to a collaboration.**

Sandy: Emily and I used to work at the same university in the same medical school so we used to go to lunch together and became friends. During our meals together, and at other times, I sought Emily’s “take” on my experience and theory that women’s CF symptoms got worse during certain parts of our menstrual cycles and that female hormones contributed to lung exacerbations, GI issues, arthritis issues



**SANDY SUFIAN**

and sinus congestion. I’ve believed, since about 1999, that female hormones caused CF symptoms to get worse, but no doctor of mine, save one, believed me. I also knew friends who got hemoptysis before or during their periods. Because Emily is a family physician who specializes in women’s health, I turned to her to get her thoughts. Over the years, we continued to discuss this issue. When Emily spent a year at the CDC, she became interested in a project about contraception and its various effects upon women with chronic illness. She talked to me about this work and how CF was one of the diseases the CDC was looking at. After that, Emily approached me with a grant opportunity to work on questions about CF and the reproductive life course. Since her expertise is in family planning, we began with articulating a set of questions about contra-

ception and pregnancy, but then we expanded the list of important topics to include other issues like female hormones and menstruation, dating and sex, peri-menopause and menopause, body image, motherhood and decisions not to become a parent, among other issues. The grant was funded and out of our conversations with women with CF about these subjects, the Cystic Fibrosis Reproductive and Sexual Health Collaborative (CFReSHC) was born.

Emily: As a family physician and fellowship-trained family planning specialist, Emily Godfrey did not take care of patients with CF and thus she did not see how her work intersected with Sandy’s curiosity about issues related to menstruation and hormones and CF symptoms. This started to change when Emily served with the CDC, as stated in Sandy’s story above. The idea of bringing women with CF together came after Emily attended a two-day seminar funded through the Eugene Washington PCORI (Patient-Centered Outcomes Research Institute, a non-governmental institute) Engagement Awards Program about patient-engagement. It was at the workshop that Emily experienced the rich environment in which patients participate as equal partners as researchers and clinicians, and found out about the Pathway to Proposal funding mechanism through PCORI. Emily had her “Aha!” moment! She finally figured out how she could connect her work as a primary care reproductive health specialist with Sandy, a woman with CF and disability specialist. Together, Emily and Sandy could bring their



expertise to build a collaborative of women with CF to help change their lives through the development of research questions and proposals that matter to women with CF!

Emily, moving forward to help her friend and seek answers to their longtime conversations, applied for and received funding from PCORI. A non-governmental institute, PCORI was created as part of a modification to the Social Security Act by clauses in the Patient Protection and Affordable Care Act.

Our project seeks to understand and identify reproductive and sexual health issues important to women with CF and develop, from patient priorities, questions that may lead to research.



**LAURA MENTCH AND EMILY GODFREY AT THE CFRI CONFERENCE, 2016.**

#### **The objectives are to:**

**ENGAGE** patients as equal partners with researchers at every step of the project.

**DESCRIBE** areas of reproductive and sexual health that need to be improved for women with CF.

**DEVELOP** Comparative Effectiveness Research (CER) questions that address the priority areas chosen by patients and researchers.

Women with CF will help frame and guide the development of the project. This is a departure from our singular role as research subjects and calls forth the disability rights movement saying: “Nothing about Us, Without Us.”

We began by creating a core team and research advisory panel (RAP), each including women with CF, to guide the project. We also created a CF patient task force (CF-PTF), consisting only of women with CF and core team members to help facilitate discussion sessions. We selected the World Café format to conduct patient task force meetings. World Café brings people from invested communities together in small groups to discuss their concerns related to their common problem. This allows the voices of the community to

be expressed and heard. The World Café design was adapted to allow women with CF from all over to come together in a virtual environment, free from cross-infection concerns.

Women from the CF community were invited to participate through announcements in a *CF Roundtable* blog\* and CF Facebook groups.

\* “Announcing a Patient-Engagement Study for Women with CF about Their Experiences throughout the Reproductive Life-course.” *CF Roundtable* blog published 8/15/16.

#### **Our World Café Experience**

We met from August through April, 15 women with CF in five World Café sessions. Each meeting began with a group introduction to the agenda followed by smaller sessions in “rooms” facilitated first by researchers, then by participants. Participants visited each room with facilitated discussion of issues within the broader topic. Returning to the larger group, we reviewed the discussion and determined priorities from the concerns raised. The group then voted on the

most important issues to include in future work. The World Café model supported us in exploring our questions and encouraged everyone’s contribution as each of us listened for and shared common insights. Honoring the experience and voice of each participant was essential.

Our World Café discussions brought forward the following issues we’ll continue to develop toward comparative effectiveness research (CER) to improve sexual and reproductive health for women with CF.

- Improving care coordination between CF Centers and women’s health providers
- Discerning effective and satisfactory treatments for menopause and perimenopause symptoms in women with CF
- Improving pregnancy planning for women with CF
- Improving and maintaining the health status of CF mothers
- Evaluating health risks and benefits for women using hormonal contraceptive methods

Continued on page 28

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# USACFA Announces A Speakers Bureau

By Reid D'Amico

**I**ncorporating the patient voice into CF discussions is an important and critical move toward inclusivity and understanding of our disease. Not long ago, people living with CF had very different outlooks on life. Many of the life-extending medications available today weren't yet discovered, and individuals with CF found themselves more likely to experience exacerbations and eventually respiratory failure. However, for the first time, we find ourselves at the brink of a tipping point in CF. With growing early diagnosis, increased access to healthcare and life expectancies on the rise, CF will no longer be concretely defined as a pediatric illness. Currently, 50 percent of people with CF in the U.S. are over the age of 18. The advent of adulthood in CF presents itself with a wonderful opportunity for awareness and advocacy.

In the CF community, we find ourselves to be a growing microcosm of society. Our community has teachers, engineers, lawyers, scientists and physicians all with the common thread of our experience with our unique disease. Our disease makes our stories powerful. We understand and go through struggles few can understand, and we learn to navigate through the world in origi-

nal ways. For this reason, the United States Adult CF Association (USACFA) has announced the start of its new speakers bureau program.

The purpose of the speakers bureau is to send a patient living with CF to speak at fundraisers, education days and other CF-related events. With our

strongly rooted in our identity with CF. Having the person who lives with CF present in conversation opens the exchange to increase awareness and also puts a face to an illness many do not understand. I've run into a few misconceptions about CF in my advocacy journey, and it's always been a joy to discuss

“With our community of adults growing, it's important for us to introduce the patient voice in all aspects of discussion.”

community of adults growing, it's important for us to introduce the patient voice in all aspects of discussion. Perhaps one of the most exciting parts of USACFA's speakers bureau is that it comes at no cost to the hosting organization. USACFA has budgeted its monthly opportunities to cover the cost of travel and lodging of all CF advocates it sends.

From personal experience working as an advocate in CF, I find it critical to have an individual with CF present in every dialogue. The way we word our thoughts, our outlook on life and our approach toward curing our disease is

these topics with those outside of CF. We offer a perspective few understand, and I firmly believe that there's little reason to not have an advocate present in any discussion.

USACFA has outlined a list of topics that speakers are willing to discuss. These range from clinical trials, education and working, to parenting with CF. However, the more than a dozen advocates who are interested in speaking can cover a wide range of topics.

If you or your organization are interested in having a CF speaker present at your event, please contact [rdamico@usacfa.org](mailto:rdamico@usacfa.org) or [lmentch@usacfa.org](mailto:lmentch@usacfa.org). ▲

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**RUSSELL** continued from page 9

Since my face is fat, those dents tend to stay all day. I really don't like having those marks on my skin. So I did something about that. I took a small scrap of fabric that came from a T-shirt and rolled it around the hose. It is just enough to "soften" the profile of the plastic hose and prevent it from making such an unsightly dent in my face. Voila!

Another problem with my oxygen use is the cannulas, themselves. A regu-

lar adult cannula is too large for my nose. The horns of the cannula stick into the sides of my nostrils. To avoid this, I use pediatric cannulas. They are narrow enough that they don't stick into my nose and they still are large enough that they fit around the septum. However, this has proven to be a problem when I ordered more cannulas. They kept sending adult cannulas, even though we ordered pediatric! I think

that this is finally resolved, too. Ah, the joys of using supplemental oxygen. But I won't complain, because it is nice to be able to breathe and do things.

I hope all of your problems are easily fixed.

Kathy ▲

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

## THROUGH THE LOOKING GLASS

### Therapy

An endless rhythmic beat  
A sort of faint knocking in the distance.  
The hands of a mutual therapist  
Pounding hard on the delicate chest  
Beating delicately on the hard chest.

A cruel and merciless treatment  
Squenching our precious time on this earth.  
Depleting such valuable energy from our souls  
As the timer ticks away - anyway.

Faster! Faster!  
Harder! Harder!  
Day in, day out  
A mindless routine sets in.

The anxiety of every breath grows stronger  
With each cupped pound.  
The aching back. sore muscles.  
The bleeding hands gloved for protection  
The cup overflows.  
What meaning gives this bizarre and  
undignified practice  
To the core of our existence?

"I love you!" says the beating rhythm.  
With each beat a reminder that you are no burden  
A cherished moment of togetherness  
Of deep connection between you and me.  
A synchronicity of beating arms  
Is a synchronicity of love and affection.

*-I. Stenzel, 1997*



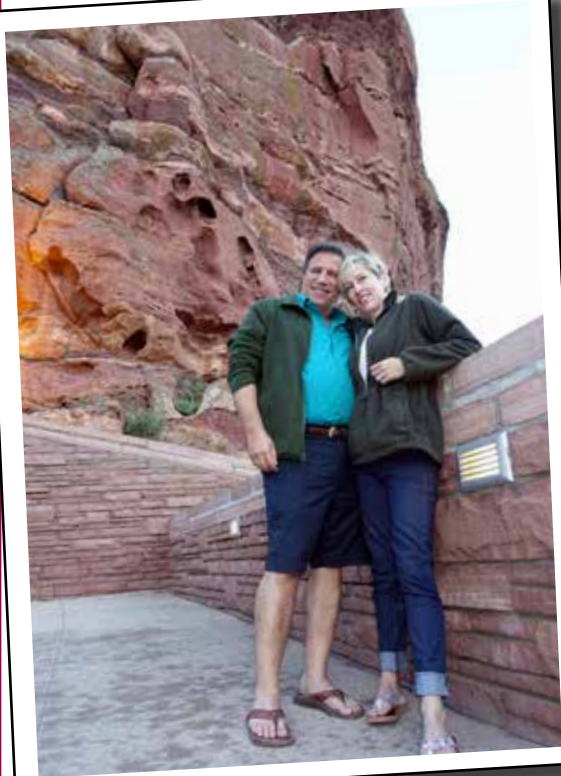
PHOTO BY DAVID MYERS

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"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...



**STEVEN BARSHOV AND SUZANNE JOYCE  
AT RED ROCKS IN COLORADO.**



**ZACK AND FARREL DEBALTZO ON THEIR HONEYMOON  
ROAD TRIP TO NASHVILLE.**



**JENNIFER KYLE, TAKING HER FIRST  
VACATION AFTER HER DIVORCE, IN  
DISNEY WORLD IN 2016.**



**JOHN AND JEANIE HANLEY AND PAUL AND KATHY RUSSELL  
ENJOYING AN OUTDOOR SUMMER LUNCH IN TUALATIN, OREGON,  
IN JUNE 2017.**





**THE HANLEYS FROM LEFT TO RIGHT - JESS, JOHN, MARIA & CARL (HENDRICKSON), JEANIE AND KEVIN HANLEY. AT MARIA'S WEDDING IN APRIL 2017 AT SOUTHERN OAKS PLANTATION IN NEW ORLEANS.**



**ALEXA GOLDMAN, LISE COURTNEY D'AMICO AND EMILY MEYER AT THE NATIONALS VERSUS ORIOLES GAME IN JUNE 2017.**



**ADAM KEYS AND SYDNA MARSHALL.**



**ERIN EVANS (MIDDLE) WITH HER OLDER BROTHERS ROSS AND JACKSON EVANS ON VACATION IN MAINE IN 2016.**



# ACTIVE FOR LIFE

## Meet Team Debo

By Aimee Lecointre

**F**or this issue I interviewed power couple Zack and Farrel of Team Debo. I asked them to share a bit about themselves and how nutrition and exercise play a role in managing their health with CF. They also share about their social media project Team Debo, which they created to help the CF community through food, fitness and faith.

Hey guys!

### **1) Can you share with us a little bit about yourselves and your Team Debo project?**

We are Zack and Farrel DeBaltzo and we're newlyweds who both have cystic fibrosis. We are in Florida and are on a mission to help change lives with faith, food and fitness! We created The Team Debo Project in early 2016, primarily as a social media project aimed at sharing our passion with other people who have CF, in hopes that they will be encouraged to implement new strategies into their own healthcare as well. I (Zack) have been a CrossFit trainer for four years now and I tackle the exercise aspect of our duo. While I (Farrel) focus more on sharing recipes, nutritional information and facts about foods and how they affect our health. Together, we are openly outspoken about our faith in Christ, our love for others and the things outside of the proverbial box that have gotten us this far in our fight. Ultimately, our goal is to continue to defy the odds for as long as God grants us on His earth, to continue to look for ways to serve those around us and to prove every day that love really is the cure for everything!

### **2) How has being physically active improved or played a role in your health?**

For myself (Zack), physical activity

decreased dramatically as I graduated high school and got away from organized team sports, which led to a steady decline in lung function well into my mid-twenties. At the end of 2011, I hit my "CF rock bottom." My PFTs had dropped to 28 percent and I found myself being evaluated for transplant at Cleveland Clinic. It was then that I decided to make numerous life changes, one of the most impactful being the choice to reintroduce daily physical activity. For a couple of years, I took up weightlifting and I noticed a dramatic weight increase but only small increases in lung function. In 2013 I found CrossFit, and in a matter of months, my lung function had jumped back up to my new baseline of 45 percent, where I continue to hover.

As for me (Farrel), I grew up in a family that dove headfirst into nutrition after my diagnosis, with the idea that if my body was fueled in a way that would efficiently combat infection, my care would be more preventative and

less damage control. I was raised on lean proteins, healthy carbs, every vegetable on God's green earth and healthy fats, while refined sugar was kept completely at bay. Contrary to how many clinics lead people with CF to believe, it was this kind of diet that kept me between a heftier 130-140 pounds throughout middle and high school. Then I fell off of the bandwagon in college. I adopted the easy, cheap, fast food lifestyle; dropped most of my weight, and found myself either hospitalized or on home IVs every few months with constantly fluctuating PFTs in the 60s and 70s for years. Fast forward to October of 2014—I had played this game for over a decade and I was sick and tired of being sick and tired, so I began to shift my focus to cleaning up my diet again. Eventually, I settled into a healthy nutrition plan again and as a result of God's grace and those efforts, my lung function climbed back up to my current baseline in the high 80s and I have few issues with weight management.

### **3) Zack, can you share some tips for those new to CrossFit, who may want to try it out for the first time?**

The thing that I love about CrossFit is that it truly is for everyone. It doesn't matter what your skill level is; the foundation of it is basic movements performed at high intensity. Functional movement, not strength, is the prerequisite to CrossFit and that can be scaled to any ability, age or body type. My advice is to shop around for a gym atmosphere that suits you. CrossFit prides itself on community, so every gym has its own personality, in most of which even introverts find themselves drawn out of their shells and able to fit in. Another big piece of



**ZACK AND FARREL DEBALTZO.**



advice is to do your best to focus on YOUR performance. It's easy to look around and compare yourself to others, but keep in mind that you're in there for you. So take notes, write down your scores and strive to be better than YOU were yesterday. In a good gym, you will notice how much everyone encourages each other. Seek that out. And don't be afraid to engage the coaches. That is why they are there.

**4) Farrel, I know you are passionate about nutrition, something we have in common outside of CF, can you please share one of your favorite post-workout snacks with us?**

I am a fan of pre-made shakes after workouts because they're easy to grab when you're sinking and you can create virtually any flavor that you like. One of mine and Zack's favorites is our Clean Apple Cinnamon Oatmeal Shake! Don't be fooled into thinking that "clean" means boring or not nutrient-dense enough for people with CF. This puppy is packed with 830 calories, 35g of protein, 50g of beneficial fat and 45g of healthy carbs (give or take moderate amounts, depending on choice of protein powder). Also, the avocado oil may sound daunting in a sweet shake, but the flavor is non-existent! The benefits, however, are countless!

To make Clean Apple Cinnamon Oatmeal Shake, simply blend the following ingredients with some ice and enjoy!

12 ounces water  
½ cup old-fashioned rolled oats  
3 scoops protein powder  
15 almonds  
1 medium apple  
3 tablespoons avocado oil  
Cinnamon to taste

I also asked Zack to share a few different workouts, for all levels, that could be done at home. Remember, go

at a pace that works for you, push yourself enough to work up a sweat, but not so much you'll set yourself back.

**Zack's Workouts**

**First Timer:**

8 round Tabata  
(20 seconds of exercise, 10 seconds of rest, 4 minutes total)  
Rounds 1 & 2 = jumping jacks  
Rounds 3 & 4 = lunges  
Rounds 5 & 6 = windmills (fingers touching opposite toes)  
Rounds 7 & 8 = running in place (heels to rear)

**Intermediate:**

"Cindy"  
(20-minute workout)  
5 pull-ups  
10 push-ups  
15 air squats  
As many rounds as possible (AMRAP)

**Seasoned (needs equipment):**

"Fight Gone Bad"  
3 rounds of -  
1-minute wall ball shots (20/14lbs)  
1-minute sumo deadlift high pulls (75/55lbs)  
1-minute box jumps (20/18")  
1-minute push press (75/55lbs)  
1-minute row (Calories)  
1-minute rest

Thank you Zack and Farrel for sharing with us!

You can follow along with Zack and Farrel on Instagram at @teamdeboforcf ▲

*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](mailto:alecointre@usacfa.org).*

**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](http://www.youcannotfail.com) to share your story, inspire others, and to become a part of this official program of the Boomer Esiason Foundation.





## PARENTING

# Labor And Delivery As A CF Woman

By Dana Giacci Rogers

**B**efore I begin this column on CF and parenting, I want to take a moment to remind our readers that this parenting column is a space created so that those CF patients interested in family planning can begin the conversation. That includes using this space for questions as well. I will always sign my articles with my e-mail for contacting me directly if there are questions you are curious about or topics on parenting you would like to see addressed in the next column. Again, this space has been created as an opportunity for our adult CF population to learn and grow from each other in an area that before wasn't talked about much. My experiences as a CF parent are singular. I have only my life to draw from. All CF adults beginning in family planning, becoming stepparents, desiring to pursue pregnancy or IVF et cetera, feel free to use this space as the beginning of that discourse in our larger CF community. If I do not have personal experience in it, I promise I will do my best to search it out.

With that said, this article is special to my heart because my husband and I welcomed our son into the world on May 31, 2017. Considering his birthday was only a week ago, I thought this might be the perfect time to talk about labor and delivery with CF. All stories that include bringing a human into the world are different and unique. There is no way to talk about ours, which is so personal, without excluding all the other stories which were/are so different. However, I do have a couple of suggestions based upon our experiences that hopefully some might find helpful. As a CF patient, you will

be considered high risk from an OB perspective. There are many things that can go awry when creating a human, let alone if you have a chronic lung disease that can impact the health and stability of the baby about to be born. Needless to say, you will be watched very carefully. Every six weeks you will need to be seen by either your CF doctors or your OB, I think is the general rule. Through my last pregnancy, I was very stable from a CF perspective. Therefore, I followed up mostly with my OB with the quarterly visit still to

the CF clinic. For us that included checking in every three to four weeks to check on the baby's growth. This is mostly due to the issues of CF and malnutrition as well as CFRD for some.

This brings me to my first suggestion: think about your birth plan. Both of my pregnancies were a planned induction. Our goal was always to make it to 37 weeks. At that point, the baby is considered fully grown and from there continues to put on a pound a week until delivery. Our doctor

felt that the risk did not outweigh the benefit of a chunky baby considering my lungs at that point were already crowded. When you are that big with CF, no matter what, your O<sub>2</sub> saturation needs to stay above 93 percent for the safety of your baby.

I was okay with this decision, it made logical sense to me and it was nice to know when my pregnancies would end. I found it easier to prepare and more realistic to plan pre- and post-doctor's appointments. Also, with more control over the situation, I was sure to have my doctor that I trusted on the delivery day, as well as my birth plan respected because of it.

When the day finally arrived, we were sent up to the labor and delivery floor. The first thing the nurses did was put sensors on the baby to monitor his heart rate, his movements, my oxygen saturation, my heart rate and my contractions. By the time you reach delivery however, you will be used to this process because of all the time you spend in fetal monitoring the last two months of your pregnancy. This is only to ensure there is no traumatic stress

*The most important thought perhaps  
as if anyone had any doubt.  
Make sure your staff and nurses  
are aware of your CF.*



DANA GIACCI ROGERS



on the baby during the labor process that might indicate the baby needs a be delivered more quickly via caesarian section.

My husband and I felt very strongly that a caesarian birth needed to be avoided. Considering the number of airway clearance and breathing treatments I do daily, we knew that abdominal surgery was no way to have success after our children were born. If you cannot take care of your CF, you will not be able to take care of your tiny person. This is always the basis of CF and parenting and it is true in all situations. This, and primarily this, is what our birth plan consisted of: No C-sections as long as it could be avoided. Not to mention, let's be honest, we CF women have strong abdominal muscles. We can make light work of delivery as long as our lungs are healthy.

Second suggestion: consider whether you will want pain management during the labor and delivery process. Once on the labor and delivery floor of our hospital which we knew very well and where our plan was well established ahead of time, we began the process of our induction. In both of my birthing processes my doctor and I discussed an epidural. I know a lot of women prefer not to have drugs the day of delivery, which I under-

stand. Whatever mom takes baby takes, and that is no small deal. However, because our primary goal was to avoid a C-section our doctor felt we might increase our chances of success if I am not already exhausted from pain by the time it came to finally deliver. I agreed, and both times I did have an epidural placed. Take the time to discuss whether or not an epidural is a part of your birth plan. Ask whatever questions you need ahead of time so that when you get to that moment the decision is not one you have to think about while already uncomfortable.

Final suggestion during the labor process: oxygen. The most important thought perhaps as if anyone had any doubt. Make sure your staff and nurses are aware of your CF. Certainly, your doctor will be aware of this, but the nurses and staff there to support you in the meantime might need the quick update. During the delivery of our oldest we found that each time I had a contraction my oxygen would dip. As my oxygen went down, so did our baby's heart rate. As they saw the heart rate continuing to dip, the quicker they wanted to take me for a caesarian section. However, my husband who sat there staring at the monitors the entire time, noticed this correlation, that amazing man. He requested that I be

put on oxygen, and sure enough, as soon as I was on oxygen the problem was resolved. With our second, I just asked for oxygen ahead of time. I didn't need it necessarily, my O<sub>2</sub> saturation was fine, but I figured at some point it wasn't going to hurt to have it, and it made things just that much more comfortable. Pay attention or have your birth partner pay attention to these things for you, or preemptively decide a little oxygen can't hurt. I know this experience is singular, but if you have a lung disease and you are entering a stressful situation for your body, make sure you have the tools you need to make it through safely.

From there it is a waiting game. Babies come into the world on their own time. It is your job to conserve your energy, stay as comfortable as possible and get that baby into the world safely for both of you. Easily, the two best days of my life, and I hope anyone desiring the experience will have the opportunity to do so.

Please feel free to share your stories or questions with us.

Sincerely,

Dana Giacci ▲

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*Dana is 27 and has CF. She lives in Troutdale, OR. You may contact her at: [dgiacci@usacfa.org](mailto:dgiacci@usacfa.org).*

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**TILLMAN** continued from page 11

ity, safety and pharmacokinetics (what the body does with the drug) of increasing single doses of CTP-656 showed that the drug was safe and well-tolerated by patients.

<http://tinyurl.com/ycjnz779>

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**Arcturus Therapeutics to Receive Up to \$3 Million from Cystic Fibrosis Foundation Therapeutics to Advance a Novel LUNAR-Formulated mRNA CFTR Therapeutic**

Arcturus Therapeutics, Inc., announced that it has entered into a research agreement with Cystic Fibrosis Foundation Therapeutics Inc. (CFFT), in which CFFT will pay up to \$3 million to advance LUNAR-CF, a novel messenger RNA (mRNA) therapeutic formulated with Arcturus's LUNAR™ delivery technology. If successful, it has the potential to treat all CF patients. We are now positioned to accelerate the development of our CF program and

expand our LUNAR™ platform to include pulmonary delivery of nucleic acids.

<http://tinyurl.com/yd3thnrf>

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**Celtaxsys announces full enrollment of its landmark EMPIRE-CF Phase 2b clinical trial assessing the potential of novel anti-inflammatory investigational therapy, oral acebilustat, to preserve lung function in CF patients**

Continued on page 27



# IN THE SPOTLIGHT

## With Suzanne Joyce

By Andrea Eisenman and Jeanie Hanley

**O**ur latest star comes from New York but winters in Florida, a true snowbird—and smart, as the weather benefits her health. She finds she gets sick less often and enjoys the warmth that allows her to stay more active in winter months. All of which are good CF strategies for physical and mental health. Speaking of which, we connected because she works a lot with improving her mental health by practicing and teaching yoga. I (Andrea) learned of her through her social worker who told me about techniques Suzanne is using: Hatha yoga and iRest Yoga Nidra. With others in mind who have CF and who could benefit from these relaxing modalities, I thought it was worth investigating and asked her to share her experiences. Read how she became deeply involved in yoga after having two demanding careers.

Please welcome our newest star, Suzanne Joyce. Spotlight, please!

**Age:** 54

**Diagnosed when?** I was two years old when I was diagnosed. My mother observed that I had the runs a lot and was always hungry.

**Having a successful career in communication software, what made you want to become a lawyer at 33?**

I was bored intellectually. I was investigating other careers and Steven, my husband, suggested I consider law school as there are so many ways you can use a law degree. I took a paralegal course beforehand while still working full time. I think that was helpful to get my brain back in study mode. Going back to school at 33 was a challenge and I truly felt my brain stretch.

**Do you work now?**

No, I'm not working now. I worked full time as a lawyer for two years and then started working part time. Part



**SUZANNE JOYCE WITH HER DOG, JADE, IN THE NAPLES BOTANICAL GARDENS.**

time didn't work too well in the legal field for me. After about four years the stress of either not taking care of myself so I could get my work done or not getting my work done so I could take care of myself was too much. It wasn't an easy decision but I went on long-term disability. I realized that I was not in my twenties any longer and therefore I couldn't bounce back from not taking care of myself physically. I also found it so stressful to be away from work for four weeks or more and to know that a colleague was having to do my work. I felt I couldn't be relied on.

**What led you into learning and then teaching yoga?**

I took my first yoga class in 2000 at New York Sports Club (NYSC) and

noticed how chill I was after each class. I then tried some classes at a yoga studio, which I liked even more. I tried vinyasa, Hatha, yin, Sivananda, Ashtanga, and even laughter yoga at various studios. I also began studying Vedanta (Indian philosophy) and meditation at the NYC Sivananda Yoga Vedanta Center. When I moved out of NYC up to Rockland County in 2012, I thought taking a teacher training program at a local studio might be a good way to make new like-minded friends as well as deepen my personal yoga practice.

Around the same time my teacher at the Sivananda Center suggested I take the Sivananda teacher training. I ended up doing both courses and received my 200-hour certification from Sivananda in 2013 and my 300-hour certification from Yoga Mountain in 2014. I never had any intention of teaching but, as part of the certification courses, I had to and found I actually liked teaching yoga. I enjoy teaching Hatha, Sivananda, restorative and gentle yoga. I don't really like the power yoga style, and it is often too difficult for me to keep up physically.

**Where do you teach?**

In winter, in Florida, I teach a Sivananda class at a studio in Naples, which is open to the public, and a lunchtime Hatha class to the staff of Grace Place, a nonprofit that supports education for children in need. When I'm back up north, I teach at studios in NY and NJ. Proceeds from my classes go to Coach-Ed, a nonprofit that supports fitness and health for people with CF and their families.

**And then what led you to iRest Yoga Nidra?**

I was introduced to iRest by a Pilates teacher in Santa Fe, NM. I was

taking some private lessons with her and she mentioned iRest and I thought it sounded interesting and so I tried it. I had a very powerful experience of calmness for several days afterwards. It is not that way for everyone. Some people experience the effects over time and some people might not enjoy the experience at all. It is a guided meditation practice where you begin by deeply relaxing your body. Only when the body is relaxed can the mind relax. It is a practice that you do with your eyes closed lying down or sitting in a chair. No yoga poses are involved. At that point you can simply stay with that relaxation, which has great health benefits itself, or you can go deeper into a guided, yet completely private self-inquiry. For example, I use iRest to help me with anxiety. During the self-inquiry part, I might focus on my anxiety and where I experience it in my body. Then I find anxiety's opposite, say calmness, and where I experience that in my body. Then I go back and forth experiencing each opposite. It is amazing because the anxiety dissipates. It doesn't last forever; iRest is not a cure. But having done this practice multiple times, I can "call it up" in times of anxiety wherever I am, on a train, in bed at night, getting a PICC line placed, or before teaching a yoga class to a new group.

I took an iRest Level I teacher training course in 2011 to deepen my own practice and to learn how to share it with others. I then took a Level II course in 2015.

The benefits can be similar to a physical yoga class in that you may come away feeling more calm or relaxed than when you started. This calmness or relaxation can be felt in both your body and your mind.

**How do you feel doing yoga has helped you?**

There are a number of ways yoga has helped me. I always feel in a better place after the class than when I went in. I'm now able to do poses I never thought I could and it gives me a sense of accomplishment. I've become friends with some amazing people. I can see anyone having any or all of these same experiences through yoga.

**What is your current health status?**

Last summer I was having my first conversation with my doctor about the possibility of transplant and what was involved with getting on the list etc. My lung function was low and I began to use oxygen during any cardio exercise. I had a tune-up and returned to using TOBI after many years of not being able to tolerate it. Subsequent to the tune-up I developed a massive blood clot from the PICC line. I don't know what happened to me over the

following three to four months, but I can honestly say I feel much better than earlier in 2016 and my lung function has improved. I still need oxygen for any cardio exercise.

I have spent the past two winters in Naples, FL, and have not gotten sick. Is it the absence of dry air, coats, boots, scarves, people coughing and breathing on me in over-crowded public places? Or is it sunshine, salt air, humidity, blue skies? Probably both!

**What is your exercise regimen?**

Currently each week I try to: take a Pilates reformer or tower class, take one or two yoga classes for myself, teach two yoga classes, follow the pulmonary rehab exercises (walk on the treadmill for 45 minutes with O<sub>2</sub> and do their crazy hand-held weights routine each three times a week). I also walk my dog 20 minutes two times a day. I don't always get all this done but I really try. I used to do a lot of weight lifting and I keep thinking I'm going to fit that in as well, but it doesn't seem to be happening.

**What do you attribute your longevity to?**

My mother was religious about doing all my treatments since I was four years old (once we moved from Florida to Ohio she found a doctor who knew what was necessary). She

Continued on page 28

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**TILLMAN** continued from page 25

Celtaxsys, Inc., announced full enrollment of its Phase 2b clinical trial testing its flagship anti-inflammatory drug candidate, once-daily oral acebilustat (formerly CTX-4430), in adult CF patients (NCT02443688). The study is investigating the potential of acebilustat to reduce lung inflammation and preserve lung function over the course of 48 weeks in CF patients at a high risk for rapid lung function decline. In total,

200 CF patients of all CFTR genotypes across North America and Europe have been enrolled for 48 weeks of treatment comparing 50 mg and 100 mg doses of once-daily oral acebilustat against placebo. In this trial, acebilustat or placebo are being given concomitantly with standard CF care, including CFTR modulator therapies (Orkambi® or Kalydeco®). In addition to investigating the effect of acebilustat on lung func-

tion (FEV<sub>1</sub> percent predicted, a standard efficacy readout for CF trials), this trial will also assess the effect of acebilustat on pulmonary exacerbations and patient-reported outcomes.  
<http://tinyurl.com/y7336kg8>

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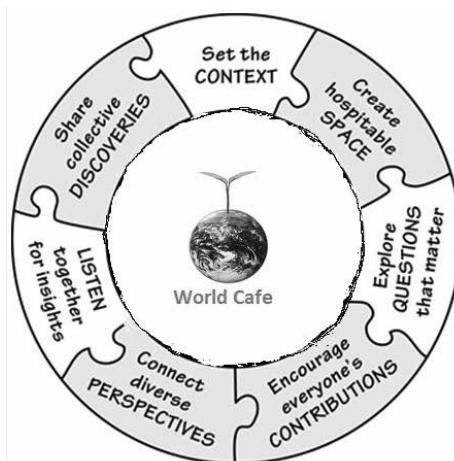
**Novoclem's Candidate Therapy Shows Promise Against Nine Pathogens Associated with Cystic Fibrosis**

Continued on page 29

- Adequately addressing mental health concerns stemming from body image and fertility issues for women with CF

Our activities have successfully recruited patients with CF to the CF-PTF and researchers and clinicians to the RAP. Our groups included advocates, social workers, CF researchers and care providers along with reproductive health researchers. Women with CF participated in each group. A member of the Patient Task Force joined RAP meetings to report on World Café sessions.

Moving forward, we will be bringing more women with CF into the collaboration. The CFReSHC will consist of a Governance Board, with members from the CF-PTF, core team and RAP groups. The Board will guide the overall direction and vision of the project and be in charge of major decision making. The CFReSHC will also consist of three subcommittees, in which there will be



CF-PTF, core team and RAP members. The subcommittees include the Communications Committee, Meeting Planning Committee and the Group Norms and Governance Committee. We are looking for enthusiastic members to join our collaborative! An ideal candidate would be someone who is passionate about voicing the research needs related to reproductive and sexual

health of women with CF and who wants to help shape our newly developed CFReSH Collaborative with written documents that define who we are and what we are hoping to achieve, how often we meet, and how we maintain an equal voice in the Collaborative, regardless of who you are. Will you join us?

If you are interested in joining us, please contact Kelly Gilmore at: [cfreprohealth@gmail.com](mailto:cfreprohealth@gmail.com).

### Acknowledgment

Project activity reported in this article was funded through a Patient-Centered Outcomes Research Institute (PCORI) Pipeline-to-Proposal Award (5140022). ▲

*Sandy Sufian is 49 and has CF. She lives in Chicago, IL. Laura Mentch is 64 and has CF. She lives in Bozeman, MT. Emily Godfrey is a physician who practices in Seattle, WA.*

### JOYCE continued from page 27

also made sure I ate well and exercised. I was on the swim team.

I've kept up exercising, which for the most part I enjoy. I firmly believe it has helped me keep most of my body in a very good physical condition. I also firmly believe that exercise builds confidence and is something you can do for yourself.

#### Do you know your mutations?

I have F508 delta and G542X.

#### What is your hope for the future for CF research?

I'm not sure if future treatments will help me. I don't often qualify for new treatments because my FEV<sub>1</sub> is not always above 40 percent. I also wonder if some of the newer drugs will prove to be helpful only for younger people with less damaged lungs than mine. I do have a lot of hope in the "personalized" medicine and genetic-driven medicine idea.

#### What do you do for fun?

A lot of the exercise I do I actually find fun. Down in Florida I love to walk on the beach for an hour or so at sunrise. I enjoy taking my dog to the dog park and see her run around like crazy or wrestle with other dogs. I've traveled to many places such as the Galapagos Islands, Hawaii, India, Caribbean Islands, England, Ireland, Spain, Belgium, Scandinavia and Italy.

#### How do you cope with CF?

In addition to yoga and iRest I find I can cope with CF-related difficulties by listening to music. Loud usually. I like rock and the blues. I usually read in bed every night to turn off my mind and it helps me fall asleep.

#### What would you say to someone who is younger and has CF to give them hope?

Be diligent on a daily basis with all you need to do for your body. I know it

sucks and it is the day-after-day-ness that wears you down. Try to find some way to make the tasks more tolerable if not pleasurable. Do something you like while you do your Vest or an aerosol. Use the time to learn something new and see after so many days what you've accomplished. But accept that you have to do these things every day and hopefully by accepting it and not fighting it you'll get it done and out of the way. Then on to the things you really want to do! ▲

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





# 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](mailto:cfroundtable@usacfa.org)**

## ANNIVERSARIES

### **Birthday**

**Kathy Russell**

Gresham, OR

73 on April 17, 2017

**Mike Schnitzer**

Vernon Hills, IL

60 on June 4, 2017

### **Wedding**

**Kathy & Paul Russell**

Gresham, OR

52 years on March 27, 2017

**Mike & Shari Schnitzer**

Vernon Hills, IL

32 years on April 28, 2017

### **Transplant**

**Mike Schnitzer, 59**

Vernon Hills, IL

Bilateral lungs

2 years on May 28, 2017

## NEW BEGINNINGS

### **Transplant**

**Tanya Cunningham, 53**

Sandy, OR

Bilateral lungs

May 14, 2017

**TILLMAN** continued from page 27

### **Infection**

Novoclem Therapeutics' lead drug candidate, a nitric oxide-releasing biopolymer, can eliminate nine of the most prevalent microorganisms responsible for severe lung infections in cystic fibrosis (CF) patients. Nitric oxide, or NO, is an important player in the normal immune system response against microorganisms that can cause disease. It also regulates the inflammatory process. Novoclem's NO-releasing biopolymer makes it possible to regulate both the immune system and inflammation with just one treatment. The drug candidate can effectively tackle several disease-associated microorganisms including methicillin-resistant *Staphylococcus aureus*, methicillin-sensitive *Staphylococcus aureus*, *Pseudomonas aeruginosa*, *Burkholderia cepacia*,

*Mycobacterium Avium-Intracellulare Complex (MAC)*, *Mycobacterium abscessus*, *Achromobacter xylosoxidans* and *Stenotrophomonas maltophilia*. Novoclem will soon apply to the FDA for Investigational New Drug status and begin its first clinical trial in early 2018. <http://tinyurl.com/ycfghh2g>

### **Polyphor Secures \$40 Million for Late-Stage Development of Murepavadin for CF Airway Infections**

Swiss-based Polyphor has secured financing for late-stage development of Murepavadin (POL7080), an antibiotic targeting antibiotic-resistant strains of *Pseudomonas aeruginosa*. The drug is about to enter a Phase 3 trial. In addition to the antibiotic, Polyphor is developing POL6014, an inhaled drug that blocks an elastase enzyme in neutrophil

immune cells. Intended for the treatment of CF and other severe lung diseases, it is in Phase 1b clinical trials. Murepavadin belongs to a class of compounds called precision Outer Membrane Protein Targeting Antibiotics (OMPTAs). They are the first new class of antibiotics targeting Gram-negative bacteria that have reached the late stages of the clinical trial process in 40 years.

<http://tinyurl.com/yc5ljhps>

### **Hypertonic Saline Treatment No More Efficient Before than During Airway Clearance in CF**

Treatment with hypertonic saline administered before airway clearance is no more effective than when administered during airway clearance in cystic

Continued on page 31



## WELLNESS

# Look For The Helpers

By Julie Desch

**T**here is a difference between living an easy life and living a life with ease. Nobody would say that a life lived with cystic fibrosis is “easy.” Even when healthy, there is the need for treatments, extra food, enzymes, extra vitamins, exercise, extra sleep etc. And when sick, don’t get me started.

With four hospitalizations in as many months, not to mention the addition of two body parts, my life of late has most definitely not been easy. First came spinal fusion surgery, which sounds awful, but was only at one disk level (L5-S1). After several months of needing to literally stop everything and either immediately sit or squat in order to cough due to excruciating pain, I decided that I had no choice. Not coughing was clearly not an option. And the pain was seriously cramping my exercise style and, therefore, my quality of life. The intervertebral disc at this level was demolished, and the bone on either side angry with inflammation. Enter my new titanium cage (I call him Titus). When the bone fuses through this sucker, I should be able to lift small automobiles.

But of course major surgery comes with risks, and I was not spared. Without going into graphics (and who needs to, given this audience), three episodes of pneumonia ensued (hereby termed P1, P2 and P3), culminating in a brand new port named Pauline.

So it was not an easy four months. But in retrospect, I don’t feel traumatized. Maybe that comes with the “seasoning” of living 56-plus years with CF. Or perhaps it is because I’m starting to learn a few things? Many a “problem”

needed solving, to be sure. But with each, I grew in what I guess would be best termed an “attitude” toward my current life situation.

I’ve never been great at gratitude. I say thank you, of course, but when I see people who literally exude gratitude about this life, I want to be them. I want to feel it in my bones, and for some reason this has been hard. Perhaps a layer of survivor guilt has blanketed the seeds of gratitude buried deep in my soul. But with each recent hospitalization, I’ve started to sense a growing authentic gratefulness.

*“With each recent hospitalization, I’ve started to sense a growing authentic gratefulness.”*



JULIE DESCH, MD

It began with the pouring out of support for me after my surgery. I literally needed a “squad” to help, and was amazed by its appearance. From my partner, to my sister, to a great friend who flew out from Texas, I was literally “handed off” for a week at a time for the daily help with moving, making sure I was eating, doing treatments and keeping me company. That’s not to mention food and grocery deliveries by friends, two sons eager to help in any way at any time, or the encouraging texts and phone calls.

I remember one very poignant time when I was helping my mother with getting dressed very near the end of her life when she was very weak and could barely see. Our faces were extremely close, and she suddenly stopped everything, looked right at me and laughed, saying, “I love you so much!” This is exactly how I felt as so many people came to my aid. Of course I was too shy to say it quite that way, but I think they know.

Gratitude began to germinate and grow from that point forward. As fate would have it, a few days after the PICC pull just after P2, I began a two-year mindfulness teacher training program with a five-day retreat in one of the most beautiful locations in California: deep in the redwoods midway between the beaches of Santa Cruz and my home in Palo Alto. How could I not be grateful to be there? I was there with Tara Brach and Jack Kornfield, two of the leading mindfulness teachers in the West, along with 300-plus other trainees all on the same path with me. I was able to interact with many of the attendees in small groups, exploring our practices and lives

and sharing our deepest joys and suffering. Yes, my body was coughing and short of breath, and I knew that there would certainly be more allopathic medicine in my very near future. But the “medicine” of the connections made and the environment absorbed was every bit as powerful as the antibiotic cocktails that would bookend this retreat. Each day, I could not believe my good fortune.

It’s no secret that years and years of antibiotics breed multiple resistances as well as multiple allergies. P3 set in as soon as I set foot back in my house following the retreat. It followed P2 so closely due to a new antibiotic resistance discovered after-the-fact. P2 was treated with two drugs, one of which was no longer effective. But nobody picked this up until I started to have fevers when the IV course was over and the one drug that *did* work was stopped.

Yes, I was a bit upset when I learned that this happened, but my irritation was replaced by relief and appreciation when my old friend, ceftazidime, became a player again after desensitization was attempted for the first time. As I sit here, infusing ceftazidime, currently able to breathe and not in anaphylactic shock, I bow to the desensitization gods and immunology scientists who figured out how to defeat an allergic reaction.

Finally, eight of my closest friends dating back to junior high school (now that is a long time!) will descend upon my house tomorrow as I am in my second week of IV therapy. Our annual gathering does not stop for anything and is timed perfectly to deliver the world’s best medicines, connection and laughter. The nine of us do nothing when we meet except reminisce, act like we are teenagers again, and laugh

and laugh and laugh some more. They all know I am not at my best. But they are all mothers (and many, grandmothers) and I am certain I will not be allowed to lift a finger. Could I be more fortunate?

So problems arise. Some hit pretty hard psychologically, and some are just temporary nuisances. I am under no illusion that I will not face more as my body continues to dance with cystic fibrosis. My secret weapon has become what used to be one of my biggest weaknesses, gratitude for the people in my life, from junior high friends, to family, to new connections with mindfulness teachers. As Mr. Rogers used to say about troubled times, “Look for the helpers. You will always find people who are helping.” There are helpers all around me. ▲

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*Julie is 56 and is a physician who has CF. You may reach her at [jdesch@usacfa.org](mailto:jdesch@usacfa.org).*

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## **TILLMAN** continued from page 29

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fibrosis (CF) patients. The study showed no significant difference in either the lung clearance index or spirometry readings before and after treatment. Nor were there differences in the weight of the patients’ sputum, perceived ease of clearance or satisfaction. However, treating with hypertonic saline during airway clearance took significantly less time than treating before airway clearance.

<http://tinyurl.com/ydabw5k4>

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### **PMD Healthcare Launches Spiro PD 2.0 and Health Platform in May, Cystic Fibrosis Awareness Month**

PMD Healthcare launched its second-generation spirometer, Spiro PD 2.0, as well as its Wellness Management System (WMS) – a lung health monitoring and management digital health platform for people with cystic fibrosis (CF). PMD’s remote monitoring pro-

gram and its WMS platform will monitor biometrics, symptoms, medication adherence and care plan compliance outside of the provider’s office. The WMS platform includes validated symptom tools, personalized care plans and customizable alerts that enable physicians and nurses to intervene in real-time via a secure, HIPAA-compliant portal. Spiro PD 2.0 and WMS introduce the use of 24-hour telemonitoring technology via Wi-Fi to connect patients, providers and caregivers. This helps keep track of a patient’s progress without trips to the clinic or hospital. <http://tinyurl.com/y8zyu3s7>

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### **Sound Pharmaceuticals announces \$1.6 Million Award from Cystic Fibrosis Foundation Therapeutics to prevent and treat the hearing loss and tinnitus associated with antibiotic use**

The Cystic Fibrosis Foundation

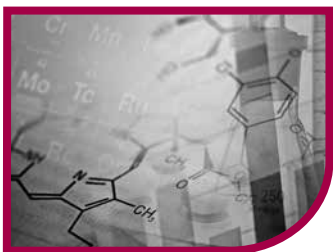
Therapeutics Inc. (CFFT) development award will support the initial testing of SPI-1005 in people with CF who are receiving tobramycin for the treatment of acute pulmonary exacerbations or lung infections. A leading side effect of tobramycin involves damage to the inner ear and is called ototoxicity. The Phase 1b STOP Ototoxicity clinical trial will examine the incidence and severity of hearing loss, tinnitus, dizziness or vertigo due to IV tobramycin treatment. SPI-1005 is a novel oral drug that is being developed to reduce the incidence and severity of sensorineural hearing loss and tinnitus. <http://tinyurl.com/y8hajnlrt>

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### **Genomics Reveal How Bacteria Evolve in Cystic Fibrosis Patients**

Scientists have shown how long-term infection leads to genetic and

Continued on page 35



# SEARCHING FOR THE CURE

## My Clinical Trial Experience: One Year Update

By Meranda Sue Honaker

One year ago I made the decision to enroll in a new clinical trial targeting my CF mutations on a cellular level. At that point I had been on Orkambi for over two years. Initially, I had symptomatic improvement of my CF symptoms; however, despite my best efforts on Orkambi, my lung function continued to decline. Additionally, I endured unpleasant side effects (abnormal hormone levels, hair loss, severe chronic fatigue etc.), which were motivating factors in my decision to discontinue Orkambi and pursue an alternative therapy. The Orkambi alternative came to me in the form of yet another clinical trial. I enrolled eagerly in my current study, and undoubtedly made the right decision for my health.

It has now been one year since moving to open label in my current CF study. Going to “open label” in a clinical trial means you are no longer blinded to whether you are receiving active drug or placebo. I am in fact on active study drug. I am followed closely in my study, which means there are frequent trips to have laboratory tests conducted (PFTs, blood work, EKGs etc.). I am typically exhausted after research study visits. However, participating in this clinical trial is absolutely worthwhile given the improvements I have experienced thus far.

The last year has felt like a whirlwind. I reflect upon my life in ways I had not before. I consider what my future looks like knowing there are potentially groundbreaking CF thera-

pies on the horizon. I also realize the study medication (which I am not allowed to discuss by name) that I am on could prove life changing for some with CF, including many of my beloved friends. I am carefully pushing boundaries that CF previously imposed upon me. I continue to dedicate myself to remaining diligent in my healthcare regimen. My overall routine has remained unchanged other than the addition of my study medication, but

my quality of life has certainly improved.

Four months into open label, I had a CF clinic visit where I was finally able to see my PFTs. For the first time in years my FEV<sub>1</sub> and FEF<sub>25-75</sub> increased! I was so surprised by my lung function improvement that I asked if the PFT machine in CF clinic had been calibrated recently. I considered the PFT device could be malfunctioning, before I processed reality—my lung function increased! I became

overwhelmed with gratitude and joy. Seeing a meaningful improvement in my FEV<sub>1</sub> was a small but profound victory. The increase in my lung function serves as inspiration for me as I continue exploring the world and contemplating my own future. I am

more hopeful now than ever about the future of CF therapies.

I encourage everyone with CF to get off the sidelines and get involved with CF research. Every person participating in research is contributing toward future advancements in treating CF. Although some may be ineligible for drug studies, there are likely other types of studies in which you can become involved (i.e., observational, early intervention etc.). Our entire community benefits with each step forward in the research process. I am a 15-year veteran of clinical trials and living proof that participating in research is worthwhile. ▲



MERANDA SUE HONAKER

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



# Got AFLAC?

By Bill Coon

When speaking to Jeanie Hanley recently, I brought up AFLAC. Everybody has seen the duck advertisement on TV, but how many of you with CF have one of its policies to help with out-of-pocket expenses? I have been a member since 1996 when it was offered at the company I worked for. Today it offers more than 25 different policies, but I'm going to talk about only the two that I have and how I have benefited from them. Everyone is approved and there are NO medical questions to qualify. AFLAC is a "Supplemental Insurance" and cash is paid directly to you! Please check with your employer, most offer them, but you can also get individual policies if they do not. And, your premium will never go up! There is a 30-day waiting period.

My first policy, which I still have to this day, is a Hospital Indemnity Plan. I signed up for this plan because, with CF, I spent many days in the hospital receiving IV treatment for pneumonia or other illness. My cost at the time I signed up was about \$27 per month and remains the same today.

During the years leading up to my lung transplant, I was averaging about three hospital stays a year.

This policy paid \$100 per day for the first seven days and \$200 for days eight to 30. This policy also pays for surgical benefits up to \$1,000, an ambulance benefit of \$100 and a \$50 per year wellness benefit. One of the surgical benefits pays \$250 for each bronchoscopy. I've had so many I can't count them.

If you have these medical situations, please contact a local AFLAC representative as cost and policies may change per state. But for me, AFLAC easily pays for itself.

Four years later, in 2000, AFLAC came out with an "Intensive Care" policy. This also was a no brainer for me because it offers \$25,000 for human organ transplant. Having CF, I knew it was a high possibility that this would be needed. It also paid \$600 per day for days one through seven, then \$1,000 per day for days eight to 15. After that I was paid \$250 per day for my step-down unit care up to 30 days. Unfortunately for me I was in ICU for 38 days, my recovery post-trans-



BILL COON

plant did not go well. This policy costs me today \$8.70 per month. Once again, AFLAC easily paid for itself. Because it is separate from my health insurance, AFLAC payouts accumulated and eventually helped to pay off my house.

I hope this information helps. Look into AFLAC and see the many plans they have to offer. It pays to stay healthy and stay covered! ▲

*Bill Coon is 59 and has CF. He lives outside of Houston, TX. He is seven years post-bilateral-lung transplant.*

## 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](http://www.cfmothers.com).

If you would like to join our Facebook support group, please e-mail Karen Vega at: [kvega@usacfa.org](mailto:kvega@usacfa.org).

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# Call For Nominations From The CF Community

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**N**ominations are being accepted for the 2017 Jacoby Angel Award and the 2017 USACFA Founders Award. These awards are made every two years by the United States Adult CF Association. Readers of *CF Roundtable* may nominate individuals for each award. The USACFA Board of Directors then vote on the nominees. Directors of USACFA cannot be nominated to receive an award.

Nominations should be e-mailed by September 4, 2017, to [bsufian@sufianpassamano.com](mailto:bsufian@sufianpassamano.com).

The USACFA Awards have gained significance in the community. The last awards were presented at a USACFA Awards Breakfast held at the North American CF Conference, with more than 120 people in attendance. Those who were nominated for the awards were also recognized and presented with crystal plaques. If you know someone who exemplifies the qualities discussed below, please send a nomination.

## Jacoby Angel Award

The Jacoby Angel Award is presented to a person with CF who is making a difference in the lives of one or more people with or without CF.

The award is named in memory of Dr. Jack Jacoby who was a CF physician at the St. Vincent's CF Center in New York City for more than 15 years from 1982-1997. Dr. Jacoby had CF and used his own experience living with the disease to provide exceptional medical care to his patients. Dr. Jacoby's patients still talk about the significant impact he had on their lives and still refer to him as an angel sent to help them fight the effect of CF on their lives. Dr. Jacoby always put the needs of others above his own needs and worked tirelessly to bring comfort and relief to his patients. He was the medical advisor for USACFA and wrote a medical column for *CF Roundtable* for many years.

We look forward to receiving nominations for the Jacoby Angel Award for people who have followed in the footsteps of the award's namesake, Dr. Jack

Jacoby, by being people with CF who dedicate themselves to helping others.

Past Jacoby Angel Award winners include: Michelle Compton, Susan Burroughs, Robyn Petras, Pammie Post, Jerry Cahill and Isabel Stenzel Byrnes.

## USACFA Founders Award

We also welcome nominations for the USACFA Founders Award, which recognizes a person who has made an outstanding contribution to the adult Cystic Fibrosis community. The nominee can be a person who does not have CF or a person who does have CF. The award was named in honor of the group of adults with CF who founded USACFA and worked tirelessly to bring information to the adult CF community at a time when there was no Internet and no efforts to connect adults with CF and provide information and support to the adult CF community.

Past recipients of the Founders Award include Dr. Jerry Nick, Dr. James Yankaskas, James Passamano, Robyn Petras and Dorothy Hello. ▲

## POETRY

## CORNER

### Burden

*By Lauren Hunsaker*

Fine mists gulped with zeal  
Pernicious goo gone, for now  
Respite from rhonchi.

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*Lauren is 30 and has CF. She is a Director of USACFA. Her contact information is on page 2.*

physical changes in *B. cenocepacia*. Their study, provides the first comprehensive genome-phenome analyses of *B. cenocepacia* infection in cystic fibrosis lungs. After using whole genome sequencing, they found that long-term infection leads to significant genetic changes, as well as physical changes, such as progressive declines in bacterial motility, and changes in biofilm formation over time. The researchers also observed broad phenotypic and genotypic variation for samples obtained from the same patient at the same time. The study increases the number of genome sequences for the species by more than 10-fold, and by coupling these genomes with extensive phenotypic tests, it offers a unique resource for understanding how the species evolves in the context of chronic lung infection. What researchers found most surprising was that some bacterial traits typically associated with pathogenesis (namely motility and biofilm formation) actually became less pronounced over time, which suggests that one way

the bacterium may be adapting to long-term chronic lung infection is by becoming less aggressive and thus potentially more able to avoid the immune system. <http://tinyurl.com/y8lhyyye>

AND

<http://tinyurl.com/y7tbpnww>

#### Cystic fibrosis study offers new understanding of silent changes in genes

Since 1989, when CFTR was first identified, more than 2,000 changes have been reported in its gene; 1,700 of these changes lead to cystic fibrosis. Among the remaining 300 replacements are a group of silent changes, so called because they alter the gene without changing the composition of the CFTR protein. Such silent changes have long been considered without effect on how proteins are made and how they work in cells. Researchers have investigated the impact on the CFTR protein of a silent change in its gene called T2562G.

T2562G changes how the CFTR protein is made by cells. It causes the

CFTR pathway for chloride ions to become narrowed, slowing chloride movement across cell membranes. The change in how CFTR is made is the result of how the cell reads genetic information. T2562G causes protein-producing machines called ribosomes to slow down the speed with which CFTR is made, resulting in an altered protein with impaired chloride transport. This finding reveals a new unexpected way by which silent changes in genes alter how proteins are made and how they work in cells. The findings provide new understanding of the impact of silent changes in genes and highlight the genetic complexity of cystic fibrosis, which in turn affects disease severity and an individual's response to treatments targeting the root cause of cystic fibrosis.

<http://tinyurl.com/yanlvnjx>

#### Scientists uncover interactions between bacteria that infect the lungs in cystic fibrosis

Continued on page 36



## Bene factors

### BRONZE

Eileen Burker  
Pauline Dinello  
Davie Ebacher  
Marge Fye  
Lauren Hamburg  
Cindy Hayes (In honor of Zach Hayes)  
Lynda Jensen (In memory of sister, Lisa Cockrum)  
Carol Keenan  
Amy Rung  
Alice Todd (In honor of Cheri DeWilde's 50<sup>th</sup> birthday)

*In the Spring 2017 issue we inadvertently listed a gift from Cindy Hayes as being "in memory of" rather than "in honor of" Zach Hayes. We apologize for our error.*

### GOLD

Aileen Vizel

### PLATINUM

Genentech

### SUSTAINING PARTNERS

Kroger Specialty Pharmacy

### PEARL SUSTAINING PARTNERS

Marshall & Margherite McComb Foundation

### DIAMOND SUSTAINING PARTNERS

Nancy Wech

Substances produced by *P. aeruginosa* in the lungs of cystic fibrosis patients may enhance the growth of other bacteria that, in turn, inhibit the harmful bacterium's biofilm. Recent studies suggest that bacteria known as streptococci might inhibit *P. aeruginosa* and improve lung function. The scientists found that a carbohydrate substance known as alginate, produced by a particular strain of *P. aeruginosa* known as FRD1, promotes the biofilm of the streptococcus species *Streptococcus parasanguinis*. At the same time, biofilm formation by *S. parasanguinis* restricts biofilm formation by *P. aeruginosa*. The team also found evidence that molecules known as adhesins, which are produced by *S. parasanguinis*, play an important role in this process. Adhesins help cells stick together in a biofilm or attach to surfaces, and they appear to be necessary for enhanced biofilm formation by *S. parasanguinis* in the presence of alginate. These findings suggest a potential mechanism by which *S. parasanguinis*, which is normally found on the surface of teeth, might colonize the lungs of a cystic fibrosis patient and inhibit *P. aeruginosa*. <http://tinyurl.com/yeh232ha>

### CF Patients' Bacterial Gut Diversity Decreases as Disease Worsens, Study Shows

Cystic fibrosis (CF) patients have a poorer diversity of gut bacteria than healthy people, and it gets worse as the disease progresses.

In addition, patients with severe lung impairment have significantly fewer bacterial species in their gut than those with mild lung problems. Because frequent intravenous antibiotic courses are known to decrease bacterial diversity, it is obvious that disease management contributes to the abnormal gut-microbiome state.

Researchers suggested CF-specific probiotics should be developed in order to minimize the risk. Mutations in the CFTR gene cause CF.

<http://tinyurl.com/yctsgvay>

### Protein in Stool Could Be a Marker of Disease Severity in Cystic Fibrosis

The levels of a protein called calprotectin in the stool could not only be a marker of intestinal disease but could also help determine disease severity in cystic fibrosis. This marker could be useful to monitor clinical worsening of the disease over time. Calprotectin is a calcium-binding protein that is found in high levels in a type of immune cells called neutrophils. The presence of calprotectin in stool indicates the migration of neutrophils to the intestines. This occurs in cases of inflammation. In CF patients, the levels of calprotectin were significantly higher than in healthy volunteers. Moreover, the levels of calprotectin were significantly higher in CF patients who were older than 18 years, who had pancreatic insufficiency, were underweight, had an infection in their airways with *Pseudomonas aeruginosa*, had CF-related diabetes, had reduced lung function or had had a high number of pulmonary exacerbations.

Levels of calprotectin in stool are already in use as a clinical biomarker to assess the presence of bowel inflammation. Recent studies suggested that in CF patients there may be a correlation between bowel and lung disease.

<http://tinyurl.com/y7rgeg78>

### TGV-Inhalonix's New Drug to Help Save Cystic Fibrosis Patients with Resistant Fungal Lung Infections

TGV-inhalonix announced that Mul-1867 has shown tremendous potential against clinical isolates of fungi from patients with cystic fibrosis and other severe lung infections. The authors report that there was high activity of Mul-1867 against multidrug-resistant strains of *Candida* spp and *Aspergillus* spp, which are especially risky for people with cystic fibrosis and patients with compromised immune system including those with lung trans-

plants. Broad spectrum activity of Mul-1867 against multi-resistant bacterial isolates such as *S. aureus* and *P. aeruginosa* has also been confirmed. Its high anti-infective efficacy could make it a breakthrough new agent for addressing a variety of lung infections starting with those in cystic fibrosis patients.

<http://tinyurl.com/y78egmsx>

### Cystic fibrosis patients' tube feeding could be source of infections

Multi-drug-resistant nontuberculous mycobacteria (NTM) can be found in the gastric juice and tubes that are used to feed some cystic fibrosis patients. Tube feeding (gastrostomy) can also be used to drain the stomach when there's a blockage. Scientists have found a gastric juice reservoir of organisms involved in CF lung infection in patients fed by a tube, a procedure whose scientific name is percutaneous endoscopic gastrostomy (PEG). The researchers looked for NTM in gastric juice and sputa from 16 adults with moderate to severe CF lung disease who were being fed by a tube. They found bacteria or fungi in all gastric juice, sputa and PEG samples. They discovered that MABSC can be isolated from the gastric juice and PEG tubes of patients with CF, in addition to sputum. In addition, the team said it was the first time MABSC had been found in a gastric sample in these patients. Thus, the team concluded that gastric juice and PEG tubes may be sources of MABSC in CF patients.

<http://tinyurl.com/y9tcqgs7>

### CF Patients May Have Improved Quality of Life with Early Palliative Care

Researchers have developed a primary palliative care framework to improve cystic fibrosis patient care and reduce the symptom burden. This model aims to integrate palliative care with standard CF therapies to promote a positive effect on the long-term quality of life for these patients. Palliative care is a multidisciplinary approach for



patients with serious diseases to relieve their suffering by improving their pain, physical symptoms and psychological distress. For many patients, CF is associated with distress due to chronic symptoms such as pain, difficulty breathing, cough, fatigue and insomnia. This can negatively impact CF treatment adherence and overall quality of life. Palliative treatments have been shown to help CF patients cope with distressing factors associated with the disease. In many cases, however, patients are introduced to palliative care in later stages of the disease, which may not be the best way to manage the condition. Based on previous assessments and implementation of a primary palliative care training curriculum tailored to CF care, the authors developed a pilot model of routine implementation of a CF-specific primary palliative care intervention. Among the available post-assessment services provided during the study, CF patients requested mainly cognitive-behavioral therapy and acupuncture/acupressure. About 95 percent of the patients agreed that discussing the challenges of living with CF was helpful, and 85 percent found that the assessment identified most of the patient's "bothersome CF-related symptoms." The majority of patients believed this process helped

them identify and prioritize the symptoms that needed to be addressed. Focusing on early assessment and treatment of CF-associated symptoms, as well as distress and coping, this palliative care model is expected to provide caregivers information to support an individualized symptom management plan, which more likely will enhance self-management, health outcomes and quality of life for cystic fibrosis patients. <http://tinyurl.com/yalhbbhlu>

#### **FYI**

**The CF-CARES primary palliative care model: A CF-specific structured assessment of symptoms, distress, and coping.** Deborah Friedman, Rachel W. Linnemann, Lily L. Altstein, Suhayla Islam, Kieu-Tram Bach, Chelsea Lamb, John Volpe, Caitlin Doolittle, Anita St. John, Patricia J. O'Malley, Gregory S. Sawicki, Anna M. Georgiopoulos, Lael M. Yonker, Samuel M. Moskowitz. *Journal of Cystic Fibrosis*. Article in Press

Current palliative care tools do not address distressing chronic symptoms that are most relevant to cystic fibrosis. Individuals with CF, regardless of disease severity, face challenges managing symptom burden. Frequently reported symptoms are not consistently associated with distress, suggesting the impor-

tance of individualized evaluation. The CF-CARES (Coping, goal Assessment, and Relief from Evolving CF Symptoms) primary palliative care assessment model provides a framework for patients experiencing chronic symptoms to explore interventional options with their clinicians.

**Cystic fibrosis pulmonary exacerbations attributable to respiratory syncytial virus and influenza: a population-based study.** Somayaji R, Goss CH, Khan U, Neradilek M, Neuzil KM, Ortiz JR. *Clin Infect Dis*. 2017 Mar 9

Characterization of the role of respiratory viral pathogens on cystic fibrosis pulmonary disease is needed. The authors aimed to determine the association of influenza and respiratory syncytial virus (RSV) activity with risk of pulmonary exacerbation (PEX) in persons with CF in the United States. Results indicated that the large CF population-based cohort demonstrated a significant association between PEX risk and influenza activity in children and adults and with RSV activity in children.

<http://tinyurl.com/yce3bmfx>

**Complications of long and intermedi-**

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**ate term venous catheters in cystic fibrosis patients: A multicenter study.** Teresa L. May, Alex H. Gifford, Thomas Lahiri, Adam Black, Janet Trang, Alexandra G. Cornell, Karyll Gonzalez, Scott Morin, Mark Napier, Christine W. Duarte, Jonathan B. Zuckerman. *Journal of Cystic Fibrosis*. Article in Press

Totally implantable venous access devices (TIVADs) or peripherally inserted central venous catheters (PICCs) are commonly used in the care of patients with cystic fibrosis (CF), but they are associated with various complications, including thrombosis, infection and insertion site symptoms. Specific patient- and catheter-related characteristics were associated with increased risk of complications. Center effects on complication rates were observed for PICCs. <http://tinyurl.com/ycoagp44>

**Is there an association between back pain and stress incontinence in adults with cystic fibrosis? A retrospective cross-sectional study.** Jane E. Ashbrook, Carol Shacklady, Sue Johnson, Gillian Yeowell, Peter Charles Goodwin. *Journal of Cystic Fibrosis*. Article in Press

Back pain and stress urinary incontinence (SUI) are common in adults with cystic fibrosis (CF). This study aimed to establish whether there is an association between back pain, lung function and stress urinary incontinence and its relative risk. An association was found between back pain and SUI in adults with CF. This information is important when developing management strategies in the CF population. <http://tinyurl.com/y9lj7prj>

## BACTERIA

**Co-colonization with *Aspergillus fumigatus* and *Pseudomonas aeruginosa* is associated with poorer health in cystic fibrosis patients: an Irish registry analysis.** Reece E, Segurado R, Jackson A, McClean S, Renwick J, Grealley P. *BMC Pulm Med*. 2017 Apr 21;17(1):70

Pulmonary infection is the main

cause of death in cystic fibrosis. *Aspergillus fumigatus* (AF) and *Pseudomonas aeruginosa* (PA) are the most prevalent fungal and bacterial pathogens isolated from the CF airway, respectively. The study's aim was to determine the effect of different colonization profiles of AF and PA on the clinical status of patients with CF. The authors established that CF patients co-colonized with AF and PA had poor clinical outcomes comparable to patients persistently colonized with PA, emphasizing the clinical significance of co-colonization with these microorganisms. <http://tinyurl.com/yazl4xoz>

## TREATMENTS

**Immediate effects of lumacaftor/ivacaftor administration on lung function in patients with severe cystic fibrosis lung disease.** Natalia Popowicz, Jamie Wood, Anna Tai, Sue Morey, Siobhain Mulrennan. *Journal of Cystic Fibrosis*. May 2017 Volume 16, Issue 3, Pages 392–394

Safety data for lumacaftor/ivacaftor (LUM/IVA) combination therapy in patients with severe lung disease (percent predicted forced expiratory volume in 1 s [ppFEV<sub>1</sub>] <40) remain limited. The authors report immediate post-dose respiratory-related adverse events in 12 patients with severe cystic fibrosis lung disease prescribed LUM/IVA. All patients experienced a decline in ppFEV<sub>1</sub> from baseline at two hours that persisted at 24-hours but recovered in most patients at one month. No pre- and post-differences in bronchodilator response were observed. Ten patients reported non-severe respiratory-related adverse events within 24-hours of LUM/IVA initiation. At one month, eight patients had persistent symptoms and six were treated for a pulmonary exacerbation. These results highlight that LUM/IVA respiratory-related adverse events are common in patients with a ppFEV<sub>1</sub> < 40. <http://tinyurl.com/ybcz6e2w>

**A treatment evaluator tool to monitor**

**the real-world effectiveness of inhaled aztreonam lysine in cystic fibrosis.** Barry J Plant, Damian G Downey, Joe A Eustace, Cedric Gunaratnam, Charles S Haworth, Andrew M Jones, Edward F McKone, Daniel G Peckham, R. Ian Ketchell, Diana Bilton. *Journal of Cystic Fibrosis*. Article in Press

The authors felt that studies are required that evaluate real-world outcomes of inhaled aztreonam lysine in patients with cystic fibrosis (CF). They concluded that in patients with CF in routine practice, inhaled aztreonam lysine is associated with improved lung function and weight, and reduced hospitalization and intravenous antibiotic use. <http://tinyurl.com/y7wgewpy>

**Desensitization to inhaled aztreonam lysine in an allergic patient with cystic fibrosis using a novel approach.** Guglani L, Abdulhamid I, Dittouras J, Montejo J. *Ann Pharmacother*. 2012 Oct;46(10):e25

Desensitization of a patient with CF who is allergic to intravenous aztreonam was successfully accomplished with the novel approach of rapid intravenous desensitization followed by inhaled therapy. As inhaled antibiotics are being increasingly used for patients with CF, this novel strategy can be used for desensitizing allergic patients with CF to ensure that they can continue to receive these medications safely. <http://tinyurl.com/ycypdxmm>

**Allergic bronchopulmonary aspergillosis in patients with cystic fibrosis.** Janahi IA, Rehman A, Al-Naimi AR. *Ann Thorac Med*. 2017 Apr-Jun;12(2):74-82

Allergic bronchopulmonary aspergillosis (ABPA) is a pulmonary disorder that often occurs in patients with asthma or cystic fibrosis (CF) and is characterized by a hypersensitivity response to the allergens of the fungus *Aspergillus fumigatus*. In patients with CF, growth of *A. fumigatus* hyphae within the bronchial lumen triggers an immunoglobulin E (IgE)-mediated hypersensitivity

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ity response that results in airway inflammation, bronchospasm and bronchiectasis. Since the clinical features of this condition overlap significantly with that of CF, ABPA is challenging to diagnose and remains underdiagnosed in many patients. Management of ABPA involves the use of systemic steroids to reduce inflammation and modulate the immune response. In patients who do not respond to steroids or cannot toler-

ate them, antifungal agents should be used to reduce the burden of *A. fumigatus* allergens. Recent studies suggest that omalizumab may be an effective option to reduce the frequency of ABPA exacerbations in patients with CF. <http://tinyurl.com/y8mjt4p>

**Impact of azithromycin on the clinical and antimicrobial effectiveness of tobramycin in the treatment of cystic fibro-**

sis. Dave P. Nichols, Carrie L. Happoldt, Preston E. Bratcher, Silvia M. Caceres, James F. Chmiel, Kenneth C. Malcolm, Milene T. Saavedra, Lisa Saiman, Jennifer L. Taylor-Cousar, Jerry A. Nick. *Journal of Cystic Fibrosis*. May 2017 Volume 16, Issue 3, Pages 358-366

Concomitant use of oral azithromycin and inhaled tobramycin occurs in approximately half of U.S. cystic fibrosis

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**TILLMAN** continued from page 39

patients. Recent data suggest that this combination may be antagonistic. The authors discovered that azithromycin appears capable of reducing the antimicrobial benefits of tobramycin by inducing adaptive bacterial stress responses in *P. aeruginosa*, suggesting that these medications together may not be optimal chronic therapy for at least some patients.

<http://tinyurl.com/y9r6c9vy>

### TRANSPLANT

**A clinical tool to calculate post-transplant survival using pre-transplant clinical characteristics in adults with cystic fibrosis.** Stephenson AL, Sykes J, Berthiaume Y, Singer LG, Chaparro C, Aaron SD, Whitmore GA, Stanojevic S. Clin Transplant. 2017 Mar 7

The purpose of this study was to develop a clinical tool to estimate the risk of death after transplant based on pre-

transplant variables. Four pre-transplant factors most predictive of poor post-transplant survival were older age at transplantation, infection with *B. cepacia* complex, low FEV<sub>1</sub> percent predicted and pancreatic sufficiency. A nonlinear relationship was found between risk of death and FEV<sub>1</sub> percent predicted, age at transplant and BMI. The authors constructed a risk calculator based on their model to estimate the one-, three-, and five-year probability of survival after transplant. The risk calculator quantifies the risk of death associated with lung transplant using pre-transplant factors. This tool could aid clinicians and patients in the decision-making process and provide information regarding the timing of lung transplantation.

<http://tinyurl.com/y8gtpo8>

**Increased risk of PTLT in lung transplant recipients with cystic fibrosis.**

Erin M. Lowery, William Adams, Shellee A. Grim, Nina M. Clark, Leah Edwards, Jennifer E. Layden. Journal of Cystic Fibrosis. Article in Press

Post-transplant lymphoproliferative disease (PTLD) is an important cause of morbidity and mortality following lung transplantation. Recipients with cystic fibrosis may have an increased risk of PTLT. The primary aim of the authors is to examine PTLT in an adult lung transplant population by utilizing the International Society for Heart and Lung Transplantation Registry. They concluded that CF recipients have a higher risk for PTLT compared to non-CF recipients.

<http://tinyurl.com/ycjaslw5> ▲

*Laura Tillman is 69 and has CF. She is a former Director and President of USACFA. She and her husband, Lew, live in Northville, MI.*