

Defining My Path To Success — A Glimpse Into The Life Thoughts Of A Young Woman Living With Cystic Fibrosis

By Ella Balasa

Decision making is one of the hardest tasks that we humans face. Decisions determine our futures and our fate. There are some individuals who are able to make decisions by “flipping a coin,” meaning they do not put much forethought into their decisions, rather they rely on gut instinct, and never look back with regret or uncertainty. I see both the positives and negatives of this decision-making style, and sometimes I wish that were my personality. I may, however, be inching towards that impulsive lifestyle, as I’ve come to understand the enormous complexity of life with this “special” gene mutation called cystic fibrosis (CF).

I have never thought CF would slow me down or halt my advancement in the professional world.

Unfortunately, I received a call about a month ago that revealed the limitations associated with this disease. I think that success is defined by power: being able, being driven and being more confident in oneself than others

“I am trying to find a way to achieve my definition of success, despite the limitations in my way.”

around you. Success is having a title. I realize that everyone has different ideas of success that can be applied to many different situations, but I am defining success as it relates to my future, my career and my life. I am trying to find a way to achieve my definition of success, despite the limitations in my way.

I graduated with a Bachelor of

Science in Biology and minor in Chemistry from Virginia Commonwealth University (VCU) in December of 2014. I have been interested in the science of water quality since I was an undergraduate, and have worked in

the field as an intern. After graduation, I began working at a water treatment plant, and then eventually took my current part-time job as a lab manager for a microbiology professor at VCU. These jobs have been stepping stones that have given me time to work in the field. They have also given me the opportunity to think about

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CF ROUNDTABLE
FOUNDED 1990
Vol. XXVI, No. 2

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

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EDITOR'S NOTES

Well, here we are in another spring season. Here in the West, we have been having a lot of rain, while the East has been getting SNOW! Maybe the weather will settle down sometime soon. I certainly hope so.

There have been changes to the USACFA board of directors, once again. **Chris Kvam** has resigned from the board and we thank him for his service. **Amy Braid** has joined the board and we look forward to working with her. See her introduction on page 34. Also, we have a new columnist. **Aimee Lecointre** will be writing about exercise and nutrition. Her column is called "Active For Life" and she introduces it and herself on page 24.

As always, this issue is filled with good information. **Beth Sufian** answers questions from our readers regarding SSI and SSDI benefits from the Social Security Administration, in "Ask The Attorney." "Spirit Medicine" has **Isabel Stenzel Byrnes** recounting her experience with CF boot camp.

Our Focus topic is Managing Various Conditions (Under The CF Umbrella). I talk about some of these conditions that accompany CF in "Speeding Past 50." **Laura Mentch** and **Kori Tolbert** both discuss incontinence in their Focus articles. **Kasey Ream** continues the topic with a discussion of CF-related diabetes (CFRD). **Julie Desch** wraps up the topic in "Wellness" as she tells of how her "CF-umbrella" is burgeoning into more of a tent!

Laura Tillman has, once again, compiled a wonderful list of "Information From The Internet." "Searching For The Cure" finds **Meranda Honaker** discussing the importance of participating in clinical trials. A sidebar has a list of studies that currently are enrolling participants.

In "Parenting," **Megan Parker** tells of lessons learned in the first year of life with her twins. The "Poetry Corner" has some thoughts on spring written by **Linda Stratton**. **Ella Balasa** is on the front page talking about career choices with CF. **Susie Baldwin** answered questions from **Andrea Eisenman** and is the feature of "In The Spotlight."

In "Voices From The Roundtable" be sure to read **Devin Wakefield's** article about the CFRI Retreat. He invites everyone to attend.

We invite everyone who has CF to check out the upcoming Focus topics that are listed on page 3. See if any of them pique your interest. If so, please consider writing something for the newsletter. We love to receive articles from our readers. You can find all the information in the box marked "Looking Ahead."

Until next time, stay healthy and happy.

Kathy

Publication of *CF Roundtable* is made possible by donations from our readers and grants from Sustaining Partners - AbbVie, Boomer Esiason Foundation, Cystic Fibrosis Services/Walgreens, Foundation Care, Gilead Sciences, Hill-Rom, TLC Rx/Modern Health, The Estate of Pamela Gordon Beaton, and Vertex Pharmaceuticals; Diamond Sustaining Partners - In Memory of Lauren Melissa Kelly.

Information From The Internet...

Compiled by Laura Tillman

PRESS RELEASES

Baby Bottle Steam Sterilizers for Disinfecting Home Nebulizers Inoculated with Non-tuberculous Mycobacteria

Non-tuberculous mycobacteria (NTMb) can contribute to respiratory infection in patients with chronic pulmonary disease. Contaminated nebulizers are a potential source of respiratory infection. Baby bottle steam sterilization was compared with vigorous water washing for disinfecting home nebulizers inoculated with NTMb mixed with cystic fibrosis sputum. No NTMb was recovered from any nebulizers after steam treatment whereas viable NTMb grew after water washing, demonstrat-

ing that steam sterilization effectively disinfects NTMb-inoculated nebulizers. <http://tinyurl.com/zlbqacb>

Two New Cystic Fibrosis Studies to be Stepping Stones to Future Research

A Case Western Reserve University researcher is starting two studies, based on angiotensin signaling and gene "correction," to advance research into more effective ways of treating cystic fibrosis (CF) and to better understand the ideal timing for such interventions. The first study will examine the potential benefits of medications, such as ACE inhibitors, designed to lower blood pressure and regulate fluid balance. The second study aims to help design more effective

clinical trials by uncovering important new data. Researchers will use CF mouse models treated with a drug to try and modify – "correct" – the genetic mutations that cause the disease. There are three specific issues they hope to determine: 1) which respiratory symptoms are reversible or preventable with genetic correction; 2) the ideal timing for the genetic correction (i.e., in infants or adults); and 3) the amount of genetic correction required (i.e., would partial correction suffice to alleviate or prevent symptoms, or must the gene be completely corrected?). <http://tinyurl.com/zv4ol65>

ProMetic's PBI-4050 To Target Cystic Fibrosis As Its Next Orphan Indication

ProMetic Life Sciences Inc. announced plans to initiate a double-blind placebo controlled Phase 2 clinical trial in patients suffering from cystic fibrosis (CF) and related diabetes and liver steatosis. With a longer life expectancy, it is estimated that 70% of CF

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LOOKING AHEAD

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

Spring (Current) 2016: Managing Various Conditions (Under The CF Umbrella).

Summer (August) 2016: Living With Anticipatory Grief And Survivor's Guilt. (Submissions due June 15, 2016.) Do you live with anticipatory grief or survivor's guilt because of your CF or have you dealt with it in the past? How do/did you handle it? Please share any good suggestions you may have for managing these feelings.

Autumn (November) 2016: Advocates And Advocacy. (Submissions due September 15, 2016.) Do you have someone (family member, spouse, caregiver etc.) who knows how to advocate for you if you can't (whether mildly or seriously ill)? Do they have a list of current medications, clinic visits, contacts etc.? How do you keep your advocate up-to-date? Tell us about your successes and pitfalls in making sure that your healthcare continues optimally when you are too ill to advocate for yourself.

Winter (February) 2017: Using Non-traditional Medicine Or Treatments. (Submissions due December 15, 2016.)



ASK THE ATTORNEY

Answers To Readers' Questions

By Beth Sufian, JD

In the past three months many readers have asked questions related to their ability to work part-time while receiving SSI benefits or SSDI benefits. A compilation of the answers to readers' questions is below.

People with CF, their family members and members of their healthcare teams can contact the CF Legal Information Hotline® sponsored by funding from the CF Foundation at 1-800-622-0385 or CFLegal@sufianpassamano.com if they have questions about the information provided or if they have other questions about the legal rights of people with CF. All contacts are free and confidential.

Questions and Answers

Question: How much money can I make from part-time work activity and still keep my Social Security benefits?

Answer: It depends.

A. General Information on Benefit Programs.

First, it may be helpful for some readers who may have little knowledge about Social Security benefits to understand some basic information.

Supplemental Security Income (SSI) is a program that provides a monthly cash benefit and Medicaid coverage. There are three groups of individuals who may be eligible for SSI benefits. First, a child under the age of 18 who meets the SSA medical criteria and whose parents meet the low income guidelines. Second, an adult who meets the SSA medical criteria but has not worked enough to receive SSDI benefits. Lastly, an adult who meets the SSA medical criteria and receives an SSDI benefit that is less than the SSI benefit amount in her or his state.

SSDI is a program that provides a monthly cash benefit and Medicare cov-

erage. Medicare coverage starts 29 months after the person's date of disability, which is the last day of full-time work.

SSDI is a benefit that a person who is unable to work full-time can receive after being approved for the benefit by SSA. The person must meet one of the SSA medical criteria and meet certain work requirements. In 2016, the person cannot be working making more than \$1,130 per month from part-time work in order to be eligible for SSDI. A person who has never worked cannot receive SSDI. A child cannot receive SSDI.

When applying for SSA benefits, only one application needs to be filed with SSA. SSA will determine if the applicant meets the criteria for SSI or SSDI.

B. Working and Benefits

It is important to understand that there are different rules regarding how much a person can work and still be eligible for SSI and how much money a

person can make from work and still be eligible for SSDI.

Sometimes a person with CF may give advice to others on-line about the amount a person can make from work activity and still keep Social Security benefits. Often the information is not correct because the answer depends on the type of benefit a person receives from SSA. The work rules differ depending on the Social Security benefit program.

C. SSI Work Rules

Under the SSI rules, for every \$2 a person makes from work activity their SSI check is reduced by \$1. For example, if a person receives \$300 a month from SSI and works making \$300 in a month then the SSI check will be reduced by \$150 that month. So instead of getting an SSI check for \$300 that month, the person will receive an SSI check in the amount of \$150.

If a person's work earnings reduce the SSI check to zero, then the person loses SSI and loses Medicaid. For example, if a person receives \$300 a month from SSI and works making \$700 in a month, then the SSI check will be reduced by \$350 that month. If you subtract \$350 from \$300 you get -\$50. Since the person is not eligible for SSI benefits at all the person will lose his or her Medicaid coverage as well.

A monthly SSI check can be different amounts for different people. Some people have worked a small amount and so are entitled to an SSDI check that is less than the SSI amount in their state. In such a situation, the person will get some SSDI benefits a month and some SSI benefits.

For example, if a person gets an SSDI check of \$400 a month, the person would be entitled to \$330 in an SSI benefit if the state SSI amount is \$730. SSA deducts the SSDI amount



BETH SUFIAN

from the SSI amount, \$730 minus \$330, and determines the SSI check amount is \$400. Some states provide the base SSI amount of \$730, but a few states add some funds to make the SSI check larger.

D. SSDI Work Rules

The work rules are very different for a person receiving SSDI benefits. Under the SSDI work rules, a person can make up to \$1,130 from part-time work each month and still meet the SSDI eligibility criteria. The SSDI recipient is given nine months of a Trial Work Period where he or she can make more than \$1,130 from work activity. A confusing part of the Trial Work Period is that when a person works making over \$780 in a month, that month is counted as one of the nine Trial Work Period months.

A person can be reviewed for SSA benefit eligibility at any time even if he or she has not used all of the Trial Work months. When SSA conducts a Continuing Disability Review, it can consider the part-time work activity and the person's health. SSA may determine the person can work full-time and terminate benefits. In most cases if SSDI benefits are terminated then Medicare coverage stops.

E. Continuing Disability Reviews

Based on calls to the CF Legal Information Hotline®, it seems that in the past six months SSA is reviewing individuals who are working making close to \$1,130 a month and in many cases SSA is terminating benefits for those individuals.

SSA conducts a Continuing Disability Review and finds that given the person's ability to work part-time SSA thinks the person is able to engage in full-time work. This is especially true for people who are working part-time in jobs that are considered heavy work. For example, working five to six hours a day standing or lifting heavy objects repeatedly is considered heavy work.

Individuals with CF need to be aware that the climate at SSA right now is not favorable for people who are working at heavy jobs and making close to \$1,130 from part-time work.

For a person who receives only SSDI, working making under \$780 a month from part-time work activity rarely results in a review of SSDI benefits.

If a person loses benefits because he or she has worked too much or has worked at a job that is considered heavy work, the person will be able to reapply for benefits, but it can take a long time to be approved. Currently it can take five to nine months for a person to be approved for SSI or SSDI benefits. Social Security is also scrutinizing applications for SSI or SSDI benefits.

F. Loss of Benefits Equals Loss of Insurance

Many people tell me, "I just need to keep the Medicaid." However, if a person loses SSI benefits, the Medicaid benefit also stops.

In 19 states the only way for an adult to obtain Medicaid coverage is to also be eligible for SSI benefits. A loss of SSI and Medicaid can be a disaster if the person does not have access to other health insurance coverage. In 31 states an adult who has low income and assets can be eligible for Medicaid even if the person does not have SSI benefits.

The Affordable Care Act (also known as Obamacare) expanded Medicaid to low income adults, but the U.S. Supreme Court said that the states could not be forced to offer Medicaid to low income adults. The Supreme Court decision made the state expansion of Medicaid benefits voluntary and that is why some states do not offer Medicaid to low income adults unless the adult is also receiving SSI benefits or is eligible for Section 1619b.

G. SSI and Section 1619b

If a person is able to work and make more than the amount of their SSI check times two, then the person

may be eligible for a program called Section 1619b.

Section 1619b is basically a section of the Social Security regulations that allows a person to work making a certain amount of money per month and lose his or her SSI check but still keep the Medicaid benefits.

A person needs to make sure that the Social Security office has indicated the person is enrolled in Section 1619b on the person's Social Security file. If Section 1619b status is not noted on the person's file, then the person will lose SSI benefits and Medicaid if the person goes over the allowable work activity income amount.

Recently, many people with CF have found that there is trouble with their classification as a person who is eligible for Section 1619b status. If there are issues with Section 1619b status, the person can lose her or his Medicaid benefit because there is no SSI check received.

A visit to the local SSA office to correct the problem is usually necessary. Many SSA offices are crowded with people who have a variety of serious medical conditions. It is important to be cautious and if possible wear a good mask when going to an SSA office. Arriving before the office opens in order to be one of the first in the office usually reduces the wait time to speak to an SSA representative.

H. Correct Understanding

People who receive benefits should make sure they understand the work rules before starting to work part-time. Loss of benefits is a serious matter, especially when a person is relying on Medicaid or Medicare benefits for insurance coverage. ▲

Beth is 50 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.



SPIRIT MEDICINE

CF: Boot Camp For The Spirit

By Isabel Stenzel Byrnes

Lastly, like some of my post-transplant CF friends, I've thrown myself into heavy-duty training mode for the Transplant Games of America, which will be held in June 2016 in Cleveland, Ohio. Training for the Games is a mental and emotional roller coaster, in addition to being a physical ordeal. So, in this article I thought I'd muse about how CF itself is boot camp for the spirit.

What is boot camp? Boot camp is a colloquial phrase for basic or recruit training that is used as initial indoctrination of new military personnel. Boot camp usually engenders images of men doing push-ups and jumping jacks, running through tires or doing pull-ups, while someone on the sidelines screams incessantly to do more. In the traditional sense, the military mindset usually won't accept failure, weakness, giving up or expressions of exhaustion.

For me, I've been attending boot camp because I have a goal to perform at the best possible ability for this 44-year-old, twelve year-post lung transplant anemic diabetic with arrhythmia. The mental game of boot camp begins when I look at the gym calendar to see when boot camp is scheduled. I am filled with a dread, a deep "I don't want to do this" message that permeates my body. My mind ignores it and packs my gym bag, sets the alarm and plans on attending.

I owe my discipline to a lifetime of CF. This was my training. I remember

all those years when I didn't want to do treatments or didn't want to go into the hospital but had to do it. I had to distance my mind and spirit from my body and follow Nike's advice to "just do it."

In the CF world, our bodies are constantly being confronted with physical obstacles. For some, there is reprieve; but what remains is a con-

stant cloud of uncertainty ahead. For others, there is one CF symptom after another that rears its ugly head. Often we think, "What's next?" Like the idea of boot camp, there is dread, anxiety and a "world of worry" that we carry around all the time. We train our minds to cope with this. We distract, withdraw, adapt, plan, compensate or we motivate ourselves to take care of our bodies.

In boot camp, the instructors set an agenda for what to do. They yell and scream for us to keep it up, go faster; don't give up. There are repetitions of plank jacks, squat jumps, jumping jacks, wall squats... whatever it takes to build muscle and cause pain. Repetitions increase strength. They work the muscle to its max, so that

muscle fibers tear and grow over time.

In CF, our health-care team sets the agenda for what to do. They, or our family and friends, scream on the sidelines for us to do more. The longer we live, the more our physical ailments repeat. There is repetition in our exacerbations, our weight loss and gain, our exercise tolerance and loss; even hemoptysis comes and goes. While some healthy people might freak out about pneumonia or a lung bleed, over time, we develop comfort with this "routine" because we know no better and have no choice. Like in boot camp, CF causes repetitions that increase our strength. We gain experience, comfort, familiarity even with physical trials. In other words, we gain resilience: the ability to endure and be flexible during

I owe my discipline to a lifetime of CF. This was my training. I remember all those years when I didn't want to do treatments or didn't want to go into the hospital but had to do it.



ISABEL STENZEL BYRNES

times of challenge.

Boot camp causes breathlessness, even with my normal lung capacity. My muscles burn. I experience pain. I constantly monitor my body, what hurts, what is strained, whether I have proper form. My mind is overcome by “how much longer.” I want to quit. But I don’t. I look at all the people around me, who are of different ages, shapes and sizes. I am especially inspired if I’ve dragged my husband to join me and I observe his agony. But really, most people are stronger than I, and I am usually the slowest and weakest in boot camp. I set aside my shame and joke that I’m “special needs.” But I also embrace that at least I’m here; doing my best alongside others who are doing their best. If they can do it, so can I... so I keep going, even if I need to pause when I am at the brink and start up again when I can. I tell myself, “Just keep breathing.”

And so it is with CF. We monitor our symptoms with hyper-vigilance 24/7. We endure pain, breathlessness and discomfort. Often we may ask when this struggle will be over. CF requires teamwork, just like boot camp. I can do far fewer push-ups in a minute by myself than when I’m in a group. My CF peers gave me the sense of belonging, the will to endure, the will to survive. Even now, I need to know we are all persevering together.

We need to “train the brain” to cultivate a mental drive to endure boot camp and CF. We need to work out our minds as much as our bodies. We need mantras like the little engine that could: “I think I can... I think I can.” Sometimes it helps to have spiritual faith that you can handle it or to say a prayer that strength will be sent to you. For me, when I push myself to the brink, I often look upwards and dedicate myself to my donor or my sister. It

“My CF peers gave me the sense of belonging, the will to endure, the will to survive. Even now, I need to know we are all persevering together.”

can help to dedicate our efforts to something bigger than self, such as God, someone who has died or our caregivers.

Intrinsic to my drive to be part of boot camp is a very important value: that I’m worth it. I deserve the time, energy, expense and focus to take care of myself. This sense of worth came from my upbringing as well as a deep-seated sense that I can own and control a part of my life. My motivation comes from believing in myself, wanting the best for myself and for having confidence that I can achieve. And isn’t that the same in life with CF, with all the treatments we have to do? We need to have a profound sense of worth to exert this much self-care. We need motivation that comes from within to do all of our treatments. We need hope that this effort will one day pay off. That we’ll win.

The coolest part about boot camp is that practice makes a difference: there are results. And so it can be with CF. The more we work at this disease, the more we pack our gym bags and “just do it,” for the most part, we can see results. Fitness does get easier; managing routine CF can get easier once we gain confidence. Our results are longevity and higher quality of life.

Unfortunately, CF is the kind of crummy disease where at some point, not everyone sees results. Some of you might be reading this article thinking, “boot camp is what other people can do, but not me.” Today, at our “transplant” boot camp, we had a woman with 27% lung capacity, waiting for lungs, walking slowly, carefully and deliberately around the track. Her

“boot camp” was harder than any of ours. My friend Julie Desch taught me, “If we can move, if we can breathe, we can exercise!” All of us, at some point, may see a decline in our strength, our lung capacity, our functioning. We have to acknowledge and grieve our losses. We have to recognize that the more we once were able to do, the more grief we’ll experience as we lose our abilities.

Finally, I tell my husband that the best part about boot camp is when it’s over. That’s a terrible attitude for someone who is blessed enough with health to be able to be in a boot camp. But really, the spiritual and emotional gifts of boot camp are priceless. I have earned a nap, rest, relaxation, freedom. My real reward for one hour of misery is my own sense of accomplishment and pride that I’ve endured. I have no one else to inspire except for myself. Isn’t it the same for living with CF and enduring the boot camp of CF life? At 44, I am filled with a sense of awe, gratitude and pride that I’m still around and that I’ve survived. Maybe you feel the same. Maybe you’ve come a long way, too.

We CF adults have been in boot camp for decades. We have been indoctrinated, like military personnel, for the fight for life. We are masters of training. We have mentally trained ourselves for the hurdles of CF; we have lifted the heaviest weights; we have lunged ourselves forward, down the track of our lives. I don’t say this lightly: that if CF has any blessings, one is that it has shaped our characters to become endurance athletes. ▲

Isabel is 44 and has CF. She lives in Redwood City, CA. She works as a bereavement counselor. You may contact her at: Isabear27@hotmail.com



SPEEDING PAST 50

And Another Thing

By Kathy Russell

Wahoo! We made it through another winter. That statement is no big deal to many people, but for most of us who have CF it is a very big deal. Winter brings so many risks of respiratory infections and other ills. Just making it through is a victory. I am grateful that I made it to another spring.

Having lived more than 70 years with CF, I have discovered many aspects of protecting me from winter's ills. (Not to mention the rest of the year.) I am among the first to get my flu shot every year. I also have had pneumonia shots, including the newer Prevnar shot, and I got my shingles shot a few years ago. I try to avoid getting any of these bugs that could try to do me in.

Another way that I try to protect myself is by using good hand washing technique. So many pathogens can be transferred on our hands. I wear gloves – leather ones, not plastic – when I go away from home. They serve two purposes. They keep my achy hands warm, and they protect my hands from all the “bugs” that are on all the surfaces I may touch. I wash my hands as soon as I get home and I wash the gloves periodically. I think it helps.

I mentioned my achy hands. CF-related arthritis is one of our maladies that can cause us great difficulties. My thumbs are tender and often are quite useless. They sometimes swell up and get really sore. Since opposable thumbs are supposed to be one of the things that separates us from other animals, having them not work is a real inconvenience. Fortunately, there are many days that my thumbs work quite well.

The arthritis in my feet is helped

by the custom orthotics that I wear in my shoes. I don't wear cute or fancy shoes, but my feet feel much better than they used to. I still have the sensation of having a sock wadded up beneath my feet but at least it isn't as painful as it was before I got orthotics.

My shoulders, ankles, hips and knees are bothered by intermittent pain of arthritis. I usually try to take it easier on whichever joint is giving me pain. Being nicer to the painful one seems to help alleviate the pain. My back, however, is a pain of a different color. The pain in my spine always is there. Sometimes it is hardly noticeable, but

most times it is screaming at me. I spend a lot of time sitting in a comfortable chair with heat at my back. It does give me some relief from the worst pain. Since I am unable to take most pain meds, I am grateful that heat helps. I am unable to sit in a chair that has a hard back. When we go out, where I may have to sit for some time, I take a small pillow that I can put behind my back. That way I am able to tolerate sitting in the types of chairs that usually are used in hotels and restaurants.

Blood pressure problems can go along with CF, too. I first knew of my high blood pressure when I was 12. No

one did anything about it, because they didn't know what was causing it. It wasn't until I was in my 50s that a cardiologist took the time to figure out what meds would help me the most. Since then, I have been able to keep my

B.P. in a much better range.

Another problem that many of us who have CF may face is gastro-esophageal-reflux disease (GERD). It is characterized by frequent reflux of our stomach contents back up our throats and into our mouths and/or lungs. When it goes into our lungs, we may experience pneumonia. Just having those stomach acids travel back up our throats is painful, unpleasant and can cause long-lasting damage to our throats (the esophagus). I dealt with this discomfort for most of my life until I found esomeprazole magnesium (Nexium was the brand that started it). Between taking that med, eating a low fat diet and using polyethylene glycol, which is the generic version of MiraLAX®, my GI problems have all but disappeared. I am a much happier

“One side-effect of hard coughing is incontinence. As a rule, people don't talk of incontinence in polite company.”



KATHY RUSSELL

person than I used to be. I don't miss the burning throat or the terrible cramping of my intestines.

As I think of all the things that are part of CF, I must remember sinus disease. For most of us, sinus disease is a given. My sinuses were bad from infancy. I had recurrent sinus infections and the concomitant pain. At one point, I could not tip my head forward because of the pain that caused. I had invasive sinus surgeries, where holes were cut into my sinuses from inside my mouth. Windows were cut into the interior of my sinuses. They were supposed to help the sinuses drain. The windows "healed up" in about a year and I had to have another surgery done.

My last sinus surgery was done by endoscope and used a laser to clean everything out. There was much less bleeding and the packing was much easier to remove. That surgery was more than 21 years ago! Why was that one so successful as compared to the others? Good question. I believe that it is because I have irrigated my sinuses with sterile normal saline and a small amount of sterile glacial acetic acid every day since about two weeks after that surgery. I have no more growth of polyps, and I have had only one or two slight bouts of sinus infection in all those years. Whatever the reason, I am delighted to be able to breathe through my nose and to not feel as if my eyeballs are being shoved out by the pressure of infected sinuses.

I did lose my sense of smell with the last surgery. Although I really miss being able to smell good things and to taste fine flavors, it was worth it to get rid of the pain. Fortunately, I have regained some of my sense of smell. It isn't as good as it once was, but it is a price I am willing to pay for the lack of pain.

I haven't mentioned anything about lung involvement in CF. Most of us do experience troubles with our lungs at

some time. Some, like me, go through periods of less lung involvement followed by periods of major lung involvement. I started out with frequent lung infections and pneumonia. Then I had some time where my lungs seemed to behave a little better – for a while. Then it was back to frequent infections and pneumonia. Although my lung capacity stayed fairly good for many years, when it started to decline it just kept going down. First I was on supplemental oxygen (O₂) at night. Then I went on O₂ 24/7/365. Thankfully, I have a portable concentrator so I still can get around.

For many people, the next step is lung transplant. Those who do that have outcomes that may be wonderful. Some are not so fortunate but most are. Since I have not gone through that, I will not say any more about transplants.

One thing that goes with lung involvement is hard coughing. Most of us are accustomed to that. One side-effect of hard coughing is incontinence. As a rule, people don't talk of incontinence in polite company. We do talk about it here. Although we do Kegel and other exercises, after about three really hard coughs our muscles give up and we wet our pants. At my advanced age, wetting my pants isn't so odd. A lot of old people wet their pants. I just never thought that I would be one of those old people. I have found something that helps with the problem. I buy TENA® panty-liners. They have a super absorbent material in them and give me some protection from ending up with wet panties. They have many products for different needs.

A problem that once plagued me but no longer is a problem for me is temporomandibular joint disorder or TMJ. The temporal mandibular joint is what connects the jaw to the skull, just in front of the ear. The muscles around the joint can become inflamed and very painful. I dealt with TMJ for many

years. I used various treatments, including a night bite-guard, to treat it. I didn't get full relief until after I started using supplemental oxygen. Apparently I had been holding my mouth in an odd configuration in order to breathe when I slept. Once I started using O₂ I no longer had to hold my mouth that way and the muscles were able to relax. I was able to get rid of the night guard and sleep relaxed. I feel that this definitely was a part of my CF.

When I was young, in my 20s and 30s, I had a lot of trouble with super sweaty feet. I was working as a hospital nurse so I was on my feet for eight hours each shift. My feet poured out sweat that "ate" the leather of my shoes. I had to replace my work shoes about every six weeks. My docs suggested many different methods to try to lessen the sweating, but nothing did any good. Once I stopped working, I started wearing open sandals and my feet were so much happier. Now I wear cushiony men's sox that help to keep my feet warm and dry or at least a little drier. This problem definitely is related to having CF, as is sweaty hands. I remember a time when I was about 13 and I had a part in an American history play. Each of us was given a 3x5 card with our lines on it. I held mine and the card practically dissolved from my perspiration. How embarrassing!

So, as you can see, there are many conditions that are related to CF and can cause us all sorts of problems. These are not all of the conditions that can accompany CF. Other people will tell you of theirs, elsewhere in this issue.

It's time for me to go out and enjoy the beautiful spring that is just starting. I hope that all of us have a happy spring, without too many allergy troubles. But that's another story for another time. ▲

Kathy is 72 and has CF. She is Managing Editor of CF Roundtable. Her contact information is on page 2.



WELLNESS THANKS CF!

By Julie Desch

I had a little chuckle when I read the focus topic for this issue, “Managing Various Conditions (Under The CF Umbrella).” It’s funny only in a sick kind of way, because I’ve found that as I get older and (thankfully) older, this umbrella is expanding into more of a circus tent. Kathy warned me of this. At age 28, my sister, Kathy, looked at me and said, “Julie, weird things start to happen when you get old!” She had CF, and three years later would die from lung failure. I was a mere teenager at the time and, of course, didn’t take her seriously. But she was spot on. That pesky little CFTR protein is a little bit like Kevin Bacon, except it usually doesn’t take six degrees of separation to find the link to whatever health issue is under consideration. Here are a few examples:

I HURT MORE: Maybe this is just getting old, I’m not sure. But now I make noises when I get out of a chair that I have never made before. My back...my knees...my feet, my shoulders, my freaking thumbs (what’s up with that?). I guess I’m partly to blame. After all, snatching a kettlebell repeatedly does place undue burden on one’s thumbs. But when your knees don’t let you jog anymore, your back hates bicycles and swimming pools bring on that unpleasant drowning sensation, what is a fitness addict supposed to do?

I blame CFTR. Years of antibiotics to rid my lungs of pseudomonas has converted my tendons to withered and worn shoestrings. Decades of coughing to clear my lungs of tenacious mucus doggedly determined to cling to my airways has caused my posterior chest

wall to morph into a balloon-like structure. Since the back of my chest is necessarily connected to the front of my chest as well as the rest of my back, these anatomic changes create havoc elsewhere. It’s an “ankle bone connected to the knee bone” scenario. Everything is connected. It sounds very Zen, and it’s true.

I AM ALLERGIC TO EVERY-

Whether it be hives and red-man syndrome, or near liver failure, the list of meds I can’t take is growing like the tapeworm my seventh grade biology teacher was certain I had.

THING: Again, thanks CFTR. After being exposed to probably every known antibiotic by now, my body has just said, “No.” Whether it be hives and red-man syndrome, or near liver failure, the list of meds I can’t take is growing like the tapeworm my seventh grade biology teacher was certain I had. This induces a lot of head scratching when I need IVs, of course.

MY INTESTINAL TRACT IS GIVING WAY TO GRAVITY: I’ve written about this before, so I will be brief. Sitting on a toilet for hours at a time is not what the anal sphincter was designed to accommodate. Fifty-five years of this and, well, you get the point.

...AS IS EVERYTHING ELSE: But we

won’t go there.

MY SKIN IS BARELY CONTAINING THE REST OF ME: Okay, so maybe those summers of lying on aluminum foil slathered with baby oil to get the coveted teenage tan is partially to blame. But the addition of occasional prednisone tapers have left my skin the thickness of toilet paper...single ply toilet paper. It doesn’t help that I have very little subcutaneous fat (thanks CF). With five dogs, many of which love to paw for attention, I am often a bloody mess, fascia and muscle visible beneath the growing pools of blood. My partner has become so exasperated that she literally wants me to wear soccer shin guards all the time (I said no). That, and those forearm sleeves that basketball players wear to look cool. Of course, even the size extra-small arm sleeves are too big for even my thighs, so that isn’t happening.



JULIE DESCH, MD

So I wear a lot of sweat pants and long-sleeve shirts. This doesn't always work well with menopausal hot-flashes, but who am I to complain?

I SMELL NOTHING: This is not always bad, as those of us with CF understand well.

I FORGET WHY I'M HERE: This one takes a bit more of mental gymnastics to connect to CF, but, it can be. Experience has proven to me that as one gets older, it becomes more difficult to multitask. Memory prowess naturally fades. This doesn't really bother me that much these days. Who cares about the steps of the Krebs cycle? But it helps to be able to remember why you walked into a room. I blame CF. My brain has been taxed daily for decades with remembering treatments, pills, airway clearance sessions, exercise etc. I think it's worn out. Thanks CF.

I HEAR THINGS THAT AREN'T THERE: Ringing, mostly. This is, of course, due to the years of aminoglycosides coursing through my body to rid my lungs of bacterial invaders. My inner ears rebel. Occasionally, usually at night, one ear will go completely dark (hard to imagine, I know). Without warning, all sensation of "hearing" will disappear from one ear, like it suddenly is submerged in water. Then, a new ringing will appear, in a different key, to join the chorus of other tiny hair cells singing their songs. In my mind's eye, I see another inner ear cell dying...choking on tobramycin residue. Oh, to be a pathologist with cystic fibrosis!

THE BEST PART OF THE DAY IS BEDTIME: It sounds more depressing than it really is, but I so look forward to going to bed! Life is good, to be sure. There are so many things I still love to do while awake...play and snuggle with my dogs, chill with my partner, walk outside, talk to my kids, go to the

gym, write articles for *CF Roundtable*, stay involved in CF research, watch with equal parts fascination and horror the incessant coverage of presidential election politics, glue my eyes to the television on TGIT to get my Shondra Rhimes fix. But, the best part of the day is when it's all over and I get to sleep. Maybe that is just about getting old, but I blame CF. It's tiring.

A NAP TOPS MY "TO DO" LIST: See above. Once I wake up, I sometimes can't wait until bedtime to sleep again. In fact, I usually schedule in "nap time" when I create my to-do list in the morning. Thanks, CF. Sometimes, I literally go back to bed after my morning treatment.

I "WARM UP" AT THE GYM FOR LONGER THAN I ACTUALLY WORKOUT: What is that snap, crackle and pop sound coming from that tiny old lady wearing the shin guards and arm sleeves? Oh that's just me warming up for my workout. You'll find me slowly moving over my foam roller and lacrosse balls moaning and groaning as I slowly work through the parts of my body that are supposed to move. After the pops and cracks are out, I then go through various contortions to make sure each joint is able to move in the direction it is designed to go. There are occasional surprises that arise from this regimen, which is why I must do it before I decide which movements are safe to load with resistance. This routine goes on for at least 45 minutes before I dare pick up a weight. Would this be needed without the barrel chest and resulting lumbar lordosis and hip flexor tightness? Yea, no.

I could go on, but it is nap time. This is what y'all get to look forward to. Welcome to my circus! ▲

Julie is 54 and is a physician who has CF. You may contact her at: jdesch@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 to share your story, inspire others, and to become a part of this official program of the Boomer Esiason Foundation.





FOCUS TOPIC

MANAGING VARIOUS CONDITIONS (UNDER THE CF UMBRELLA)

Preventing Incontinence As An Adult Living With Cystic Fibrosis

By Kori Tolbert

Anyone who's had a CF pulmonary exacerbation knows that coughing can be a full body experience. I have, on more than one occasion, pulled a stomach or other core muscle in the throes of a coughing fit. For women, this "full body coughing" can also affect our pelvic floor muscles and unfortunately lead to the possibility of incontinence or, to be blunt, wetting ourselves.

Incontinence is definitely something I would like to prevent. We certainly have enough else to deal with as adults living with CF. So, based on some research, and the work I already do to keep my whole body flexible and strong, here is what I have found to be helpful in preventing this problem.

Kegel exercises: I've been experimenting with these exercises since about the age of 18. These exercises aim to improve muscle tone of our pelvic floor by strengthening the pubococcygeus muscles (the muscles we use to stop the flow of urine). There are lots of other benefits of these exercises including sexual function for both males and females. There are several resources on the web and even applications that can be found on "smart phones" that can help with different types of Kegel exercises. Once I discovered how to engage these muscles, I could, and can, do these exercises almost anywhere. It's a good way to pass the time on the chair lift, at a clinic visit, in the grocery store check-out line.

Now, when I cough, especially if I'm having a coughing fit or especially hard cough, I can engage these mus-

cles. This helps to secure my pelvic floor and sort of stabilizes that part of my body to deal with the force of the cough. I have also found that tucking my pelvis under slightly helps support my lower body when I'm trying to get through those abrasive, full body coughs. When I remember to do this, it prevents cramping and pulled muscles that sometimes accompany prolonged, heavy coughing.



KORI TOLBERT

Some other tools in my tool bag for preventing incontinence have been Myofascial Release and Continuum. I came across these therapies in my exploration for enhancing my own whole body health. These forms of body work are great for both body awareness and true strength. I have

found that keeping my tissues free from adhesions and moving smoothly is not only helpful in preventing incontinence, but in having a healthy, free, strong body in general.

Myofascial Release: Myofascial Release, MFR, is a form of body work that frees adhesions that form in the fascia (connective tissue) of our bodies. Our fascia wraps around all of our muscles, bones, organs, everything right down to the cellular level. Adhesions can form from previous trauma, stress patterns, areas of holding, or repetitive motion (coughing). Making sure I take care of any restrictions in my pelvic floor keep it strong and free so that it can do the job of supporting me, keeping things intact and functioning well. (This, of course, also helps with my reproductive health, digestive health, respiratory health, spinal health... you get the picture.)

Continuum: Continuum is a form of self-movement that uses breath, micro-movement, sounding and attention as tools of inquiry. I have found it to be helpful in developing full body strength, flexibility and freedom. It's a practice that teaches us how to both listen to and communicate with our bodies. Again, it's not only helpful for dealing with and preventing incontinence, but also working with these bodies that we have been given on a larger scale. The ability to draw breath and sound through my lower body has helped deal with the beginnings of, and then preventing reoccurrence of, incontinence for me.

So there you have it: My two cents on preventing incontinence in our adult bodies. The tools I have mentioned above have helped, not only in

the area of incontinence prevention, but also in the health of my whole body. I hope that some of my experience is useful for you or your loved ones living with CF, or, maybe has inspired you to find something else for yourself that is.

Keep going. Keep growing.

Helpful websites for exploring MFR or Continuum in your area:

www.myofascialrelease.com

www.continuummovement.com ▲

Kori is 36 and has CF. She is a licensed massage therapist and health coach in the State of New York where she lives with her

husband and “fur babies.” She is passionate about the quest for optimal whole health for herself and others. She also is president of the Cystic Fibrosis Family Connection and runs *BoosterJots*, a patient empowerment company. Kori can be reached at kdt4@live.com or www.cffamily.org or www.boosterjots.com.

TILLMAN continued from page 3

patients will have an abnormal glucose tolerance and 35-40% of adult CF patients will have developed cystic fibrosis-related diabetes (CFRD). PBI-4050 is an orally active lead drug candidate with excellent safety and efficacy profiles confirmed in several in vivo experiments targeting fibrosis. Fibrosis is a very complex process by which continuing inflammation causes vital organs to lose their function as normal tissue is replaced by fibrotic scar tissue. The proof of concept data generated to date confirms this drug's antifibrotic activity in several key organs including the kidneys, the heart, the lungs and the liver. This double-blind placebo-controlled Phase 2 clinical trial will investigate whether PBI-4050 can reverse fibrosis in the pancreas and help manage the glucose intolerance and related diabetic complications experienced by the CF patients. The clinical trial will also allow us to monitor the effects of PBI-4050 on other important parameters in CF patients, especially lung and liver function.

<http://tinyurl.com/gsxr2em>

Cystic Fibrosis Phase 2 Trial of Drug Aiming to Stabilize Key CF Gene Underway

Nivalis Therapeutics, Inc., a Boulder, Colorado-based clinical stage pharmaceutical company, announced that the first patient has been dosed in the Phase 2 clinical trial of its lead investigational drug, N91115, a stabiliz-

er of the cystic fibrosis transmembrane conductance regulator (CFTR) protein. The purpose of this double-blind, randomized, placebo-controlled, parallel group study is to investigate the efficacy and safety of N91115 in adult patients with CF who have two copies of the F508del-CFTR mutation and are being treated with the CF drug Orkambi (lumacaftor/ivacaftor). According to Nivalis, N91115 works through a novel mechanism of action, called S-nitrosoglutathione reductase (GSNOR) inhibition, that is presumed to modulate the unstable and defective CFTR protein responsible for CF. GSNOR inhibition restores GSNO levels, thereby modifying the chaperones responsible for CFTR protein degradation. In preclinical studies, N91115 was shown to increase the function of F508del-CFTR, the mutant protein that is estimated to be present in almost 90 percent of CF patients. This stabilizing effect was shown to increase and prolong the function of the CFTR protein and may lead to an increase in net chloride secretion – an effect both complementary and agnostic to other CFTR modulators, like Orkambi.

<http://tinyurl.com/zdjwn7t>

AND

<http://tinyurl.com/je3uhtt>

Galapagos Starts SAPHIRA Phase 2 Study with GLPG1837 in Cystic Fibrosis Patients

GLPG1837 is a candidate CFTR

potentiator drug in clinical development for the treatment of Class III mutations in cystic fibrosis. The SAPHIRA Phase 2 program will explore the safety, tolerability and efficacy properties of GLPG1837 in CF patients with a G551D (SAPHIRA 1) or S1251N (SAPHIRA 2) Class III mutation.

<http://tinyurl.com/j4rccgj>

AND

<http://tinyurl.com/gquobzg>

Capsugel and Pulmatrix Enter Collaboration to Develop and Manufacture Novel Inhaled Therapeutics

Capsugel and Pulmatrix, Inc., have formed a collaboration to develop novel inhaled therapeutics to treat serious pulmonary diseases. Capsugel will combine its spray drying process development with Pulmatrix's iSPERSE (inhaled small particles easily respirable and emitted) technology.

Pulmatrix's iSPERSE dry powder technology is a small, dense and dispersible engineered particle technology that enables pulmonary delivery of drugs with high delivery efficiency, dose reproducibility and flow-rate independence. Several products leveraging the iSPERSE platform are in development, including an inhaled anti-fungal (PUR1900) for cystic fibrosis (CF). Capsugel and Pulmatrix have played leadership roles in the development of spray drying as an enabling technology to address the

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Living With CF And CFRD

By Kasey Ream

We all know what cystic fibrosis is, but what is CFRD? CFRD is cystic fibrosis related diabetes. CFRD develops when destruction is done to the islet cell, the cell in the pancreas that makes insulin as well as decreased sensitivity of the liver and muscles which help with making insulin naturally in the body. Not all people with CF will develop CFRD though.

My diagnosis:

I am a 29-year-old female living with CFRD. I was diagnosed with cystic fibrosis at the age of five weeks, after my parents started noticing I wasn't eating and passing food like a normal baby should at that age. That's when my parents made an appointment at Children's Hospital of Philadelphia (CHOP). The doctors at CHOP found I had a blockage, so I had surgery and was diagnosed with cystic fibrosis.

At the age of 12, I was diagnosed with CFRD, after having many glucose and hemoglobin A1C levels checked that kept coming back abnormal. That's when it became even more of a challenge and a daily struggle for me to keep both illnesses managed and to stop them from triggering each other since they go hand in hand.

My daily routine and managing both diseases:

Typically, a day for me managing two illnesses and trying to live as much of a normal life as possible starts early. First I need to check my blood sugars and do an insulin check by carb counting the breakfast I'm going to eat. When sugar levels are above 150, I need to use a sliding scale for my insulin to correct the levels and get them back under control. If levels are lower than 80, I need to adjust my insulin accordingly so I don't continue to go

low the rest of the day. I also drink or eat a little more to get them back up to baseline. If levels are high or low, I will have to check two hours after eating to make sure sugar levels have been cor-



KASEY REAM

“When I am sick, everything such as treatments doubles and sometimes blood sugar checks along with other antibiotics that may be needed.”

rected properly.

Next, I will do my morning CF treatments, which consist of four nebulizers such as Cayston/TOBI, Albuterol, Pulmozyme and 7% hypertonic saline solution. Along with those treatments I have to do airway clearance for 20 minutes with the Vest. All of this takes about an hour and a half.

I also take numerous vitamins and oral antibiotics on a daily basis and Creon with my meals.

My midafternoon lunchtime routine is to check blood sugars again, count carbs and do insulin according to what I'm going to eat for lunch. And then it's my midafternoon treatment time, which consists of two nebulizer treatments of Cayston and albuterol and a 20-minute airway clearance session with the Vest. This takes about 45 minutes. Dinnertime is the same for me once again as I check blood sugars, count carbs in the meal and plan the insulin accordingly. Nightly treatment times consist of three nebulizer treatments of Cayston/TOBI, albuterol and hypertonic saline 7% and a 20-minute airway clearance session with the Vest. I also check my blood sugars and do a once-a-day long-lasting insulin shot of Levemir (done at the same time every night). Also I do a second dose of vitamins and oral antibiotics.

Since exercise is a good way to manage both CF and CFRD, I try to

walk and/or do yoga a few times a week. This helps to keep my sugar levels good and my airways opened up and my lungs strong. When I am sick, everything such as treatments doubles and sometimes blood sugar checks along with other antibiotics that may be needed.

Insight/advice:

While managing two diseases is hard and takes a lot of time and self-discipline, don't ever forget to also live as normal a life as possible. The more active you are in managing your illness, health and everyday life, the better you are at staying strong and healthy. Take it day by day, challenge by challenge, and never let it bring you down. Make CF/CFRD fit into your life not the other way around. ▲

Kasey is 29 and has CF and CFRD. She has two siblings, one older and one younger, and neither has CF or diabetes. She lives in Pennsylvania right outside of Philadelphia. Her biggest passions are her family and friends. She enjoys going on vacation and living life to the fullest. You may contact her at: Kclyn1986@gmail.com



In Memory

**Mary Catherine
"Katie" Kelly, 26**
Chicago, IL

Died on January 20, 2016

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

Send to:

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PO Box 1618,
Gresham, OR 97030-0519.

E-mail to:

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IN MEMORY OF
LAUREN
MELISSA KELLY



Stress Urinary Incontinence In Cystic Fibrosis – Can We Talk?

By Laura Mentch

Urinary incontinence, the involuntary release of urine, is a common problem for girls and women with cystic fibrosis. Men and boys with CF also experience incontinence, but less frequently. An October 2015 study reported the prevalence of incontinence in CF for ages 12 and over as 68% in women and 11% in men¹. This article will focus on stress incontinence in CF with specific suggestions and resources for women; many of which may be helpful for men.

Earlier *CF Roundtable* articles have brought our attention to incontinence, repeating the guidance to do Kegel exercises to strengthen the pelvic floor muscles. Other types of incontinence, urge and overflow, are most often associated with menopause and aging.

Coughing is a significant cause of incontinence. The abdominal muscles contract with coughing or sneezing, putting pressure on the bladder, which forces open the urinary sphincter releasing urine in drops or gushes. The muscles of the pelvic floor are pushed outward. Can you feel the action of these muscles when you cough?

Pelvic muscles support the pelvic organs and bladder, like a hammock. With frequent and severe coughing, these muscles weaken and become less effective. The pelvic floor muscles surrounding the urethra, vaginal opening and anus are also stressed. Strengthening these muscles can reduce incontinence.

Our Experience with Incontinence
Though it can be part of our every-



LAURA MENTCH

day lives, we seldom talk about incontinence with family, friends or health-care providers. However, this topic comes up in Internet discussion among women with CF. When the door is opened, we share our frustrations along with practical and creative ways we address this problem. What women with CF say about incontinence: "It's so upsetting and embarrassing." "This is definitely one of the more humiliating things about CF." "I always wear panty-liners because I pee myself if I cough too hard." "I ALWAYS have pads in my purse." "Sometimes I wear two pairs of underwear." "I cough harder at home and while I'm doing breathing treatments and usually that is when it happens. Usually I just change clothes." "I often pee when I vest." "If I'm walking and I have to cough, I stop walking so I can concentrate on holding it in." "The only thing I have known to try is Kegels and so far it hasn't worked. Maybe I haven't

done it right. Sometimes I feel like I need Depends. It's pretty embarrassing." "My research nurse tells me this happens because Kegels aren't enough for us to be doing and we need to do pelvic floor exercises, whatever those may be." "This is the reason I do not work out in a gym." "I love to run but I leak the entire time, it is uncomfortable." "I swear it happens more during that time of the month." "My hospital bathroom is a laundry room." "I feel like it is one of those annoying side effects of CF that no one tells you about." "I haven't talked to my doctor about it yet. I never think it's a big enough issue or I have too many other CF-related things to worry about." "I guess it's not bad enough yet to say anything." "ANOTHER thing for me to bring up at clinic."

Incontinence can be distressing, affect our quality of life and further complicate our days. We compensate by wearing panty-liners, pads, incontinence underwear; by altering our activity, reducing fluid intake and limiting the strength of our coughing. We often carry this problem (along with pads or extra clothes) silently without telling

¹Gina Reichman, Veerle De Boe, Johan Braeckman & Dirk Michielsens (2015): Urinary incontinence in patients with cystic fibrosis, *Scandinavian Journal of Urology*, DOI: 10.3109/21681805.2015.1096826

PRACTICAL SUGGESTIONS

- Avoid bladder irritants such as alcohol, coffee and tea
- Maintain healthy fluid intake, especially water
- Support your pelvic floor when coughing with muscle contraction or placing your hand under pelvic floor muscles; some women sit on their foot or heel to support the pelvic floor during airway clearance.
- Keep coughing. Effective airway clearance is vital in cystic fibrosis care.
- Don't stop at every bathroom, "just in case."
- Avoid "hovering" above the toilet seat while urinating. Sorry, Mom.
- Adopt healthy posture for urination: feet on the floor, lean forward with breasts lifted up.
- Urinate before treatments or PFTs.
- Maintain fecal continence and avoid straining with bowel movements.
- With trampoline exercise, "jog" instead of jumping with both feet.

those close to us, including our CF care providers. If you experience incontinence, bring it up at your clinic visit. It will increase an awareness of the problem and encourage our nurses and doctors to learn about resources that can help. The good news is there is help.

Support for Incontinence

Kegels. Kegels are the incontinence mantra. Begin by finding and contracting the muscles that stop the flow of urine* and passing gas. Contract only these muscles, not those of your legs, buttocks or abdomen. Sitting with a folded washcloth under your vulva can help you isolate these muscles.

(*avoid squeezing the pelvic muscles while urinating; doing so will compro-

mise the normal process of urination)

There are two ways to perform Kegel exercises:

(1) quickly contract and squeeze these muscles, and

(2) contract, squeeze and slowly lift up, up, up and hold for a count of five.

Practice and repeat ten times at least three times each day.

If you are unsure if you are correctly practicing Kegels, search for instruction online, visit a gynecologist or find a women's health physical therapist.

I recommend these websites and their newsletters for help with Kegel exercises:

<https://www.pelvicexercises.com.au/>

[kegel/](https://urogyn.coloradowomen-shealth.com/patients/kegel-corner/)

<https://urogyn.coloradowomen-shealth.com/patients/kegel-corner/>

"The Knack." Tighten and lift your pelvic floor muscles BEFORE sneezing, coughing, laughing or lifting to avoid leaking urine during these activities that put pressure on the pelvic floor.

Posture. Keep your spine straight and upright during airway clearance to support the muscles engaged in coughing.

Incontinence Products. Before reaching for Depends, consider Poise products (panty-liners and pads) or protective underwear designed to hold urine.

Women's Health or Incontinence

Physical Therapy. Women's Health Physical Therapists are specially trained to help women of all ages and use specific techniques for incontinence. This therapy is especially helpful during and after pregnancy. Make an appointment with the Physical Therapist at your CF clinic or find a Women's Health or Incontinence PT at www.apta.org.

Urogynecology. Urogynecology is a specialty within obstetrics and gynecology. Urogynecologists have highly specialized training in surgical and non-surgical treatment of pelvic floor disorders, including incontinence. <http://www.findurogynecologist.com/> ▲

Laura Mentch is 62 and has CF. She is a health educator who is a USACFA Director. Her contact information is on page 2.

FACTORS ASSOCIATED WITH URINARY INCONTINENCE IN WOMEN

- Pregnancy and childbirth
- Age
- Obesity
- Changes at end of the menstrual cycle
- Medication
- Disease, e.g. diabetes
- History of pelvic or abdominal surgery

ADDITIONAL FACTORS FOR WOMEN WITH CF

- Persistent and violent cough
- Coughing during airway clearance & PFT
- Pulmonary exacerbation
- Prednisone
- Increased hydration with IV antibiotics
- CFRD

continuing my education. I have been considering the best road towards making good money and being successful. In an effort to further my career, I applied for a job as a chemist for an international water engineering consulting firm.

I got the job. This is my ticket to success ... my opportunity to work my way up the corporate ladder. I have a lot of experience in this field; I can succeed. I can prove to my family and my friends, who are in professional schools and working in their careers already, that I am just as successful. CF doesn't make a difference, and I am just like everyone else. This is seemingly amazing news, until I realize the realities associated with this job opportunity and my CF. This news then brings stress, anxiety, sadness and anger.

I have been receiving Social Security benefits along with Medicaid for a number of years. It has been a highly beneficial situation for me thus far. Free health insurance, along with the ability to earn a little money through work, all while living at home with my parents, has equated to having no major expenses. I have a little extra spending money for vacations and fun events with friends. I also have the most amazing man that I've recently met who has supported me with his whole heart, but I digress.

There is a maximum on how much one can earn through a work salary while receiving Medicaid. The salary offered with my job exceeds the maximum allowed by Medicaid. This gives me many facts to consider: First, I would lose Medicaid. I would never be able to get it back, because after working a while I would be eligible for disability, which isn't free. I would have to pay for my own health insurance, which comes with premiums, copays and medication costs. This could cost me upwards of ten thousand dollars a year. This would equate to 22% of my net pay ... quite a substantial amount. The largest change would be working the extra 13 hours a week plus any overtime. Needless to say, this is a

change from my current routine.

I am forced to face the facts. I have 30% FEV lung function. When I'm free of infection and flareups, I feel good. I hiked an elevation of about 700 feet in Virginia this past November. Despite feeling good, it was extremely difficult. When I am feeling bad, walking to and from the parking deck proximal to my lab leaves me short of breath. I have to keep up with many therapies to maintain my lung function and keep



ELLA BALASA

mucus accumulation at bay. Three times a day, I have a one-hour session that consists of nebulizing, VEST therapy, and chest physio therapy that has to be done by my mother, father or boyfriend. This is my routine: morning, afternoon, and late at night. I have a very accommodating professor (shout out to Dr. Franklin!) who allows me to work the 5-6 hours per day, midday, between my two therapy times. Once I get all my treatments and dinner over with at the end of the day, I have free time to spend scrolling through Facebook, going to yoga class, cooking up new dinners with my boyfriend and planning my upcoming trip to Spain. I am not stressed about my job, and I have the necessary

time to relax and rest.

So after receiving the call for the offer, I was overcome with dread. Making this life altering decision felt unimaginable. Why me? Why the hell do I have to be the one who has to deal with this? Why can't I just take this job and not worry about what's going to happen with Medicaid? Why can't I accept and advance my career and finally feel like I am all grown up? I want to have big-girl job status. I could ... but with it comes sacrifices.

I decided that I am not willing to make the necessary sacrifices. I would have to wake up at 6 a.m. five days a week, spend a lot of my energy during the day, come home tired, do my treatments, cook dinner, sleep and repeat in order to make a living. I could take this job, but then I would only have two weeks of vacation time during the year. I just want to make memories while I still can. I want to take care of my health and enjoy my life while I have it. If my passion for this field of work were stronger, yes I would have taken the job. I am a motivated individual and I put all my effort into my current work, but I have other passions and aspects of my life that I do not want to lose or ignore.

For example, I love drawing portraits of people, as it brings me joy. When I capture the likeness in a face, whether it be a smile extending to the twinkle in one's eyes, or the solemn eyes of a man staring into a far distance, I feel a sense of accomplishment. I feel a sense of success.

Exercising is also a crucial part of maintaining lung function, and I have been slacking in that department for months. I have fortunately been able to hold steady at 30% FEV for years, with the ability to do a minimal amount of physical exertion before my heart starts pounding and my breathing becomes rapid. I am quite winded after two flights of stairs and walking up a hill from the riverbank after an afternoon on the Belle Isle rocks is a workout. Remember that mountain hike? I want

to be able to climb that elevation and not have the feeling I might collapse. I wonder why I can't focus my energy on working towards being able to walk farther, faster, stronger ... that is success. That is success that matters to me. I have decided that this is where I want to place my focus.

My definition of success remains the same, but I am going to try to redirect myself. My prize will be one of a fulfilled life. I will be present in each moment, and take joy in small things. Honestly, I never really understood this cliché phrase until this situation hit me in the face. My life hasn't changed from one day to the next, and I am still the same person with the same mind, job and place. Through time and continued effort to succeed at new goals I have set for myself, I will become the person I want to be.

I have thoughts and unanswered questions about my future life because of these realizations. Will I ever make enough money to support myself and live the lifestyle I would like? Living with my parents and having their support has allowed me to have that extra money for

spending on trips and activities that I desire to do, but will I be able to have that if I'm still around and they aren't? Will I need someone to rely on financially? Being a woman doesn't make me feel any less obligated to make a living and contribute to a household income, in fact I want that. But can I feel like I am contributing if I stay on Medicaid and make very little money? How will it all work? Am I too lazy and that's why I don't want to work full-time? I've made enough excuses here as to why I can't work to last for days. I realize I am a bit of a worry wart, but these are all valid and rational thoughts.

I hope you've come to have a better understanding of just this one woman's thoughts on life, and the physical and mental obstacles that everyone can apply to their own lives in some way. I have realized that I am not a failure because I chose not to take the job position. I am not giving up on success, for I already am successful. And one day when I have stronger, more stable lungs, whether that be through transplant, rigorous exercise and therapy or new

gene therapy drugs, I still have the opportunity to climb that corporate ladder and find that kind of success. I am choosing to be positive, not to be complacent, and to move forward in the aspects of my life that I can control now.

I began writing this piece on a day I took off from work because I was fever-ridden and congested. See the perks of this job? Why would I ever want to change? Oh, the convoluted thoughts that go through my mind ... but it's all because I live and breathe. For that, I'm thankful every day. ▲

Ella is 23 and has CF. She lives in Richmond, VA, where she was born and raised. She is first generation Hungarian and proudly speaks the Hungarian language. She has two older siblings and is an aunt of three. Ella is the only one in her entire extended family to have CF. Her favorite season is summer. She loves the ocean, traveling, spending time outdoors, doing makeup and eating her favorite dessert, crème brûlée. She can be contacted via e-mail at balasag@vcu.edu. She would be happy to talk to others about CF or any other topics.

TILLMAN continued from page 13

limitations of conventional lactose blending dry powder inhaler (DPI) and metered dose inhaler (MDI) technologies. Spray drying technology provides fewer formulation dependencies, the ability to deliver either crystalline or amorphous drugs, and tighter particle size distribution than that provided by lactose blend technology. This engineered particle approach and expertise applied to the iSPERSE technology translates into significantly improved delivery efficiency, flow-rate independence and drug dosing for patients. <http://tinyurl.com/hf3mu3j>

Potential CF Treatment for Antibiotic-Resistant Infections Seeks EU Orphan Drug Designation

Arch Biopartners' AB569 is a novel CF Roundtable ■ Spring 2016

therapy for pulmonary infections with mucoid and non-mucoid *P. aeruginosa* bacteria that are resistant to traditional antibiotics. AB569 received orphan drug designation from the U.S. Food and Drug Administration (FDA) in November 2015 for its combined active ingredients in an inhaled solution. The drug combines two active ingredients, sodium nitrite and ethylenediaminetetraacetic acid. <http://tinyurl.com/guu9o4s>

Potential Cystic Fibrosis Therapy Gets FDA Fast Track Approval, Clinical Tests to Follow

Proteostasis Therapeutics, Inc., announced that it recently received Fast Track Designation from the U.S. Food and Drug Administration (FDA) for PTL-

428, which is an oral modulator in the amplifier class. Amplifiers are CFTR modulators that selectively increase the amount of an immature form of the CFTR protein, providing additional substrate for other CFTR modulators (e.g., correctors or potentiators) to act upon. <http://tinyurl.com/j5nthvp>

Concert Pharmaceuticals Announces CTP-656 Solid Dose Phase 1 Results Confirmed Superior Pharmacokinetic Profile to Kalydeco®

Concert Pharmaceuticals, Inc., announced new results from Part 1 of its multiple ascending dose Phase 1 clinical trial in which a single-dose tablet formulation of CTP-656 was compared to the commercial tablet formula-

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Mailbox

You all do amazing work and your positive magazine is just wonderful and inspirational. Thank you. *CF Roundtable* is just

what we CF families need. I have two kids ages 20 and 16.

Cheryl Baum
Lake Zurich, IL

In memory of all my fellow CF "Cysters and Bros" (over 20 CFers I've known and/or were close to) that this breath-taking disease has taken in my 57 years of CF life including Larry Culp who, if I remember correctly, was a founder of *CF Roundtable*.

Lynn Pancoast
Allentown, PA

Continue the great work!
Joan Finnegan Brooks
Osterville, MA

Many thanks for the years of *CF Roundtable* we have received. While our daughter with CF was a child and teen, we read it for motivation and inspiration. Now that she is 25, it means even more to us. Wishing you all every blessing in 2016.

From Maureen Marlow (no address)

Just a note to tell you our son, Ken O'Brien, was one of the original group who started *CF Roundtable* along with Kathy Russell and four others. Our Ken left us at age 45 on March 2, 2008. Happy New Year to all of you. Hello to Kathy & Paul Russell.
Jim & Carol O'Brien
Frankfort, IL

I am a 40-year-old with CF and I have a beautiful healthy 2-year-old daughter, Maislyn Lana Ostensen. We are making this donation through my husband's engineering company in the name of Gratitude! We are so thankful for my health and our amazing daughter! I am also thankful for *CF Roundtable*. Every issue is filled with inspiration!

Sonya Ostensen
Inverness, FL

Much appreciation to Beth Sufian and her colleagues!

Norman Young Jr.
Falls Church, VA

TILLMAN continued from page 19

tion of Kalydeco®, the current standard of care for cystic fibrosis patients with gating mutations. The results confirmed CTP-656's superior pharmacokinetic profile compared to Kalydeco, including a longer half-life, reduced rate of clearance, substantially increased exposure and greater plasma levels at 24 hours. CTP-656's half-life of approximately 15 hours supports the potential for once-daily dosing.
<http://tinyurl.com/ho2vncw>

Phase 3 Trial of Inhaled Antibiotic to Treat CF-related Lung Infections Supported by \$20M in New Financing

Savara Pharmaceuticals will advance a pivotal Phase 3 clinical trial of AeroVanc (vancomycin hydrochloride inhalation powder), the first dry powder inhaled antibiotic being developed for the treatment of methicillin-resistant Staphy-

lococcus aureus (MRSA) lung infection in cystic fibrosis (CF) patients. By delivering vancomycin directly to the site of infection, AeroVanc has the ability to improve its clinical efficacy and reduce adverse effects caused by broad-spectrum antibiotics. The purpose of the research trial is to evaluate the effectiveness, safety and pharmacokinetics of AeroVanc compared to placebo in subjects with CF and a chronic MRSA lung infection.
<http://tinyurl.com/hlhef3m>

XOR-Labs Receives \$2.6M to Advance Work in Improving the Quality of Donor Lungs

Currently, fewer than 1 percent (6,000 procedures per year) of lung patients benefit from lung transplantation, and an estimated 20 percent of the patients registered on waiting lists die before donors are found. The problem

is less a lack of replacement lungs than the suitability of donated lungs. XOR-Labs Toronto developed a process – based on Toronto Ex Vivo Lung Perfusion, or Toronto EVLP – to access and repair low-quality and unusable donor lungs, making them suitable for transplantation.
[success.http://tinyurl.com/hhdl9bl](http://tinyurl.com/hhdl9bl)

CF Patients Seen as More Likely to Reject Lung Transplants

A study found that cystic fibrosis (CF) patients, especially younger patients, may be at increased risk of acute cellular rejection of transplanted tissues compared to other transplant patients. Researchers compared the records of 44 patients who underwent lung transplants as a CF treatment and 89 patients who received lung trans-

Continued on page 29

THROUGH THE LOOKING GLASS

Going Through Hoops

*Sometimes it seems that we always are going
through hoops to just be.
There are meds to be taken and treatments to
do, you see.*

*Then phone rings. I answer and someone says,
"Let's get together."
I say, "I'd love to, but what do you think about
the weather?
Will it be too warm or too cool?"
(Having to ask that makes me feel like a fool.)
"At the place we are going, do they allow
smoke?"
(You know that most 'No Smoking' areas are
such a joke!)*

*"Do they have good air circulation there?"
(Or will we all be re-breathing each other's air?)
"Will everyone follow guidelines for cross-
infection protection?"
(Or do we have to worry about coming home
with some new infection?)
Before you can invite someone new you
must say,
"Do you have pan-resistant bugs, B. cepacia
or MRSA?"*

*Going to a conference requires even more work
Sometimes I feel like I am a jerk.
First it's a trip to a doc to collect sputum for culture
Then it may mean a visit to a blood-drawing vulture.
Forms to be filled out, releases to sign
We may risk our lives on the dotted line.*



PHOTO BY STEPHEN BOYER

*Seeing friends may be less work for you than for me
Since I must go through so many hoops, just to be.*

-K. Russell, 2000

"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...



CARL ROBINSON MARRIED KARA MCCANN ON NOVEMBER 22, 2015, AT ASILOMAR IN PACIFIC GROVE, CALIFORNIA.



THE FOUR MENTCH SISTERS (JULIE, LAURA, MARTHA AND SUE).



ISA STENZEL BYRNES ON TOP OF MOUNT WATZMANN IN THE GERMAN ALPS LAST JULY (2ND HIGHEST PEAK IN GERMANY).



SUSIE AND ADAM BALDWIN IN CAPE POINT, SOUTH AFRICA, IN DECEMBER 2015.



KORI TOLBERT "IN THE SWIRL" TAKEN FROM THE FILM THREE LIVES.



ELLA BALASA IN OREGON.



LINDA STRATTON WITH FELLOW CHURCH VOLUNTEERS, JENNY WINKLER (MIDDLE) AND BETTY FERRIN (RIGHT).



KASEY REAM'S FAVORITE PLACE TO BE IS AT THE PHILLIES GAME WITH FAMILY AND FRIENDS. CLOCKWISE FROM TOP LEFT: COUSIN LACEY, COUSIN MANDI AND KASEY, COUSIN NIKKI AND HER SISTER KOREY.



MEGAN PARKER WITH SADIE IN KANANASKIS, ALBERTA, CANADA.



ACTIVE FOR LIFE

Welcome And Hello!



By Aimee Lecointre

Welcome to CF Roundtable's new exercise column, "Active For Life"! I'm really excited about this opportunity to share my passion and knowledge for living an active lifestyle. Being active plays a very important role in helping me to maintain my lung function and overall health.

When I was younger, movement and being active came naturally for me. I loved riding my bike around the neighborhood, rollerblading, playing basketball with my dad and brother in the driveway. I learned to ski at a young age and kept it up until my first back surgery for scoliosis at age 16. I would wake up early just so I could jump on the trampoline before school. Sometimes I even got to jump around for a bit before school instead of using the Vest—what a treat! I enjoyed hiking with my family and friends. I was in dance classes for a few years as well as tumbling/gymnastics. I took swimming lessons most summers. In school I took electives like dance, weight-lifting and aerobics. Being active was always fun for me and I never felt like I was exercising. I truly enjoyed it. At the time, I never realized the significant impact it had on my health.

After graduating high school, I lost touch with being active. Exercise was no longer a priority or really even a part of my life. I don't really know what happened, but I slipped away from an active lifestyle without even realizing it. When I reflect back on those years, I can see the negative impact not being active and a lack of exercise had on my health, physically and mentally. My health definitely suffered.

Thankfully somewhere along the way I found my way back to an active lifestyle. It took me a while to get my groove back. I started out slowly with

things like walks around the neighborhood, then decided to get a gym pass where I started walking on the treadmill or riding the bike. I. Was. Bored. Treadmills just weren't my jam. I tried running but my knees hated it and mentally I didn't enjoy it all. I finally started to find the things that interested me and held my attention—activities that I enjoyed, made the time fly by and didn't feel like exercise. I began to realize the significant impact being active had on my health, which moti-



AIMEE LECOINTRE

vated me to keep going.

These days I do yoga almost daily and just recently started my 200-hour yoga teacher training! I typically stick with Vinyasa yoga; I like the flow and the challenge. It's amazing to me the heat we can build with our own bodies! Occasionally I like to mix things up and try different styles of yoga like

Hatha (restorative), Bikram (heated) or Kundalini (spiritual). I also really enjoy SUP yoga, which is yoga on a stand-up paddleboard on a body of water. I try to hike the local trails weekly. I lift weights three to four days per week, nothing crazy and it's usually in my own living room or the gym at our apartment complex. Living in sunny southern California, I love to take advantage of being outside and trying new things. I love ocean kayaking (total upper body workout) and stand-up paddleboarding (talk about a FULL body workout). I have also recently taken up skateboarding, which is really testing my balance and is fun. As you can probably tell, I like to mix things up and keep it fresh! Most of all I like to have FUN!

I absolutely believe that those of us living with cystic fibrosis need to be active. Our health depends on it. Our lives depend on it. Being compliant with our medications and treatments is a must and I truly believe that exercise and activity need to be a part of our treatment plans.

I have found that yoga has really helped me with my breathing. I can breathe deeper, slower, and I can catch my breath more easily. Yoga has helped me with my physical strength. It has helped me slow down my racing thoughts and lessen the anxieties that come with living with chronic illness and life in general. It's helped me to accept myself where I am and for who I am. It's really helped me to truly love myself, and that in turn has motivated me to continue to take the best care of myself that I can.

Living an active lifestyle has helped me in a number of ways. It's helped me strengthen my immune system and bounce back quicker when I do get sick

(which is much less often these days), reduce chronic pain (from scoliosis, coughing, CF-related arthritis etc.). It's made me happier and helped me fight years of depression; it's helped increase my energy levels and be more productive throughout the day. It's helped me to maintain my lung function. I sleep better. I feel healthier, happier and stronger. It truly has improved my quality of life all around.

After realizing the positive impact being active has had on my health, I have become passionate about sharing what I've learned over the years with the CF community and being a source of encouragement. I also want to change the way so many of us think about exercise. It doesn't have to be boring and it doesn't have to be a chore. No matter where an individual is in their health, there is something he

or she can do. I'm thankful and very much looking forward to writing for *CF Roundtable* about exercise with a dash of nutrition thrown in there! ▲

Aimee Lecointre is 31 and has CF. She is a Nutritional Therapy Practitioner and soon-to-be certified yoga teacher. She currently lives in Orange County, CA, with her husband. She can be reached by e-mail at: alecointre@usacfa.org.



Pay It Forward

BRONZE

Paul Albert (in honor of my 22nd lung transplant anniversary)
K. Gerald & Ruby Balls (in memory of Gregory Ted Baxter and Tamera Balls)
Bridget & John Barnes (in honor of Lucy Barnes)
Cheryl Baum (in honor of *CF Roundtable*)
Lauren Brenneman (in celebration of my 37th birthday and 13th wedding anniversary)
Joan Finnegan Brooks
Isabel Stenzel Byrnes (in memory of Ana Stenzel)
Maureen Cokelet
Edward & Elaine Corr (in honor of Kevin E. Corr's 58th birthday)
Mike Darrar (in celebration of my 50th birthday and 5th lung transplant anniversary)
Janie Davies (in honor of my 69th birthday)
Ruth Dunafon
David & Barbara Ebacher
Mary Lou Figley (in honor of my 56th birthday)
Ed Fleischman (in honor of my 74th birthday)
Doreen Gagnon (in memory of

Joseph Kowalski - founder and first president of USACFA)
Meranda Honaker (in memory of Jennifer Hale)
John Jacoby
Tom Jenkins (in honor of Steve Jenkins)
Theodore Kowalski
Gay Lazur
Maureen & Thomas Marlow
John & Susan McMurry
Jim & Carol O'Brien (in memory of our son, Ken O'Brien)
Ben & Donna Olsen
Lynn Pancoast (in memory of Becky Leonard)
Tia Schweizer
Juanita Serfoss
Larry & Ruth Shaw (in honor of Briana Robin Esther Shaw)
Janice Siegel
Robert Tate (in memory of Janice Tate and in honor of my 51st birthday)
Lloyd Wilder (in memory of beloved wife, Lisa Marie Wilder)
Lucie Wiseman (in honor of daughter, Caitlin Tarnof, celebrating 2nd lung transplant anniversary)
Melony & Steve Xynidis (in memory of Jennifer Hale)
Pamela & Dave Yanero

Norman Young Jr. (in celebration of 4 years cancer free, 32nd wedding anniversary and 61st birthday)

SILVER

Scott Adamson (in memory of Anabel Stenzel)
Sonya Ostensen/Ozomatic

GOLD

Phyllis L. Kossoff (in cherished memory of Stephanie Lynn Kossoff)

PLATINUM

Genentech
James Passamano & Beth Sufian

SUSTAINING PARTNER

Vertex Pharmaceuticals Inc.
Boomer Esiason Foundation

\$1 - \$249 BRONZE
 \$250 - \$499 SILVER
 \$500 - \$999 GOLD
 \$1,000 - \$4,999 PLATINUM
 \$5,000 - \$9,999 SUSTAINING PARTNERS
 \$10,000 - \$19,000 PEARL SUSTAINING PARTNERS
 \$20,000 and up DIAMOND SUSTAINING PARTNERS



IN THE SPOTLIGHT

With Susie Baldwin, MD, MPH

By Andrea Eisenman and Jeanie Hanley

When we started the “In The Spotlight” column, we hoped to feature a variety of people with CF who would inspire us and with whom we could relate; others who are going through situations similar to our own with our disease, cystic fibrosis, and also post-transplant life. In this interview, Susie fills the bill perfectly.

Susie took time out of her busy schedule of work and health-related issues to answer these questions posed to her. And she is busy—doing important things, as you will learn when you read her answers. You may have seen her previous articles in our newsletter about her transplant journey and her breast cancer diagnosis. She has also been through quite a lot post-transplant with struggles from sinus surgeries and many rounds of IV antibiotics to a pericardial cyst and peripheral neuropathy. Or as she puts it, three-plus years after transplant, “head to toe” issues. But it doesn’t seem to faze her. She, like most people with CF, is driven by her desire to help others and truly live life fully. See a photo of her and her husband, Adam, on page 22 on a vertiginous peak in South Africa! Speaking of Adam, he is involved in her healthcare and helps her emotionally as well as physically to keep her going. She has a great attitude about life that we find inspiring. We hope you do, too. Please welcome our newest star. Spotlight, please!

Age: 49 by the time this is published
Diagnosed at what age? Eight. I had what they called a “mild” case of CF, at a time when no one yet understood the full spectrum of this disease, both genotypically or phenotypically, the way they do now. I was pancreatic-sufficient so I didn’t have failure to thrive or GI issues, just a little, persistent

cough that no one could diagnose for many years. An astute pediatrician in our neighborhood, Dr. Edith Chen, sent me for a sweat test. I wound up having three sweat tests at three different medical institutions, because the results were equivocal. We had a test on Long Island, another in Manhattan, and then had to go up to Boston before



SUSIE BALDWIN

they made the diagnosis. My parents made that trip a mini family vacation and apparently I didn’t even ask why we were stopping at the hospital on the way to Plymouth Rock.

Where do you live? I live in Los Angeles, in a lovely little neighborhood in the hills not far from downtown.

Where did you grow up? Long Island, NY.

How has moving around affected your CF care? It’s been hard leaving the amazing doctors that have taken care of me in different places. The late Dr. Jack Gervoy, a kind, brilliant man with a

deep gravelly voice, who was a pioneer in CF care, took care of me at Long Island Jewish/Schneider Children’s Hospital from the time of my diagnosis until I left NY at age 29. Then my husband and I moved to Tucson, where I trained and worked for a total of eight years at the University of Arizona Medical Center, which had a fabulous CF center – Drs. Mark Brown (now in Colorado), John Mark (Stanford) and Wayne Morgan (still co-leading the U of A center) took wonderful care of me. The transition to LA CF care was the hardest – that’s another story in itself. Suffice it to say for now that my post-transplant care at UCLA with Dr. John Belperio and the transplant team has been terrific, but that much of the CF care I accessed in LA before that was not. In the years before my transplant, I actually traveled back to Tucson a few times to be hospitalized there instead of somewhere in LA – there was just no comparison. But the quality of care at U of A started worsening for me, when the adult patients were transferred from the pediatric pulmonary team to the adult service.

Was medical school a challenge for you with your CF care? Amazingly, it was not. I mentioned earlier that I was considered to have a mild case of CF. My first hospitalization was at age 19, then next at 23 and then not again until I was 31, when things started getting more difficult on a more permanent basis. So my life in medical school was, in retrospect, incredibly normal. I didn’t take any extra precautions in my clinical rotations and compared to my time in college, when I studied all the time, I managed to have a good social life too. I realize that I have been incredibly fortunate to have CF and still have been able to do all this.

How did you meet your husband,

Adam? We've been committed for 20 years, and will be legally married 20 years in October. It's gone by very fast. We met during the beginning of my third year of medical school, at the beginning of my surgery rotation. I was off for the weekend and went out to hear a band my college friend, Mark Ambrosino, was playing with at CB's Gallery, a little club next to CBGB. Adam was on bass. I had a thing for musicians (and other artist types). We chatted afterwards and I liked him immediately, but he didn't talk to me much. We met again nine months later at a surprise party for Mark and apparently were also being set up by Mark's wife, Rozan – she just had to get us in the same room.

How does Adam help you when you are not feeling well? Adam helps me every day, whether I am feeling well or not. I could not have my career without him. He feeds me, takes care of the household, including most of the animal chores. He “pounded” me every day, every night, for many years, often while watching Jon Stewart and Steven Colbert. Before my transplant three years ago, he helped me clean my nebulizer cups and packed my respiratory medicine and equipment when I traveled, which was frequently. He removed my PICC line for me once when the nurse couldn't come, and remains haunted by that – I guess he thought it was going to be a much shorter tube. He's helped me to be a calmer and better person, and taught me how to live in the moment. He comforts me. Other than the first few days I was going through opioid withdrawal, he made me laugh every single day throughout my difficult transplant recovery.

Being diagnosed with breast cancer, what went through your mind? I just thought it was ridiculous. I knew there was no such thing as a one-disease-per-person rule, because I had worked at the VA, taking care of

women with incredible stories of survival from multiple diseases or trauma – but somehow I believed in the one-disease-per-person rule for myself. Fear and frustration came later, as I realized that every step of the management of my cancer, which was stage 1, was being complicated by CF. It was actually my breast cancer that caused me to reach out for a CF support group, and that's how I found *CF Roundtable*.

How was it diagnosed and what was your recovery like? I was sitting on the couch, absent-mindedly watching TV, when I felt a firm lump on my side. It was about a centimeter long on the outer underside of my left breast – side boob, if you will. That was December 22, 2008. USC squeezed me in for a mammogram on Christmas eve and the radiologist came into the room immediately to tell me the lump was highly suspicious for cancer. He did the ultrasound right there and then, too. They squeezed me in again for a biopsy the week between Christmas and New Year's. We woke up first thing January 2nd to a call from the primary care doctor confirming the diagnosis.

Did that worry you about being a candidate for a transplant, having had cancer? Of course! When I went into respiratory failure I was only three years beyond my cancer diagnosis, and my understanding is that you're generally supposed to have survived cancer a minimum of five years to even be considered for a transplant. My case had to be carefully considered by the transplant board. Another amazing doctor I've had the good fortune to be treated by, breast oncologist Sarah Hurvitz, apparently made the case (as I've been told – wish I could have been a fly on the wall in that board room) that my hormone receptor positive cancer, which responds well to hormonal treatment, was not going to be what killed me. She believed that my prognosis from the breast cancer, following sur-

gery, radiation treatment and ongoing hormonal therapy, was very good.

When did you get your new lungs? December 21, 2012.

How much time did you take off after transplant and what issues did you have? I took off six months during the first half of 2012. I managed to get back to work for almost three months to try to wrap some things up, had to stop, got on the list and then was off another three months before I got “the call.” I thought I would be one of those people who goes back to work six months post-transplant, but I didn't go back until 15 months later. I had head-to-toe problems, one thing after another going wrong. But the lungs were great throughout. I lucked out with a solid match.

How was it going back to work? It was very emotional. When I left there in September to go on disability, they kept my office waiting for me, but I knew there was a chance I might never return. So I was excited to go back but also nervous. I started off really slowly with only eight hours a week at the end of March. Three weeks later I had an emergency appendectomy – totally unrelated to having a transplant – just a freaky, bad luck thing. It was another incident that just struck me as ridiculous. Because of the immuno-suppressants, my abdominal pain was mostly masked, so I wasn't a typical appendicitis patient. I was ready to walk out of the ER, thinking they were going to give me a diagnosis of constipation or something, when the attending hurried over and told me the CT scan showed that my appendix was severely inflamed and had to be removed immediately so that it wouldn't burst. “Are you kidding me?!” I asked. He was not. The surgeons rushed me off to the OR. Of course, I had to stay in the hospital longer than the average patient for IV antibiotics, and the recovery took longer because everything healed very

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slowly... and it took me until August to build back up to full-time work.

When I did get back to my old work routine, though, it started to feel like I'd never left. In a good way.

I should add, though, regarding going back to work, that I did a lot of volunteer work while I was recovering at home. When I recovered my capacity to read and think, about six months post-transplant, I immediately jumped back in to the health and human trafficking work that I'd been immersed in before I went into respiratory failure. I worked whenever I could the summer after the surgery to create a new website for people working at the intersection of health and human trafficking, because we were all very isolated from each other at the time. Then I connected with an East Coast colleague who had started a list-serve on the same topic. This led to the creation of our organization, HEAL Trafficking, which is actually making a difference – it's so gratifying to have lived to do this.

What do you do now? Since last April I've been the Sexually Transmitted Disease Controller for the Los Angeles County Department of Public Health, in the Division of HIV and STD Programs. My background and passion are in women's health and reproductive and sexual health, so this is a good fit for me. Now I also like to work in anti-violence public health and trauma-informed care. I co-lead HEAL Trafficking on the side.

Is being a physician hard when you are immune-suppressed? Yes. I haven't been able to return to patient care since my transplant. It's changed

my life profoundly to not be doing *any* patient care. I've done many other things in my career, like research, teaching, clinic administration, epidemiology, but taking care of patients even only one day a week kept me grounded. Until January 22, 2012, I had a twice monthly free clinic for human trafficking survivors and also worked at Planned Parenthood in San Bernardino on days off from regular work. Losing my ability to provide patient care was a great loss for me. I plan this spring to start doing some sexual health clinical work and would love eventually to also get back into working with trafficking survivors and youth at risk for trafficking. One step at a time...

What thrills you most about your work? The best part is being able to help people, which of course is most immediate in one-on-one direct medical services, but working at the systems level in public health is exciting too because you can fix things that can impact and help a lot of people. I also love the intellectual challenges and learning and getting to work with a lot of smart, dedicated people.

What do you do for fun? Spend time with animals, my husband, hiking, reading, visiting friends; TV, going to rock concerts and doing political activism – though that is more out of compulsion than for fun.

Do you know your CF mutations? Homozygous 3849 +10kbC. Rare Ashkenazi Jewish mutation associated with a small amount of functioning CFTR and conferring more mild disease. Until it's not mild anymore and at

44 you quickly start dying.

Are you interested in new medications targeting specific CF mutations to help with underlying CF issues like sinus or GI stuff? I will be very, very excited if there is ever a drug to help with my sinuses. I've had two sinus surgeries since my transplant and right now am completing a fourth week of IV antibiotics, the third course in four months. It's one of my new chronic problems and it's not easy.

Do you have advice for those with CF? Learn to contemplate death at a young age and live your life to make every day count, whenever you can. Plan for the future, even though you will struggle for survival. Take deep breaths when possible and try to slow down, enjoy beauty. Find your force or your faith. Find a trusted friend or family member and learn to ask for help when you need it. Cry when you need to. Allow yourself to feel bad for yourself sometimes, or angry. But be loving and open and kind to other people or animals or plants (or aliens) whenever you can. Helping other people is a great way to spend hours not thinking about your own situation or condition. ▲

Andrea Eisenman is 51 and has CF. She is a Director of USACFA and is the Executive Editor of CF Roundtable and Webmaster. Her contact information is on page 2. Jeanie Hanley is 53 and is a physician who has CF. She is a Director of USACFA and is the President. Her contact information is on page 2.

If you would like to be interviewed for "In The Spotlight," please contact either Andrea or Jeanie.



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Blogspot: <http://www.cfroundtable.com/blog-2/>



MILESTONES

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

ANNIVERSARIES

Birthday

Lauren Brenneman

Southbury, CT
37 on September 11, 2015

Scott Cherry

Hillside, IL
45 on March 7, 2016

Amanda Cokelet

Hazlet, NJ
36 on June 6, 2015

Kevin E. Corr

Taunton, MA
58 on July 29, 2015

Mike Darrar

Post Falls, ID
49 on August 25, 2015

Ed Fleischman

Plainview, NY
74 on December 24, 2015

Delayne Santos

Gulfport, FL
49 on February 14, 2016

Robert Tate

Elgin, IL
51 on October 20, 2015

Wedding

Lauren & Jehrod Brenneman

Southbury, CT.
13 years on March 29, 2016

Scott & Wendy Cherry

Hillside, IL
15 years on September 9, 2015

Kathy & Paul Russell

Gresham, OR
51 years on March 27, 2016

Delayne & Gerry Santos

Gulfport, FL
23 years on September 19, 2015

Carol & Robert Shepherd

Ft. Worth, TX
34 years on July 11, 2015

Transplant

Paul Albert

Catasauqua, PA
Bi-lateral lungs
22 years on February 10, 2016

Scott Cherry

Hillside, IL
Bilateral lungs
3 years on March 17, 2016

Amanda Cokelet

Hazlet, NJ
Kidney & bi-lateral lungs
16 years on June 16, 2015

Mike Darrar

Post Falls, ID
Bi-lateral lungs
5 years on May 3, 2016

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plants for other reasons, and found no difference between the two groups regarding acute cellular rejection events. Patients by group, however, differed in many respects. Those in the CF group were younger and had more pre-transplant infections or bacterial colonization, a lower body mass index (BMI) and fewer co-morbidities.

The team then stratified the patients into age groups, and compared patients with similar levels of cellular rejection. When taking age into account, they observed that CF patients had a higher risk of rejection. Moreover,

younger age was a risk factor for rejection in its own right. The study also looked at how often CF patients receiving lung transplants died of bronchiolitis obliterans syndrome (BOS) — an increase in fibrosis that closes the airways. No difference was found in BOS occurrence between CF patients and other lung transplant patients one year after surgery.

<http://tinyurl.com/jo4t6z9>

Infections May Increase CF Patients' Risk of Death in Lung Transplant Surgeries

A recent Dutch study found preoperative infection or inflammation to be associated with worse outcomes in cystic fibrosis patients undergoing lung transplantation surgery. This finding might help clinicians better estimate risk before such surgery. The study enrolled 53 patients scheduled for lung transplantation surgery at University Medical Centre, in Utrecht, the Netherlands. The team assessed their preoperative status using high-resolution computed tomography (HRCT) in combination with the severe advanced

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PARENTING

CF Mother Of Twins: Lessons Learned In The First Year

By Megan Parker

As I sit here writing this and doing my hypertonic saline, Desmond is happily perched beside me on the couch with a maraca in his hand. Shake, shake, shake. Smile. Shake, shake, shake. Smile. Charlie alternates between being mesmerized by my typing while standing up holding onto my leg and raising his arms way up above his head and waiting for my “Sooooooo big!” Such is life as a mother with cystic fibrosis (CF) during treatment time.

Let me back up. On September 30, 2013, I was introduced to two of my absolute favorite people in the whole wide world. After years of infertility and many failed attempts at becoming pregnant with assistance from the fertility clinic, I became pregnant following my second try with in-vitro fertilization. Although we only transferred one embryo, I became pregnant with identical twin boys. I was induced at 36 ½ weeks along and five days later we got to bring our new family home.

As any new mom can attest to, life at home with new babies is a bit of a steep learning curve. Being a new mom with CF is an entirely different game. For me, thriving during this first year really came down to having an amazing husband, incredible family support and being very adaptable and creative.

I am the number one biggest fan of my fabulous husband. He has always been my rock, my support, my encouragement, the kind of person that you are so proud to know. The kind of husband that makes you stop and think, “How did I get so lucky”? This was true before having children and has especially been true in the times since. The first few weeks after bringing our babies home are full of such

happy memories, despite being exhausted and not always feeling like we knew what we were doing. One of my most fond memories of these times is of my husband with one or two babies asleep on his chest, blanket snuggled around them. I’m pretty sure I have about 100 pictures of them asleep together, in various clothing and blankets, because it was so completely adorable and I want-



MEGAN PARKER WITH HER TWINS DESMOND AND CHARLIE.

ed to make sure to always remember how precious this was. These were also the days where my husband and I would alternate who did night feeds. Although the babies were getting only breast milk, it took a while before they were efficient at breastfeeding, so most of their milk was pumped breast milk. This gave us the opportunity to alternate night feeds so that we each could get a longer stretch of sleep during the night. One

particular night will always stand out for me. I was starting to develop a cold and my husband graciously told me that he would take night duty for a whole night so I could just rest (minus the 2 a.m. date I had scheduled with my breast pump). Keep in mind that we had to set the alarm every three hours to feed the boys to make sure they were gaining appropriate weight since they were still pretty little. Feeding the boys meant (1) going to the fridge and getting breast milk, (2) warming up breast milk bottles, (3) moving two sleeping boys from our bedroom into the living room where we fed them (this step and previous steps got pretty intense when one/both boys were awake and screaming for food!), (4) fighting to keep them awake long enough to finish their bottles, which often involved stripping them down to their diapers and blowing on their faces to keep them awake), (5) cleaning up the bottles once done, (6) changing diapers, doing up all those millions of snaps on their sleepers, re-swaddling babies, and (7) returning them to bed in our room. When I did this, I also added the extra steps of trying to breast-feed (it took them awhile to figure out the whole concept), pumping, transferring pumped milk into sterile containers, labeling containers and cleaning pumping supplies. Some nights this whole process could take a whole hour. So stepping up for night duty was a *big* deal. Unfortunately, that night the boys decided that they didn’t want to sleep because they were starting to catch the same cold that I had. So my remarkable husband spent the whole night alternating which baby he would cuddle and rock back to sleep, repeat with the other. Switch. Switch. Bottles. Diapers. Switch. Switch... And then go to work for a full day. And come back home to

care for three sick family members. Without a word of complaint.

Family support has been essential to us this first year. I am truly grateful and thankful that I have such an amazing family that steps up without me even needing to ask. My mom graciously wakes up at the crack of dawn and forces her way across the city through rush-hour traffic to get here at 7 a.m. to babysit so that I can get to the CF clinic. She sometimes would send me photos and videos so that I could see my babies while stuck in clinic. I had to do two courses of IVs during my boys' first year, and my mom came over

every single day to help out, keep me company and play with the boys while Homecare Nurses would come over to either change my PICC dressing or infuse one of my medications. My dad came to stay with us for a couple of weeks shortly after the boys were born to help out, and I have had my incredible in-laws come over countless times to help out and do errands and chores for us. They have even stepped up for night duty and let us (gasp!) sleep in from time to time. I'm not even sure the last time that my husband or I had to clean the house because family took care of it.

I learned early into the game that being adaptable and creative are essential traits. Not only did I have to learn to adapt to my children's ever-changing needs, but I had to learn to adapt my expectations of myself. Some days can be more challenging than others. Some days, I might feel short of breath. Other days, I might have to cough for 20 minutes in the morning before feeling okay. Other days, I might have no energy. And that's okay. That is CF. But what is harder to accept is how these things might impact my day and my interac-

tions with my children. When I feel short of breath, I might not be able to sing them their lullaby without sounding out of breath before their nap by the time I lug two babies upstairs, tuck them into their sleep sacs and put them in their cribs. When I have to cough in the morning before I feel okay, I accept

pounds of it (plus an extra 10). Despite wanting to exclusively breastfeed my children, to keep myself healthy, I had to partially wean my boys off of breast milk and onto formula until I reached a point where I could maintain and even gain some weight back. I have found that I need to be very creative in how to

entertain babies while completing my medications. I do not skip treatments. This is so important to me, because without my health, I have nothing, so I will do everything I can to keep my lung function as high as I can. Finding the where and when has been something that I have had to regularly change as the boys change and their schedule changes. It was easy

“As any new mom can attest to, life at home with new babies is a bit of a steep learning curve. Being a new mom with CF is an entirely different game.”

to get treatments done when the boys first came home and basically slept for 90% of the time. And then it was easy because they had several predictable naps throughout the day. And then it got harder because they would be awake during my medication times. It is *much* more difficult to commit to treatments when you have two adorable babies rolling around smiling at you, waiting to hear your voice while you sing or read a story. However, we adapt. I can easily sit on the floor while I complete my nebulizer medications and pass a ball back and forth, bounce a baby, stack cups, push a toy car or tickle little tummies. Books, songs, and conversations can wait until after. Chest physio time is independent play time for the boys so that I can really focus on getting the most out of it. Perhaps because they have to, or maybe because they are used to it, I find that they are great at entertaining themselves without me during this medication time. Completing treatments with awake and curious babies has made me learn a few things. Did you know that flutters make the greatest

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SEARCHING FOR THE CURE

Participating In A Clinical Trial: Benefits Versus Risks

By Meranda Sue Honaker

Cystic fibrosis (CF) research is progressing faster than ever with several therapies coming down the drug development pipeline. However, patient participation in clinical trials is an essential and required component of moving this research forward. Currently, the Cystic Fibrosis Foundation has several studies listed that are enrolling patients. These include drug studies and observational studies.

On a personal note, I have been participating in CF research as a patient for over two decades. Most recently, I completed the Phase 3 Orkambi clinical trial after two years of being enrolled in the study. Prior to the Orkambi study, I participated in other drug studies, observational studies, an early intervention study and studies for genetic markers. My contribution to CF research, even as a child giving blood for genetic markers, has advanced scientific knowledge.

Every person with CF who participates in research is making a positive impact on the future of thousands of people living with CF. That, alone, should be enough reason to encourage each person with CF to participate in research in some way.

Before a new therapy or medication can be introduced to the CF market, it must be put through at least three phases of clinical trials and these phases involve CF patients. In theory, these trials prove that the new medications and therapies are safe and efficacious in treating CF. However, this does not mean a new medication is devoid of side effects. Patients must be

aware that participating in a clinical trial carries risks in addition to potential benefits.

I choose to participate in clinical trials for a few reasons. First, the data collected from me advances knowledge of CF and gives researchers valuable information that may ultimately lead to a cure. Second, I want to gain access to potential therapies prior to marketing. For example, I chose to participate in the Phase 3 Orkambi trial because I knew I had the option of enrolling in the open-label extension study. This allowed me to have access to Orkambi for 18 months prior to FDA approval.

Third, I have always hoped that someday a child born with CF would not have to endure the challenges we from older CF generations have. This will be made possible only by research breakthroughs that are impossible without clinical trials and patients who participate in CF research.

When considering a clinical trial, you should first find out if you meet the inclusion criteria. This means you will need to meet a basic set of qualifications such as age, lung function, culture (for antibiotic studies), genetic mutations (for CFTR studies) etc. If you meet the basic inclusion criteria

and are screening for a drug study, you will then have testing at a screening visit. For example, when I screened for the Orkambi study I had to have blood work (which included genetic mutation testing), a vision exam, PFTs and

EKG. After all of my tests were within normal limits and I was confirmed to have homozygous f508del, I was able to schedule my randomization visit. The randomization visit was most exciting for me. I knew I would receive Orkambi or placebo with hope of receiving the former and not the latter.

Participating in drug studies means subjecting oneself to known and unknown potential side effects. It is imperative that each study participant thoroughly reads and understands every sentence on the consent form. Consent forms consist of information to include known side effects, who to contact during the study in case of emergency and limitations of the drug company's responsibility should you experience complications. They state that a patient may drop out of the study, by his or her

“Patients must be aware that participating in a clinical trial carries risks in addition to potential benefits.”



MERANDA SUE HONAKER

own choice, at any time.

I join drug studies with hope of symptomatic improvement, but consider the possibility of unpleasant side effects. For example, Orkambi reduced my CF symptoms. However, I also experienced side effects that were known and others that had not previously been reported. It was at this point I logically weighed the benefits versus side effects and decided it was in my best interest to remain on Orkambi from a pulmonary perspective.

You should consider participating in a clinical trial; however, you must assess potential risks while considering potential benefits as well. I encourage you to speak to your CF doctor about participating in a study. Something seemingly small such as donating blood for a genetic marker study may prove to have a positive impact on therapeutic advances in the near future. It will take

the majority of the CF population to advance scientific research. The more of us who participate in research, the faster data will be collected and analyzed. Subsequently, drugs will get to market faster than in years past.

Meranda is 32 and has CF. She is a Director of USACFA and is the Vice President. Her contact information is on page 2.

Clinical Trial Spotlight – CF Studies Currently Enrolling

Pharmaxis: A Safety and Efficacy Trial of Inhaled Mannitol (Bronchitol) in Adult Cystic Fibrosis Patients
<https://clinicaltrials.gov/show/NCT02134353>

2. Abbvie: Long-Term Study in U.S. Cystic Fibrosis Patients Receiving Digestive Enzyme Supplements to

Assess Narrowing of the Large Intestine Causing Adverse Intestinal Symptoms (Fibrosing Colonopathy)

<https://clinicaltrials.gov/show/NCT01652157>

3. Prevention of Cystic Fibrosis Related Diabetes (CFRD) Using Sitagliptin

<https://clinicaltrials.gov/show/NCT00967798>

4. Improving Treatment of Non-tuberculous Mycobacteria Infection in Cystic Fibrosis

<https://clinicaltrials.gov/show/NCT02372383>

5. Bayer: Early Signs of Efficacy Study with Riociguat in Adult Homozygous Delta F508 Cystic Fibrosis Patients

<https://clinicaltrials.gov/show/NCT02170025> ▲

PARKER continued from page 31

rattles ever? Did you also know that nebulizers are hands down one of the most interesting things ever made? And that all those tiny pieces of medication vials are so absolutely not baby-friendly? Sigh.

This first year I have learned some very valuable skills and lessons that I did not necessarily expect going into motherhood. I have learned how to hide IV tubes out of the reach of tiny grasping hands. I have learned how to breastfeed while giving myself IV meds (and how to tuck the IV pump out of the way of the breastfeeding pillow). I have learned how to entertain babies while tethered to my nebulizer. I have learned how to take care of my babies' needs while making sure mine are taken care of as well. I have learned the art of quick and healthy meals to satisfy myself and my babies. I have learned how to breastfeed two babies at the same time so that we actually have time to get out

and do things! I learned to make a packing list on the computer so that I didn't feel like we were missing anything whenever we left the house for a night (especially because it feels like we are packing up half the house). I learned how many calories you burn while breastfeeding two little hungry boys and how much work it can be to keep weight on. I learned that energy is precious, and that sometimes, spending your energy on tossing babies into the air or dancing around the kitchen is a way better use of energy than making sure your bed is made every day. I learned that it's incredible how much your medical team cares about your kids, even before meeting them. I've learned how showing off new pictures of my kids is an expectation and a top priority when it comes to CF Clinic visits. I learned that Homecare Nurses are really great at cuddling babies while IVs are being infused. I learned

that babies develop fake coughs to mimic mine and it is both hilarious and adorable. I have ascertained how incredibly fortunate I am to have such an unbelievable support system including my husband, family and friends. Most of all, I have learned that life as a CF mother is beautiful and rewarding beyond belief, despite at times being hectic and quite challenging. I have learned a great deal about myself this past year as I continually strive to be the best mother, wife, CF patient and person that I can be. ▲

Megan is 30 years old and has CF. She is happily married to her husband, Jordan. They have twins, Desmond and Charlie, who are 2 years old and a dog, Sadie. They live in Calgary, Alberta, Canada, and love to get out to the mountains as often as they can for camping, hiking and snowshoeing. Megan works as an Occupational Therapist.

Meet A New Director Of USACFA

Hi everyone. My name is Amy Braid and I am excited to be a part of USACFA. I am 35 years old and was diagnosed at age 6 after my mom was told many times that I had allergies. I was fortunate to have a very “vanilla” childhood and CF didn’t really affect my life too much until college.

I earned an Associate degree in interior design in 2001 from the Art Institute of Philadelphia and a Bachelor degree in interior design in 2007 from the Art Institute online. I received a Master’s degree in history in 2014 from Salem State University.

Currently I live in a small town, Wakefield, MA, just north of Boston with my husband and two stepchildren. However, I was born and raised in a

suburb north of Philadelphia. I moved up to the Boston area in early 2010 after meeting my now husband, and at that



AMY BRAID

time decided to stop working. Instead, I have focused on my health and staying busy with crafts and volunteer work.

In 2011 I was referred to Brigham and Women’s transplant clinic to be evaluated for a bilateral lung transplant. I was told I was too healthy to be actively listed and I have continued to stay that way!

Besides CF, I also have Lynch Syndrome and I have been learning how to manage both together. In my free time I love to read, crochet, draw/paint, bake bread, play with my dog and visit my niece and nephews (okay, my other family members too).

I am honored to be a director of USACFA and look forward to being more involved in this great CF community. ▲

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lung disease score (SALD), allowing it to analyze infection/inflammation, air trapping/hypoperfusion, normal/hyperperfusion and bulla/cysts. Patients also underwent a physical examination, a diabetes screening and a detailed overview of disease history. The team found that patients with a dominant preoperative infection or inflammatory disease, according to the SALD score, were more likely to die from postoperative complications. The authors of the study believe that if the results are confirmed in other studies, HRCT can be a useful tool for assessing preoperative and postoperative risk in lung transplant patients with CF. The technique is currently not widely used for perioperative risk estimations. <http://tinyurl.com/glvwr7v>

U of T Researchers Discover Mysterious Fungus That Defends Against

Neighboring Bacteria

Researchers at the University of Toronto examined fungi in the mucus of patients with cystic fibrosis and discovered how one particularly cunning fungal species has evolved to defend itself against neighboring bacteria. A regular resident of our microbiome – and especially ubiquitous in the lungs of cystic fibrosis patients – the *Candida albicans* fungus is an “opportunistic pathogen.” This means it usually leaves us alone, but can turn against us if our immune system becomes compromised. *Candida albicans* is a particularly wily fungus. Its signature maneuver is shapeshifting – it can morph from a round, single-celled yeast into a long stringy structure, allowing it to adapt to different environments and making it exceptionally harmful. What surprised the researchers was that some of this fungi began shifting into its stringy shape with-

out any environmental cue – usually this transformation (called filamentation) doesn’t happen spontaneously, but is triggered by the presence of certain substances, such as blood. To see if there could be a genetic explanation, the researchers sequenced the genomes of these samples and found a common denominator. All but one had genetic mutations in a gene known to repress the change shape – called *NRG1*. To find out why certain strains of this fungus would have developed this genetic variation, researchers looked to neighboring bacteria. As part of an ongoing battle between microbes, certain bacteria, which are also found in cystic fibrosis patients, secrete molecules preventing the fungus from changing into its stringy shape. The researchers tried exposing the mutated fungus to these bacterial rivals. Instead of responding to the bacterial signals, the fungus kept to its

Another Spring

By Linda Stratton

The bleakness of winter has passed,
turning into the hope of spring.
Tips of green slowly break Earth's surface,
a promise of beauty to come.

Warm air swirls through budding trees,
awakening the sense of change.
Newness, a rebirth, a revival born,
after a long winter's rest.

Spring is in the air for animal and human
alike,
both drawn to the warmth of the sun.
Always cautious of possible late winter's frost,
where ice covers tender bloom.

Small bodies, winged and bushy tailed,
build nests for this year's young.
While neighborhood gardeners gather tools
of their trade,
anxious to begin.

All is well for a moment in time,
before the heat of summer arrives.
Brown turns green and all through the land,
new attitudes emerge.

As for me, I'm thankful for life-giving spring,
put another notch on my belt.
I'm alive—I've made it one more year,
defying cystic fibrosis still.

Linda is 61 and has CF. She lives in Denver, CO, and enjoys the change of seasons that living in the "mile-high city" provides. In addition to dealing with CF, she stays busy caring for her father, Jim, her cat, Missy, and doing volunteer work through her church.

stringy form. The researchers believe these fungi have evolved to counter the tactics of their bacterial rivals.

<http://tinyurl.com/gpvzph9>

CF Infection Can Be Safely Treated by Daily and Continuous TOBI Use, Study Reports

A study comparing two continuous treatment regimens of tobramycin inhalation (TOBI) for *Pseudomonas aeruginosa* infection in patients with cystic fibrosis (CF) found that administration of both once a day and twice daily treatment was safe and well-tolerated.

Tobramycin has been shown to be effective against the bacteria. Since drug resistance might become an issue, a treatment schedule of 28 days on treatment

followed by 28 days off was introduced to minimize the risk of resistance. While the therapy has proven effective during the active treatment period, benefits are lost during the off-phase. This study investigated if continuous TOBI treatment could present a safe option. The authors concluded that TOBI inhalation treatment might be a safer option for *P. aeruginosa* treatment compared to systemic administration, as well as to the on/off treatment regimen. Longer studies including more patients are needed to confirm the results of this pilot study. <http://tinyurl.com/htz99de>

TREATMENTS

Inhaled therapy in cystic fibrosis: agents, devices and regimens. Breathe

(Sheff). 2015 Jun; 11(2): 110–118.

This comprehensive review discusses recent advances in inhaled therapies for the management of CF, including devices such as intelligent nebulizers, drug formulations and supporting evidence for inhaled antibiotics (for the management of chronic *Pseudomonas aeruginosa*) and muco-active drugs. Included is practical advice for clinicians regarding the optimization of inhalation technique and education. The influence of adherence on the use of inhaled therapies in CF is also reviewed.

<http://www.ncbi.nlm.nih.gov/pmc/articles/PMC4487378/>

Positive expiratory pressure physio-

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CFRI Retreat

By Devin Wakefield

Last August, I picked up a big, inflated ball with a cylindrical hole wide enough for me to shove my torso inside. Inside the hole, I found straps to carry this big ball, and put them on my shoulders like one would a backpack. I grabbed onto straps going down the inside of the cavity I was in, and ran. I crashed into another person inside a similar ball, and we bounced off each other. I rolled upside down before coming to my feet, but my friend was not so lucky: she fell on her butt. And so began the CFRI summer retreat's fun with bubble balls!

Every year in August, Cystic Fibrosis Research Inc. (CFRI) hosts a retreat for adults with cystic fibrosis and their families and friends in Menlo Park, California. The retreat provides a safe and welcoming environment aimed at enhancing positive coping skills, social support and education for people who share common experiences with CF. We gather at a retreat center called Vallombrosa, with its green lawns, happy trees, stray bunnies and California sunshine.

The venue has individual rooms for CF patients (one of our cross-infection protection guidelines, about which I will go into more depth later), a glorious snack room filled to the brim with favorites, a dining cafeteria, chapel, arts and crafts room, a big hangout room where we all meet together and extensive grounds for outdoor games and activities.

Last year, we started with a game of Hide and Go Seek, a way to get into small teams and start to learn about each other. Each team has to take a picture of as many items on the list with as many team members as possible. Unfortunately, I have to report

that my team did not win, but I SWEAR that bunny was RIGHT THERE behind those leaves in the picture! Honestly, it was! It's the camera's fault, clearly.

Each day, the retreat follows a general pattern: first breakfast at 8 a.m. (we have a snack room if you miss it snoozing!), one or two activities before lunch, a few more activities, and then

dinner. We leave time in between activities in case you need a break, or have to do medication.

The organizing committee tries to include the activities that people with CF would find fun, educational, healthy and inclusive. To stay fit, we have yoga and UJam (a dancercise similar to zumba), but we discuss with each instructor the varying levels of ability of CF patients and make sure there are plenty of breaks, and fluids (including water and Gatorade). I really appreciate these, as my case with CF requires a lot of exercise to stay well and have clear lungs. Oh my GOSH does exercise get up gunk!! (Ew, speaking of, where are my tissues?)

For emotional support, we have group "rap" sessions, with topics as diverse as work and careers, sex, grieving or coping with loss of health. These sessions allow time and a safe space to process some difficult challenges that come from CF. For example, at the last retreat, I began to really process the loss of a friend. I still miss her dearly, but I was finally able to start to let go.



DEVIN WAKEFIELD



BOUNCING AROUND IN BUBBLE BALLS ON THE LAWN.

And, of course, we have other activities just for fun. Last year we had a movie night, a comedy night (an improv troupe came, and together we made all the silly jokes), and every year we have a hilarious talent show! We get to see plenty of attendees do something cool or silly. No talent needed! Each attendee can watch, or join in to his or her heart's content.

As with any event with CF adults, CFRI has strong cross-infection protection guidelines that make the event safe for people who attend. We all take cross-infection controls seriously; without these guidelines, we would not have this retreat. Each CF patient attending has his or her own room to stay in, and gets a "hygiene kit"—a bag that comes with gloves, Purell, a mask and tissues. We instruct attendees to Purell at least before and after every meal, and when entering the snack room. CF patients use the tissues to cover their coughs and sneezes and reuse the bag to store the used tissues. The gloves and mask are available as an extra precaution. CF patients keep a safe distance from each other, especially when one is coughing, as there have been multiple studies showing that spray from a cough or sneeze may carry CF-risky airborne pathogens, and can travel up to six feet. And, of course, we remind everyone to use hand gel, use hand gel, and use hand gel! I keep saying we should do a Purell slip and slide one year, but that keeps getting voted down for some reason. I mean, what better way to fight cross-infection than getting Purell absolutely everywhere?

We also have medical advisors come to help during the day. They attend so when a medical issue arises, we have an experienced healthcare professional knowledgeable about CF. Usually, these medical advisors are nurses from Stanford, the CF center only a town away.

When we come together, we all

know we have a connection through CF. We also know that everyone has used different means to deal with CF and live their lives as they want. Each year I look forward to learning all that I can from each new member who comes, and to build deeper connections from past members I may not have had as much opportunity to get to know. These are the people I text and message on Facebook throughout the year as a new challenge comes up, and I seek their wisdom.

As fears of cross-infection have

physician showing that they have had a sputum culture at a CFF-accredited treatment center laboratory within six weeks of the event, and that they have:

1. Never had a confirmed positive culture for an organism belonging to or related to *Burkholderia cepacia* complex;
2. Have not cultured MRSA within the past 12 months;
3. Have not had a positive culture for Nontuberculous mycobacteria (NTM) in the past 12 months;
- and 4. Do not currently culture positive for any bacteria resistant to multiple antibiotics and potentially a



JESSICA MARTENS, ELLI HILL, LINDA SHAK AND ANNA MODLIN.

intensified, CF patients face a selected isolation. While my non-CF friends and family may do all they can to support me, I have consistently found that the people who best understand what I go through and how to deal with it are the ones who have had to face this themselves: other people with CF. Talking to someone with a perspective that intuitively understands the world of CF makes all the difference.

Register today for 2016! The retreat will be August 16th to August 21st. Prior to registering, CF adults should review the cross-infection guidelines: cfri.org/pdf/CFRI_Infection_Control_Policy.pdf. We require patients to get a signed medical release form from their

risk to other individuals with CF. While this unfortunately means that in a given year, some people cannot come, we would not be able to have this event without these rules.

For answers to questions, you can visit us at cfri.org, call (855) 237-4669 or e-mail CFRI at cfri@cfri.org.

I hope to see many of you there! ▲

Devin is 24 and has CF. He lives in Palo Alto, CA, with his family. He loves playing soccer, running around like a five-year-old, saying silly things and who knows what else. Currently he's funemployed, but whatever! He can roll around in the grass any time he wants. You can contact him if you like, at: devin.wakefield@gmail.com.

therapy for airway clearance in people with cystic fibrosis – The Cochrane Library. Wiley Online Library. Maggie McIlwaine, Brenda Button, Kerry Dwan. Editorial Group: Cochrane Cystic Fibrosis and Genetic Disorders Group. Published Online: 17 JUN 2015

Following randomized controlled studies in which positive expiratory pressure (PEP) was compared with any other form of physiotherapy in people with CF, this Cochrane review determined that there was a significant reduction in pulmonary exacerbations in people using PEP compared to those using high frequency chest wall oscillation (HFCWO) in the study where exacerbation rate was a primary outcome measure. The effectiveness of PEP is similar to other methods of chest physiotherapy such as postural drainage with percussion, active cycle of breathing techniques, autogenic drainage, oscillatory PEP devices such as the flutter and acapella, thoracic oscillating devices such as the “Vest,” and BiPaP, which is a type of PEP system delivering both positive inspiratory and expiratory pressure. No difference in lung function, the amount of mucus cleared from the airways or its related effects on the health of people with CF between PEP and other forms of chest physiotherapy was found. Although PEP seems to have an advantage in reducing flare-ups (based on the combined results of a few studies), different physiotherapy techniques and devices may be more or less effective at varying times and in different individuals during baseline function and chest flare-ups. <http://tinyurl.com/oxmz3j8>

Killing effect of nanoencapsulated colistin sulfate on *Pseudomonas aeruginosa* from cystic fibrosis patients. E. Sans-Serramitjana, E. Fusté, B. Martínez-Garriga, A. Merlos, M. Pastor, J.L. Pedraz, A. Esquisabel, D. Bachiller, T. Vinuesa, M. Viñas. *Journal of Cystic Fibrosis*. Published Online: December 18, 2015

The aim of this study was to explore

antimicrobial activity of nanoencapsulated colistin (SLN-NLC) versus free colistin against *P. aeruginosa* clinical isolates from CF patients and to investigate their efficacy in biofilm eradication. The authors found that colistin nanoparticles had the same antimicrobial activity as free drug against planktonic bacteria. However, nanoencapsulated colistin was much more efficient in the eradication of biofilms than free colistin. Thus, these formulations have to be considered as a good alternative therapeutic option to treat *P. aeruginosa* infections. <http://tinyurl.com/hxzgzs>

Reinforcement of the bactericidal effect of ciprofloxacin on *Pseudomonas aeruginosa* biofilm by hyperbaric oxygen treatment. Kolpen M, Mousavi N, Sams T, Bjarnsholt T, Ciofu O, Moser C, Kühl M, Høiby N, Jensen PØ. *Int J Antimicrob Agents*. 2016 Feb;47(2):163-7

This study demonstrates that biofilm reoxygenation by hyperbaric oxygen treatment (HBOT) can significantly enhance the bactericidal activity of ciprofloxacin on *P. aeruginosa*. Combining ciprofloxacin treatment with HBOT has potential to improve the treatment of *P. aeruginosa* biofilm infections. <http://tinyurl.com/h7jl7hl>

One-year safety and efficacy of tobramycin powder for inhalation in patients with cystic fibrosis. Kinston MW, Flume PA, Galeva I, Wan R, Debonnett LM, Maykut RJ, Angyalosi G. *Pediatr Pulmonol*. 2015 Dec 27

The authors aimed to determine one-year safety and efficacy of tobramycin powder for inhalation in patients with CF. Efficacy of tobramycin powder for inhalation (TIP) was maintained for up to seven cycles. Long-term treatment with TIP was generally safe and well tolerated with no increase in adverse events (AEs). <http://tinyurl.com/znh6b8q>

Activity of Tobramycin against Cystic Fibrosis Isolates of *Burkholderia cepacia* Complex Grown as Biofilms.

Kennedy S, Beaudoin T, Yau YC, Caraher E, Zlosnik JE, Speert DP, LiPuma JJ, Tullis E, Waters V. *Antimicrob Agents Chemother*. 2015 Oct 26;60(1):348-55

High-dose tobramycin can effectively decrease the thickness of *B. cepacia* complex biofilms, even in the presence of sputum, suggesting a possible role as a suppressive therapy in CF. <http://tinyurl.com/jd8lh9u>

A Phase 3, multi-center, multinational, randomized, double-blind, placebo-controlled study to evaluate the efficacy and safety of levofloxacin inhalation solution (APT-1026) in stable cystic fibrosis patients. Flume PA, VanDevanter DR, Morgan EE, Dudley MN, Loutit JS, Bell SC, Kerem E, Fischer R, Smyth AR, Aaron SD, Conrad D, Geller DE, Elborn JS. *J Cyst Fibros*. 2016 Feb 4

For patients with cystic fibrosis (CF), the use of inhaled antibiotics has become standard of care to suppress chronic *Pseudomonas* airways infection. There are limited antibiotic options formulated and approved for inhaled use and antibiotic efficacies attenuate over time, making additional inhaled antibiotic classes desirable. APT-1026 (levofloxacin inhalation solution, LIS) is a fluoroquinolone in development for management of chronic *P. aeruginosa* airways infection in patients with CF. It was determined that LIS did not demonstrate a difference in time to next exacerbation when compared to placebo. An improvement in FEV₁ (% predicted) at 28 days was observed and LIS was well tolerated. LIS is safe and has a potential role in the management of CF patients with chronic *P. aeruginosa*. <http://tinyurl.com/z5ccd2n>

Gaseous nitric oxide to treat antibiotic resistant bacterial and fungal lung infections in patients with cystic fibrosis: Phase 1 clinical study. Deppisch C, Herrmann G, Graepler-Mainka U, Wirtz H, Heyder S, Engel C, Marschal

M, Miller CC, Riethmüller J. *Infection*. 2016 Feb 9

Individuals with cystic fibrosis (CF) receive antibiotics continuously throughout their entire life which leads to drug resistant microbial lung infections which are difficult to treat. Nitric oxide (NO) gas possesses antimicrobial activity against a wide variety of microorganisms in vitro, in vivo in animal models and a Phase 1 study in healthy adults showed administration of NO to be safe. In none of the patients were serious drug-related adverse events observed which caused termination of the study. The intention-to-treat analysis revealed a significant mean reduction of the colony forming units of all bacteria and all fungi, while mean forced expiratory volume 1 s % predicted (FEV₁) relative to baseline increased.
<http://tinyurl.com/zn87t6q>

Omalizumab Treatment for Allergic Bronchopulmonary Aspergillosis in Cystic Fibrosis. Emiralioglu N, Dogru D, Tugcu GD, Yalcin E, Kiper N, Ozelik U. *Ann Pharmacother*. 2016 Mar;50(3):188-93

Allergic bronchopulmonary aspergillosis (ABPA) in cystic fibrosis (CF) is characterized by destructive changes in the airways. Long-term treatment with oral corticosteroids is often required for repeated exacerbations. Because elevated total IgE is a cardinal abnormality of ABPA, omalizumab has been used sporadically to decrease corticosteroid dose or totally replace corticosteroids. This study showed steroid-sparing effect, decreasing IgE levels, and improvement in respiratory symptoms in six CF patients with omalizumab treatment. Although this is a small sample of the population, omalizumab may be an alternative therapy for ABPA in CF patients who fail to respond to systemic corticosteroids or have serious adverse effects.
<http://tinyurl.com/hk8qscu>

PATHOGENS

Clinical implications and characteriza-

tion of Group A Streptococcus infections in adults with cystic fibrosis. Skolnik K, Nguyen A, Somayaji R, Thornton CS, Waddell B, Surette MG7, Rabin HR, Parkins MD. *BMC Pulm Med*. 2015 Dec 12;15(1):161.

Streptococcus pyogenes (Group A *Streptococcus*; [GAS]) was an uncommon lower respiratory pathogen of adults with CF. Identification of GAS in sputum was frequently associated with pulmonary exacerbation PEx, particularly when numerically dominant. However, transient GAS infection did not result in chronic infection nor appreciably change long-term disease trajectory.
<http://tinyurl.com/nbqx4do>

Detection of airway colonization by *Aspergillus fumigatus* using electronic nose technology in patients with cystic fibrosis. de Heer K, Kok M, Fens N, Weersink EJ, Zwinderman AH, van der Schee MP, Visser CE, van Oers MH, Sterk PJ. *J Clin Microbiol*. 2015 Dec 16

Objectives - Currently there is no non-invasive test that reliably detects early invasive pulmonary aspergillosis (IA). An electronic nose (eNose) can discriminate various lung diseases through analysis of exhaled volatile organic compounds. Results indicate that *A. fumigatus* colonization leads to a distinctive breathprint in CF patients. The present proof of concept data merit external validation and monitoring studies.
<http://tinyurl.com/ps24x9y>

Epidemic spread of *Pandora* *pulmonicola* in a cystic fibrosis center. Degand N, Lotte R2, Decondé Le Butor C, Segonds C, Thouverez M, Ferroni A, Vallier C, Mély L, Carrère J. *BMC Infect Dis*. 2015 Dec 26;15(1):583.

Pandora spp. are recently discovered bacteria, mainly recovered from cystic fibrosis (CF) patients, but their epidemiology and clinical significance are not well known. This study, which is the first to describe an epidemic spread of *P. pulmonicola*, notes the potential transmissi-

bility of this bacterial species and the need for infection control measures.
<http://tinyurl.com/j5btuj4>

Culture-based and culture-independent bacteriologic analysis of cystic fibrosis respiratory specimens. Mahboubi MA, Carmody LA, Foster BK, Kalikin LM, VanDevanter DR, LiPuma JJ. *J Clin Microbiol*. 2015 Dec 23

Cystic fibrosis (CF) is characterized by chronic infection and inflammation of the airways. Sequence analysis indicated that in a considerable proportion of samples, species not reported by selective culture constituted a relatively high proportion of the total bacterial load, suggesting that routine CF culture may underrepresent significant segments of the bacterial communities inhabiting CF airways.
[analysis.http://tinyurl.com/hhvghjw](http://tinyurl.com/hhvghjw)

Changing Epidemiology of the Respiratory Bacteriology of Patients with Cystic Fibrosis. Salsgiver EL, Fink AK, Knapp EA, LiPuma JJ, Olivier KN, Marshall BC, Saiman L. *Chest*. 2015 Jul 23

A retrospective analysis using data reported to the CF Foundation Patient Registry (CFFPR) from 2006-2012 was performed in order to determine the annual percent changes in the prevalence and incidence of selected CF pathogens. Pathogens included *P. aeruginosa*, methicillin-susceptible *S. aureus* (MSSA), MRSA, *Haemophilus influenzae*, *B. cepacia* complex, *Stenotrophomonas maltophilia* and *Achromobacter xylosoxidans*. Changes in nontuberculous mycobacteria (NTM) prevalence were assessed from 2010-2012 when the CFFPR collected NTM species. In 2012, the pathogens of highest prevalence and incidence were MSSA and *P. aeruginosa*, followed by MRSA. The prevalence of *A. xylosoxidans* and *B. cepacia* complex were relatively low. From 2006-2012, the annual percent change in overall (as well as in

Continued on page 42



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Cost: \$85 per person for the entire week. Daily fees are \$15 per day for visitors or \$10 per meal for those who drop in for a meal only. Overnight accommodations and transportation are the

responsibility of participants. Scholarships are available.

Safety: All people with CF are required to comply with cross-infection behavioral precautions. A medical advisor is available at all times, and volunteers are available to assist with respiratory treatments.

* Attendees with CF must have a medical release form signed by their physician showing they have had a sputum culture within six weeks of the event, and that they have: 1. Never had a confirmed positive culture for an organism belonging to/related to *Burkholderia cepacia* complex (B.cc); 2. Have not cultured MRSA within the past 12 months; 3. Have not had a positive culture for Nontuberculous mycobacteria (NTM) in the past 12 months; and 4. Do not culture positive for any bacteria resistant to multiple antibiotics and potentially a risk to other individuals with CF.

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most age strata) prevalence and incidence significantly decreased for *P. aeruginosa* and *B. cepacia* complex, but significantly increased for MRSA. From 2010-2012, the annual percent change in overall prevalence of NTM and *M. avium* complex increased.
<http://tinyurl.com/h98sqnu>

Clinical outcomes in cystic fibrosis patients with *Trichosporon* respiratory infection. Charles R. Esther Jr., Rongpong Plongla, Alan Kerr, Feng-Chang Lin, Peter Gilligan. JOURNAL of CF. (Article in press)

Respiratory cultures from CF patients were screened for novel pathogens *Trichosporon* and *Chryseobacterium*.

Patients with positive *Trichosporon* cultures had a greater decline in FEV₁ over time, whereas *Chryseobacterium* did not influence lung function. Treatment of *Trichosporon*-infected patients was associated with improved lung function.
<http://tinyurl.com/h2zkop3>

CF Patients with Uncommon Lung Bacteria at Initial Risk of Exacerbations

A recent study reported that the presence of transient Group A streptococcus (GAS) in patients with cystic fibrosis increases the risk of pulmonary exacerbations. GAS infections are uncommon in CF, but can contribute to potentially dangerous community-acquired pneumonia. The researchers also tested if the bacteria were sensitive to commonly used antibiotics. They found that GAS was susceptible to anti-streptococcal antibiotics and anti-pseudomonal antibiotics often used in CF, with the exception of azithromycin. The study reported that none of the patients developed chronic airways infection or other complications such as bacteremia, necrotizing pneumonia, or empyema following GAS infection. The authors concluded that transient GAS infections do not lead to chronic infections or altered long-term outcomes in CF

patients exposed to the bacteria.
<http://tinyurl.com/gl59pkd>

FYI

Recovery of resistant bacteria from mattresses of patients under contact precautions. Viana RE, Santos SG, Oliveira AC. Am J Infect Control. 2015 Dec 28.

Microorganisms may contaminate hospital mattresses even after terminal cleaning. The authors investigated the recovery of resistant bacteria from the mattresses of patients under contact precautions at a university hospital. They evaluated 51 mattresses. A total of 26 had resistant bacteria on the surface; the predominant species were *Acinetobacter baumannii* (69.2%), *Klebsiella pneumoniae* (11.5%), and *Pseudomonas aeruginosa* (11.5%). The median length of hospital stay was 41 days; the bed occupancy for patients under contact precautions and the time at which the patient was diagnosed as a carrier of resistant bacteria was 18 days. These results highlight the importance of controlling the potential spread of microorganisms through hospital mattresses.
<http://tinyurl.com/hmt46jc>

Hepatopulmonary Syndrome in Patients With Cystic Fibrosis and Liver Disease. Breuer O, Shteyer E, Wilschanski M2, Perles Z, Cohen-Cymerknoh M, Kerem E, Shoseyov D. Chest. 2016 Feb;149(2):e35-8

Hepatopulmonary syndrome (HPS) is a liver-induced lung disorder defined as a triad of liver disease, pulmonary vascular dilatation, and a defect in oxygenation. It is most commonly associated with portal hypertension. Severe liver disease with portal hypertension is present in 2% to 8% of patients with cystic fibrosis (CF). The authors speculate that HPS is underdiagnosed in patients with CF because of their coexisting respiratory morbidity, and advocate routine screening for every patient

with CF who has liver disease and portal hypertension.
<http://tinyurl.com/gp2zmm4>

Colonoscopic screening shows increased early incidence and progression of adenomas in cystic fibrosis. Niccum DE, Billings JL, Dunitz JM, Khoruts A. J Cyst Fibros. 2016 Feb 2

Colorectal cancer is an emerging problem in cystic fibrosis (CF). The goal of this study was to evaluate adenoma detection by systematic colonoscopic screening and surveillance. The authors concluded that early screening and more frequent surveillance should be considered in patients with CF due to early incidence and progression of adenomas in this patient population.
<http://tinyurl.com/h2p5u6x>

Maternal and perinatal outcomes of pregnancies in women with cystic fibrosis – a single center case-control study. Aude Girault, Julie Blanc, Vanessa Gayet, François Goffinet, Dominique Hubert. Respiratory Medicine. Article in press.

Due to increased survival, more women with cystic fibrosis become pregnant. However, studies on the specificities of pregnancy in CF versus healthy women are lacking. This study indicates that multidisciplinary care of pregnancy in women with CF resulted in maternal and perinatal outcomes similar to those found in women in the general population.
<http://tinyurl.com/zwqwj2c>

A Prospective, Randomized, Double-Blind, Parallel-Group, Comparative Effectiveness Clinical Trial Comparing a Powder Vehicle Compound of Vitamin D With an Oil Vehicle Compound in Adults With Cystic Fibrosis. Hermes WA, Alvarez JA, Lee MJ, Chesdachai S, Lodin D, Horst R, Tangpricha V. JPN J Parenter Enteral Nutr. 2016 Feb 22

There is little consensus on the most effective vehicle substance for vita-

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min D supplements. Fat malabsorption may impede the ability of patients with cystic fibrosis (CF) to absorb vitamin D in an oil vehicle. In adults with CF, vitamin D3 is more efficiently absorbed in a powder compared with an oil vehicle. Physicians should consider prescribing vitamin D in a powder vehicle in patients with CF to improve the absorption of vitamin D from supplements. <http://tinyurl.com/zjphocq>

Cystic Fibrosis (CF) Patients Suffer from Infection and Inflammation, Exercise Can Enhance Quality of Life

Cystic fibrosis patients suffer from infection and inflammation, but exercise has been shown to help enhance quality of life. There are numerous benefits from exercise for overall good health, but moreso for those with cystic fibrosis. A study has found that exercise could have anti-inflammatory effects in

cystic fibrosis patients. Exercise has been shown to exert anti-inflammatory effects and is associated with a reduction in disease incidence and viral infection susceptibility.

<http://tinyurl.com/jzuc94h>

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

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Published by the United States

Adult Cystic Fibrosis Association, Inc.

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