CF Roundtable
A NEWSLETTER FOR ADULTS WHO HAVE CYSTIC FIBROSIS
SPRING 2015

Selling Hope: The Façade Of Commercial Stem Cell Clinic Regenocyte

By Stephanie Woodward, the Skeptic Cystic

There's been a lot of buzz lately centered around a commercial stem cell treatment center called Regenocyte. I wanted to voice my concerns for people who may have a sparked interest in seeking out this company and summarize the research I have compiled on it. Based out of Florida, Regenocyte, which is also known as Intercellular, is run by beleaguered doctor Zannos Grekos. Or perhaps I should say *former* doctor.

After years of trouble and reprimand, Dr. Grekos was finally relieved of his license to practice medicine in April 2013, and his appeal was lost early in January of this year. The decision by the Florida Board of Medicine to revoke his license was made after a judge found him guilty of committing medical malpractice, when a patient he was treating in 2010 with his unproven stem cell treatment died shortly after the procedure. Her cause of death was a massive stroke due, allegedly, to grossly filtered particles of bone marrow injected directly into her carotid artery. After the patient's death, Dr. Grekos's license was put on emergency restriction and he was strictly told not to perform any stem-cell related procedures.

Unfortunately, this information was not relayed to the family of a five-year-old Texas boy, who paid Dr. Grekos his $57,000 up-front fee for stem cell therapy to treat the boy's primary pulmonary hypertension. Despite calling desperately for three months, the treatment was never scheduled, and the boy died. The family received only a $10,000 refund. Months later, Dr. Grekos was found to have performed yet another stem cell procedure in his clinic, during which the patient also died. That case is still under review.

Regenocyte's website still claims that Grekos is a licensed cardiologist with extensive experience in stem cell science. Cystic fibrosis is one of the conditions for which it specifically advertises the treatment as a panacea. It asserts that the blood samples the patient gives are sent to Israel for the cells to be "educated and activated" to become a target organ. He calls the final product "regenocyte cells" (which he admits is a marketing term, not a biological one) and claims that his clinic has observed patients circumvent transplant and reduce hospital admissions and the need for supplemental oxygen, steroids and inhalers.

However, experts and leaders in the field of stem cell research have

Continued on page 13

INSIDE THIS ISSUE

CF ROUNDTABLE
FOUNDED 1990
Vol. XXV, 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.

CF Roundtable now is free for everyone. Tax-deductible donations are gratefully accepted to help defray the costs of printing, production and mailing of the newsletter as well as the costs of Website maintenance. Please subscribe online at: www.cfroundtable.com or by mailing in the subscription form at the back of this newsletter.

CF Roundtable does not give medical advice. Any medical opinions represented in these articles are those of the writer and do not represent the views of USACFA, any of our Sustaining Partners, or any other group or individual. We strongly suggest you consult your doctor regarding any medical references and before altering your medical regimen in any way. USACFA does not endorse any products or procedures.

United States Adult Cystic Fibrosis Assn., Inc.
PO Box 1618
Gresham, OR 97030-0519
E-mail: cfroundtable@usacfa.org
www.cfroundtable.com

USACFA Board of Directors

Jeanie Hanley, President
Manhattan Beach, CA
jhanley@usacfa.org

Meranda Honaker, Vice-President
Fayetteville, NC
mhonaker@usacfa.org

Lisa Cissell, Secretary
Bardstown, KY
lcissell@usacfa.org

Stephanie Rath, Treasurer
Brownsburg, IN
srath@usacfa.org

Mark Levine, Subscription Manager
West Bloomfield, MI
mlevine@usacfa.org

Andrea Eisenman, Executive Editor/WEBmaster
New York, NY
aeisenman@usacfa.org

Laurel Avery, Director
Manassas, VA
lavery@usacfa.org

Klyn Elsbury, Director
San Diego, CA
kelsbury@usacfa.org

Paul Feld, Director
Florissant, MO
pfeld@usacfa.org

Chris Kvam, Director
Rochester, NY
ckvam@usacfa.org

Laura Mentch, Director
Bozeman, MT
lmentch@usacfa.org

Beth Sufian, Director
Houston, TX
1-800-622-0385
bsufian@usacfa.org

Karen Vega, Director
Cortlandt Manor, NY
kvega@usacfa.org

Kathy Russell, Managing Editor
Gresham, OR
krussell@usacfa.org

EDITOR'S NOTES

I hope that spring has come to your area. I know how hard this winter has been on so many. We can only hope that the rest of this year will be better than the beginning was. It is beautiful here in Oregon and we finally are getting some snow in the mountains. We won't get as much as normal, but at least we will get some. Maybe we won't have to ration water in the summer.

This is another jam-packed issue of *CF Roundtable*. We are so fortunate to have so many people who will share their experiences with all of us. Remember that you are what makes this newsletter.

We introduce a new column in this issue. **Meranda Honaker** writes "Searching For The Cure." You will find a list of current clinical trials there. Check it out on page 32. **Jeanie Hanley** has a letter for you on page 11 that talks of our survey results. It also tells of how we use those results and a new survey that is coming.

By now, I hope you have read the article that starts on the front page, written by **Stephanie Woodward**, the Skeptic Cystic. She talks of dubious claims of "stem cell" cures of CF. We must all be aware of the dangers inherent in such claims. Check out "Voices From The Roundtable" on page 36, to read what five respected physicians—**Michael Boyle**, **Jerry Nick**, **Daniel Weiss, Moira Aitken** and **Patrick Flume**—have to say about stem cells.

The Focus topic is: Transitions – Many Types Of Changes and several people had something to say about transitions. **Lisa Cissell**, **Andrea Eisenman**, **Klyn Elsbury**, **Laura Mentch** and **Jessica Newport** all wrote of various changes in their lives and how they handled them. **Jennifer Hale**, in "Coughing With A Smile," writes of getting back up. **Isabel Stenzel Byrnes** suggests that we use our inner strength to deal with change, in "Spirit Medicine." In "Protecting What Matters," **Mark Manginelli** writes of protecting one's finances and property in advance of changes. **Karen Vega** has a guest writer for "Parenting," where **Megan Murray** writes of how she handled the differences between her dreams of being a parent and what really happened. I continue the transitions theme in "Speeding Past 50."

Beth Sufian answers questions about insurance and co-pays in "Ask the Attorney." As always, **Laura Tillman** has done an outstanding job of gathering and compiling "Information From The Internet." "Transplant Talk" is by **Alison Lynch** and tells of her experience with Nissen fundoplication. "In The Spotlight" features **Janine Ullyette**. I think you'll find her life interesting.

On page 41 there is information about nominating people for the two USACFA awards. Be sure to take a look and think about people who deserve to be recognized.

Until next time, stay healthy and happy.

Publication of *CF Roundtable* is made possible by donations from our readers and grants from Sustaining Partners - AbbVie, Boomer Esiason Foundation, CF Services, Foundation Care, Gilead Sciences, and Hill-Rom.

Information From The Internet...

Compiled by Laura Tillman

PRESS RELEASES

U.S. Lawmakers Pass "Ensuring Access to Clinical Trials Act of 2015" for Rare Disease Research

U.S. lawmakers recently introduced new legislation that could greatly benefit thousands of Americans suffering from CF by helping them gain access to and participate in clinical trials without having to worry about their health coverage. The new bill, called the "Ensuring Access to Clinical Trials Act of 2015," was sponsored by a bipartisan group of senators and legislators. The bill was constructed with the goal of bolstering and making permanent the "Improving Access to Clinical Trials Act of 2009 (IACT)," which was due to expire come October 2015. Aside from ensuring participants receive adequate compensation, the original bill mandated that compensation received from participation not be considered a part of one's income when computing for Supplemental Security Income (SSI) and Medicaid.
http://tinyurl.com/q5rry7p

Experimental Drug Shows Dramatic Improvement Against Bacterial Infection In CF Patients

A new therapy based on engineered cationic antimicrobial peptides was shown to be more efficient than antibiotics when used against bacteria that have become drug-resistant. In this study, the research team sought to determine if a possible alternative to traditional antibiotics known as "eCAPs" (engineered cationic antimicrobial peptides) are effective in treating both gram-positive and negative bacteria as well as against several strains that are highly resistant to treatment. The research team found that eCAPs stopped growth in 87% to 91% of the samples. In particular, the bacteria will develop resistance to these new cationic antimicrobial peptides with more difficulty compared to traditional antimicrobial treatments.
http://tinyurl.com/mxph9gt

Cystic Fibrosis Drug Quinsair For Pseudomonas Aeruginosa Treatment Given Positive Opinion By CHMP

The Committee for Medicinal Products for Human Use (CHMP) issued a positive recommendation for the commercialization of Quinsair (240 mg) as a therapeutic for adult

Continued on page 37

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) 2015: Transitions – Many Types Of Changes.

Summer (August) 2015: What To Expect Post-transplant. (Submissions due June 15, 2015.) Have you had a transplant? Can you share your post-transplant experiences to help others? Help others avoid pitfalls and trouble spots. Tell us how to handle it.

Autumn (November) 2015: Incorporating Work Into Our CF Care. (Submissions due September 15, 2015.) Are you working full time? How do you make time for all your treatments and have time for work? Tell us your techniques.

Winter (February) 2016: Dealing With Gastrointestinal Issues. (Submissions due December 15, 2015.)

ASK THE ATTORNEY

Answers To Readers' Questions

By Beth Sufian, JD

In the past three months, readers have asked many questions about private insurance co-pays, Medicare coverage and Social Security benefits. A compilation of questions is below. No confidential information is shared in the questions below, and all questions were asked by two or more readers. CF Roundtable *readers with additional questions can contact the CF Legal Information Hotline at 1-800-622-0385 or CFLegal@sufianpassamano.com. The Hotline is sponsored by the CF Foundation. All contacts are free and confidential. The attorneys who run the Hotline are not employees of the CF Foundation. Nothing in this column is meant to be legal advice about your specific situation and is meant only as information.*

Q: My employer is self-insured. This means my employer pays all healthcare expenses for employees out of the company profits. Is this a problem, since I have CF and high medical costs?

A: Some employers are self-insured. Typically the employer pays healthcare expenses up to a certain amount and then has a policy often called a "Reinsurance Policy" that pays healthcare expenses over a certain amount. For example, the company will pay healthcare expenses up to $150,000. After $150,000 has been spent by the company, the Reinsurance Policy takes over and pays all healthcare expenses. The Reinsurance Policy may charge the employer more for the Reinsurance Plan in a subsequent year if some employees had high medical expenses. For example: in 2014 the employer had an employee who had $150,000 in healthcare expenses. After $150,000 was paid for expenses incurred by the employee, the Reinsurance Policy started paying claims for the employee. The Reinsurance Policy paid $300,000 in healthcare expenses for that employee in 2014. The Reinsurance Policy is allowed to charge the employer a higher price for the Reinsurance Policy in 2015. Thus if an employee has high costs in one year due to high-priced healthcare costs such as medication or a transplant, the Reinsurance Policy may cost the employer more in the following year.

Q: I have Medicare and I need to buy a Medicare Supplement policy to pay for my Medicare Part B 20 percent co-pays. However, every time I apply for a Medicare Supplement policy I am turned down. Does an insurance company have to sell me a Medicare Supplement policy?

A: The answer depends on the state in which you live. Approximately half of all states do not require an insurance company to sell a Medicare Supplement policy to a person who has Medicare, if the person is eligible for Medicare because he receives Social Security Disability Insurance benefits (SSDI). In states where there is no requirement that an insurance company sell a Medicare Supplement policy to a person who receives Medicare because he receives SSDI benefits, it is difficult (and in many states impossible) to find an insurance company that will sell such a policy. All people who are eligible for Medicare because they are over 65 are eligible to purchase a Medicare Supplement policy regardless of the state in which the person lives. An insurance company cannot deny Medicare Supplement policy coverage to a person who is eligible for Medicare because he is over the age of 65.

BETH SUFIAN

Q: Does Medicare require a person pay 20 percent of ALL medical charges billed to Medicare?

A: No. There are different parts of Medicare and each part covers different healthcare services and treatment. Each part of Medicare has a different cost-share between what Medicare pays and what the Medicare recipient pays.

Medicare Part B is the section of Medicare that provides coverage for outpatient services, such as physician office visits, blood work, x-rays etc. Medicare Part B also provides coverage for a limited number of prescription medications. Medicare Part B pays 80 percent of the allowable charges and the patient must pay the other 20 percent. If a person has a Medicare Supplement policy, that policy will typically pay the Part B 20 percent portion that Medicare does not pay. There is no cap on how much a person can pay out of pocket in terms of the 20 percent cost-share under Medicare Part B.

Most CF medications that are covered under Medicare Part B are made by drug companies that have Patient Assistance Programs that can provide assistance paying the Part B 20 percent co-pay for the medication. However, most Patient Assistance Programs have income eligibility guidelines. Those individuals whose household income is over the allowable amount will not be eligible for help from the Patient Assistance program. Each Patient Assistance Program has its own household income eligibility guidelines.

Recently, some Patient Assistance Programs have excluded help to those who have Medicare. It is unclear why some Patient Assistance Programs have excluded Medicare recipients from receiving help with the Part B 20 percent co-pay. If a Patient Assistance Program denies help to a person with CF who has Medicare, the person should ask the Patient Assistance Program for an explanation of why the program will not provide assistance.

Medicare Part A does NOT have a 20 percent co-pay for services. Medicare Part A covers charges related to a hospital stay. There is a Medicare Part A deductible, but after the deductible is paid there is no additional co-pay for hospital charges during that hospital stay. Each hospitalization requires the Part A deductible be paid. Medicare Supplement policies typically pay the Medicare Part A hospital deductible. However, some physicians or services provided in the hospital are billed separately. For example, a physician may charge separately for physician services provided while a person is hospitalized. Such charges are often billed under Part B and may have a co-pay that must be paid by the Medicare recipient. If a person is unable to pay the co-pay, a person can always ask the provider to waive the co-pay. Typically the person will have to show he/she is financially unable to pay the co-pay. The provider has discretion to waive the co-pay.

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

A: Marriage will result in the new spouse's income and assets being considered by Social Security for purposes of determining eligibility for SSI benefits. If a person loses SSI benefits then he/she will lose Medicaid coverage. In 27 states a person who meets certain low income eligibility guidelines will be able to enroll in Medicaid even if the person does not have SSI benefits. The Affordable Care Act (also known as Obamacare) provides that states can offer Medicaid to low income adults. The United States Supreme Court held that the states could not be required to provide Medicaid to low income adults. In 23 states the only way for an adult to be eligible for Medicaid is to also be eligible for and receiving SSI benefits. If a young adult with CF receives SSI benefits and is thinking about marriage, the young adult should check to see if his/her future spouse's income will make him/her ineligible for SSI benefits once the person is married. If the person will lose SSI benefits if he/she marries, he/she will also lose Medicaid coverage. The person should explore the possibility of obtaining Medicaid coverage because he/she meets certain low income guidelines in a state that has expanded Medicaid to adults who are low income. If a person is not eligible for Medicaid, the person can purchase a health insurance policy on the Healthcare Exchange (www.healthcare.gov) and possibly receive a subsidy to help pay the insurance policy premiums.

Q: Can I still enroll in the Healthcare Exchange and purchase an insurance policy?

A: Open enrollment for the Healthcare Exchange or state healthcare exchanges has closed for 2015. However, if a person has a change in certain life circumstances, the person may be eligible for special enrollment in a Healthcare Exchange policy. Special circumstances include, but are not limited to, a change in income, change in household size or a change in the state where a person lives. The person must enroll in the Healthcare Exchange within 60 days of the life change occurring. Go to www.healthcare.gov to see if you may be eligible for a special enrollment period. The information on special enrollment is easy to access and understand.

There are bills pending in Congress that could offer a special enrollment period near the time IRS tax forms are due (April 15, 2015). If you are in need of special enrollment but have not had a life change, you can check www.healthcare.gov to see if additional special enrollment periods have been added by Congress or your state.

Q: I received SSI until this month. I live with two roommates. My roommates put money in my account and then I pay the rent. Now I have lost my SSI benefit because Social Security says I had over the allowable $2,000 in assets last month. What can I do?

A: It is not a good idea for anyone on SSI to have money from room-

Continued on page 19

SPIRIT MEDICINE

Changing One's Mind

By Isabel Stenzel Byrnes

This issue's topic is "Transitions – Many Types of Changes." In the cystic fibrosis (CF) experience and in life in general, change happens. Some change is invited, such as attending college, getting married or changing jobs. Many more changes are involuntary interruptions in our lives. Most importantly, change forces us to search for inner resources. In this Spirit Medicine article, I'd like to explore how our minds and our thinking are our greatest allies to change.

For those with CF, uninvited change can permeate our lives. The need for a tune-up, a new diagnosis such as CF-related diabetes, getting laid off work after too many absences, or noticing it's harder to climb the stairs are just some examples of changes in the life of a person with CF. These changes are hard. They naturally invite reactions and responses by the person experiencing them.

Change is a form of loss. It robs us of our comfort level. It creates new demands for situations we haven't encountered and skills we haven't developed yet. Change means we give up how we see ourselves and are forced to adopt new roles and identities. All change needs to be mourned. Clearly, change causes stress. I heard recently, "Happiness is defined as the absence of stress." And stress is defined as one's reactions and responses to things that happen in our lives. Sonja Lyubomirsky, author of *The How of Happiness*, states that, "50% of our happiness is in our genes, 40% is in our attitude, and 10% is based on what happens to us." So we have around 40 percent of control for our own stress. That means how we react and respond to *change*—our attitude—can contribute to 40 percent of our happiness. Wow.

So, can we be happy with change? The other day, my father said, "Variety makes life interesting." This is the genetic component of my happiness: I, too, like a lot of controllable change in my life. I find the status quo boring and need trips, activities and social engagements to provide the spice and excitement to my life. And yet, then there is the unwanted change. Does unwanted change have to create unhappiness? Only if we label it as such.

For me, unwanted change is usually health-related. Lately, I've had a broken bone (osteoporosis), onset of chronic arrhythmia, and skin cancer—all requiring interruptions from my daily routine. At times, I've felt a surge in stress—with thoughts like, "If I can't exercise, I'm going to get weaker. If I have heart issues, I can't travel or hike safely. If I get my skin cancer removed, I'll look weird at work. How can I work with all these appointments?!" These thoughts start the cycle of downward negative thinking about my life, myself and even the irritating medical team who are imposing these inconvenient appointments on my life. My own thoughts make me feel bad.

ISABEL STENZEL BYRNES

And then I stop myself. Whoa. I remember the trilogy of Buddhism: impermanence, non-judgment, and non-attachment. Nothing stays the same. No need to grasp a whole, healthy body, because that is not possible and just adds to my suffering. I remind myself of thoughts that calm me: *In the big picture, this is nothing. I am not my body. I am spiritually on fire, no matter what happens to this body. These things happen and will keep happening, so don't react so much. Accept. Life with illness is just this.*

My recent ruminations for my medical problems are remnants of rehearsed patterns of thinking I've had since childhood. When I was younger, as my health took a turn for the worse, my usual thought was, "This is the beginning of the end." This was unhealthy catastrophic thinking, to say the least. (Of course, there were many CF peers who were facing the end with similar exacerbations so it wasn't all hyperbole.) I remember key phrases I used to say when I had a CF exacerbation. One was, "What's my problem? Why can't I get up and (blank)…?" Or I'd say, "I should've done more to stay well…" When I was on oxygen, and too sick to work, I thought of myself as "useless," unless I volunteered profusely to "contribute." These thoughts were all self-judgmental and just making things worse. My attitude was getting in my way.

Though 50 percent of our happiness is in our genes, we are not victims of our heredity. The field of epigenetics shows that our thoughts and behaviors can actually turn on genes that can then activate proteins that harm or help us. For example, feelings of love can turn off genes that suppress the immune system, or early trauma can turn on genes that predispose us to lifelong depression. The point here is that how we think about our lives impacts our health. Here's another accessible scenario: we are evolutionarily wired to have stress responses when our security and lives are threatened—this is the natural "fight, flight or freeze" impulse. When we have a chronic illness, where our security and lives are *routinely* threatened, we are at greater risk of chronic stress, which then can negatively impact our physical health. Talk about a dangerous cycle!

Dr. Amit Sood, a professor of Medicine at the Mayo Clinic, writes about chronic stress and the impact it has on our brains. He says, "Your brain's neurons fire the same way to events whether imagined or real; it cannot distinguish physical pain from emotional hurt. Simply put, thoughts can damage your brain just as surely as an unhealthy lifestyle or physical trauma." In other words thinking thoughts like, "This is the beginning of the end," starts the neural spiral of cortisol, adrenaline, anxiety, shallow breathing, rapid heart rate and slower gut, that all exacerbate the symptoms of CF disease anyway. Dr. Sood's prescription is beautifully simple: Each day, practice thoughts that include gratitude, compassion (toward self as well as others), acceptance, purpose and forgiveness.

The truth is: CF is a hard and sucky disease. If only I pointed fingers *at the CF*, and not myself. If only someone told me that I was worthwhile as a human being, even a sick one, whether I "did" something or not, because I was intrinsically loveable and being sick could even make me a valuable teacher, so that I could've internalized this wisdom at a younger age.

It is very, very difficult to change our thinking patterns. In health education, providers often use the "Readiness to Change" tool. This tool outlines the steps needed to make permanent behavioral change, such as becoming adherent to treatments or quitting smoking. The steps are: precontemplation, contemplation, preparation, action, maintenance and termination. I believe changing our minds takes all of these steps as well. We have to think about our thoughts—unconsciously, then consciously. Then we have to prepare ourselves to change unhealthy thoughts. Then do it, practice it and maintain it. It takes awareness, deliberate intention and lots of practice. We have to constantly pay attention to thoughts that make us feel bad. Living with CF gives us all long-term, insidious and unrelenting training on how to think differently about our disease so that we can integrate CF positively into our sense of self, and live better, happier lives.

So how do we change our minds? This is all really complicated and everyone has their own way. First, it takes time. If we're lucky, we'll have the time for growth and experience to help us figure this stuff out. Change of mind comes when we intentionally embrace the "serenity prayer" and accept that we have some, but not all, control. We have to stop our negative thoughts and reassess the statements we make to ourselves, asking constantly if they are true or irrational. We also have to trust in life's "seasons"—being very sick is a chapter in our lives, and that our sense of self must come not only from that chapter, but the rest of the story. We also can learn that being self-critical can be a way to avoid the really painful truth—that we have a terminal disease that can take over our lives. We have to stop overthinking and getting in our own way. And most importantly, we have to challenge any unhealthy thought habits we've developed over the years and say, "Enough is enough."

Lastly, in my job as a bereavement counselor, I work with people undergoing major change—the loss of a loved one. I am inspired by my faithful clients who, when overwhelmed, give up their fears, frustrations and anxieties to God. That is the power of surrender—knowing that we can't always control whether change happens, that we can't always contain all of our responses and reactions to change, but we can give it all up to God. Through prayer, mindful release and relinquishment, and a deep spiritual trust that "all shall be well," our spirituality can be an anchor in times of transition. We have to trust that, despite it all, change is a gift—the Lakota people say *taku skanskan*—that change creates spiritual vitality. Our spiritual energy gets stronger with movement and change. ▲

Isabel Stenzel Byrnes is 43 and has CF. She lives in Redwood City, CA, with her husband, Andrew. You may contact her at: isabear27@hotmail.com.

SPEEDING PAST 50...

Change Is The Only Constant In Life!

By Kathy Russell

Our lives are filled with transitions. Each year we live through the transitions of the seasons. In some places, those transitions are quite dramatic, while they are less so in other areas. Much of the USA is hoping for a peaceful transition from winter to spring this year. We here in the Pacific Northwest have been much more fortunate where winter weather was concerned. We had the warmest meteorological winter on record. February was the warmest February since records have been kept. Although we had a couple of cold weeks early in the season, most of our winter was quite pleasant. Spring flowers started to bloom in February. As long as we don't get a hard freeze, we should be okay. We hope the rest of the country will have as easy a transition.

That's enough about the weather. How about some life transitions? Those start at the moment of birth, when we make the transition from living in liquid to breathing air. For some, this can be a difficult transition. For most of us, the change is smooth and we learn how to breathe and how to clear our airways.

For some, clearing the airways becomes a very difficult transition. Inhalers, nebulizers and medicines become necessary and "the norm." Actually, I think that we are more fortunate when we start all of those treatments while we are young. For some people, it can be very difficult to make the transition from not having to do any meds or treatments to doing all that we have to do.

Many of my friends who have been healthy all of their lives now must learn how to deal with chronic ailments. It sounds to me as if they have a lot of trouble getting used to their "new normal." Many are taking pills for the first time in their lives. Some do whine about that. Others have to use inhalers or even oxygen because of lung disease and they really don't like it. We must try to be understanding of them and not scoff at their pain. Just because we are accustomed to taking pills by the handful and doing hours of treatments, doesn't make their pain any less real. Heaven knows that we have been given much support by our friends and families for most of our lives. Now it's time for us to transition to being supportive friends.

Let's go back to a discussion of our early years. We learned to crawl, then toddle and then walk. Each change had its own difficulties. We managed. We went from charming preschoolers to kindergartners and then "real" school kids. Again, we managed.

The next big change for me was leaving a familiar pediatrician and going to a doc who dealt mostly with adults. I think I was 15 or so when I decided that I was ready to make that change. There were no CF clinics when I was young, so I saw a private physician. I loved my pediatrician, but I realized that he still thought of me as sweet, little kid. I was tired of being in rooms that had fairy tale characters on the walls and had furniture that was sized for little children. I needed a doc who would see me as a person, not a child, and who would include me in all decisions relating to my health. Fortunately, my parents were agreeable with this change. That made it much easier.

KATHY RUSSELL

Making the transition from pediatric CF clinic to adult CF clinic can be extremely traumatic for some. I have known people who had great difficulty making this change. They or their parents (or maybe both they and their parents) were so attached to the pediatric clinic that they fought the change. They didn't want to leave a group where they felt comfortable. They wouldn't even give it a chance.

By fighting the transition from kid to young adult, normal development can be hampered. Everyone must make that change at some time. My feeling is that it is easier to make the transition from pediatrics to an adult clinic earlier rather than later. If we live long enough, each of us will be attending a clinic that is for adults. Since we will make the transition eventually, why not make it at our earliest convenience and by our choice?

The next transition in my life was going from high school to nursing school. Gone were the familiar halls of my high school and I found myself in unfamiliar classrooms and hospital settings. I was only 18-and-a-half and felt as if I had entered another world. I had to learn a new language and ask questions of people that usually weren't asked in polite society. For instance, how often would you ask people when they had their last bowel movement? I got over my uneasiness pretty quickly.

> *"My next transition was the most difficult one for me. I stopped working for pay."*

I had to adjust to being the caregiver rather than the patient. I had the experience of being a patient so I had empathy for those who were hospitalized and for their families. That whole experience can be quite traumatizing at first, and caregivers can ease the way for others with understanding.

It didn't take long for me to feel at ease in each new hospital setting in which I found myself. Every different ward had its own personality. I found that I really gravitated toward pediatrics and maternity. My first long-term job was with school-age pediatric patients. Later, I moved to the maternity ward and worked mostly in the newborn nursery. That was my favorite place to work.

My next transition was the most difficult one for me. I stopped working for pay. My docs wanted me to stop working because I kept getting sicker and sicker. I applied for Social Security Disability Insurance (SSDI). That was a years-long battle, which I lost. I still did not go back to work, because I was much healthier when I stayed home. That caused another transition.

When I stopped getting a paycheck, we had to readjust our budget and our plans. Cutting my pay meant that we no longer could do many of the things that we had been doing. We just couldn't afford to do them. We found new ways to enjoy life.

One big transition for me, when I quit work, was that I felt I had no "self-worth." It seemed that everyone measured people's worth by how much money they made. Since I made no money, I had no worth. In retrospect, I doubt that my friends measured me in that way, but I felt that was what was happening. It took me a long time to realize that money is not our only measure of worth.

I discovered that although I couldn't hold down a traditional job and maintain my good health, I could volunteer without causing me too much trouble. I volunteered with my city in crime prevention, with the juvenile court system in neighborhood accountability and with USACFA and *CF Roundtable*. None of these endeavors took too many hours of my days, and they did give me a sense of accomplishment. I stayed with city things for a few years. The accountability board lasted a couple of years. The long-term gig has turned out to be USACFA and *CF Roundtable*.

I have been volunteering with USACFA for 25 years. I was the original treasurer and set up the bookkeeping system for the organization. In the beginning, all the work was done by hand. When I got a computer, I was able to transfer the first three years of records to the computer in only a couple of days. What a nice transition that was.

I also served as president. That transition became necessary after our president and secretary died within a day of each other. That was a difficult time for all of us, but we got through it. Later I spent another couple of years as treasurer, before being able to step down once again. Since then, I have made the transition from director to just a helping hand. A very pleasant transition, I might add.

My most recent transition is being thought of as "old"! I know I'm old. My friends and family know I'm old. Getting my brain to accept that I am old has been more difficult than I imagined it might be. I was watching TV one night recently, and the reporter was talking about an "elderly woman." When the story played out, that "elderly" woman was several years younger than I! Talk about a wake-up call! When someone who is younger than you is elderly, then you are elderly! It's official.

My husband, Paul, and I recently celebrated our 50th wedding anniversary. Who would ever have thought that someone who has CF would live to be married for 50 years? No doctors ever gave me any reason to think that I might be able to do that. The interesting thing is that the doctors who were giving me such gloomy forecasts of my life did not live to be married 50 years. They died of various maladies in their 50s or 60s. So much for CF being a "death sentence."

Now that I am officially a "geezer" I will make a concerted effort to not whine about my aching back, shoulders, knees, ankles, feet, thumbs or whatever. I will try not to wheeze and rattle when talking. I will try to remember that no one wants to hear an "organ recital" of my troubles. I will try to be a "happy old geezer" and share a smile with someone who needs one.

The only thing that has been constant in my life is change…it has been, and ever will be, thus.

Stay healthy and happy. ▲

Kathy is 70 and has CF. She is the Managing Editor of CF Roundtable. Her contact information is on page 2.

COUGHING WITH A SMILE...
Get Back Up

By Jennifer Hale

Hello *CF Roundtable* readers! Hope all of you have been doing well and had a great holiday season! Today I wanted to write about a quote from ESPN announcer, Stuart Scott, who lost his battle to cancer recently and passed away. Mr. Scott said, "When you die, it does not mean that you lose to cancer. You beat cancer by *how* you live, *why* you live and *in the manner* in which you live." When I heard this quote it really resonated with me. Obviously, I substituted CF for cancer and, with that, it really had a lot of meaning for me.

CF is a tough walk, I always say, and everyone's walk is unique. CF also has no rhyme or reason to why things go downhill or why things suddenly improve. It is a mysterious and unrelenting beast that I fight daily and even more so as my health has been declining in recent years. But how true it is what Mr. Scott said about coping and living with a terminal disease. It is all about how, why and in what manner you look the devil in the eye and persevere with all the odds against you.

I think it would be easy to walk around pissed off, in a bad mood and generally being unpleasant when you are fighting for your life each and every day with each and every breath. But what good would that do? It doesn't make you feel better. It doesn't get rid of CF. All it does is poison your body, mind, spirit and those around you. And who wants that? I sure don't and I have never lived my life that way. I have always tried to find the silver lining in the midst of the dark clouds. By doing this, it makes for a more fulfilling and loving life with what life we have left.

I think having CF or any terminal disease makes it hard to keep moving forward sometimes. I feel we sometimes judge ourselves based on what we think we should be doing or feeling. Or we see others doing better and we think to ourselves, well why am I not doing well? I am taking all my meds and doing my treatments. I am being compliant. Why am I not seeing the same good results? Well, that is just it. We cannot compare.

JENNIFER HALE

We cannot compare ourselves to others or to who we once were. I am constantly comparing myself to how I was when my FEV_1 was 65% and I expect sometimes to feel that same way now, but how could I at 29%? It is just not possible and that is okay. There is a quote from John Wooden, who said, "Don't measure yourself by what you have accomplished, but by what you should have accomplished with your ability." Wow! That hits the nail on the head. To be present in your current circumstances and to expect out of yourself what you CAN do with WHAT you have going on now. For instance, I am still going to the gym to work out even though it is incredibly hard and I need O_2 in order to move these days. But the thing is, I am going. I may not be able to do it as hard as I used to or as often. But I am still going and doing the best I can with what I've got. I have accomplished a lot with my current, and I stress *current,* ability!

It all goes back to how, why and the manner in which we live with CF. You gotta keep moving forward with a smile on your face, no matter how much you want to frown. This quote came from an unknown source, "Life has knocked me down a few times… It has shown me things I never wanted to see… I have experienced sadness and failures… But one thing for sure… I ALWAYS get back up!" Get back up my fibros and cysters! Get back up!

Be an example for all those to see. Be the light that stays on even in the darkest of moments. Keep fighting your CF battle with a light heart, a smile on your face and a spark in your eye. This will feed your soul and spirit and give you strength to battle on. Lastly, I leave you with this quote by Henry David Thoreau, "Things do not change, we change."

Until next time readers! ▲

Jennifer is 43 and has CF. She and her husband, Mark, live in St. Petersburg, FL. You may reach her at: jhale@usacfa.org.

You Asked & We Answered – From The USACFA President

Dear Valued Readers of *CF Roundtable*,

On behalf of the Directors of the U.S. Adult CF Association (USACFA), the publisher of *CF Roundtable*, I want to thank you for responding to our survey in March 2014. We were delighted so many people contributed their ideas and suggestions. This helps us produce the best newsletter possible to serve the CF community. Your input allows us to bring new information and support that enhances the daily lives of adults with CF.

The survey consisted of ten questions about who you were and asked for your critiques, comments and suggestions for future issues of *CF Roundtable*. We received hundreds of completed surveys – all online via Survey Monkey – and listened to your needs. We appreciate the multitude of kind, positive remarks and constructive comments about *CF Roundtable*.

The following lists what you requested and how we responded.

First, we asked for ideas for future Focus topics. There were more than 70 responses that included parenting, mental health challenges, transplantation, dealing with grief/death, nutrition, specific medical diagnoses, career challenges, disability, financial issues and more. As a result we created Focus topics to address your requests, e.g., in the summer 2014 issue we covered "Dealing with Conditions That Are Part of CF"; in autumn, "Dealing with the Death of a Loved One with CF"; this past winter 2015, "Ways to Become a Parent When You Have CF"; and in this spring 2015 issue, "Transitions – Many Types of Changes." Future Focus topics will include many of your other requests.

We dealt with some of your top suggestions for future Focus topics a bit differently. There was so much interest in financial and parenting issues that new columns were begun to address these. Fortunately, we had two adults with CF who were well positioned to be columnists for these topics. Mark Manginelli, a financial adviser, launched his new column, "Protecting What Matters," in the spring of 2014. Karen Vega, a USACFA Director, parent extraordinaire raising three children and overseer of a parenting Facebook support group (www.cfmothers.com), stepped forward to create her new column entitled "Parenting" in the autumn 2014 issue.

JEANIE HANLEY

In response to the numerous requests about clinical studies and research, our Vice President, Meranda Honaker, launched a special committee that collects information on future clinical trials. Her compilation of studies along with their information is disseminated on our social media sites including Facebook, Twitter and the *CF Roundtable* blog. All of these sites can be accessed through our website, www.cfroundtable.com. In addition, beginning with this issue of *CF Roundtable*, Meranda is overseeing a new column entitled "Searching For The Cure" that will include a listing of research trials that are actively recruiting.

The USACFA Board of Directors, an all-volunteer group consisting of adults with CF, meets regularly, and we work to continually improve communications with you, our readers, through our newsletter *CF Roundtable*, the website, blog and social media sites such as Facebook, CF Connect and Twitter. In addition, we appreciate your letters and respond to them in as timely a manner as possible. We also want to give a shout out to all of you who have contributed articles over the years, sharing your experiences with the rest of the CF community – keep them coming!

Another annual survey is on its way, and you should be receiving it very soon by e-mail if you are subscribed online. If you'd like to receive the survey by postal mail, then please send us an e-mail or letter request. We appreciate your participation and look forward to hearing your ideas and comments.

Kindly,
Jeanie

P.S. If you would like to fill out a survey for us, e-mail: CFRoundtable@USACFA.org or write to:

USACFA
P.O. Box 68105,
Indianapolis, IN 46268-0105 ▲

FOCUS TOPIC

TRANSITIONS – MANY TYPES OF CHANGES

An Incomparable Life

By Jess Newport

Growing up with chronic illness, I was never told that I couldn't experience all that life had to offer. In high school, when I began to feel how "sick" my lungs were, it contributed to thoughts that I did not want children in my future. Life was going to be hard enough—I didn't want the extra worry of meeting "life's benchmarks." My mother always told me that I shouldn't compare my life with others, because I'm not a normal twenty-something. Internet staples like Facebook and Instagram have certainly turned how we compare our lives on its mountain-shouting head. I decided that I would not expect babies, a spouse, or buying a house; that way, I couldn't be disappointed.

I wouldn't say that I had much success in relationships until I met my current boyfriend. Ten years of relationships led me to the man I want to marry. Marriage for those of us who have CF can be very complicated. I'm a disabled adult according to my insurance and I would lose that if I married. Trying to get insurance under my future husband's work...I don't know that that's really an option. My current insurance policy covered my transplant and has worked very well for me. I finally find a husband and I cannot marry. My future may be a commitment ceremony that I will actually treat as a wedding. What matters is that our love is forever.

In the meantime, my boyfriend has moved in with me. I've never lived with a man before and I'm both excited and apprehensive. Financially, I could never afford to buy my own place, but I really like the townhouse I started renting in July. To keep my SSI (Social Security Income), I have to keep them (the Social Security Administration) abreast of my living arrangements. I can't afford to have it decreased because there's been a change in who I'm keeping house with. Honestly, the Social Security regulations were not tailored to an individual with CF who wants to live an independent life. The same federal institution that gives money to those trying to live autonomously, keeps them chained to others. It is so frustrating.

JESS NEWPORT AND JEREMY LAMBERT AT A HURRICANES HOCKEY GAME.

> "I'm a disabled adult according to my insurance and I would lose that if I married."

In September 2013, I had a hysterectomy because the HPV I contracted at some point was causing bleeding and cellular changes in my cervix. Previous treatments, including a LEEP (Loop Electrosurgical Excision Procedure), did not improve this and I was even once misdiagnosed with cervical cancer in 2012. The only answer seemed to be a hysterectomy.

I don't know whether it was the surgery, but afterward I began to feel a draw toward being a mother. I fostered two dogs, desired pets and took strongly to my friend's baby boy. I had never before desired to be a mother.

Now that I'm in such a loving relationship, I've felt great moments of sadness that I cannot give him a child. Once I'm gone, a child would be the one physical connection he would have to me. But at the same time, I do not want to leave a child parentless. I do not mean to offend, but I have always felt it is selfish to have a child, knowing the hurt they will feel when I'm gone. I attribute my maternal confusion to the loss my potential child would feel.

Despite the fact that my life is, and will be, very different from those around me, I am the happiest I've ever been. Life with CF is short. We must make it a point to fill it with many smiles and happy memories. Those small moments make for a life well-lived. ▲

Jess is 28 and has CF. She lives and crafts in Durham, NC. You may reach her at: nu6586@gmail.com

WOODWARD *continued from page 1*

called Dr. Grekos "wrong" and "deplorable," preying on families and individuals who desperately need a little hope. They have criticized the data Grekos has proclaimed to collect as being poorly designed and poorly conducted, yielding unreliable results. There has been no objective oversight to the data gathered by Regenocyte, which has refused to participate in independent regulatory programs designed to protect patients and expand research in regenerative stem cell science.

Despite all of this, Dr. Grekos maintains an almost cult-like following of patients who believe their lives have been transformed by him, even though many of these patients generally do not measure significantly better in quantifiable tests. Much of the improvement falls within the parameters of a powerful placebo effect. As Dr. Norman Edelman, chief medical officer of the American Lung Association, puts it: "I do not doubt the sincerity of patients who believe they have been helped by stem cell therapy, but there is an enormous placebo effect in almost all of these cases. I have looked for the scientific efficacy of this approach, and can find none."

The typical procedure costs upwards of $54,000 or higher. This is not covered by insurance. In the state of Florida, being a cash-only clinic provides some measure of regulatory protection from consequences of unproven claims. The patient will also need to travel to the Dominican Republic for the procedure, which is performed by Dr. Grekos's Dominican partner, as Grekos is not licensed to practice on the island. Regenocyte reports it has treated more than 200 patients, which puts its gross revenue at nearly $11 million.

Combining this with Dr. Grekos's other streams of income, which include being the owner/ operator of a popular Greek restaurant, a Greek Orthodox church, several real estate and development companies (one of which was at the center of a protected land preservation dispute), personnel and consulting firms and marina-based businesses, Grekos and his wife, who is also a part-time doctor, have made quite a comfortable living for themselves. According to the Florida Department of State, there are 18 corporate entities filed under Zannos Grekos's name.

STEPHANIE WOODWARD

Knowing this, it would seem that Dr. Grekos keeps himself very busy, which makes his seemingly impressive CV all that more incredible. Upon further inspection, one might notice that some of the distinguished honors for which he purports to take credit are either really hurried mistakes (less likely considering how important a doctor's resume is to his or her reputation) or deliberate falsifications designed to fool inattentive and uneducated readers. For example, changing a single letter in an abbreviation can mean the difference between a merit-based, peer-nominated distinction and an invented mark of respect bought and paid for with the intent to deceive.

Dr. Grekos has shrugged off accusations that he is practicing unethically, charging instead that he is cutting edge and helping people who can't afford to wait for the government to catch up with his methods. However, he has done little to provide evidence independent from patient testimonials that could hasten research efforts to progress treatments for the public. Ironically, he accuses the FDA of conspiring to line the pockets of "Big Pharma" with new patents.

You see, when a doctor truly believes she or he has made a breakthrough discovery in medicine, she or he doesn't incorporate a for-profit private business to proprietarily treat patients for the accumulation of their life savings account. They carefully, meticulously put it through trials and record every data point, positive or negative, and submit it for review and replication by other research scientists and to the FDA for approval. They want to share that information far and wide to help as many people as possible, not keep it to themselves as a business opportunity. There's nothing wrong with making money, but without the scientific method, there can be no self-correction or progress made in actual treatment options. Zannos Grekos is a gunslinger taking advantage of an unregulated market and the mystery and awe surrounding public perception of stem cell technology. ▲

Please read a more detailed account of Dr. Grekos and Regenocyte's stem cell promise at my website, http://abreathofreason.com. A Breath of Reason was started in 2014 to provide evidence-backed rebuttals to dubious health claims related to cystic fibrosis. I'm a 30-year-old mother, wife, artist, and nature lover, living with CF. I am not a scientist, but am well educated in many areas of health science, logic and source verification. You can find my Facebook page at: facebook.com/abreathofreason.

365 Days—The Road From Ruin

By Klyn Elsbury

I stayed in my bedroom for an entire day, sobbing uncontrollably and afraid of the world outside. At 26 years of age, I just hit send on an e-mail single-handedly destroying my career as a biotech pharmaceutical associate director. I had sold my house in Orlando, Florida, and moved to San Diego, California, for this opportunity. I hoped to take my new city by storm and make a life here where people could see my success, read my story, and think, "One day, I want to be like her."

It was a career I was more than proud of, until lung infections and declining PFTs took over. The medical bills quickly outpaced the salary. Several times in my new city, the decision to go out with friends or go out for groceries had to be made. But my love for my new career was worth the sacrifice. That hunger of chasing the sale and succeeding was more than my hunger for date night, for friends or family, for myself. I wanted (at the cost of my health) to surpass quota and prove that I was somebody that made it in this life, despite cystic fibrosis.

A couple of months into the chase, one hospitalization turned into two…then three. My PFTs plummeted from 87% to 52%. Weeks on antibiotics were stretched out as the doctors tried new intravenous combinations. When the doctors said I could go back to work, I lost personal balance and attempted to make up for the time in the hospital by putting in extra hours.

It was eight months into my career, 15 pounds down and 40 percent less lung function, when a social worker brought up going on disability. Going on disability, to me, meant that I was a complete failure, I was cowardly scum, I (frankly) didn't deserve to live. It meant I couldn't succeed in the real world so I took an easy way out. It meant my parents should never have spent all that money to keep me alive as a child and make sacrifices for my success, if I couldn't even survive as an adult. I was worthless.

KLYN, LEFT, TEACHING AT A ZUMBA MASTER'S EVENT. 90 MINUTES OF EXPERIENCING A VARIETY OF RHYTHMS INCLUDING SALSA, MERENGUE, REGGAETON AND CUMBIA.

I hesitantly started to research, contacting an old childhood friend who is a very successful and big-hearted attorney specializing in disability services for cystic fibrosis patients. We chatted about my options and as I hung up the phone after talking with her, I realized it was time to hang up my career.

On a conference call the next day, I was called out for not having enough heart, not wanting to succeed. I cried until I coughed. And the cough turned into blood. And I called the hospital, which was the only place I knew as home in California. I sent my resignation e-mail a few days later.

My lung function continued to plummet. I had no insurance now, so I put off care until the paperwork went through. I had no income. I had no family within 1,500 miles. I flew home and when my parents rented a wheelchair for me because I couldn't keep up walking around Six Flags, I knew no amount of money was worth risking more of a decline and checked myself into the local hospital with an FEV_1 in the 30s. I needed oxygen at night and thought to myself, "This is truly the end." This is how I will pass. And with the exception of family and a few friends scattered around the world, nobody will really care.

I left the hospital a few weeks later, deciding to check things off my bucket list in the amount of time I had left. It was important to me to visit friends back in the Midwest, so I spent six weeks driving through farmland, couch-surfing and laughing. When I got back to my parents' house, I looked at my list and saw one word that terrified but excited me. Dance. My mom and I went to the local gym, and I decided to hack my way through an hour of Zumba.

I went to the back of the studio, kept my eyes to the floor, my coughing tissues next to my water bottle on my immediate left. That first song, the cough started. People stared. My face was flushed with embarrassment. As the cough continued, the people next to me moved away. The instructor, noticing the embarrassment, turned the music up to mask the sound of the plague coming from the back of the room. I made it only half an hour until I started crying and left.

I went back. And back again. On the third attempt, the instructor grabbed my hand and pulled me to the front. I knew the routine by then and I led part of the class, hack and all. And for a moment, full body chills enveloped my body and soul, and I knew…this was where I was meant to be. Front of a studio, dancing my heart out, helping other people forget their struggles to an eight-count I created. I said to my family, "I'm going to become a Zumba instructor."

I got back to California, committed to Zumba several times a week. To be strong enough to get through the hour-long intense cardio class, I started lifting weights. Over time, I noticed people would stand behind me during the class to watch my moves. I started making friends. When I wasn't dancing, I fell back on my sales roots by fundraising for CF, and was the top fundraiser for 2014. I became heavily involved with Cystic Fibrosis Lifestyle Foundation and was asked to use my recruiting talent as the Chairwoman of Board Development.

Even though I was still on IV antibiotics every six weeks, my lung function started to improve. The day I went for my Zumba license, I was actually on an IV. I told the lead instructor, every three hours I needed to sit in the back of the dance studio and infuse my medication. I got some odd looks, but then I would hit the floor afterward and people replaced the smirks with smiles. I left the studio eight hours later, with my license and my new mission.

I have since taught at Petco Park, in a Master's class and in several boutique gyms throughout San Diego. This week, I was offered an opportunity to lead a Corporate Wellness Program on Coronado Island…with the main client being a biotech company. My lung function has stayed stable for two months, long enough for me to go back to school and get a certificate through National Academy of Sports Medicine in Personal Training.

I found out a few days ago I was nominated for and won the award "Women Who Take Our Breath Away" through the Cystic Fibrosis Foundation. I have fellow instructors with whom I regularly get together to dance, and a few of my students have requested me on Facebook. Most important, my parents sent me a bouquet of roses, with the simple message, "We're proud of you."

I once read that ruin is the road to transformation. After my dream career failed, I felt like I had nothing. But the reality is, I had everything. I had a chance of a lifetime…a chance to start over. I've learned to focus less on monetary gains and more about loving and helping others. I learned to listen to my body and look at my treatment time not as a negative thing I have to do because I have cystic fibrosis, but as time to reflect on my journey and feel gratitude that the medications are helping my body perform feats I never thought would be possible. I may not know what my future holds or how long my body will cooperate with me, but I do know that I will continue to crank up the music when I dance. ▲

Klyn is 27 and has CF. She is a Director of USACFA. Her contact information is on page 2.

YOU CANNOT FAIL

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

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

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

My Journey Back To Independence

By Lisa Cissell

My first taste of independence began when I left for college in the early '80s. I had not yet been diagnosed with CF and my symptoms mostly mimicked seasonal allergies. As I was the oldest of seven children, my parents didn't have much time to hover over me while I was at college; I was pretty much sent on my way. With the help of grants and part-time jobs, I financed nearly all of my college expenses.

Upon graduation, I wasn't able to land a full-time job immediately out of college and lived at home for a brief time. Being back home was frustrating for me and my parents. So, a few months after being offered a job with the USDA, I moved to the small town of Columbia, Kentucky, 60 miles south of my home.

About one year into my career in agriculture, I was diagnosed with CF, at the age of 25. The news was a shock as I pretty much exhibited no symptoms. I basically tried to carry on as usual with my job and social life. I moved a year after my diagnosis to another small town, Monticello, in southern Kentucky. Two years later I was promoted and moved back to central Kentucky, closer to my hometown. In all of these relocations I was single and established myself in these new places, making friends and memories along the way. Being on my own allowed me to choose my own path and get out of my comfort zones in order to fully enjoy my life.

Around the time I turned 40, my health started to decline, slowly at first and then speeding up after I got past 45. My lung function dropped, I had more hospitalizations and working a full-time job was becoming very demanding. At 47, I was evaluated for a lung transplant at Barnes-Jewish Hospital in St. Louis, Missouri, and was told I wasn't quite ready yet, but that I would be in the near future. To say that the time following that information was difficult would be an understatement. I went on oxygen 24/7, my lung function dropped to the low 20s and my lungs just seemed to stay full of mucous.

LISA CISSELL AND HER ICELANDIC HORSE LOGI IN DECEMBER 2014.

I became a hermit and felt like a prisoner in my own home. I was able to work some from home, but those days I had to go to the office completely wore me out. Even with all that physical difficulty, one of the hardest things for me to have to do was ask people to help me. I had someone come clean my house, family members (especially my brother-in-law, Mike) would go get my groceries, and my aunts and other relatives would cook me food. For me, it was so hard to let others do the chores I had done for myself over the years. I knew it was not my fault, but I felt embarrassed and helpless.

After living this way for a year, I was re-evaluated early in 2011 and was told by my transplant team that it was time to be listed. My friends, Paul and Kristi, had graciously offered to let me live with them near the hospital while I waited. Within a few weeks, my family helped me to pack and take care of my affairs at home. Then my sisters, Shannon and Renee, moved me five hours west. Being able to take my dog, Sherman, with me helped, but it was very hard to see my sisters leave.

Paul and Kristi were wonderful hosts, making sure I had what I needed to be comfortable. Just as Sherman and I were getting settled in, we all were shocked when I received a 3:30 AM call. Only eight days after my move, there were lungs available for me.

April 16, 2011, is mostly a blur for me; I do remember being in a pre-op room for a long time. My surgery began around 2 PM, and the next thing I remember is waking up late the next day in the ICU at Barnes-Jewish. By all accounts, my surgery was a success, and for the first several days I surprised the doctors by how well I was doing. That was short lived however, when it was discovered that I had a thoracic duct leak. Three more surgeries later, along with the discovery that I actually had two thoracic ducts, and the leak finally was stopped. This scenario made what should have been seven days in the hospital blossom into a month.

Over those 30 days, I became very dependent on and accustomed to the continuous attention from the hospital staff. Morning and night, there were only tiny spaces of time when someone was not in my room. Much of the time I spent in the hospital, I felt pretty miserable, and the presence of people, both staff and loved ones, was very comforting to me. I felt more secure and reassured that I was going to be okay when others were with me.

Exactly one month from the date of my transplant, I was released from the hospital. It definitely was a day of mixed emotions for me. I was elated to be finally leaving the hospital but also terrified to be back out into the world without the constant medical care I had been receiving.

My sister Renee was in town at that time to stay with me at the extended-stay hotel my family had reserved for us during my next two months of rehab. Despite her encouraging presence, I was very frightened to be out of the hospital and worried that something would go wrong in my recovery. I remember us watching a Harry Potter movie that first night to help distract me with something I enjoyed.

After a few days at the hotel, Renee had to go back to Kentucky to her family, and my dad and his sister arrived. So began a rotation of caretakers I had in town that we called the "changing of the guard." Starting with Renee's departure, I went through cycles of despair each time someone had to leave. I also did not ever want to be alone, mainly, I believe for fear that something terrible would happen to me.

Those two months of recovery in St. Louis, accompanied by visits from family and friends, sometimes seemed to go by slowly. But with each week and new visitors, I was going to rehab and getting out and seeing the sights. I gradually started to lose the anxious feelings and believed that I might be okay.

Exactly three months after my surgery, I moved back to Kentucky. It was a bit scary to leave the security of my home-away-from-home and the excellent medical care, but it was wonderful to be back to my home, bed and pets. Two months after returning home, I went back to work part-time. Then, two months later, I was working full-time and continue to this day.

The exclamation point to my returned liberation and health came six months to the day after my surgery, when my sister Shannon and I attended a concert by my favorite band, Duran Duran, in Washington, DC. It is still surreal to think about to this day and it began my new life of experiencing joy in every day with friends, laughter, traveling, my career and regaining my lost independence. ▲

Lisa is 52 and has CF. She is a Director of USACFA. Her contact information is on page 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

Delayne Santos
Gulfport, FL
48 on February14, 2015

Wedding

Susie Baldwin and **Adam Levy**
Los Angeles, CA
18 years on
October 13, 2014

Transplant

Susie Baldwin, 47
Los Angeles, CA
2 years on Dec 21, 2014
Bilateral lung transplant

Adjusting To A Work Environment

By Andrea Eisenman

While some transitions can seem scary, the majority are exciting and just represent a challenge to our status quo and, once conquered, become rewarding. Starting a job in the real world, 9-5, was all of the above. I was eager to start working in the field I was trained for—graphic design. Many had told me it would be nothing like the creativity I experienced in college and I would have to start at the bottom, ruling boards for layouts for the art directors (this was before computers were available), following designs from superiors or just doing paste-up mechanicals. I was ready to start my financially independent life and dive in.

I was under no illusions I would be designing much. And I started as a freelancer for a furniture catalogue doing paste-up mechanicals. It was fun and freelance meant I was not needed every day and could come in at 10:00 AM. I had days off, not necessarily when I wanted, but it was not five days a week unless the catalogue was going to press. It was a relaxed environment with cool, artsy types. It was my first job post-college. This lasted three to four months. Once the catalogue was done, I needed to get a full-time job. With freelance, I would never receive health insurance. I need that plus the other benefits of working at a company for sick leave and a steady salary.

I went on many interviews and wondered if I should tell my future employers that I had cystic fibrosis (CF) or not. I decided not to as I wasn't so sick. I only did IVs once or twice a year on breaks from school.

My first job at a monthly magazine started relatively well except that after a few months of work, I had to do IV antibiotics for pneumonia. I did two weeks on the IVs. About five to seven days were spent in the hospital and then the remainder I brought my meds with me to work. It was hard to go to work and then fit in two antibiotics, one every eight hours and one every six. I got very little sleep and, as we all know, that is really important when being on those IV drugs.

It was difficult to work with an IV drip going in. Sometimes my insurance paid for those vacuum-packed balls and sometimes I had to get bags to hang. But if I was infusing, I had to plan to sit at my desk for that period of time. This was not always easy, as I didn't have that kind of job. I had to go to the (photo) stat room, run to the mailroom for my art director and see the copy editor. It was awkward because I also didn't know my co-workers very well before I got so sick.

I felt that they resented me when I had to be in the hospital and then when I was at work, I was not as "there" as I was normally. So, it meant my co-workers had to work harder. This happened two more times about a month apart. I just kept getting pneumonia and having to do IVs. The third time was bad and I was in the hospital the whole two weeks. This was when my parents were told I only had six months to live. They told me nothing but, somehow, I had the feeling that I was doomed. I felt I might die if I didn't adjust the way I was living. I went home with an oxygen concentrator to sleep with and a new vision of how to manage my job and life. Or I would have neither.

It was not like I had been out every night partying. But there was a night or two that my friends who worked in the city went out after work. I had to stop joining them. Not only that, I had to come home after work and take a nap. Then eat and try to exercise and then get ready to sleep for the night so I could get up well-rested. The supplemental oxygen did help me stay more oxygenated at night. I woke up feeling better.

> "I went home with an oxygen concentrator to sleep with and a new vision of how to manage my job and life. Or I would have neither."

Each task I had to do, I weighed the effort against the reward. Everything exhausted me and I was only 23. This was not how I envisioned my working life. I saved my laundry to do on the weekends and eventually paid a laundromat to do it for me. It took too much energy. With shopping, I broke down and got a shopping cart to lug groceries home. My friends thought this was hysterical and made fun of me. In the end, what mattered was, I could go to work every day, get paid and get medical benefits.

This schedule went on for a while, probably almost six months. I was limited in my time to see friends or go out much. But with getting more rest and prioritizing my health over fun, I finally adjusted to working full-time and was able to start socializing more. I still had to do IV antibiotics for exacerbations, but they happened less frequently, about twice a year.

But what I didn't expect after returning to work was my managing editor taking me aside to have a serious talk. I had just come back from my third hospitalization, my worst, and he asked if "this" was going to keep happening. I was not sure what to say. I said I had hoped not but had little control over it. He told me how hard it was for my co-workers and that if this kept up, my absences, they would have to reconsider my job. I was upset but also knew on some level it was illegal to threaten me about my job when it had to do with my health. It was discriminatory. Even though this was before I knew Beth Sufian or had ever heard of *CF Roundtable*, I knew there were laws in place to keep people like me from being fired. But it certainly scared me senseless. Who wants to go through a lawsuit to keep their job? So, aside from trying my hardest to get well, I was stressed about people at work resenting me plus possibly getting fired.

Eventually, I started to feel better and was able to keep up with my workload. I was promoted and given more responsibilities. I started to design some layouts and hire photographers and illustrators for pieces in the magazine. I was promoted again and then was trained to use a computer for layout of the magazine electronically. This led me to teaching other art departments in the company to use the computer for layout. I finally moved to a different, more challenging part of the company, promotions, before leaving for greener pastures.

While my beginning at this company was uneven and scary, I feel like it was a wake-up call about what having CF and working can feel like. I didn't realize working full-time would be so hard on my health and that CF would be harder to manage. I was lucky that I was able to turn it around and continue working while I regained my health.

My PFTs never really recovered, but I guess I learned to manage what little I had (low 40s) in order to remain working. My doctor who thought I was going to die after having pneumonia three times in a row never understood how I did it. I want to say it was perseverance, determination to succeed and wanting to live my life to the fullest. It was also learning to be compliant and listening to my body. Before this, I might have taken my health for granted, as I surely did in college, pushing myself, staying up way too late. It was a valuable lesson that I wish I had learned earlier.

Working for 13 years in publishing allowed me to realize my creative potential while keeping my health benefits as I was going into my next transition, waiting for and receiving a bilateral lung transplant. This then led me to my next transition: retirement from full-time employment. And then, life as a married person. And now, into the AARP-set. Never stop growing and transforming, it is what keeps us learning to adapt and fighting to stay alive. Keep on keepin' on! ▲

Andrea is 50 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.*

SUFIAN *continued from page 5*

mates coming in and out of a bank account to pay rent. If the amount in the account goes over the allowable amount, then the SSI benefits should terminate. It will not matter that some of the money was provided by roommates to pay rent. A recipient of SSI benefits must meet the income/asset eligibility criteria set by the Social Security Administration (SSA). An SSI recipient can only have $2,000 in assets, if the person is single. SSA does not know what money in an account is for and can consider any money in a bank account to be an asset of the SSI recipient. Any money in a bank account (even money in an account for one day) may put the person on SSI over the SSA monthly asset allowable amount for a single person.

A person who receives SSI benefits is allowed to own one car and one house. The value of one car and one house will NOT be counted toward the SSI asset limit. Please see the front page article in the autumn 2014 issue of *CF Roundtable*, which discusses Special Needs Trusts that may allow an SSI recipient to have additional assets over the SSI asset limit.

A person has the right to appeal a termination of Social Security benefits or a person can file a new application for SSI benefits 60 days after the time to file an appeal has passed. ▲

Beth is 48 and has CF. She is an attorney who specializes in disability law and she is a Director of USACFA. Her contact information is on page 2. You may contact her with your legal questions about CF-related issues.

FOCUS TOPIC

TRANSITIONS – MANY TYPES OF CHANGES

November

By Laura Mentch

Halloween was my last day of work. Everything in order for the new Health Educator arriving on Monday, I remove my nametag, hand in my key, swipe my time card and close the door behind me. With apprehension I leave the familiar and cross the threshold to an uncertain place.

My keys felt lighter and it seemed so dark with the time change. Through the months contemplating then readying to leave this job after 20 years, I had not prepared myself for this transition. Suddenly it was November with a long winter before me. I recalled Art Wilmot's words and found the clipping in my cookbook:

November is the transition period between fall and winter, the month between a burst of beauty and stark barrenness. As a transition month it is dangerous to the psyche—keeping us potentially tied to the past, or living in dread of the future.

Transition periods are always difficult and dangerous. Yet they are the true times of adventure in life. If life is fundamentally an adventure for the human creature, and I believe it is, then I cast my lot, not with fall or winter, but with November. Therein lie Life and Death, Glory and Tragedy, Love and Pain. This is what it means to be Human.

I hadn't considered leaving my work life before a certain age, when my friends would also retire. But, I hadn't planned to live with CF. My diagnosis came late, years after creating a career and family. My CF care was sandwiched in with existing responsibilities. Many days I did not do this very well.

So, I left work to better care for myself and give cystic fibrosis focused attention: step up to my treatments, exercise, rest when needed and "get out" in fresh air and enjoy this beautiful place where I live.

Many, eager to fill the perceived void in my professional life, offered suggestions for things I could do next; most were unaware of the reason I left

LAURA MENTCH

my job. What was I going to do? they asked. I wondered as well and hoped for opportunities to stay involved with my craft. My wise friend told me, "Be patient, it will come to you."

Again it is November. Now I regularly go to the gym, a 10-minute walk from home. This is a big deal. For seven years I also taught Human Sexuality at the university. My office was in the gym, but I never used it; too tired, too busy or unsure. Working with a personal trainer, I've gained confidence. Caring for myself is easier. When I had an exacerbation in July, my biggest concern was finding dog care. Not feeling pressure about work during a tune-up has been a huge relief, and no meetings urge me to leave earlier than the doctors would like. I started practicing restorative yoga with the afternoon IV. My doctor says my lungs sound better than they have in our 11+ years of CF care. My nurse tells me I don't look so tired.

Blessedly, opportunities have come. "Sexuality and CF" was the title of my talk with CF care providers last May, and I have been invited by a CF center to speak with hospital nurses. Bringing the conversation about sexuality to the CF community is rewarding, fun and a way to give back. I've become a trainer for *Our Whole Lives*, helping others prepare to facilitate this comprehensive sexuality education curriculum. This year we have engaged 12 young people in our community with this program. More time has supported more active board work with AIDS Outreach, The Cody Dieruf Benefit Foundation for Cystic Fibrosis and United States Adult Cystic Fibrosis Association.

My extended family celebrated Thanksgiving together. My father would have been 100 on this day and we toasted him with gratitude. This November I reflect on the past year, appreciating those who gently, persistently and directly encouraged me to step away from my daily work life. The new balance in my days feels good. In the year ahead maybe I will learn to rest. ▲

Laura is 61 and has CF. She is a Director of USACFA. Her contact information is on page 2.

THROUGH THE LOOKING GLASS

PHOTO BY ROB ROHDE

Lucky 7

The fifteen of us gather
Yet only seven are seen
Seven double lung transplant recipients
One, twice.
Graced with another chapter after cystic fibrosis
The lucky seven have survived.
It's a gamble, this chance to live
The seven of us have beaten the odds
Because someone else didn't
Standing behind us are the shadows
Of eight saints who gave us breath
And with this breath
We are free, really free!
Free to be friends, lovers, artists, athletes… adults
We are free to laugh, love, to take chances
We are free to face each other
There is no shame, only intimacy
With this breath
We see everything
The hope, the gratitude, the amazement
The fear, the sorrow, the yearning
Our joy above all
And with this breath
We won courage
To come to the edge and roll the dice
And to know we need more courage
For the next hand
The seven of us make a promise to each other
And to the eight saints
I'll stay in the game if you stay in the game.
Please.

-I. Stenzel Byrnes, 2009

"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.
To learn more about us and view more images in the collection, please visit our website at:
http://www.thebreathingroom.org

FROM OUR FAMILY PHOTO ALBUM...

MEGAN MURRAY, THE GLOWING BRIDE, IN 2010.

JANINE ULLYETTE AND HER BOYFRIEND, ERIC, TAKE A TWIRL IN HAWAII.

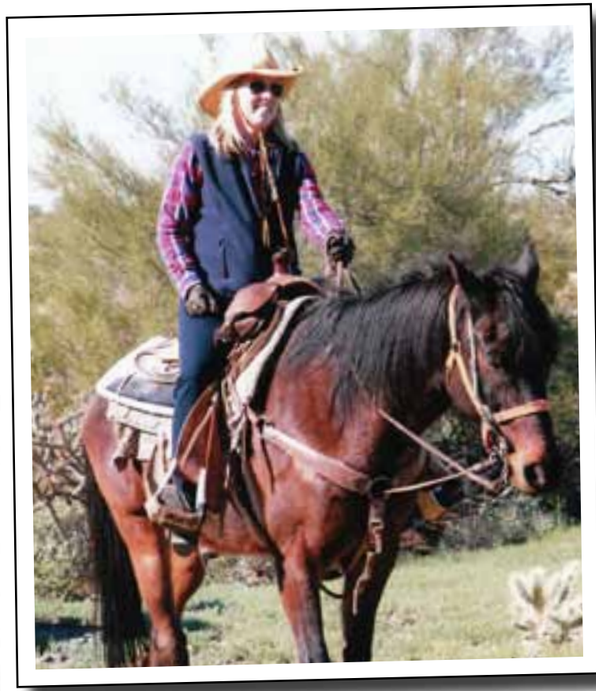
JESS NEWPORT WITH THE PADDLE SHE USED IN THE 1ST ANNUAL RALEIGH DRAGONBOAT FESTIVAL WHERE HER TEAM PLACED 2ND.

ALISON LYNCH AND HER HUSBAND, GEORGE, SHARE A WEDDING TOAST.

ANDREA EISENMAN AND HER HUSBAND, STEVE DOWNEY.

WESTON AND STEPHANIE WOODWARD ON MUSTACHE NIGHT WATCHING THE DENVER CUTTHROATS, A SEMI-PRO HOCKEY GAME.

MERANDA HONAKER.



LISA CISSELL ON A HORSE AT A RANCH IN TUCSON, AZ, IN FEBRUARY 2015.

IN THE SPOTLIGHT
With Janine Ullyette

By Andrea Eisenman and Jeanie Hanley

Meeting Janine Ullyette, formally, for the first time for lunch, she is so vivacious and exudes so much energy, she seems 30 years younger than her 59 years. I (Andrea) had been receiving her e-mails about her singing events for some time and bumped into her at a screening in Manhattan of *The Power of Two*. She was with a CF doctor whom I knew. It was good to finally put a face with a name. As I knew she was a singer and seemed so outgoing, I took her number to see if she would be interested in being featured for this column. The more I found out about her in our "pre-interview" lunch, the more I liked and found intriguing. She wears a flower in her hair to remind her of her favorite place, outside of New York, Hawaii. She is happily in a long-term relationship with the love of her life, who, it turns out, had some surprises of his own to offer. She seems to take life as it comes and makes the most of it. She is a talented singer who has benefitted from increasing her lung function doing what she loves—singing. This vocation has allowed her to travel the world and share her gift. While she is now slowing down with work, her passion is still clear, living life to its fullest and keeping Hawaii in site. Please welcome our newest star. Spotlight please.

Your age? 59 (60 this coming July and proud of it)

Age you were diagnosed with CF and why:

I was diagnosed when I was 13. First, my sister Laurie was diagnosed as a result of having been sent to an allergy specialist because of nasal polyps. It was the allergist who suspected she had CF. She then was sent to a CF specialist in Rochester, NY (Dr. Robert H. Schwartz), at Strong Memorial Hospital to have a sweat test. She was positive and because CF is genetic, Dr. Schwartz had my other

JANINE ULLYETTE

two sisters as well as me tested for CF. Sally and I were positive, but Susie was either a carrier or negative. Susie never chose to find out which, as she never wanted children.

How are your sisters now?

Two are deceased. Susie died in 2007 from uterine cancer. She was 58 years old. Sally died in 1996 at the age of 42 from complications after bilateral lung transplant due to CF. Laurie and I are doing fairly well, all things considered having CF, although, we are having more and more issues.

Where do you live?

I live in Brooklyn, NY. I'm on the border of Brooklyn Heights and Boerum Hill.

What is your relationship status?

I am in a very committed relationship with a wonderful man, Eric. We have been together for almost 15 years although we have never lived in the same state. He is originally from Massachusetts and for the past eight years, he's been living near Hilo, Hawaii.

How did you meet your boyfriend?

I was asked to sing in the chorus of an opera with the Berkshire Opera Company. The first day that the women of the chorus arrived, one of them said she needed a spool of black thread to fix her zipper. A handsome man came over with one. My colleague went off and fixed her zipper. When she came back, she said she couldn't remember who gave her the spool of black thread. I said, "Oh I do and there he is." That was Eric, who was filling in for the regular theater manager.

Neither of us was looking for a new relationship with anyone. Both Eric and I had been married before and I, at that time, was in a relationship with an older Italian man whom I was trying to dump, but he wouldn't accept that I wasn't in love with him. He loved to hear me sing, so he came to the performance. I had spent two and a half weeks in Massachusetts and had what I thought was going to be a relationship just for that period of time with Eric. I was giving Eric a hug and kiss goodbye, as I thought I wouldn't be seeing him after that. My Italian friend witnessed this and Eric thought I was a horrible person to treat my soon-to-be-ex-beau this way.

Eric didn't understand that I was trying to dump this man. I didn't know that he (my Italian friend) was coming down the stairs behind me.

Eric still wasn't sure about me, but was coming to NYC to work on a human rights issue. Instead of staying with the people with whom he was working, he began to stay with me. By the end of that year, we were in love and it's been that way ever since.

How did you feel about Eric being diagnosed with CF? How did he find out?

After falling in love with each other, we started talking about possibly having a child together. I had never wanted to have children until I met the true love of my life, Eric. He was so wonderful with children. I had told Eric everything about myself on our very first date, including having CF. So I said, "We know that I have physical problems, let's make sure that you don't." He went to a doctor in Pittsfield, Massachusetts, and after six months of testing to find out what was "wrong" with him, he was sent to a specialist in Boston. That doctor looked over all his test results and said that he had CF. He said, "Oh no, not another stupid doctor. No, it's my girlfriend who has CF."

I didn't have any brothers so I was unaware of CBAVD (congenital bilateral absence of the vas deferens). This was what led the doctor to believe Eric had CF.

Eric called me from the doctor's office and asked if I was sitting down. He told me that he had CF. Dead silence on my end. He said, "Janine, Janine, are you there?" I said, "I don't know whether to laugh or cry." It was just so unbelievable. I had noticed all along that he often had to cough and spit, but he was 40 years old at that time. I couldn't believe that he wouldn't have been diagnosed by then, so I never said anything. This diagnosis changed his life and not always for the better. The best thing that happened was the bond between us.

Has Eric having CF brought you two closer?

Eric is so incredibly caring when it comes to my health. Because we both have CF and I've known since I was 13, I've been able to inform him through first-hand knowledge. Eric never spoke with me about his digestive issues before he knew he had CF. Once he was diagnosed, I was then able to help him make sense of what had been going on during his childhood.

I know that people with CF aren't supposed to be near each other, but obviously, in our case, that's impossible. The same goes for my sister. I'm not going to stay three to six feet from her or Eric. Eric is always there for me even though he lives in Hawaii. Our love of Hawaii and our Hawaiian connection is a huge bond just like our bond with CF. Whether in NYC or Hawaii or even on the phone, we are so in touch with each other's needs. I couldn't ask for a better partner in life.

What was your CF like in childhood? What is it like now?

As a child, I didn't really understand what was going on. I just thought everything I felt was normal. I had awful stomach pains. As a child in school, whenever I had to go to the bathroom, I would wait so that no one else was in there. Or, if anyone came in, I'd wait for them to leave. I was embarrassed. The other kids would make nasty remarks because the bathroom smelled so bad.

As I got a little older, I noticed how I had to clear my throat often. All my life, all I ever wanted to do was sing. The abundance of mucus was a real problem. Because of the mucus, I decided to pursue a professional choral singing career. If I had to clear my throat, at least it wouldn't be so noticeable.

Now, as an adult, I still go through similar feelings, but I'm more accepting and do my best to prevent both the digestive and coughing issues. I'm having more CF-related problems, such as liver and needing to use oxygen when I exercise and while I sleep. Since being sick in early January, I have had to use oxygen whenever I'm walking outside as well.

What do you or did you do for work?

I've always been singing. I actually get paid for singing in a church choir. I've also sung with major world orchestras when a professional choir is hired. I've sung at Lincoln Center as well as Carnegie Hall. I've toured all over the U.S., Far East and the Scandinavian countries.

After high school and my one year of college, I worked as an *au pair* in Paris, France. While I was there, I became the soprano soloist at the American Church in Paris. Other than singing, I worked for the Council on International Educational Exchange assisting foreign students with temporary housing while visiting NYC. Later, my ex-husband and I had a real estate business in Brooklyn called Janine Realty. After that, I worked for the Legal Aid Society, Juvenile Rights Division's Special Litigation Unit. I was always singing throughout these jobs.

Are you retired?

I guess you could say I am semi-retired. I intend to keep singing as long as I am able. I am collecting SSDI, so I am not allowed to earn as much money as I used to through my singing. But then, I'm also not physically as able to do as much singing as I used to.

Where do you perform?

Continued on page 26

ULLYETTE *continued from page 25*

In addition to what I said above, I love to sing musical theater songs and standards. I like to go to piano bars to sing. Occasionally, I've done cabaret shows here in NYC.

Have you seen any benefits to CF from singing?

I absolutely feel that singing has helped my CF lungs. Breathing deeply is great exercise plus the vibration from singing loosens the mucus. When I was first diagnosed, the doctors couldn't get over my terrific PFTs. At 13, I said, "Well, I'm a singer," very proudly. The doctors said to keep singing because it would probably help me. I believe they were right. As I sing less and less, I am noticing that I have more problems with my breathing.

How often do symptoms or medication side-effects get in the way of your singing?

In my earlier years of singing, I had to clear my throat a lot. When I started using hypertonic saline, it made all the difference in the world. I wasn't constantly clearing my throat. My colleagues noticed the difference, too.

In more recent years, I've had more problems with my breathing and the ability to hold long notes or sing long lines. I do miss the days when I could sing without taking lots of catch breaths. Sometimes various medications make me hoarse and lose my voice. Years ago, I was prescribed TOBI, but I lost my voice entirely. I told the doctor I would not take anything on a long-term basis that would leave me without my voice. If I can't sing, I'd rather be dead. Whenever a new drug is prescribed, the first thing I ask is, "Will it leave me hoarse or cause me to lose my voice?"

What kind of organizations do you do volunteer work for and why?

I've sung for various events in memory of friends and family as well as for residents in nursing homes. But, the main volunteer work I have done was in memory of my sister, Sally, who died from CF. After she passed, I was trying to think of something I could do in her memory. The only thing I knew how to do was sing. For ten years, I produced "An Evening of Music" with the help of many of my very talented friends. The show featured a variety of styles of music including: classical, standards, musical theater, jazz and gospel. They were amazing performances. Although they were small events, during those ten years, we brought in over $50,000 to help find a cure for CF. Of that, I am very proud.

How do you keep in shape?

I was in a pulmonary rehab program for ten weeks, three times a week. I wanted to do this to help my singing and to become healthier. It certainly made a difference. I should have been exercising at least five days a week, but that just wasn't happening. I try my best to exercise at least three days a week. My singing is a form of exercise for my lungs as well. I'm very fortunate that there is a small fitness room in the building in which I live and it is free to all tenants. I really have no excuse not to exercise seeing how it is so convenient. But I do have a life and I have lots of paperwork that is so very time consuming. A personal trainer comes a couple of times each month, so I work with her, too. My boyfriend created an intense six-minute workout for me, which I was doing every day until just before Christmas when I got very busy singing. In early January, I got really sick with flu and pneumonia and am still recuperating. I'm very weak and tired, so it's hard to exercise at the level I was doing.

What do you do for fun?

Have lunch, dinner and/or drinks with friends. I love to watch period films with one of my best friends. Going to piano bars to listen to others as well as sing for them, although I'm not doing this as much as I used to. Go to Hawaii and visit my friends there, especially my boyfriend, Eric. I also dance hula.

What do you see yourself doing in five years?

I hope I'm still able to sing. I'd like to learn the Hawaiian language so I can sing Hawaiian songs. I also love dancing hula and hope to continue. The original intention was to spend more time in Hawaii with my boyfriend. Unfortunately, my health has deteriorated to a point where living full-time in Hawaii is out of the question. I need to be near my doctors here in NYC. I still love Hawaii and want to be able to spend time there as well as NYC, but I'm not sure if I can travel and stay for a long period of time in Hawaii.

Do you feel a responsibility to be a role model for younger people who have CF?

Absolutely. I always love to speak with parents of children with CF and young people with CF. Being 59 years old, I can give them hope for a longer life despite CF. When I tell them that I am a professional singer, they are amazed. Hope. That's what it's all about.

What is your attraction to Hawaii?

I love that my mother lived and taught in Hilo right after WWII. I feel like a part of her is with me whenever I am there. The house where she lived is still there as well as the school where she taught. I love the Hawaiian music and the feeling of "ohana," which means "family," including an extended family. I love the spirit of aloha.

Do you know your genotype for CF? Are you excited about new medications targeting CF mutations?

F508del and E60X. It is absolutely exciting to hear of the new meds targeting CF mutations. I'm so happy for those people who are being helped with them. I hope one day that my two mutations are targeted.

Do you see a cure for CF in your lifetime?

That's hard to say for sure, but I believe it is possible. I certainly hope so for the sake of the younger people with CF.

Do you have a favorite saying or quote?

Live each day as if it were your last. Tomorrow may never come. ▲

Janine is 59 and has CF. She lives in Brooklyn, NY. She can be contacted by e-mail at: janine_ullyette1@verizon.net

Andrea Eisenman is 50 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 52 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.

In Memory

Fr. Anthony Cassese, 66
Cleveland, OH
Died on December 30, 2014

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

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

SUSTAINING PARTNERS
DONATE $5,000 OR MORE A YEAR

abbvie
Supported by a grant from AbbVie
AbbVie Pharmaceuticals – It Starts with Science and Ends with A New Way Forward
www.abbvie.com

Boomer Esiason Foundation
Esiason.org

CYSTIC FIBROSIS SERVICES
a Walgreens Alliance Pharmacy
CF Services Pharmacy
www.CFServicesPharmacy.com

FOUNDATION CARE Your Full Service Cystic Fibrosis Pharmacy
Foundation Care Pharmacy
www.foundcare.com

GILEAD
Advancing Therapeutics. Improving Lives.
Gilead Sciences
http://gilead.com/

Hill-Rom
Enhancing Outcomes for Patients and Their Caregivers.™
Hill-Rom
www.hill-rom.com/usa/

PROTECTING WHAT MATTERS

Preparedness During Financial Transitions

By Mark Manginelli

"Times of transition are strenuous, but I love them. They are an opportunity to purge, rethink priorities, and be intentional about new habits. We can make our new normal any way we want." Kristin Armstrong (born August 11, 1973) is a professional road bicycle racer and two-time Olympic gold medalist, the winner of the women's individual time trial in 2008 and 2012. In 2001, at the age of 27, Kristin was diagnosed with osteoarthritis in both hips, ending her triathlon career and leading her to focus exclusively on cycling. While this diagnosis could have meant the end of the road for most people, Kristin fought through the adversities and transitioned into her new athletic life with refinement. She didn't prove to the world only that she could overcome the unfortunate odds that were stacked against her in an insanely competitive athletic field; she proved to herself that with a positive mental attitude as her foundation, she needed to rely on only the strategies she put into place throughout her career to overcome any obstacle, whether it be the good, the bad, the ugly and anything else in between.

These types of stories can be read all over social media today. We get to hear about a heroic person's journey as they go through an incredible transition and come out on the other side with a renewed sense of self and life experience. What helps these people get through these seemingly impossible feats has a lot to do with their preparedness in the event that something unfortunate can ultimately take place, and they must ensure they've already considered action steps to make these transitions more manageable.

The same mentality and preparedness is necessary when you consider your financial transitions as well. Getting married, starting a family, buying your first home, retiring can all be celebratory transitions but obviously require some money management. On the other hand, there are the life transitions we don't look forward to such as divorce or death of a loved one. Clearly, financial management is complicated enough. When you undergo a personal transition, the complexities grow. Rational long-term thinking is often replaced with confusion, emotional turbulence and inconsistent behavior, which can lead to regrettable financial decisions. Even in the absence of a life transition, most people find it challenging to apply their money effectively toward the meaningful and lasting lifestyle they would like to enjoy.

When it comes to financial preparedness before ANY of these financial transitions take place (good or bad), it is crucially important to ensure your financial strategies are deployed and up to date. There are too many families today that continue to put off important financial planning conversations. If you don't have a will drafted, get one done. It doesn't just stop there once you do, however. Once you have your will completed, revisit your estate plan at least every five years regardless of how complicated it may or may not be; laws change, relationships change, beneficiaries die or become estranged etc.

MARK MANGINELLI

After the conversation about estate documents is complete, the "preparedness" conversation should shift to your risk management strategies and ensuring your financial foundation is as solid as it can be (a.k.a., insurances). Nobody likes to buy insurance but most people wouldn't even consider driving their vehicle without auto insurance just like you wouldn't buy or rent a property without homeowner's/renter's insurance.

After personally experiencing an unfortunate transition, witnessing my family and friends lose everything in the aftermath of Hurricane Sandy, homeowner's and flood insurances are no-brainers for us. We're still transitioning but thankfully my family was prepared. So many others weren't and it's difficult to watch. Their transitions became infinitely more difficult after realizing their insurances didn't cover certain "acts of God" or that their flood insurance didn't insure any of the contents that were inside their home that just got flooded with 10 feet of water.

The conversation regarding risk management shouldn't stop there even though it usually does. Two imperative conversations I have with all of my clients involve the transitions of death or disability. Many people know and understand they need life insurance, but many people still don't ever get it or don't get enough. I have the comfort knowing that if I were to leave this earth unexpectedly, my family will not have to worry about any financial concerns after my passing. My life insurance will not only pay for my funeral expenses, but I have a strategy in place to ensure my life insurance will be used to replace my income for a number of years. I never want my wife to worry about having to change her lifestyle while she goes through her own difficult transition.

Just as important, if not more important, is a conversation that many people haven't had and that involves disability insurance. We are all much more likely to become disabled than we are to die. If one of those unfortunate transitions were to happen to you and your family and you found yourself unable to perform the duties of your current occupation, how will you continue to live your current lifestyle? How long will your savings last or how long will family members help to pay your monthly obligations? When I ask my prospective clients what they think their most valuable asset is, I will typically get, "Well my house, of course." Most people don't understand that their ability to earn an income is often their largest and greatest asset.

I pose a simple scenario, "Imagine you had a machine in your basement that printed cash all day long. Would you be willing to pay a couple pennies for every dollar it makes to ensure that, in case it breaks down, stops working or even suffers a serious jam that keeps it down for a few years, you will still get paid until it gets fixed?" When I challenge the way people think about it, people often appreciate it just a little bit more.

This analogy resonated with me when I first heard it, and it changed the way I perceived disability insurance for myself and my clients alike. I look at these two elements with such a different perspective than most people because I see them not only as important financial instruments that I can utilize to protect my client's future goals and desires, I also have a unique vantage point of having a chronic illness and not being eligible to purchase most of these products even if I wanted them.

Many of the people reading this article are either in the same boat or have a direct relationship with someone in a similar situation, be it cystic fibrosis or a long list of other chronic or terminal illnesses. Thankfully, through employer benefits and many companies' group coverages, many people can obtain life and disability insurance regardless of any pre-existing condition. I have the peace of mind that if something were to happen to me and I couldn't work for an extended period of time, my income will continue to come in.

In closing, transitions are inevitable. We will all go through important life-changing transitions many times throughout our lives. The more prepared we are for the widest array of circumstances, the better off we will turn out when we look back at it through our rear view mirror. If and when our preparations are tested and we face a transition period in our lives, we need to be cognizant of how we react and respond to them. If we can maintain a positive outlook along the way, even during the most grueling experience, we know we can get through anything.

I'm a firm believer in the power of adversity and the competitive advantages we have over "normal" people. As Kristin Armstrong said, "We can make our new normal any way we want." I take it for granted from time to time, but I feel bad for the people who never truly get to appreciate their good days because they've never really had to deal with those really, really "bad" days. I hope you all have a healthy, sunny and allergy-free spring! ▲

Mark is 28 and has CF. He lives in Edison, NJ. You may contact him at:mmanginelli@usacfa.org.

Mailbox

Please accept this donation in memory of my son, Douglas Peter Riley, age 51, who died on February 5, 2014. As a family we were comforted and appreciated the information you provided. Continue your good works to aid others.

Judith Riley
Brooklyn, CT

Please accept our donation to USACFA. We truly appreciate everything you do for the CF community.

Norman Young
Falls Church, VA

PARENTING

Managing Expectations As A Mom With Cystic Fibrosis

By Megan Murray

The morning my daughter entered this world, my life changed instantly and beautifully. I know it's a cliché statement, but it's true. I suddenly had this precious miracle of a life in my arms and I knew I would do anything and everything in my power to protect her. I knew I wanted to give her the very best life possible, and I knew I wanted her to experience the world in a safe yet exciting way.

I started dreaming of the adventures we would go on, the homemade meals that would grace our immaculate dining table and the hours we would spend playing and learning together. I knew I would breastfeed until at least a year and make her baby food from scratch; I would never use television as a babysitter and each night I would make sure the house was clean and calm for the next day. I would be the mom that our culture holds up on a pedestal; the one who can do it all and still have time to bake pies and decorate the house with Pinterest ideas. This perfect dream filled my head and I hoped desperately to achieve it. And then reality hit.

MEGAN MURRAY WITH HER HUSBAND, DAVID, AND DAUGHTER, SYDNEY.

It began the very first night of her life, to be honest. Due to being born at 36 weeks through a long and difficult birth, my daughter spent her first nine days in the neonatal intensive care unit (NICU). Her exhaustion from birth kept her from being able to nurse. A physician's assistant on my daughter's care team informed me this was due to my cystic fibrosis-related diabetes and it could take weeks for her to gather the strength to breastfeed. I worked on pumping as much as I could, but it was obvious she needed formula. I felt that I had failed her already. It took months of hard work to get her to nurse correctly, and right about the time we finally figured it out, we discovered my daughter had a dairy allergy. This meant giving up cheeses, butter and milk myself to be able to continue nursing. In that crushing moment, I knew our nursing journey was over, because I simply could not afford to lose the calories those foods provided. I had to come to terms with the fact that I could not uphold my goal to nurse her a full year, and I found myself furious with my disease for making me give up that goal.

In my mind, it wasn't just breastfeeding that my disease had taken away from me and my daughter. I also found myself too exhausted to make homemade baby food when jarred food was so much easier. I was a stay-at-home mom who was going through the drive-through or eating frozen meals on a daily basis because dinner was an impossible feat while taking care of an infant and doing my daily treatment requirement. I was the mom who resorted to turning on the television so that my daughter could focus on something else while I nebbed and vested. And my house was a disaster.

At some point, around seven or eight months of being a mom, I had a meltdown. I realized that I was not even close to being the mom I hoped I would be. I cried and blamed my CF because instead I was the type of mom I *never* wanted to be. I didn't have it all together, and I wasn't giving my daughter the type of childhood I had naively wished I could. CF had taken many expectations I held for myself and utterly crushed them. It had robbed me of the goals I felt were so desperately important in those early days, and I hated this disease so much for doing that to me and my daughter.

Over the next few days, the months of anger and shame were replaced by the slow realization that it wasn't so much that CF had taken these things away from me, but rather that I needed to manage my expectations and reexamine my priorities as a mom. There is no such thing as the perfect mom and the perfect childhood (even for healthy parents this goal is totally unrealistic). Obviously, I knew that deep down, but I had to force myself to come to terms with that fact during the first year of my daughter's life. I had to shift what mattered in my mind to focus on what I could actually control and what was important in the long run.

Slowly, I found myself realizing that the things I thought were so critically important in the early days of her life were actually rather trivial. The only fact important enough to focus on, I realized, was that I had a healthy daughter, and I was staying healthy myself. Yes, my daughter drank formula, but I maintained a healthy weight because of it. Yes, my daughter ate jarred baby food, but I was able to nap when she napped because of that choice. Yes, my daughter watched (and still watches) more than her fair share of television, but I used it as a strategic tool so that I could get my treatments done and be healthy for her. To be honest, we still eat too much fast food, but we're working on it. As for the clean house, some days it is pretty tidy and others it is a disaster because I have felt miserable all day long. Is this the ideal world that I imagined giving to my daughter? No, of course not. But it is my reality of making parenthood work while having cystic fibrosis.

For all of the things I have failed to be able to give my daughter because of my disease, and for the many hundreds more I will fail to be able to give her as she grows, I am now focusing on what truly counts. Because of my CF, I have been given the luxury of understanding how precious time is, and I am fully aware of that fact every single day. With each hug and kiss my toddler gives me, I hug and kiss her back with matching intensity. I understand that tomorrow is never a given, and so I take the time to write down the moments that make my heart melt like when my daughter comes up and touches my face so gently just to examine it. I enjoy the beautiful times when she spends an hour picking up leaves outside and generously giving each one to me as an unconditional gift. My heart melts when she fake coughs during my own coughing fits so that she can be like me and thus she makes me feel less alone. I write these moments down, and I cherish them because CF has taught me to do that.

As my priorities have shifted, I know that my daughter already has and certainly will continue to miss out on numerous play dates and activities because I don't feel well or I don't have the energy to do them. But I decide to focus instead on the fact that she has my attention all day, every day. If it weren't for my CF, I would most likely be working alongside my husband for 40 hours a week and she would be in daycare where I wouldn't get to share all the special moments that occur each day. Instead, I'm a stay-at-home mom who is able to read countless books to her, to do art projects, and to take slow walks around the neighborhood as she discovers the vast world in front of her.

The struggle to manage my expectations of the life I want my daughter to have and the life I can actually give her is a daily battle. There are days when I do better at it than others. The days when I have the energy to take her places and I feel well enough to run around the yard chasing after her while she laughs happily, I know I'm creating a beautiful and memorable childhood for her. On the days when all I can do is lie on the floor or couch while she begs me to chase her up the stairs or push her around in an empty box but all I have the energy to do is just to stay awake, I wonder at the many ways I am probably failing her.

And yet, I try to focus on the fact that each day I have loved my little girl unconditionally. I have given her more kisses, hugs and encouragement than many mothers are able to because I'm blessed with time to be with her, day in and day out. I have cherished and recorded hundreds of memories, because I am aware of how short our time in this world can be. I consistently provide my daughter with a safe, happy and loving environment for her to flourish in and I am proud of that fact. Even though all of those other unrealistic expectations would have been nice to achieve, I know that each day I give her what is really important. I give her my love and, no matter how hard the day is, CF can't take that away.

As each day turns to night, I remind myself of this fact, and I know that my priorities have changed. I understand CF keeps me from being that unobtainable "perfect" mom and yet I know I'm doing okay. I know that by taking the time to focus on me and my health, I am in turn giving her the best chance to have me around in her life for a long time. That, to me, is much more important than the other expectations I once held for me and my family. Managing my expectations as a mom has allowed me to get in one to two hours of treatment time a day while not making me feel guilty for taking care of myself. I hope, by staying as healthy as I can, my daughter will have countless happy memories of her childhood. At the end of the day, that matters far more to me than being what our culture deems the perfect mother, and I take comfort in my new perspective that allows me to feel proud of the mom I have become. ▲

Megan is 27 and has CF. She lives in Minooka, IL.

If you are interested in writing for "Parenting," you can contact Karen Vega at: kvega@usacfa.org

SEARCHING FOR THE CURE
Updates In CF Research

By Meranda Honaker

"Searching for the Cure" is a new and exciting column in *CF Roundtable* that will have an emphasis on clinical research trials. Thanks to scientific advancements, we are watching history unfold as CF research progresses faster than ever! Moreover, CF is at the forefront of the development and implementation of personalized medicine. Our goal is to provide our readers with updates on cutting-edge research, breakthrough therapies and information on actively enrolling clinical trials to bridge the gap between patients and researchers. Information on clinical trials can also be found under the "Clinical Trials" section of our website.

The CFF Drug Development Pipeline currently lists 13 medications in development for CF. Additionally, clinicaltrials.gov has over 100 studies listed for cystic fibrosis, which includes drug studies and observational studies. Patient participation in clinical trials is critical to the advancements made in CF research. Every CF research study is in need of patient participation, and without our participation, research is significantly slowed. Consequently, this delays implementation of improved CF therapies and new medications going to market. CF research will advance faster by improving patient participation in research studies. The first step in understanding the clinical trials process is knowing its phases, which are described below.

Phases of Clinical Trials: Clinical trials are conducted in phases designed to answer specific research questions. Below is a simple breakdown of the phases of clinical trials.

Phase I: Researchers study a new drug in a small group of patients to test for safety and identify possible side- effects.

MERANDA HONAKER

Phase II: The drug is studied in a larger than phase I population of patients with the targeted condition, to further test safety and efficacy.

Phase III: The drug is studied in a larger than phase II group of patients to confirm efficacy, continue to monitor side effects and compare it with equivalent or standard treatments, while continuing to collect information allowing the investigational therapy to be used safely in the targeted patient population.

Phase IV: Studies conducted after the drug or treatment post-marketing to continue gathering information about the efficacy and side effects associated with long-term use. *Source: National Institutes of Health*

I am 32 years old and have participated in CF research for nearly two decades. For the last 18 months I have been enrolled in a phase III drug study with two medications targeting my genetic mutations on a cellular level. I am living proof that CF research not only improves the quantity of CF lives but also quality of life. Medications such as Cayston, TOBI, Pulmozyme, hypertonic saline and, most recently, Kalydeco were once studied in clinical trials. Now these medications have improved the lives of thousands of people who have CF, including mine. "Clinical Trials: A Vital Part of Cystic Fibrosis Treatment Advancements," which appeared in *CF Roundtable* Winter 2014, was the catalyst for this column. I am confident that the research being conducted today will prove to be valuable in the future of all who have CF.

Updates in CF Research and Clinical Trials

Visit http://www.cfroundtable.com/announcements/clinical-trials/ for a comprehensive list of currently enrolling CF studies.

Ataluren: Phase III Study of Ataluren in "Nonsense" Cystic Fibrosis Mutations
www.actcf.com
Trial Sites: North America, South America, Europe, Australia http://ClinicalTrials.gov/show/NCT02139306

Hypertonic Saline: A Phase I Study of Lung Clearance After Hypertonic Saline Delivery Using the tPAD (transnasal Pulmonary Aerosol Delivery) Device
http://ClinicalTrials.gov/show/NCT02141191

Phase I Study of N91115 in Patients With Cystic Fibrosis Homozygous F508del-CFTR Mutation
http://ClinicalTrials.gov/show/NCT02275936

Vitamin D for Enhancing the Immune System in Cystic Fibrosis (Phase III DISC Study)
http://ClinicalTrials.gov/show/NCT01426256

Vertex: A Randomized, Double-Blind, Placebo-Controlled, Parallel-Group Phase III Study to Evaluate the Efficacy and Safety of VX-661 in Combination With Ivacaftor in CF Patients Homozygous F508del
http://ClinicalTrials.gov/show/NCT02347657

Source: Clinicaltrials.gov ▲

Meranda is 32 and has CF. She is a Director of USACFA and is the Vice President. If you are a CF clinic and would like to contact her regarding listing a CF study on the USACFA website, e-mail her at: MHonaker@usacfa.org

Cystic Fibrosis Mothers

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

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

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

Pay It Forward

Again, our readers continue to amaze us. Our annual fund-raising campaign continues on —beyond our expectations! We are overwhelmed by your generosity and support of our efforts to make *CF Roundtable* available for free. We thank you for your continuing endorsement of our endeavor.

Below is a list of those who continue to "pay it forward":

BRONZE

Colleen Adamson
Michelle Allen
Shirley Althaus (In memory of daughter, Janice Kessinger, and son, Stewart Kessinger)
Tanya Cunningham (In memory of Pat Hannegan)
Ed Fleischman
Charles Hawkins
James Henry
Douglas Hornick
Paul Feld
Joanne Jacoby
Bonnie Larner-Langer
Gay Lazur
Mark Levine
Adi Loebl
Kim Nunnari
Jim & Carol O'Brien (In memory of son, Ken O'Brien)
Ben & Donna Olsen
Mary Pasquesi
Judith Riley (In memory of son, Douglas Peter Riley)
Stephanie Rath (In honor of Beth Sufian for all her hard work helping those of us with CF)
Stephen Scheu
Michael Schnitzer
Sheila Schnitzer
Reid Seilheimer Family Trust
Laura Tillman
Alice Todd (In honor of daughter, Cheri DeWilde)
Alex True
Norman Young, Jr.

SILVER

Sean Sanford (In honor of Lisa Marino)
Karen Scott
Robert Neville

GOLD

Jeanie Hanley (In memory of Theresa Lopez-Ponaman-Boujie)

PLATINUM

Nancy Wech (In loving memory of Lauren Melissa Kelly)

TRANSPLANT TALK

The Power Of The Almighty Nissen Fundoplication

By Alison Lynch

Initially I heard about the Nissen fundoplication from a fellow CFer and transplant recipient and I knew right away that I wanted this surgery. However, when I was first tested for acid reflux, five years after my bilateral lung transplant, I was informed that I was not a candidate because my pH score was too low.

On June 16, 2014, I celebrated the tenth anniversary of my bilateral lung transplant. I continued to have intense reflux episodes, but I brushed them off because I was living a dream. I was teaching high school Spanish and was now the department head. I was about to celebrate the first anniversary of my marriage to a wonderful man, and I was on my way to compete for the fourth time at the U.S. Transplant Games.

Acid reflux had become a tolerable nuisance. I went for my usual visit to my transplant pulmonologist where I had an x-ray, a pulmonary function test, and blood work. My PFTs showed a slight decline and I knew there was a possibility that something might be wrong, but it was only a few days before I had to leave for the Transplant Games. I was not going to bail out now because of a mere hiccup in my lung function. I was off to the Games and ready to compete.

My first event was the 5K. I woke up not feeling so great and couldn't run the whole race, but whatever, I placed eighth and I vowed to do better next time. It was over 100 degrees in Houston, Texas, and the next day was my favorite part of the Transplant Games, swimming! I swam in five events and medaled in all of them. However, every time my hands hit the wall I gasped for breath like never before. I was so winded and weak, in fact, that I needed help getting out of the pool after each race. Luckily, there was a robust, friendly Texan man with a big sombrero waiting for me at the end of every race and, without prompting, he noticed me struggle and graciously lifted me out of the pool.

By the third race, reality hit me. Something was terribly wrong. I knew as I walked away from the pool that my donor lungs had been discovered by my immune system. I knew even before I went home and back to see my pulmonologist that I was "rejecting." No one needed to tell me this, my body could sense it. Upon arriving home from the Games I had a bronch with biopsies and, sure enough, bad news followed, but it was much worse than I expected. It was a double whammy.

ALISON WITH FIVE SWIMMING MEDALS FROM THE 2014 TRANSPLANT GAMES IN HOUSTON, TEXAS.

On July 16, 2014, ten years and one month after my lung transplant, I was diagnosed with two forms of rejection, acute and chronic. The acute was no big deal; in fact I had already beaten acute ten months post-surgery. They gave me the same treatment—three big bags of IV Solumedrol and then a steroid taper. But as the days went by and my body swelled from all the "ROIDS" being pumped into me, instead of feeling better, I just started feeling worse.

Chronic rejection is no joke and on top of not breathing well, a deep depression came over me. I desperately shrugged it off. Although my lung function had gone from the mid-90s to only 70 percent, I had to remind myself that I could still breathe, I could still walk and I could still function, and I had a life to live, too. I had already planned to resign from my job as a Spanish teacher, because I had been accepted into a master's program in Educational Technology and I was scheduled to begin in August. I was excited for my next adventure and nothing, not even chronic rejection and shortness of breath, would hold me back.

However, I was not prepared for what happened next and there was no way to predict how I would be able to handle it. The dreaded heartburn suddenly began to surge out of my stomach and it was erupting like lava coming out of a volcano and I could feel it splashing on my "donor lungs." It was enveloping me like a raging sea with no mercy. All the medicine for chronic rejection was making the symptoms worse. I couldn't sleep, I couldn't eat and the heartburn was just following me everywhere. Even long walks around the block at 3 AM did not help. My stomach, chest and donor lungs were in a blaze. I was in trouble and the pain just kept getting more and more severe. It was so unbearable that I even had thoughts of suicide.

In the meantime, something very unexpected happened. I found my donor family while doing a search online and within a few days I was in contact with the husband and mother of my donor. It was emotional and also stressful. How could I tell them that I wasn't well? I wanted to be able to show them my gratitude, to meet them and hug them. But it wasn't a good time for me. I was struggling to get well again. Thoughts of life before transplant resurfaced. I thought about how weak I was, how short of breath I felt, how every moment I struggled to breathe. I feared going back to what life was like before my bilateral lung transplant.

> *"I knew as I walked away from the pool that my donor lungs had been discovered by my immune system."*

Meanwhile, my doctor decided it was time for the next medical intervention, and I was asked to report to the hospital for RATG, also known as Rabbit Antibodies or thymoglobulin. I was told that this treatment, given every day, over four to six hours, would completely wipe out my T-cells and hopefully slow down the chronic rejection. It seemed to work, but then my lung function dropped more. It was time for another endoscopy and to have a small pill with a camera inserted into my esophagus. It is called the Bravo test. This test revealed a triple whammy! I now officially had severe reflux disease! It was also determined with the endoscopy and a manometry study that not only was my pH level triple what it should be for a normal person, but that I also had Barrett's esophagus, a disease that makes your esophagus look like your intestines and can lead to cancer. In addition, as if that weren't enough, it was also discovered that I had a hiatal hernia. Finally, it was clinically determined that I was now officially a candidate for the Nissen fundoplication, but it would take several months before the procedure could be scheduled.

On January 8, 2015, I received the Nissen fundoplication, also known simply as, "the stomach wrap." The surgery was performed laparoscopically and consisted of five small incisions. I am very impatient and I was up and walking within a few hours after the surgery. However, the bloating was very intense. I did not feel much pain, but instead I felt a tremendous amount of discomfort. My belly was bloated with gas that was not moving at all. It was as if I had swallowed a watermelon.

Since I have trouble sitting still, I decided to return to work in five days, but I was not able to eat. I could only drink liquids and this would continue for eight solid weeks. My body could only handle mushy things and even the slightest bulky item, such as a pill, was not able to pass through the newly tightened wrap around my stomach.

However, the reflux, which had been plaguing me for years, was now finally subsiding and within a week or two after the surgery, it disappeared and my stomach took on a new mission — growling and gurgling. I was now producing uncontrollable gas. The grumbling and gas roared like thunder inside my newly formed stomach. I had to warn anyone sitting near me that my stomach had undergone a transformation and was now very verbally hostile toward everyone and everything that came within a few inches of it. It was difficult and stressful not to be able to eat.

Thankfully my husband had already purchased the NutriBullet and he was able to mix up any concoction that I wanted in a jiffy. Furthermore, it was wintertime and soups were plentiful. I even began to make my own soups, and when I was tired of cooking, nutritional shakes and baby food came in handy. Probably the most frustrating thing about the surgery was not being able to swallow pills comfortably. Fortunately, I was given a 30-day supply of all my transplant meds in liquid form. The only liquid I found unbearable to drink was the prednisone; I strongly recommend crushing that pill up and mixing it with pudding or baby food.

I am happy that while on a liquid diet I was able to lose a few pounds. The purpose of the Nissen fundoplication is not just to cure acid reflux but to also slow the progress of chronic rejection in transplant recipients. My transplant surgeon explained to me that about 30% of bilateral lung transplant recipients get chronic rejection due to acid reflux. However, this determination is based on recent data that was not available when I first had my transplant.

The hardest part about recovering from the fundoplication procedure is not the actual surgery but the fact that you have to completely change not only what you eat but also how you eat. As someone with cystic fibrosis, I was accustomed to overeating and with my newly wrapped stomach, this is just not possible. In fact, it was even

Continued on page 36

Voices from the Roundtable

Concern For CF Patients

Michael Boyle, M.D.
Professor of Medicine, Johns Hopkins School of Medicine
Director, Johns Hopkins Adult CF Program
Jerry Nick, M.D.
Professor of Medicine, National Jewish Health
Director, National Jewish Health Adult CF Program
Daniel Weiss, M.D., Ph.D.
Professor of Medicine, University of Vermont College of Medicine
Chairman of Stem Cell Working Group for the American Thoracic Society
Moira Aitken, M.D.
Professor of Medicine, University of Washington
Director, University of Washington Adult CF Program
Patrick Flume, M.D.
Professor of Medicine and Pediatrics, Medical University of South Carolina
Director, MUSC Adult CF Program

We are writing as long-time members of the CF medical and research community because we are increasingly concerned about the absence of accurate information surrounding "stem cell treatments" for CF. While we are always open to new approaches that might improve the health and longevity of individuals living with CF, we have serious safety and ethical concerns about these "stem cell treatments." Each of us has been asked recently by our own patients about commercial companies that are offering such treatments and requiring large amounts of self-pay from patients based on claims that these interventions are clinically beneficial to people with CF.

We want to make the CF community aware that these claims have never been substantiated in the medical, research or regulatory fields, and the companies prescribing these "treatments" have avoided any of the standard available methods to assure safety of CF patients and scientifically evaluate if the "treatments" work. These procedures, often requiring travel to foreign countries to avoid safety and clinical trial regulations of the United States, do not meet the safety standards required of FDA-approved therapies or safe and well-designed clinical trials.

The possible risks associated with these procedures include rapid worsening of lung disease, development of serious complications including stroke and death, and lack of proper patient follow-up should any medical complications arise. Because of the potential harm and the lack of medically proven benefit, we strongly caution people with CF about participating in stem cell therapy treatments outside of FDA-approved clinical trials.

We encourage individuals with CF and their families to discuss the potential risks and benefits of all CF treatments with a physician and care team with specialized CF training.

For those who would like to learn more about stem cells and the potential use of stem cells for treating lung diseases like CF, reliable information can be found at:

The International Society for Stem Cell Research (ISSCR) patient handbook:
http://www.isscr.org/home/publications/patient-handbook
The National Institutes of Health website:
http://stemcells.nih.gov/info/pages/health.aspx
The International Society for Cellular Therapy website:
https://www.cirm.ca.gov/our-progress/concerns-about-stem-cell-tourism
The California Institute for Regenerative Medicine website:
http://stemcellfoundation.ca/en/toward-treatments/treatment-abroad ▲

LYNCH *continued from page 35*

difficult to hydrate initially because even water goes down slower. The Nissen fundoplication requires that you take small bites, chew your food really well and never overdo. Never eat beyond the feeling of fullness and be careful when food gets stuck on the way down. When food does get stuck, do not panic, you are not having a heart attack, although it may feel like you are. Instead, relax and drink a nice warm broth to get whatever is stuck to pass. If you are preparing for the surgery, make sure you stock up on protein shakes because you don't want to lose all the nutritional benefits you normally get from food while on a liquid diet.

I am happy to report that a month after receiving the stomach wrap, I had no decline in lung function. My stomach still makes loud protests whenever I eat, but I am no longer experiencing painful bouts of reflux and the great hope is that my "donor lungs" will no longer suffer from acid erosion. Perhaps in time my "donor lungs" may even improve. ▲

Alison Lynch is 43 and has CF. She lives in NYC with her husband. She is 10 years post bilateral-lung transplant. You may contact her at: spanishclasses@aol.com

TILLMAN *continued from page 3*

patients with cystic fibrosis with recurrent infections by Pseudomonas aeruginosa. Quinsair has levofloxacin, a broad spectrum antibiotic from the fluoroquinolone antibacterial drug class, as an active substance. Specifically, the drug acts by inhibiting important bacteria enzymes for DNA replication, the bacterial DNA gyrase and topoisomerase IV enzymes. The drug improves the respiratory function of CF patients, as indicated by data from three clinical trials, Phase 2 (MPEX 204) and two Phase 3 studies (MPEX 207-209). The most commonly registered adverse side effects included cough/productive cough, dysgeusia (a distortion of the sense of taste) and fatigue/asthenia (weakness).
http://tinyurl.com/mwzn4cn

Celtaxsys Lead Candidate Granted Orphan Designation in Cystic Fibrosis by FDA

Celtaxsys, Inc., announced that the U.S. Food and Drug Administration (FDA) has granted orphan drug designation (ODD) to its once daily, oral leukotriene A4 hydrolase inhibitor (CTX-4430) to treat the inflammatory component of cystic fibrosis.
http://tinyurl.com/kh82s47

Proteostasis Therapeutics Announces a New Class of Agents for Cystic Fibrosis Called CFTR Amplifiers and Selects PTI130 as a Development Candidate

Proteostasis Therapeutics, Inc. (PTI), a company developing novel therapeutics to treat diseases of protein folding, trafficking and clearance, unveiled a new class of agents, CFTR AMPLIFIERS, for the treatment of cystic fibrosis (CF). CFTR amplifiers represent a new drug class able to enhance the effect of known cystic fibrosis transmembrane conductance regulator (CFTR) modulating agents, such as potentiators and correctors. The amplifiers are effective across CFTR mutation classes and form the basis for Proteostasis's strategy to develop a broad acting combination therapy able to serve CF patients with most mutations. Proteostasis Therapeutics also announced that it has nominated PTI130 as a clinical development candidate for the treatment of CF. PTI130, an amplifier, was found to have excellent pharmacologic properties amenable for oral dosing.
http://tinyurl.com/kxb5ey3
AND
http://tinyurl.com/nfypuea

Galapagos initiates first Phase 1 study in cystic fibrosis and will receive milestone payment from AbbVie

Galapagos NV announces the initiation of the first Phase 1 study with GLPG1837. This novel potentiator is designed as a CFTR targeted therapy for cystic fibrosis patients who carry class III/IV mutations (e.g., G551D). In combination with corrector GLPG2222, this potentiator will also be developed for patients affected by the F508del mutation, the most prevalent mutation in CF patients. The aim of the Phase 1 study is to evaluate the safety, tolerability and pharmacokinetics of oral single and multiple ascending doses of GLPG1837.
http://tinyurl.com/kcmw47l

Advanced Inhalation Therapies Receives Orphan Drug Designation of its Proprietary High Dose Formulation of Nitric Oxide for Adjunctive Treatment of Cystic Fibrosis

Advanced Inhalation Therapies Ltd (AIT) announced that the U.S. Food and Drug Administration (FDA) granted Orphan Drug Designation to AIT-CF, the company's proprietary high dose formulation of nitric oxide (NO) for adjunctive treatment of cystic fibrosis (CF). AIT-CF is a proprietary nitric oxide (NO) formulation and delivery system designed to deliver a high dose formulation (160 ppm) to the lungs using positive air pressure and integrated monitoring parameters. The Company's novel system has the potential to eliminate microbial infections including bacteria, fungi and viruses. NO is produced naturally by the body as a highly effective antimicrobial defense mechanism, but to date no delivery system has been able to deliver an effective and non-toxic antimicrobial dosage to the lungs. AIT's unique and proprietary system continuously monitors safety and efficacy parameters in the patient and is adaptable to treat a wide range of lung infections.
http://tinyurl.com/mtdsl6v

FDA Approves Pulmozyme with eRapid Nebulizer; CF Patients See 2 - 3 Minute Treatment Times

The eRapid Nebulizer System (eRapid) from PARI has been approved as the first electronic nebulizer by the Food and Drug Administration to deliver Genentech's Pulmozyme for cystic fibrosis treatment. eRapid is able to reduce average treatment times with Pulmozyme from 6-8 minutes down to 2-3 minutes.
http://tinyurl.com/kgheb35

UNC spinout looking to one-up Kalydeco with one-size-fits-all cystic fibrosis therapy

University of North Carolina spinout Spyryx Biosciences is developing a peptide-based therapy that, in theory, could be effective among all CF patients. The peptide, called SPLUNC1, may stop the epithelial sodium channel from becoming hyperabsorbative – upsetting the fluid regulation in the lungs and drying out the mucus channel – normalizing the excretion of mucus in the lungs. The synthesized peptide, called S18, will be developed as an inhalant peptide replacement therapy that's meant to work for all CF patients – it won't be pheno-type dependent.
http://tinyurl.com/kmdtnet

Depression Shown To Impact Lung

Continued on page 38

TILLMAN *continued from page 37*

Function in Cystic Fibrosis Patients

A recent study entitled "Symptoms of depression impact the course of lung function in adolescents and adults with cystic fibrosis," published in the journal BMC Pulmonary Medicine examined depression as a potential predictor of a decline in lung function in patients with cystic fibrosis (CF). Average rates of annual decline in FEV1% are reported to be between one and three percent. Results from this study indicated an association between depression and lung function at baseline. After two years follow-up, a marked decline in lung function was observed in those patients with depression at baseline. At the present time, the researchers underscore the fact that patients with CF with co-morbid depression are not receiving psychotherapeutic and psychopharmacological treatments to address their symptoms. Findings from this study indicated that clinicians should screen patients with CF for depression and treat this severe condition along with CF.

http://tinyurl.com/lo5gjr6

AND

http://tinyurl.com/nnhtlbq

Savara Pharmaceuticals' Aerovanc meets primary endpoint of MRSA reduction in Phase 2 trial in people with cystic fibrosis

Savara Pharmaceuticals announced today positive results from its Phase 2 clinical trial of AeroVanc, the first inhaled antibiotic being developed to address the growing problem of methicillin-resistant Staphylococcus aureus (MRSA) lung infection in people with cystic fibrosis (CF). AeroVanc is currently being developed as a treatment for persistent MRSA lung infection in people with CF. By delivering vancomycin directly to the lungs, higher vancomycin concentrations are achieved at the site of infection, which is expected to lead to improved clinical efficacy. In addition, direct delivery of the drug into the lungs reduces exposure to the drug elsewhere in the body, and is thereby expected to reduce the risk of systemic drug-related side effects. AeroVanc has received from the FDA Fast Track and Orphan Drug designations as well as Qualified Infectious Disease Product (QIDP) status providing 12 years of market exclusivity.

http://tinyurl.com/pafyg8b

Mucus Retained in Cystic Fibrosis Patients' Cells Leads to Potentially Deadly Infections

One of the key signs of cystic fibrosis is that mucus lining the lungs, pancreas and other organs is too sticky, which makes it difficult for the organs to work properly and, in the lungs, attracts bacteria and viruses resulting in chronic infections. Researchers recently found that cystic fibrosis mucus actually gets stuck inside some of the cells that create it, rather than simply becoming stuck on the outside linings of organs. The characteristics of mucus stored within the cells is not as acidic as in normal cells. Finding that cystic fibrosis mucus granules are not acidic is important because the lack of acidity slows the release of products from other secreting cells.

http://tinyurl.com/lp897b5

AND

http://tinyurl.com/p87nnyv

Cystic Fibrosis Discovery May Lead to New Treatment Strategy to Help Patients Breathe Easier

Researchers have discovered why mucus in the lungs of people with cystic fibrosis (CF) is thick, sticky and difficult to cough up. In CF—contrary to previous belief—inflammation causes new molecular bonds to form within mucus, transforming it from a liquid to an elastic sludge. The scientists also made headway in the lab in exploring a potential new therapeutic approach to dissolve those bonds and return the mucus to a liquid that is easier for the lungs to clear. Polymers—naturally-occurring molecules in mucus that form long chains—are the key to the discovery. CF mucus consists of a dense core of mucin with a layer of DNA wrapped around it, like a thin blanket draped over a solid pillow. While Pulmozyme makes mucus less stiff by eliminating DNA, N-acetylcysteine (NAC) succeeds in liquefying it by breaking up the mucin.

http://tinyurl.com/qb6f3h8

AND

http://tinyurl.com/p6rvoy4

Researchers pin down genetic pathways linked to CF disease severity

Researchers have identified genetic pathways—or clusters of genes—that play major roles in why one person with CF might never experience the worst kinds of symptoms while another person will battle severe airway infection for a lifetime. When these pathways or groups of genes are highly expressed, CF patients have less severe symptoms. When these pathways are expressed in lower amounts, patients experience a more severe form of the disease and are more likely to be hospitalized.

http://tinyurl.com/m6ovzb6

Garlic Extract Could Help Cystic Fibrosis Patients Fight Infection

A chemical found in garlic can kill bacteria that cause life-threatening lung infections in people with cystic fibrosis. The chemical, known as allicin, could be an effective treatment against a group of infectious bacteria that is highly resistant to most antibiotics. Researchers found that allicin—which can be extracted by crushing raw garlic—inhibits the growth of bacteria and, at higher doses, kills the plant pathogens. Allicin kills Burkholderia cepacia complex (Bcc) bacteria by chemically modifying key enzymes. This deactivates them and halts important biological processes within the pathogens' cells.

http://tinyurl.com/q9eal6x

AND

http://tinyurl.com/nbldyhx

TREATMENTS

Long-term treatment with oral N-acetylcysteine: Affects lung function but not sputum inflammation in cystic fibrosis subjects. A Phase II randomized placebo-controlled trial. C. Conrad, J. Lymp, V. Thompson, C. Dunn, Z. Davies, B. Chatfield, D. Nichols, J. Clancy, R. Vender, M.E. Egan, L. Quittell, P. Michelson, V. Antony, J. Spahr, R.C. Rubenstein, R.B. Moss, L.A. Herzenberg, C.H. Goss, R. Tirouvanziam. Journal of Cystic Fibrosis. March 2015. Volume 14, Issue 2, Pages 219–227

The effects of oral N-acetylcysteine (NAC), which replenishes systemic glutathione, on decreasing inflammation and improving lung function in CF airways was studied. It was found that NAC recipients maintained their lung function while placebo recipients declined. However, no effect on sputum human neutrophil elastase activity and other selected biomarkers of neutrophilic inflammation were detected.

http://tinyurl.com/lsrhw4l

Evidence for the efficacy of aztreonam for inhalation solution in the management of Pseudomonas aeruginosa in patients with cystic fibrosis. Hansen C, Skov M. Ther Adv Respir Dis. 2014 Dec 3

This article focuses on the use of inhaled antibiotics in chronic P. aeruginosa infection in CF, and specifically on studies including the use of inhaled aztreonam lysine in P. aeruginosa infection. Inhaled aztreonam is an important new treatment option for chronic P. aeruginosa infection in CF. Long-term studies have shown that the drug is safe and superior to inhaled tobramycin in these specific infections.

http://tinyurl.com/ouhr677

Enhancement of Pulmozyme activity in purulent sputum by combination with poly-aspartic acid or gelsolin. Bucki R, Cruz K, Pogoda K, Eggert A, Chin L, Ferrin M, Imbesi G, Hadjiliadis D, Janmey PA. J Cyst Fibros. 2015 Feb 12

DNase (Pulmozyme) effectiveness in cystic fibrosis treatment is in some cases limited by its inability to access DNA trapped within bundles in highly viscous fluids that also contain actin. Dissociating DNA-containing bundles using actin depolymerizing agents and polyanions has potential to increase DNase efficacy. Research indicates that the addition of low concentrations of p-ASP or gelsolin can increase the therapeutic value of Pulmozyme (DNase 1).

http://tinyurl.com/lryh3d4

Pharmacokinetics and tolerability of oral sildenafil in adults with cystic fibrosis lung disease. J.L. Taylor-Cousar, C. Wiley, L.A. Felton, C. St. Clair, M. Jones, D. Curran-Everett, K. Poch, D.P. Nichols, G.M. Solo-mon, M.T. Saavedra, F.J. Accurso, J.A. Nick. Journal of Cystic Fibrosis. March 2015. Volume 14, Issue 2, Pages 228–236

Airway inflammation is central to cystic fibrosis (CF) pathophysiology. Pre-clinical models have shown that phosphodiesterase inhibitors (PDEi) like sildenafil have anti-inflammatory activity. It was found that subjects with CF may eliminate sildenafil at a faster rate than non-CF subjects. Sildenafil administration was safe in subjects with CF and decreased sputum elastase activity.

http://tinyurl.com/n43s8yj

Factors associated with response to treatment of pulmonary exacerbations in cystic fibrosis patients. Waters VJ, Stanojevic S, Sonneveld N, Klingel M, Grasemann H, Yau YC, Tullis E, Wilcox P, Freitag A, Chilvers M, Ratjen FA. J Cyst Fibros. 2015 Feb 14

Pulmonary exacerbations are associated with significant lung function decline from baseline in cystic fibrosis (CF) and it is not well understood why some patients do not respond to antibiotic therapy. The objective of this study was to identify factors associated with lung function response to antibiotic treatment of pulmonary exacerbations. It was found that inadequate reduction of inflammation during an exacerbation is associated with failure to recover lung function and increased risk of subsequent re-exacerbation in CF patients.

http://tinyurl.com/q5yzzrn

Eradication of respiratory tract MRSA at a large adult cystic fibrosis centre. Hall H, Gadhok R, Alshafi K, Bilton D, Simmonds NJ. Respir Med. 2015 Mar;109(3):357-63

Combined antibiotic therapy, particularly Rifampicin/Fusidic acid, is a well-tolerated and effective means of eradicating new infections of MRSA in patients with cystic fibrosis.

http://tinyurl.com/lv4vcg3

Reduced risk of nontuberculous mycobacteria in cystic fibrosis adults receiving long-term azithromycin. Nathalie Coolen, Philippe Morand, Clémence Martin, Dominique Hubert, Reem Kanaan, Jeanne Chapron, Isabelle Honoré, Daniel Dusser, Etienne Audureau, Nicolas Veziris, Pierre-Régis Burgel. Journal of Cystic Fi-brosis. Published Online: February 28, 2015

Azithromycin reduces exacerbations in cystic fibrosis (CF) patients. Our aim was to investigate its association with nontuberculous mycobacteria isolation and macrolide susceptibility. The data obtained from this study suggest that azithromycin is a primary prophylaxis for NTM infection in CF adults.

Continued on page 40

TILLMAN *continued from page 39*

http://tinyurl.com/nb99ejy

Randomized, single blind, controlled trial of inhaled glutathione vs placebo in patients with cystic fibrosis. Calabrese C, Tosco A, Abete P, Carnovale V, Basile C, Magliocca A, Quattrucci S, De Sanctis S, Alatri F, Mazzarella G, De Pietro L, Turino C, Melillo E, Buonpensiero P, Di Pasqua A, Raia V. J Cyst Fibros. 2015 Mar;14(2):203-10

In cystic fibrosis (CF) the defective CF transmembrane conductance regulator protein may be responsible for the impaired transport of glutathione (GSH), the first line defense of the lung against oxidative stress. The aim of this single-blind, randomized, placebo-controlled trial was to evaluate the effect of inhaled GSH in patients with CF. Twelve-month treatment with inhaled GSH did not achieve the predetermined primary outcome measure of 15% improvement in FEV1%. Only in patients with moderate lung disease, 3, 6 and 9 months therapy with GSH resulted in a statistically significant increase of FEV1 values from the baseline. Thus, inhaled GSH has slight positive effects in CF patients with moderate lung disease.

http://tinyurl.com/l8kry63

A phase 3, open-label, randomized trial to evaluate the safety and efficacy of levofloxacin inhalation solution (APT-1026) versus tobramycin inhalation solution in stable cystic fibrosis patients. J. Stuart Elborn, David E. Geller, Douglas Conrad, Shawn D. Aaron, Alan R. Smyth, Rainald Fischer, Eitan Kerem, Scott C. Bell, Jeffery S. Loutit, Michael N. Dudley, Elizabeth E. Morgan, Donald R. VanDevanter, Patrick A. Flume. Journal of Cystic Fibrosis. Published Online: January 13, 2015

Inhaled antibiotics are standard of care for persons with cystic fibrosis (CF) and chronic Pseudomonas aeruginosa airway infection. APT-1026 (levofloxacin inhalation solution, LIS) is fluoroquinolone in development. This multinational, randomized, non-inferiority study compared LIS and TIS over three 28-day on/off cycles. LIS was found to be a safe and effective therapy for the management of CF patients with chronic P. aeruginosa infection.

http://tinyurl.com/llo4bt8

Effects of exercise intensity compared to albuterol in individuals with cystic fibrosis. Wheatley CM, Baker SE, Morgan MA, Martinez MG, Morgan WJ, Wong EC, Karpen SR, Snyder EM. Respir Med. 2014 Dec 18

Although exercise is a vital component of the therapy prescribed to individuals with cystic fibrosis (CF), it is not a priority due to a finite amount of treatment time and the view that exercise is not as beneficial as pharmacological treatments by many individuals with CF. This study sought to compare the therapeutic benefits of exercise and their prescribed bronchodilator albuterol. Results suggest that moderate intensity exercise is the optimal intensity for individuals with CF, as low intensity exercise increases EPI less than 50% and vigorous intensity exercise is over taxing, such that airflow can be restricted. Although the duration of the beneficial effect is uncertain, exercise can promote greater improvements in gas diffusion and comparable bronchodilation when compared to albuterol.

http://tinyurl.com/mc8ezyb

Physiotherapy and cystic fibrosis: what is the evidence base? McIlwaine MP, Lee Son NM, Richmond ML. Curr Opin Pulm Med. 2014 Nov;20(6):613-7

Several long-term studies have looked at the efficacy of airway clearance techniques, including active cycle of breathing techniques, autogenic drainage, high frequency chest wall oscillation, postural drainage, positive expiratory pressure (PEP), and oscillating PEP. Each of these studies reported some efficacy of airway clearance in maintaining health with no one technique being superior to another. However, one study suggested that high frequency chest wall oscillation was not as effective as PEP in maintaining health in CF patients. Individual preference needs to be considered when selecting a technique. Recent studies have found exercise to increase mucociliary clearance peripherally. Musculoskeletal issues, including posture, bone density, urinary incontinence, and pain should be assessed and managed in individuals to improve the mechanics of breathing and overall well-being.

http://tinyurl.com/ko2rr7b

FYI

Evaluation of mold exposure in cystic fibrosis patients' dwellings and allergic bronchopulmonary risk. Steffi Rocchi, Bénédicte Richaud-Thiriez, Coralie Barrera, Frédéric Grenouillet, Jean-Charles Dalphin, Lauren-ce Millon, Gabriel Reboux. Journal of Cystic Fibrosis. March 2015 Volume 14, Issue 2, Pages 242–247

Results indicate that indoor fungal contamination could be a factor favoring allergic bronchopulmonary aspergillosis and suggest that environmental surveys could help in preventing fungal risk in CF patients.

http://tinyurl.com/lnbgoh6

Idiosyncratic reactions are the most common cause of abnormal liver function tests in patients with cystic fibrosis. Jong T, Geake J, Yerkovich S, Bell SC. Intern Med J. 2015 Feb 2

The aim of the study was to identify risk factors for elevated liver function tests (LFTs) in CF patients receiving IV antibiotics. Elevated LFTs are common during IV antibiotic treatment in CF. While specific antibiotic exposure may contribute to abnormal LFTs in a minority of cases, this study demonstrates that antibiotic-induced liver injury is largely idiosyncratic and unpredictable.

http://tinyurl.com/l65wgu8

Call For Nominations From The CF Community

USACFA is accepting nominations for the Jacoby Angel Award and the USACFA Founders Award, which are awarded every two years by the United States Adult CF Association (USACFA). Readers of *CF Roundtable* nominate individuals for each award. USACFA Directors then vote on the nominees. A USACFA Director cannot be nominated to receive an award.

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

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

We look forward to receiving nominations for the Jacoby Angel Award for people who have followed in the footsteps of the award's namesake, Dr. Jack Jacoby, by being a person with CF who dedicates himself or herself to helping others.

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

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

Past recipients of the Founders Award include: Lisa McDonough, Dr. Jerry Nick, Dr. James Yankaskas, Beverley Donelson and Dorothy Hello.

Nominations should be e-mailed by June 15, 2015, to: CFLegal@sufianpassamano.com. ▲

Pseudomonas aeruginosa in CF and non-CF homes is found predominantly in drains. M.E. Purdy-Gibson, M. France, T.C. Hundley, N. Eid, S.K. Remold. Journal of Cystic Fibrosis. Published Online: November 28, 2014

The findings implicate drains as important potential sources of P. aeruginosa infection. They suggest that maximizing P. aeruginosa control efforts for drains would reduce exposure with minimal extra burden to CF patients and families.

http://tinyurl.com/pobd463

Pneumothorax in cystic fibrosis. Kioumis IP, Zarogoulidis K, Huang H, Li Q, Dryllis G, Pitsiou G, Machairiotis N, Katsikogiannis N, Papaiwannou A, Lampaki S, Porpodis K, Zaric B, Branislav P, Mpoukovinas I, Lazaridis G, Zarogoulidis P. J Thorac Dis. 2014 Oct;6(Suppl 4):S480-7

Pneumothorax is recognized as a common and life-threatening complication in cystic fibrosis (CF) patients, especially in those who are infected with P. aeruginosa, B. cepacia or Aspergillus, need enteral feeding, are diagnosed as suffering from allergic bronchopulmonary aspergillosis (ABPA), developed massive hemoptysis, and their respiratory function is seriously compromised. Structural impairment and altered airflow dynamics in the lungs of CF patients are considered as the main predisposing factors, but also inhaled medications and non-invasive positive pressure ventilation (NIPPV) could increase the risk of pneumothorax. Clinical presentation could range from dramatic to very mild. Management of spontaneous pneumothorax occurring in patients with CF is essentially similar to that for non-CF patients. Therapeutic options include intercostal tube drainage, video-assisted thoracoscopic surgery (VATS), and medical or surgical pleurodesis. Pneumothorax increases both short-

Continued on page 43

CFRI's 28th National Cystic Fibrosis Family Education Conference

July 31 – August 2, 2015

Sofitel San Francisco Bay - Redwood City, CA

CFRI's annual conference brings together experts in the field of CF to provide the latest updates in research and care to our diverse CF community.

Speakers (To Date) Include:

Chris Kvam, Esq – Rochester, New York
Elika Rad, MS, RN NPC – Stanford Hospital and Clinics
Margaret Rosenfeld, MD - Seattle Children's Hospital
Greg Sawicki, MD, MPH – Boston Children's Hospital
Scott Russell, DPT – University of Southern California
Lisa Saiman, MD – Columbia University Medical Center
Beth Sufian, Esq. – Houston, Texas
Anna Tsang, MSO – St. Michael's Hospital, Toronto

- Early Bird Registration (on or before 7/1/15) - $180 per person
- Regular Registration (after 7/1/15) - $215 per person
- Registration includes meals, reference materials, and access to presentations and support groups.
- To ensure good health for all, please use proper hygiene practices.
- All participants/guests with CF must comply with CFRI's Infection Control Guidelines.

For more information, visit www.cfri.org or call 1.855.cfri.now

CF ROUNDTABLE SUBSCRIPTION FORM (Please Print Clearly)

PLEASE CHECK ONE: ☐ **NEW SUBSCRIPTION** **OR** ☐ **UPDATE**
NAME _____ **PHONE (** **)** _____
ADDRESS _____
CITY _____ **STATE** _____ **ZIP+4** _____ **+** _____
E-mail address: _____
I am interested in CF Roundtable because I am a: CF ADULT _____ BIRTHDATE _____
OR I am a: Parent ____ Relative ____ Friend ____ Medical/Research Professional ____ Caregiver ____

CF Roundtable is available at no cost. However, to help defray printing and mailing costs, donations are always welcome.

- Donation to defray costs . . . . . $ _____
- Please send me the back issues I have listed on a separate sheet of paper.
- Please send subscriptions to the names and addresses I have listed on a separate sheet of paper.

Please do not write in this space

SP 15

For donations, make checks payable to USACFA and mail with this completed form to:
USACFA, P.O. Box 68105, Indianapolis, IN 46268-0105
We ask that everyone keep their home and e-mail address information up to date.

OR

You can also subscribe online! Go to www.cfroundtable.com. You can now scan this QR Code with your smart phone to go directly to our online registration page. By subscribing online you have the ability to download a PDF of the latest newsletter, receive the mailed version or you can receive both. The online version will be available two weeks prior to the mailed version.

KEEPING YOUR INFORMATION CURRENT

To keep our records up to date, please be sure to complete and return a subscription form, on this page, to us or register online with any changes to your information, www.cfroundtable.com. (Any issue of the newsletter that is returned to us costs us about two-and-a-half times the first-class postage rate for that piece. Currently that runs about $3.73 per returned copy.) **Thank you for helping us with this.**

TILLMAN *continued from page 41s*

and long-term morbidity and mortality in CF patients and causes significant deterioration of their quality of life.
http://tinyurl.com/lu5bw5u

Comparing Mycobacterium massiliense and Mycobacterium abscessus lung infections in cystic fibrosis patients. Roux AL, Catherinot E, Soismier N, Heym B, Bellis G, Lemonnier L, Chiron R, Fauroux B, Le Bour-geois M, Munck A, Pin I, Sermet I, Gutierrez C, Véziris N, Jarlier V, Cambau E, Herrmann JL, Guillemot D, Gaillard JL; OMA group. J Cyst Fibros. 2015 Jan;14(1):63-9

Mycobacterium massiliense is closely related to Mycobacterium abscessus and is also a frequent cause of mycobacterial lung disease in patients with cystic fibrosis (CF). The data show a particular link between M. massiliense and malnutrition specifically in CF patients. Unlike M. abscessus, the bacteriological response of M. massiliense to combination antibiotic therapies containing clarithromycin was excellent. Distinguishing between M. massiliense and M. abscessus has major clinical implications for CF patients.
http://tinyurl.com/qbuourm ▲

Laura Tillman is 67 and has CF. She is a former Director and President of USACFA. She and her husband, Lew, live in Northville, MI.

REMINDERS

- Please notify us immediately of any address changes. Returned mail wastes money and delays mailings.
- We would like to act as a referral source for active adult support groups. Please send us your group name, leader's name and phone number, number and age range of your members and geographical area covered, and we will add you to our referral list.
- Please let us know of the major occurrences in your life (*e.g.*, *marriages*, *births*, *completion of educational degrees or training*, *career advancement*, *transplants*, *anniversaries*, *birthdays*), and we will print your information in **Milestones**.
- Share your ideas for **Focus Topics**, feature articles or any suggestions for improvements you may have to help make *CF Roundtable* more relevant and interesting to you.
- You can reach **USACFA** and ***CF Roundtable*** at anytime by e-mail at **cfroundtable@usacfa.org**
- Send your questions of a general nature regarding legal issues that relate to CF to our legal advisor: **Beth Sufian, Esq., 712 Main, Suite 2130, Houston, Texas 77005. E-mail: CFLegal@sufianpassamano.com.**
- You may subscribe at www.cfroundtable.com

PRINTED WITH SOY INK

Published by the United States Adult Cystic Fibrosis Association, Inc.
CF Roundtable is printed on recycled paper.

IMPORTANT RESOURCES

Partnership for Prescription Assistance: Phone: 1-888-477-2669 http://www.pparx.org/prescription_assistance_programs
The Partnership for Prescription Assistance brings together America's pharmaceutical companies, doctors, other health care providers, patient advocacy organizations and community groups to help qualifying patients without prescription drug coverage get free or low-cost medicines through the public or private program that's right for them.

United Network for Organ Sharing (UNOS): Phone: 1-888-894-6361 http://www.unos.org/
Call for information on transplant centers, access for all patients needing organ transplants and general transplant information.

Transplant Recipients International Organization, Inc. (TRIO): Phone: 1-800-TRIO-386 http://www.trioweb.org/index.shtml
An independent, nonprofit, international organization committed to improving the quality of life of transplant recipients and their families and the families of organ and tissue donors. For information, write to: TRIO, 2100 M Street NW, #170-353, Washington, DC 20037-1233 or e-mail them at: info@trioweb.org.

American Organ Transplant Association (AOTA): Phone: 1-713-344-2402 http://aotaonline.org/default.aspx
Helps defray out-of-pocket travel expenses for transplant recipients. Helps to set up trust funds. For more information, write to: AOTA, 21175 Tomball Parkway #194, Houston, TX 77070-1655.

ADA: To learn how the Americans with Disabilities Act (ADA) applies to you, contact the Disability Rights Education and Defense Fund (DREDF): Phone: 1-800-348-4232 http://www.dredf.org/.