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

U. S. Adult Cystic Fibrosis Assn., Inc. – The First 25 Years

By Kathy Russell

This issue marks the end of our first 25 years of publishing *CF Roundtable*. I am absolutely amazed by that. The years have rushed by very fast. So many people have volunteered their time with us and we appreciate all of them.

At the start, we wanted to do it all on our own. Although the CFF kindly offered to fund us, we respectfully declined. We wanted to see if our community of adults who have CF wanted a newsletter enough that they would fund it. With the help of a loan from one of the founders, we mailed our first letter to a select group of people asking for their assistance in getting started. Those people and others came through for us and we put out our first issue in November of 1990.

Thanks to our subscribers, we were able to pay back our start-up loan in the second year. We have managed to stay on even footing since then. In recent years, we have received generous grants and donations from several corporations and individuals, which help to keep us well-funded.

> "In the beginning, our mission was to publish a quarterly newsletter to facilitate the exchange of information among adults who have CF and foster a nationwide network of support for adults who have CF."

In the beginning, our mission was to facilitate the exchange of information among adults who have CF, to foster a nationwide network of support for adults who have CF and to sponsor regional conferences in the United States for the exchange of information between the medical and research community and adults who have CF.

We have managed to print four issues of *CF Roundtable* each year and we've come a long way from our first 12-page issue. We have added regular columnists, color photos and color in other ways, too. Our largest issue was 48 pages and we routinely publish 40 pages. Contacts that people make through the newsletter have fostered a

Continued on page 18

INSIDE THIS ISSUE

CF ROUNDTABLE
FOUNDED 1990
Vol. XXV, No. 4

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

EDITOR'S NOTES

What a fantastic summer we had here in the northwest. It was the warmest year on record. Fortunately, it wasn't too hot. Now, autumn is coming in gently and beautifully. The colors are spectacular. We know that we will get rain, but we hope that it will come a little at a time rather than all at once. We hope that winter will be kind to us.

Perhaps you saw on the front page that we made it to our 25th anniversary. Hooray! USACFA was begun in 1990 and has been publishing continuously since then. We look forward to the future and hope you will share it with us.

Inside this issue we have articles by many interesting writers. **Beth Sufian** answers questions about Medicare and Medicaid from our readers in "Ask The Attorney." In "Spirit Medicine" **Isabel Stenzel Byrnes** writes of the importance of belonging. **Jennifer Hale**, in "Coughing With A Smile," continues the journal of her journey to transplant. If you ever have had trouble sleeping, be sure to read what **Julie Desch** has to say in "Wellness" about sleep and how to get it.

"Searching For The Cure" finds **Reid D'Amico** and **Meranda Honaker** discussing therapeutic strategies post-transplant. It continues with **Brad Aaron** telling of his experience with clinical trials. It finishes with a listing of current clinical trials.

Jeanie Hanley sums up her recent experience at a CFRI conference with a discussion of exercise and activity. **Laura Mentch** continues the discussion of the CFRI conference by sharing highlights of the notes she took.

In "Voices From The Roundtable" **Andrea Eisenman** recounts the bike ride where she learned when to stop an activity. As usual, "Information From The Internet" is full of all kinds of news, thanks to **Laura Tillman**. We find **Jillian McNulty**, from Ireland, featured in "In The Spotlight." **Klyn Elsbury** relates her struggles to get Orkambi approved by her insurance company in the "Conversation Corner."

Our Focus topic is: Incorporating Work Into Our CF Care. Three people chose to tackle this topic. **Amy Braid** writes of how working affected her health and the hurdles she faced after "retiring." **Chris Kvam** tells us that working and choosing the correct type of work are important to us. **Devin Wakefield** recounts dealing with a bowel obstruction while working. OY!

Please look at the future Focus topics listed on the facing page. See if any is of interest to you. If so, please write for the newsletter. We welcome your articles.

Until next time, stay healthy and happy,

Kathy

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

Information From The Internet...

Compiled by Laura Tillman

PRESS RELEASES

Researchers Offer Insight Into Complications For CF Patients Receiving Parenteral Antimicrobial Therapy

Researchers recently reported new data regarding complications in outpatient parenteral antimicrobial therapy (OPAT) for effective treatment of pulmonary bacterial infections in cystic fibrosis patients. Antibiotics are an effective treatment for bacterial infection; however, the treatment itself can last up to 21 days to ensure the maximal reduction in pulmonary bacterial load. OPAT is an option that allows patients to return home quicker and to continue their daily life, with the antibiotic therapy being provided at home, in an office-based setting or through a hospital-based infusion program. The OPAT strategy also reduces the risk of bacteria transmission among CF patients. However, for the OPAT strategy to be successful, patients have to receive the correct amount of antibiotics at the right time in order to avoid adverse reactions. The goal of the study was to investigate the frequency of complications associated with OPAT, during and after treatment, when using catheters through a peripheral intravenous line or central lines (large veins, including the vena cava) among CF patients. Researchers found that the median lifetime of peripheral venous lines before they need to be replaced is seven days when using an infusion pump and five days when using a bolus injection. The authors suggest that this indicates a more constant flow of antibiotics as provided by the infusion pump might be less traumatic for the vein and also determined that the infusion pump minimizes complications. Regarding central intravenous lines through PICC-lines (peripherally inserted central catheter-lines), researchers found that although the lifetime of these lines has been reported to be up to six months, 39.1% (9 out of 23) of the cases employing PICC-lines in the study needed to be removed after a median lifetime of 17 days due to complications. Concerning PAC (port-a-cath) lines, researchers suggested that it can be considered a long-term solution as it was found to have a median lifetime of at least 735 days. The research team concluded that when considering peripheral venous lines for OPAT, the use of an infusion pump minimizes complications. Regarding central lines, the team reports that PICC-lines and PACs have a similar frequency of complications that lead to catheter removal,

Continued on page 5

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**

Autumn (current) 2015: Incorporating Work Into Our CF Care.

Winter (February) 2016: Dealing With Gastrointestinal Issues. (Submissions due December 15, 2015.) What kinds of gastrointestinal issues have you had? How did you deal with them? What can you tell our readers about how to handle these problems?

Spring (May) 2016: Managing Various Conditions (Under The CF Umbrella). (Submissions due March 15, 2016.) Do you have conditions such as GERD, CF-related arthritis, blood pressure problems etc. that are not purely CF-related but you must manage under the umbrella of CF? Tell us about them and how you have dealt with them.

Summer (August) 2016: Living With Anticipatory Grief And Survivor's Guilt. (Submissions due June 15, 2016.)

ASK THE ATTORNEY

Answers To Readers' Questions

By Beth Sufian, JD

In the past three months readers have asked many questions about Medicare coverage and SSI income and asset eligibility criteria. Nothing in this article is meant to be legal advice about your specific situation but is meant to be only information. CF Roundtable *readers with questions about issues related to Social Security benefits, Medicare, Medicaid, health insurance coverage, school issues and employment issues can contact the CF Legal Information Hotline at 1-800-622-0385 or CFLegal@sufianpassamano.com. The Hotline is sponsored by the CF Foundation and all calls are free of charge and confidential. The CF Legal Information Hotline now can schedule a specific time for you to speak with an attorney during the week or on the weekend.*

1. I have Medicare and my CF Center says I need to buy a Medicare supplement policy to pay for the 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?

Approximately half of all states do not require an insurance company to sell a Medicare supplement policy to a person who has Medicare because he or she is also eligible for Social Security Disability Insurance (SSDI) benefits. In states where there is no requirement that an insurance company sell such a policy to a person who receives Medicare because she/he receives SSDI benefits, it is difficult (and in many states impossible) to find an insurance company that will sell such a policy. Medicare supplement policies must be provided to a person on Medicare who is over 65 years of age.

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 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 20 percent portion of Part B expenses 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. Therefore, purchasing a Medicare supplement policy often is a good idea if such a policy is available.

2. What medications are covered by Medicare Part B and what medications are covered by Medicare Part D?

Medicare Part B also provides coverage for a limited number of prescription medications.

Most CF medications that are covered under Medicare Part B will have Patient Assistance Programs that provide assistance with paying the Part B 20 percent co-pay for the medication. Some Patient Assistance Programs have income eligibility guidelines but others have extremely high income guidelines which allow most people with CF to qualify for co-pay assistance.

BETH SUFIAN

Many people are confused about coverage for medications under Medicare because, well, it is confusing! The general rule is that Medicare Part B will provide coverage for any FDA-approved medication that is used with a piece of durable medical equipment. Examples of such drugs are Pulmozyme, TOBI and Albuterol. The exception to the rule is Cayston. When Cayston was approved by the FDA, Medicare decided that it was not a drug used with durable medical equipment. Many people think this was done as a way for Medicare to shift the cost for Cayston to Medicare Part D.

So even though Cayston is used with a piece of durable medical equipment and is FDA-approved it is covered under Medicare Part D. The Cayston coverage decision helps people with CF because coverage of Cayston under Medicare Part D allows a person with CF to quickly meet the Part D deductible and to get in and out of the Medicare Part D "donut hole" quickly. Once a person is out of the Medicare Part D "donut hole," the person is eligible for Medicare Part D catastrophic coverage. A person who is in Medicare Part D catastrophic coverage has very low co-pays for most Part D covered medications. The Cayston Access program that provides co-pay assistance for Cayston can assist most people with CF with the Part D co-pays for Cayston.

Inhaled drugs like Colistin and

hypertonic saline are NOT FDA-approved and so these drugs are covered by Medicare Part D.

Many pharmacies are not familiar with the Medicare Part D medication coverage rules. There are only approximately 30 medications that are covered across all diseases by Medicare Part B. It is important for adults with CF on Medicare to understand what drugs are covered under Part B and what drugs are covered under Part D.

3. 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?

Marriage will result in the new spouse's income and assets being considered by Social Security for purposes of determining eligibility for SSI benefits and for purposes of Medicaid eligibility.

In 21 states an adult (a person over the age of 18) is eligible for full Medicaid only if he/she is also eligible for SSI benefits. Therefore, if a person relies on Medicaid for her/his health insurance coverage, it is important to understand the income and asset eligibility rules before getting married.

Marriage may result in the loss of SSI benefits and Medicaid coverage if the spouse's income and assets are over the SSI allowable amount. People with CF should understand what their health insurance coverage options are and how much health insurance coverage will cost. The Affordable Care Act mandates coverage for all citizens in the United States regardless of health condition. However, an insurance policy costs money and there is a cost share for medical care and treatment under a private health insurance policy. Some people with CF have had Medicaid coverage their whole lives and are not familiar with the cost of health insurance coverage or the cost share involved when a person has a private health insurance policy. Individuals with CF should make sure they understand how much money they will be required to pay for health insurance and co-pays for treatment if they purchase a private health insurance policy. Some people with CF will be eligible for a federal premium subsidy to help with the cost of a policy purchased on the Federal Healthcare Exchange or purchased on a state Healthcare Exchange. In addition, there are patient assistance programs that may be able to help with co-pays for medications. ▲

Beth is 50 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.

TILLMAN continued from page 3

although PACs have a considerably longer average lifetime.

http://tinyurl.com/nn4r2u4

AND

http://tinyurl.com/pdzc2pz

Faster tests for lung infections

Cardiff scientists have developed a new diagnostic approach offering an unprecedented rapid and accurate diagnosis of lung infections in patients with cystic fibrosis. According to the researchers, the current method for diagnosing bacterial infection, which relies on culture tests, hasn't changed substantially in over half a century. As well as being time-consuming and costly, they say the method is flawed and in 11% of cases fails to detect the deadly bacteria present in infection, of which there are hundreds in any given patient lung sample. The new test uses DNA isolated from lung samples by a method routinely applied across the UK for diagnosis of viral infections. Using this readily available DNA in a joined-up testing approach could revolutionize the future of microbial infection diagnosis.

http://tinyurl.com/ogq8e4l

Tackling the root cause of cystic fibrosis

Scientists have found that a small molecule, when tested in yeast, can substitute for a protein and restore a key cellular function related to those missing in people with CF. The researchers tested a natural small molecule called amphotericin B, which was originally extracted from bacteria and is used to treat fungal infections. It was tested at low doses on a strain of yeast that is unable to transport ions in and out of its cells, and therefore doesn't grow. But with treatment, the yeast grew nearly as well as the control. The researchers say this shows that channel-forming small molecules can work in collaboration with protein pumps and channels in cell membranes to restore the flow of ions.

http://tinyurl.com/ooxqfcc

Pulmatrix announces new inhaled anti-infective therapy to treat CF-associated fungal infections

Pulmatrix, Inc., announced a new drug candidate, PUR1900, an inhaled anti-infective to treat fungal infections associated with cystic fibrosis (CF). PUR1900 is a dry powder formulation of a large, complex anti-fungal compound that can be administered at high therapeutic dose to the lung while

Continued on page 24

SPIRIT MEDICINE

The Spirit Of Belonging

By Isabel Stenzel Byrnes

Flashback to CF camp, 1987. Every cabin had to perform a skit together for the Thursday night talent show. After struggling to compromise what skit we'd do among several girls who loved heavy metal and hard rock, with a few of us who preferred folk and easy listening, the one song we finally agreed to lip sync was "We Belong" by Pat Benatar. My cabin mates and I (affectionately called "The Flamingos") doused ourselves in hairspray and put on neon tights and way too much bright makeup. Needless to say, we rocked the house that night with that song.

That moment highlights a pivotal theme in my life, literally and figuratively: the need to belong. Most teenagers feel the need to belong to a group, lest they be called "out" instead of "in." By 15, I had already spent 25 weeks in the hospital, I had cancelled many play dates, I couldn't join many evening social events because of treatments, and I looked very different from my peers. I was clearly "out." All of these things made me feel different and isolated. Now, because I was at CF camp, I found my place of belonging.

Belonging is one of the most basic human needs. In fact, it's the third need among Maslow's hierarchy of needs, after only physiologic and safety needs. We enter this world as part of a family, and then belong to this tribe. Nicely conveyed in Disney's latest movie, *Inside Out*, the foundations of our sense of belonging, also known as the "core memories," are constructed by our family. And in Western culture, it's very natural that as we grow up, we need to expand our tribe and seek out groups and communities that make us feel like we fit in.

But the need to belong expands past adolescence. Adults wear sweatshirts with their college names on them; we wear black and orange if we're from San Francisco (especially when the Giants win the World Series); we wear crosses or stars to belong to a faith and rubber bracelets to belong to a cause. Our fraternal organizations, workplaces, gangs, political parties and extracurricular activities help us feel like we matter. That's the power of branding—to help us feel connected to something bigger than ourselves.

Belonging, or having connection with others, is the opposite of loneliness. Being part of a tribe tells us, "I matter. People care about me. I'm worthy. I'm like others." Through connection, we become interdependent. Belonging helps us develop a sense of self: self-esteem, self-worth and self-acceptance. Not belonging is a form of social death, and on a primitive level, social death can lead to physical death. When we're born with a serious illness, belonging to a group means there'll be people who can take care of us when we are sick. Fitting in is a matter of survival.

ISABEL STENZEL BYRNES

Thanks to CF camp, then CF retreat, and actively volunteering with Cystic Fibrosis Research, Inc. (CFRI), the CF community has fulfilled my need to belong for 32 years. Among others with CF, I am accepted, I am promised loyalty and solidarity, and I am deeply understood. Without the CF community, I believe I would have died long ago. I would never have learned basic survival tips that my healthcare team never could teach. But more importantly, I would not have developed my spirit medicine—my coping strategies that have kept me going.

Belonging is a motive that drives human behavior. Stanford assistant professor Gregory Walton, in various studies, has demonstrated that a sense of social belonging can affect motivation and continued persistence, even on impossible tasks. That is, if you don't feel like you belong, you are both less motivated and less likely to hang in there in the face of obstacles. So who knows, maybe if I wasn't part of this community, I would've been less compliant with my treatments. I wonder if this applies to many people with CF.

I remember reading the description of CF camps in Dr. David Orenstein's first edition of his signature book, *Cystic Fibrosis: A Guide for Patients and Families* published in 1989. This was before the cross-infection concerns were well-documented, so the blurb about CF camps was positive. There was one downside, he had written, which was that going to CF camp could lead children to over-identify with CF, making the disease too dominant a part of their lives. If only the only negative of CF camps was that!

Okay, as someone who has a book and film about CF and who has been tied at the hip with CFRI, I think Dr. Orenstein was right. "Belonging" risked taking over my identity. But remember, this disease damn near killed me, so it would've probably been a dominant part of my life anyway—whether I went to camp or not. Thankfully, I tried to balance my CF world with my other life: my family, friends from school, studies, hobbies, career, travel, some exercise pursuits, Japanese cultural groups etc. But as my illness worsened, my world became smaller; it took more effort to be part of non-CF groups. Eventually, I had my family and my CF world, and that was it.

My relationship with the CF community has long been ambivalent. I've wanted to be part of exceptional people who inspire me, who love me and who show me that "if they can do it, so can I." But I also didn't want to be part of a CF community because I didn't want to have CF, and I didn't want to bear witness to the struggles and losses that this community experiences on a regular basis. Now that I've been transplanted, I feel more distant to the classic CF experience, but I still feel strongly empathetic to my CF peers struggling with classic CF. I've been blessed to have good health post-transplant, and have experimented with belonging to different groups. Having become a bagpiper, I now belong to the Stewart Tartan Pipes and Drums. Having lung capacity to swim, I now belong to the Peninsula Community Center Master's Swim Team. I now belong to Team Nor Cal and the Transplant Games of America. Having the health to work, I now belong to a workplace of hospice staff made of deep, loving, compassionate people. It feels great to have the health to experiment with being part of so many other groups!

With cross-infection concerns, fewer people with CF today have a chance to benefit from the traditional sense of belonging to a CF community that I had back in my day. However, we still have *CF Roundtable,* social media, the phone and other indirect ways to connect with those with CF. Where else can we belong, if not the kind of CF bonding I grew up with? Healthcare providers "get us" and witness what we go through, especially as the disease worsens and we may feel more in need of support. How often do we grow attached to the floor where we are hospitalized? I have fond memories of 7North or E-Ground. Even the hospital fosters belonging! Spiritual groups usually contain people who are more sensitive to struggle and may offer the compassion we need to live with CF. When health and energy permit, hobby and exercise groups, workplaces, cultural groups or family provide the fulfillment we need for well-being. If I ever feel disconnected, I have a habit of browsing "Meetup" groups and seeing which ones I'd like to belong to. The sky is the limit!

The older I get, the more I appreciate the changeability of my identity that has been so molded by my belonging to the CF world. I am more relaxed about who I am and how I want to invest my energy. I have mentally and emotionally practiced a change in belonging. I realize some day my parents will die, and my family—my tribe—as I know it will change. I will belong to fewer groups as I get older and have less energy. I will lose my job, lose more friends, lose my hobbies and lose my health. I may even lose my CF community. But even if I do, I am consoled at a spiritual level. I ask God where I belong, and He tells me, "Everywhere." I meditate on this regularly, especially when I'm alone. I believe I will always belong to God, with God and by God. I thank God for the chance—short- or long-term—to be interdependent among my various groups, sharing common aspirations and giving and receiving support. This interdependence has raised my happiness and quality of life throughout my life. Yet, truthfully, all of these worldly groups that I belong to provide the illusion that I am separate, that I have my own identity, and that this "Self" needs to be accepted, attached to or part of something else. Spiritually, if I believe in the concept of Oneness, where I am connected to everything and everything is connected to me, I don't need to belong to anything. I am hoping my beliefs will carry me through whatever losses I experience ahead. It feels better to share my beliefs with others, even in this article, thus "belonging" to the *CF Roundtable* community still brings me fulfillment. So maybe I can carry both the joy in belonging to groups with common interests, but also carry the joy that I need not belong to anything because I already belong to everything.

In summary, I hope you, too, can do a mental and emotional inventory of your places of belonging. How would you rate the value of belonging to a CF community? Do you want more or less of it? What spiritual medicine do you take, to help you belong? ▲

Isabel is 43 and has CF. She lives in Redwood City, CA. She works as a bereavement counselor. You may contact her at: Isabear27@hotmail.com

COUGHING WITH A SMILE...

Emergency Exits Are To The Right And Left – Ding!

By Jennifer Hale

I have settled into my seat. It is tight squeeze, but I am little so I can clear it. I wipe down anything that I would touch and get ready for the long plane ride to my destination. As we land I am anxious to get off the plane and I can see the gate on the horizon. I am almost there! I can see it! I am so excited! I actually have a lot of emotions whirling around inside of me and these feelings make themselves known at all hours of the day. Wait, the plane has stopped but why!? I can see the gate we are just about there, just a few more feet to go! Then the flight attendant comes on the speaker and says there has been a delay. Please be patient and we will taxi to the gate as soon as we can and you will be on your way to your next destination. Well, that is frustrating. I can see where I need to go, but I have to wait patiently to get there. Ding, goes the sound telling me I can take my seatbelt off, but I cannot get off the plane, the gate is not ready for me yet. So I will wait in my seat as patiently as I can and dream of what is on the other side of that gate...

As I have been waiting to get the call for new lungs, I thought how similar it is to waiting on the tarmac when stuck in a plane. How similar it is to see the gate (my new lungs) and to know it is right there and you can walk there faster than it would take for this airplane to get the all-clear to taxi to the gate. Waiting for the call to come for new lungs, waiting for life to begin again, waiting for a second chance, waiting and waiting...

So you ask me or wonder how is it I wait? How is it I go on in a state of health that is so very hard to endure? A simple shower leaves me out of breath, and let's not even talk about stairs. Stairs are my nemeses. I am so out of breath from walking up stairs, it is exhausting. I am so tired of being out of breath. But this is my cross to bear, this is my life right now and I will endure until that phone rings with the miracle of more life. Until my "gate" is ready, I will wait in a holding pattern for the phone to ring.

> *"Waiting for the call to come for new lungs, waiting for life to begin again, waiting for a second chance, waiting and waiting..."*

JENNIFER HALE

Some of the things that I do to endure this waiting period are to get out to the gym, stay social with my friends, keep smiling and laughing, think positively and continue to feel God changing me so I can endure what is coming my way! Don't get me wrong, I have my moments. They come at night, usually, when I am getting ready to try to go sleep. Or moments when I am at the gym working out and very much in my own world with my ear buds in and the sounds of music and oxygen flowing into my nose. It is not easy to keep up with life and activities when all you want to do is rest on the couch, when you are tired of coughing and gasping for air. It is quite amazing how hard life can be when you cannot breathe.

Getting out to the gym has been a real challenge for me. It is just so hard to work out when you cannot breathe. I am also very hard on myself and feel like what I do at the gym is pathetic compared to how I used to work out. But I am not what I used to be. I am a 43-year-old, with 28% lung function, on the transplant list. Even being in the gym is an accomplishment. So with that I do the best that I can when I am there, which includes lifting weights and walking on the treadmill. I am preparing my body for the fight of my life. I am in training! Eye of the tiger! For those of you in my same position just do the best that you can. If that means you got out of bed today and did 12 bicep curls with a can of corn, then so be it! Do not judge yourself on what you can't do, reward yourself on what you can do

today. I say today because each day is different. I can feel good one day and not so much the next. So go with the flow and do what you can. Every little bit matters and is a good job well done!

Maintaining my relationships with my friends has been very important to me. It really makes me feel good to be around them even though I am not feeling well. Even though after being with them I get in the car and cough out what seems like a gallon of brown glue. But they put a smile on my face and heart and I love them all dearly! Also, my dear friends whom I unfortunately do not see in person but talk with on the phone and text with – I love them dearly! You know who you all are, wink, wink. It is important to maintain those relationships because being alone in your house all day, alone with your thoughts all day, can create a lot of tunnel vision and depression. If you cannot make it out, have your friends come to you. I am very blessed that my friends plan their time around how I feel. I call the shots as to what we are doing, based on how I feel. I am a lucky, lucky girl.

Thinking positively is another aspect of coping with waiting for the phone to ring. I really have to give my husband props for that. He is so positive and supportive that he helps me to be positive. Things I do to stay positive are read positive quotes, watch movies that I love, stay connected with my friends and think about all the awesome things I am going to be able to do when I get my new lungs. It has also helped to talk to some post-transplant people who are doing well, have had good experiences and are positive fighters themselves. I am not saying that I am sticking my head in the sand by talking only with people who have had a positive experience; but I have chosen to be picky about whom I speak to about the transplant. I know things can go wrong, but everyone is different and I choose to speak to like-minded individuals who are positive about their lives and how they approach their disease in general. My husband, as I have stated before, is a real positive light for me. I like to say I am optimistic with a pinch of pessimism. But my hubby looks at the bright side of all problems or issues and that attitude has helped me tremendously. Sometimes the best way to describe a situation is to just say, "You know what, it is what it is." That quote I use A LOT!

Lastly, I really feel God is preparing me for this journey. When I was first told I needed a transplant it was a very hard reality to accept. Yes, I was on oxygen, and yes, my lung function was low, but I felt I could live this way. In reality, at age 43, living on 24-hour oxygen, not being able to walk up the stairs or take a shower without being overly out of breath even with 0_2 and many, many more hardships is just not normal. I think we as CF people tend to really accept our limitations and live with them and build a life around them. Our "normal" really is not normal at all. So I have found it hard to realize my normal is far from it. God has shown in me this acceptance of needing a lung transplant. He has helped me come to terms and be able to say, "Okay, I am ready. Let the phone ring and let's do this," because I have a lot more living to do! He has brought people into my life in person, via e-mail and via Facebook who have shown me such support and prayers that I get tears in my eyes just thinking of it all. God is by my side and preparing my path – a path to greatness and freedom: freedom from being trapped in my body like I am right now, freedom to do good while I have this time on earth, freedom to be with my loved ones, freedom to just make dinner at night or grocery shop, freedom for the little things in life.

Heading to transplant is a journey. It requires patience, stamina, courage, support, laughter and love. I look forward to getting back to the simple things in life that we take for granted. No more being out of breath taking a shower, going up a flight of stairs and walking around in the supermarket. The simple things like making dinner and having the energy to do more than one activity during the day and have it even last more than two hours. I look forward to being able to sleep. The little things I look forward to and I will experience all the big things in life too!

I am waiting on the tarmac, I am waiting for the call. I can see it. I can visualize my recovery and fight to get back to what will be the new me! The gate or I should say my lungs are right there and I can just see it. But I will be patient. I will settle into my life that I have now and I will wait for the miraculous phone call. Ding! ▲

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

In Memory

Daniel Alexander, 48
Bismarck, ND
Died June 17, 2015

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

WELLNESS

Sleep 101

By Julie Desch

I'm fortunate enough to set my own work hours, so incorporating health-related activities into my work life is not an issue. One thing that has become an issue of late, however, is incorporating *sleep* into my life. For that reason, I'm dedicating this issue of the "Wellness" column to the importance of sleep.

Since we biologically need to spend roughly one-third of our time as human beings asleep, logic dictates that sleep must be vitally important. Until recently, what exactly happens during those seven to nine hours of daily sleep was largely unknown. While much remains a mystery, we now have evidence that a healthy immune system requires sleep, that effective learning and storage of memory is related to sleep and that the cells of our body, in effect, "take out the garbage" during sleep. We also know that if a person can't or doesn't sleep, they die.

Clearly, living well with CF requires a robust immune system to deal with all of those bacterial invaders in our lungs. But there is most definitely a fine balance between the ability to adequately kill bugs and venturing into a "hyper-inflammatory" state, which carries with it many problems (i.e., irreversible lung damage). It seems that lack of adequate sleep (what is "adequate" varies from person to person, but with CF it is a safe bet to assume that more is better) affects immune function in a variety of ways. First, circulating immune cells including white blood cells and macrophages release chemicals called cytokines. These cytokines carry messages to other immune cells, telling them what to do. In sleep-deprived people, pro-inflammatory cytokines are released and there is a decrease of protective cytokines. This imbalance of cytokines is reflected by elevated C-reactive protein—a marker of excessive inflammation—in individuals who do not get adequate sleep. The last thing we need is a more pro-inflammatory state than we already have!

In addition, one category of immune cells, called T-cells, are diminished when sleep is not sufficient. T-cells are critical to a healthy immune response to infection, both bacterial and viral. In fact, studies have shown that sleep-deprived individuals do not mount a robust immune response to immunizations as a result of this T-cell decrease. We want our flu vaccines to work, so don't mess with your sleep!

It has long been known that sleep has something to do with learning and memory, but exact mechanisms were unknown. Recently, it has been discovered that during deep, slow-wave sleep, the brain replays over and over again activities from that day and in doing so, new connections are formed between neurons in the brain. This, in effect, "hard-wires" the task, making it easier to perform or recall. This is why a good night of sleep before an exam is much more beneficial than an all-night cram session.

A more recent finding is that physiologic changes actually occur during sleep that allow the brain cells to "take out their garbage," or flush out toxins and damaged proteins. Scientists have very recently discovered that the brain actually has a lymphatic system. Throughout the body, lymphatic channels serve as a plumbing system, carrying fluid called lymph that contains immune cells as well as toxins and other waste from the cells. This systemic lymphatic system has been understood for decades and includes solid tissues such as lymph nodes, tonsils and the spleen. Lymphatic channels in the brain were unheard of until a couple of years ago, when they were discovered in mice. It seems that during sleep, cells of the brain shrink, which increases the space between neurons allowing more fluid to be pumped in and toxins washed away. Removing damaged proteins from the brain is likely very important, as diseases such as Alzheimer's and Parkinson's are associated with damaged protein build-up. I see those eyeballs roll. "Why should I care about Alzheimer's?" Well, with the new drugs in the pipeline, it's

> *"All right, so sleep is important. What do you do when you can't sleep?"*

JULIE DESCH, MD

looking like diseases normally occurring in older populations could be coming our way soon! Hence, my menopause-related insomnia.

All right, so sleep is important. What do you do when you can't sleep? When I hit menopause, sleep went from the easiest thing in the world for me to do to something that I merely longed for routinely at 2 a.m. while my monkey mind chattered on and on. It remains a problem, so I've done some research.

There is a lot written recently about sleep "hygiene." I'll summarize:

1. TURN OFF THE SCREENS: This is probably the most important thing and is, naturally, the hardest to do! Most sources say to quit staring at your phone, tablet, computer, television etc., for at least two hours before you go to bed. The reason is that the light emitted from screens messes with your pineal gland that lives in the middle of your brain and is responsible for melatonin production. Melatonin is your body's natural sleeping pill. You can take it as a supplement at night, and I've found that it works in the short term but then after a few weeks the effect wears off. So, I have begun the "no screen light" approach recently (but only for one hour because, who are we kidding?), and I really do think it helps. I will either listen to a podcast or read a real, hold-in-my-hands book (no Kindles allowed either) for an hour or so before I try to sleep.

2. Have a regular routine: This is another biggie according to the sleep experts. Just like a bedtime routine is important for a small child, it is also important for us adults as well. Come up with a few things that generally are relaxing for you. A cup of warm milk or tea (decaf), followed by a bath or shower maybe, followed by dog snuggles, and then reading...or something like that. Then, do it every night, in the same order, without fail, before you try to sleep. Your body becomes conditioned, just like Pavlov's dogs, to think that this series of events means that sleep is near, and it responds.

3. Dark, cool room: This should speak for itself, but I'll elaborate a bit. Just like screen lighting messes with your pineal gland, any light does the same thing. So cover the LED lights from the cable box and the alarm clock. If you have a window in the bedroom, get blackout shades. Aim for pitch black. As for temperature, I sleep hot, and though I'm not sure if a placebo-controlled double blind study has confirmed this, I have heard that this is true for many with CF. What I do know is that the best sleep year of my life was during college, when I slept in an open-air dorm in Nebraska. Brrrrrrr! Electric blankets were a must, and I would sometimes wake up with my little nose hairs frozen. But I was extremely well-rested, not a common occurrence for a college student. Now, I just turn a fan on and point it at the bed.

4. Avoid caffeine, nicotine (duh) and alcohol too close to bedtime. Caffeine is in a lot of things! So although it may be a no-brainer to avoid coffee, most soda is also a no-no, as is chocolate. I learned this the hard way a few nights ago when my bedtime snack was chocolate pudding! And while alcohol may be relaxing in the short term, it definitely disturbs your sleep pattern later in the night. So that evening glass of wine may help you go to sleep, but when you wake up at 2 or 3 a.m. and can't fall back to sleep, alcohol is the culprit. I know, I am sorry, too.

5. Exercise: There are mixed reports about whether you should exercise in the evening. Some say absolutely not, that it revs up your body too much and makes it harder to come down. Other sources say high-intensity exercise is good to do at night because the relaxation that happens when you are finished (and remarkably, still alive) is a sleep tonic. It seems to be a personal choice and you should experiment. Certainly, low intensity exercise like gentle yoga would not be a problem. But *all* sources agree that regular exercise is most certainly beneficial for someone with insomnia. As you might expect, I concur.

If these tricks don't work for you and you are still having insomnia, you might want to talk with your doctor. It could be that you are desaturating during sleep, when breathing naturally slows down, and that supplemental oxygen while asleep could make a huge difference.

Finally, let's discuss the elephant in the room. If there is an elephant in your room, you probably won't sleep because you will want to play with it. But I digress. We cough. Any coughing sucks when you are trying to get some rest. When my cough is at the point that it disturbs my sleep, first, I am probably sick and I should call my doctor. Second, I will take a cough syrup that contains codeine (or a similar narcotic). This has two effects. It (1) shuts down the cough reflex, which in general is not a good idea if you have CF, but let's face it, you need to sleep, and (2) puts you to sleep because, hey, that's what narcotics do. I will always follow this cough syrup with a Miralax chaser though, because narcotics not only shut down the cough reflex, they also shut down the colon. And the last thing you need to contend with when you have insomnia is the pain of being FOS (med-speak for "full of sh*$").

I hope these tips help if/when the sleep monster decides to skip your house. If all else fails, there is nothing better than an afternoon nap, my favorite activity of all time. ▲

Julie is 54 and is a physician who has CF. You may contact her at: jdesch@usacfa.org.

FOCUS TOPIC

INCORPORATING WORK INTO OUR CF CARE

Work Matters

By Chris Kvam

Work is centrally important to my life and, for me, is an integral part of living with CF. I chose to write this quarter because of this importance but do so hesitantly. I am acutely aware of how many people with CF are unable to work due to physical disability caused by CF. I am very aware of how many people with CF strove to work in a career of their choice but were unable to maintain their health while doing so and had to make painful decisions of putting their long-term health needs above their work goals or had to make other compromises.

I also know how many barriers stand in the way of meaningful work for so many of us. Employment discrimination on the basis of disability (real or perceived), wrongful terminations related to health insurance costs, and inflexible employers unwilling to accommodate hospitalizations, workplace treatments or regular clinic appointments exist, and have hugely negative impacts on people with CF and all people with disabilities, chronic illness or complicated medical needs. (As an aside, never talk about CF in job interviews!) That being said, work remains a critical piece of adult life for many people with CF, and often, our wellness hinges on it.

I'm also writing because I fear that too often work is an ignored topic, considered either nonessential or optional to CF care teams and families of people with CF. This translates into people with CF growing up not appreciating the huge costs associated with not pursuing a work life. Many people with CF qualify for disability benefits as children. This is a wonderful thing – it provides needed support to families dealing with catastrophic costs of raising a child with CF, and may provide a path to necessary medical insurance not otherwise available.

However, the label of "disabled" should not be underestimated. Do children on disability take that label to heart? Do they interpret it to mean that they cannot pursue their dreams, work or should not try? I don't know the answer, but there are too many stories of people with CF who find they do not meet stricter adult eligibility criteria for continued disability benefits, who chose to try to do everything they could to maintain a relatively small disability check and did not build a résumé, didn't try to go to college or ever contemplate a life with CF that involved working. This limits their choices when they find themselves suddenly forced to find a job, limits their ability to plan and prepare for future health crises and takes a toll on physical and emotional well-being.

I don't incorporate work into my CF care. Instead, I incorporate my CF care into all facets of my life, including work. I think this is an important distinction. Who I am as a person is far more defined by my career choice, abilities and academic accomplishments than by my CF diagnosis. My work life is an extremely high priority, not secondary. CF has given me the drive to maximize my potential and abilities while I have them and working is important to me. The personal satisfaction I derive from doing my job well and ongoing ability to work are part of what drive my adherence to treatment. Meaningful work is centrally important to my mindset, positive attitude, independence and ability to maintain my health.

CHRIS AND CHRISTINE KVAM AND THEIR DOG, HILDY.

There is a difference between a job and a career. Jobs are taken to meet immediate needs and pay bills. Careers bring long-term stability and personal fulfillment and are enabling. I am an assistant district attorney, which is both

incredibly rewarding and demanding. I didn't just end up working as a prosecutor. Many intentional choices were made by me and my parents that led me to this position and gave me the opportunity to have this amazing career.

During my time as an undergraduate, it dawned on me that I was in fact going to graduate, death was not imminent and ultimately I would need to find a job. Exhilarating! Terrifying! In the era before the Affordable Care Act, the prospect of losing health insurance loomed incredibly large. Pre-existing condition clauses still existed and my ability to stay on my parents' insurance was limited to being a full-time student. This reality drove me to pursue a graduate degree immediately. I believed that I would have the best chance at developing a career if I had a graduate degree and knew that my time to obtain one was limited by how long I could maintain insurance. I chose a field that interested me and applied. I graduated.

My first job search after graduate school was incredibly difficult. I was paying COBRA to extend my health insurance and running out of the time and money necessary to keep it. The dire reality of my situation taught me several things. First, I needed a full-time job. Temping or part-time work were not options, given my absolute need to find a job with health benefits. This narrowed the scope of my search and focused my efforts.

Full-time "white collar" work, though demanding, often has a more predictable schedule than part-time work where shifts may constantly change. It is not as physically taxing as other work, and employers are generally more invested in their workforce and therefore more willing to accommodate health needs. When I found that first job out of graduate school, I found all of this to be true, which motivated and enabled me to develop and adhere to a strict treatment schedule – that ultimately happened "automatically" – in order get to work on time every day and minimize my need for disruptive hospitalizations.

To those thinking about their future work life, I would encourage you to think about what kind of work you not only want to do, but what you reasonably expect to maintain. I would have loved to be a park ranger in Yosemite. This is not reasonable given the circumstances of my life. I don't mourn that, it's just a fact. Fortunately, I have many other interests and abilities and have been able to apply them to meaningful work that I enjoy. And that enables occasional excursions into the wild.

Benefits matter. My salary is not impressive, but it is adequate and it enables me to meet my basic needs, eat healthy food and maintain a lifestyle focused on wellness. I work for state government and I don't get stock options. However, as a full-time employee, I'm entitled to all the benefits my colleagues receive, regardless of my CF diagnosis, including health insurance. Unfortunately, access to high-quality healthcare benefits in the United States remains closely tied to full-time employment. But, health insurance is only one of several benefits to which people with CF need access.

Short- and long-term disability insurance, life insurance, 401k and retirement benefits are also very important. I have no idea how long I will live (and neither does anybody else). However, it is perhaps more foreseeable that I will become disabled and have a shortened life-span due to having CF. These facts make disability and life insurance and retirement savings plans critically important to my ability to meet basic needs should I become disabled and protect the financial stability of my wife and family. Given the higher likelihood of needing to utilize these benefits, I care about them far more than most of my young professional colleagues. My only route to accessing these benefits was through automatic enrollment from my employer. Believe it or not, life insurance companies aren't excited about insuring people with CF.

Understandably, a lot of people with CF have a hard time with long-term financial planning and thinking about the future, and as a result they don't consider these longer-term needs. I strongly urge adults with CF to recognize the importance of these issues and encourage CF clinics to do more to educate people with CF about the importance of financial planning and the positive impact these benefits can have on their quality of life, financial stability and overall wellness.

I didn't set out to be a prosecutor. After a few years of working in Boston, my wife and I made the decision to relocate and part of that decision involved me going to law school. During law school I discovered my interest in criminal law and was unbelievably fortunate to have been selected to do this work. My initial decision to attend law school was driven by my desire to broaden my qualifications and career choices.

Life with CF is full of compromises. Law school wasn't easy, and I didn't expect it to be. When I look at my PFT history, I can see its impact. Looking back, I was willing to pay that price. I work in a demanding job, which also risks negatively impacting my health. If there comes a time where I'm unable to meet the demands of being an assistant district attorney, it is my hope that my law degree and work experience give me many job options I would otherwise not have. There are tradeoffs to every choice we make and that reality is especially true for people with CF. I try to be very mindful and intentional when making these decisions and liv-

Continued on page 18

The Misconceptions Of Not Working Because Of CF

By Amy Braid

When I was 29 years old I made the decision to stop working and focus on my health. Up until that point I had been an architectural designer at a relatively small architecture firm outside of Philadelphia. I loved my job. I was one of the small minority who actually enjoyed going to work every day. I got up each morning and did an hour of treatments before hopping in the shower, eating breakfast and racing off to spend 12 hours at the office.

I look back on those years and I smile at the designs I created and the friends I made. But working took a toll on my health, even with a reduced work week (four days a week with a day to rest in the middle), and I had a decision to make. There were two options: going to work part time, but I would lose my benefits and a large part of my salary, or I could take the leap and go out on disability and apply for Social Security Disability Insurance (SSDI). I chose the latter. It was not something I wanted to do, but it was something I needed to do in order to try to stabilize my declining health. While I do not regret my decision entirely, it is a struggle each day to remind myself why I no longer work.

Young people who cannot work often struggle with misconceptions and stereotypes about disability, and this seems especially true for young women who do not have young children at home. Looking at me you would assume that I am healthy and capable of working so I must be either lazy or married to a wealthy man. Neither is true, of course. Similarly, society dictates that women who do not work must maintain a spotless home; I call this the Donna Reed misconception. Since I stay at home all day my house should be spotless and each night I should be preparing a three-course gourmet meal for my family. Again, it's far from true. And, lastly, because body image is always in play, I should look like a supermodel since I can spend all day long working out at the gym. This one is only partially false.

"While I do not regret my decision entirely, it is a struggle each day to remind myself why I no longer work."

I do spend time at the gym, or did until I was able to secure a treadmill for my home, but it is not to look like a supermodel. Instead I am trying to keep my lungs in the best shape possible. These misconceptions, though not usually voiced, make being out on disability a struggle.

Another common misconception is that not working is glamorous and exciting. Many people make comments like "it must be great not having to work," and "you are so lucky you can stay home all day and do nothing." While the idea of not working does sound exciting, it is far from the truth. I admit that the idea of not working appealed to me, when I was trying to make the decision whether to stop. I made those comments to myself, and I look back now and laugh at my ignorance.

For the first month or even year of not working, there was a sort of vacation feel to my life. There was no routine to my day and no alarm clock jolting me out of bed at 5:30 a.m. to do breathing treatments. I did not worry about making it to work on time, if there was an accident on the turnpike. And I was saving money on work clothes. Who needed $100 dress slacks and $50 pumps when I didn't even need to get out of my pajamas?

Then reality and boredom set in. I found myself sitting around not knowing what to do with myself. There were only so many television shows to watch, or books to read, or items to crochet before I got tired of doing the same thing day after day and week after week. My pajamas and comfy clothes became my office attire. I would shower every day (okay, almost every day) but still put those pjs right back on if I was not leaving my house. I invested in yoga pants and stopped wearing make-up. And some days the only person I would talk to out loud until my husband came home was my dog. There was no routine, no need to do anything. I became what I didn't want to become...lazy. I became the misconception.

The biggest misconception of all is that people think those who cannot work chose to not work. What people do not understand is that we are unable to work without putting our health at serious risk. It is coping with the idea that we are unable to work at such a young age and the constant desire to return to work that makes this part of the journey so difficult. We lose a lot that people who do work take for granted, for instance, the social aspect

of a job. Having co-workers to talk to every day, making work friends and going to work functions are highlights of the job. Making plans with the people you see 40+ hours a week for the upcoming weekend or holiday makes up for spending so many hours in the office. But when you stop working, you lose all those things. And for me, I lost the old colleague connections as well when I deleted most of them from Facebook. I watched as they posted their promotions and new jobs on social media and it depressed me. They advanced in their careers reminding me of the stagnation in mine. To see my old colleagues move on and move up while I sat idle physically hurt me; a little piece of my heart broke each time I would read a status update or see a picture posted. They reminded me each day that I missed working. Even today, I miss socializing with people who are not family. I miss having a reason to get dressed up every day. I miss leaving my house and having an office to go to. I shake my head at the people I hear complaining about working, about their bosses and co-workers and I think how lucky they are. How they are out of the house doing something every single day, earning money doing something they (hopefully) love. Furthermore, I miss being a contributing member of society. Because I was raised to earn my living, not being able to feels like failure.

Fast forward to the present. I still deal with these misconceptions and I still get comments about how great it must be not to work. But instead of arguing or rolling my eyes, I just say, "Oh, it has its perks," and move on. There is a routine to my life now, somewhat at least. Maintaining my health is still top priority and I make sure that I fit exercise in a few days each week. My crafting has expanded and each day you can find me either sewing or crocheting something for myself or a loved one. My husband and step-daughter have benefited from my new found joy in baking and even cooking more glamorous meals. My house may not be spotless, but in these "retirement years" of mine I have embraced the Donna Reed misconception and made it my own. I became the housewife that I always loathed. But I enjoy it. I get pleasure from a clean house and folded laundry. I enjoy making curtains for the kitchen or a new dress for church. Now I feel like I get something done each day.

> "Another common misconception is that not working is glamorous and exciting."

Furthermore, to bide my time I went back to school and, in 2014, I earned my Master of Arts degree in history. What I plan to do with the degree is a mystery to me, but it was a goal I was able to cross off of my list – something I probably would not have been able to do had I continued to work. And you know what else I do each day? I put on mascara and real clothes. Even if I am not leaving the house, I put on something besides yoga pants and a tee shirt. Sure there are still days where pajamas are the only thing I wear, but now, it isn't every day. But the best part of all of this is my ability to visit my family over 300 miles away whenever I choose to. I don't have to worry about missing work, taking vacation time or forgetting deadlines. I can visit my niece and nephews any time I feel like it. And that makes my retirement totally worth it for me.

While not working can be stressful and a struggle, it doesn't have to be. Each one of us who has to deal with the decision to stop working or who listens to the misconceptions regularly has a choice to make. We can sit back and let our situation depress us. We can become the misconception, like I did. Or we can do something about it. We can make good use of our time. And that does not mean you have to expand your crafting like I did or embrace your inner Donna Reed. It means doing something that makes you feel good about yourself. If that means you want to read a book a week, then do it. Or if you like to volunteer and can do only two hours a week, find a place to do so. Anything that gives you a reason to get out of bed is not only good for your emotional health, but it will help you physically as well. So get up, get moving and do whatever it is your body can handle. And if the only thing you can do each day is put mascara on, then so be it. If it makes you happy and feeling normal, then it is worth it. ▲

Amy is 34 and has CF. She lives outside of Boston with her husband and two step-children. Amy was diagnosed at six years of age and has learned to take the uncertainties of CF in stride. She earned her B.S. in interior design and her M.A. in history and has a passion for both. Although unable to work, she concentrates on maintaining her health and expanding her crafting skills, while embracing the loving housewife role. She also has her own blog about life with CF, Mastering the Art of Breathing (www.mycfjourney.blogspot.com). You may contact her at amysilcox80@gmail.com.

Did I Get Paid To Have A Bowel Obstruction?

By Devin Wakefield

Hi everyone! I'm new to writing for *CF Roundtable*, so here goes... Before sitting down to write, I just had 17 grams of Miralax with about eight ounces of Gatorade. It's been my usual three-times-a-day cocktail ever since a several-month-long recovery from an awful bowel obstruction. That's a different and much longer story than what I want to tell you about today, but it's an important backstory.

I was a preteen, and my bowels obstructed while I played miniature golf. Don't eat golf balls, kids! (Just kidding.) We don't really know why I got it–it could have been that I forgot enzymes, it could have been the road trip and lack of bathroom availability and holding it in, it could have been CF's sadism that decided it would be fun. Right away I knew something was wrong; I had terrible pain in my abdomen that clenched continually every short while. I needed surgery, got blocked up again (what?), returned to the hospital, had several enemas, blocked up again (what the f&%#?!), more enemas and repeat (holy f&%#ing s%@#!) a few times until my lungs started to tank as well. (Can you expect someone to cough when their intestines are in full riot? The pain is unforgivable.)

I started to die; my upper-GI system just couldn't absorb food anymore; I was skinnier than I had ever been. We decided I had one chance left: I went home with an NG-tube taking Pregestimil (predigested food) by day and GoLytely by night. (I like to think I had uncovered my latent super-powers: the s%@#-sprayer! Splish-splash to the rescue, anyone?) Orally, I had only clear liquids. With luck I could still absorb some of the Pregestimil's nutrients, and if the GoLytely couldn't keep my bowels moving, nothing could. I gained a pound, and we celebrated with lots of Jell-O (the closest to solids I could get). I survived! I weaned myself back on food, and I continue to live, in large part thanks to the cocktail I mentioned earlier: Miralax and Gatorade. It kept me poo-ing when I went back on food. It saved my life, and it continues to work wonders to this day.

One particular wonder happened while I worked as a Quality Assurance Intern (that means I looked for bugs in other people's software code and then tattled on them). I had a bowel obstruction at work. Now, this wasn't going to be as awful as before, but these things are never fun either. It started as a slight discomfort, somewhere in my lower belly around the scar I have from my second bowel surgery (I had meconium ileus as a kid–basically a bowel obstruction at birth–and had had surgery then, too, so the scar has actually been opened twice). I forget which side I hurt on, but it wasn't the full-on horrible pain of a total bowel obstruction, not if I could help it. Luckily, it was lunchtime, so I drove home and took two doses of Miralax and some water (Miralax draws water in from your intestines to pack a big punch, so you can get dehydrated fast–be careful!). I shouldn't have had lunch, but I think I did, thinking it was all over. It wasn't, but I did think ahead and brought an extra double dose of Miralax to work just in case.

DEVIN WAKEFIELD

Back at work, I did my midday aerosol in the room where people who just gave birth pump milk out of their breasts while at work. I always thought it interesting I used it for a similar purpose, except replace milk with sputum and breasts with lungs. This room happened to have a bean bag chair in it. I laid myself down on it in different positions in a hope that it would help position my intestines in ways that would loosen the crap my intestines selfishly cling onto for some nefarious reason–or maybe my poop just loved me that strongly. Never mess with the power of love, kids! I think the bean bag chair helped, at least my pain lessened and I heard lots of gurgles. The gurgles let you know things are moving. Anyway, enough rolling around on the bean bag chair. Time to get back to work!

For whatever reason, probably the lack of total agonizing pain and my ability to say, "Oh, I'm not crying in the fetal position, I mustn't be all THAT sick," it never occurred to me to say to my boss, "My stomach is not feel-

ing well, I need to go home." It would have taken how many seconds and how much self-respect to say that? I guess I got a bigger paycheck by staying. Yay, money to pay for the extra Gatorade and Miralax I used that day? Please don't do that, dear reader, please go home. Learn from my misguided and overbearing hope.

It turns out I didn't have pain for the rest of the day, until the last hour or so of work. I had some mild cramping around then in my left side, and I wanted to go try out the bean bag chair again. I took the extra Miralax I had brought and drank some more water. I gritted my teeth and got through the rest of the work hour, which wasn't too bad, and then drove home. At home, I continued what I did at lunch: more Miralax and water, and rolling around. Lying on my left side, I felt the least amount of pain, and rolling and stretching did seem to help get the stuck bits of poo through. At last, I ran to the bathroom and let out a big, splashy butt-pee. That's Miralax for you! It can really get through to the other side. I returned to work the next day pain- and discomfort-free, but still a little squirty down there and my nerves certainly had a good rattle. I must've missed a few doses or something the day before my saga. I hope never to be paid to have a bowel obstruction again, whether directly or indirectly. In the end, though, I got through it! Or, should I say it got through me? ▲

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

KVAM continued from page 13

ing with them. I'm incredibly fortunate to be living my "Plan A." It's comforting to me and reduces stress and anxiety to know that I have a lot of flexible "Plan B" options. As with many things, hoping and working for the best, while preparing for the worst, has enormous value. I don't stress about this part of my future and it doesn't cause me anxiety. I'm doing everything I can to maximize my abilities and potential now and am prepared for a future that may be uncertain.

Employers make a difference. I have been fortunate to have found employers who have been understanding and accommodating of my health needs. Some of this is the good fortune of working for good people. Some of this is my effort to minimize the impact of CF on my job performance and colleagues. It is also comforting to know that, with my work experience and education, I am not trapped in any one particular job. If my employer was not understanding or if I felt having CF was seen as a negative instead of a positive in my work environment, I would leave. I have worked very hard to be measured by my successes at doing my job, independent of my CF diagnosis, and make significant effort to minimize disruptions in my work due to CF. Not every day is a good day, but severe exacerbations aside, in my view, I'm going to feel bad at home or at work, so I might as well deal with it at work.

There is no shame in applying for and receiving disability benefits. However, understanding disability criteria for both children and adults with CF, the critical implications of building at least some work history on whether or not one would qualify for Social Security Disability Insurance (SSDI) versus SSI are too important to ignore and should not require a BA in Economics, Master of Public Policy and law degree to understand. CF clinic social workers need to be educating people with CF and their families about these often harsh, unfair and arbitrary realities, which nonetheless will impose themselves on their lives if they are not understood and taken into careful consideration BEFORE they are needed. Increasingly, many people with CF who were previously disabled are now finding that new medications or successful transplants are changing their disability status. This is amazing, and people with CF should be cognizant of the ramifications of these changes on access to health benefits in a world where disability is considered binary—an all-or-nothing determination—and health insurance is tightly tied to eligibility or employment.

Work matters. Don't be afraid of preparing yourself for a career, be willing to take calculated risks and do your best to plan for the unknown. ▲

Chris is 35 and has CF. He lives in Rochester, NY, with his wife, Christine, and their dog. He is an assistant district attorney, a cyclist, a member of the CFF Mental Health Task Force and is interested in transitions throughout life with CF and living full with CF.

RUSSELL continued from page 1

wonderful support network.

In 1999 and 2001 we sponsored conferences for adults who have CF. The first one was held in Houston, Texas, and the second in Chicago, Illinois. Both were well received and a great way to share information and make important personal contacts. Sadly, due to the events of 9/11/2001 and new rules for protection from cross-infection, we no longer are able to produce conferences. The heightened security at airports makes it much more difficult for potential speakers to just "hop on" an airplane to get to a conference site, speak, get back on a plane and get home in a short amount of time. The new cross-infection guidelines make many physicians leery of having their patients participate in a conference situation. So, for now, we do not sponsor conferences. Perhaps that will change in the future.

When we began, we had no computers. Our president had a word processor on which he created the manuals and paperwork that we needed to get started. The CFF kindly purchased a computer for the editor, which facilitated production of the newsletter. A couple of years later, the CFF purchased a computer for the treasurer. I was very happy to be able to put all of our records onto that computer. It surely made my job much easier. Later, a subscriber purchased a FAX machine for us. That made it easier to communicate. We didn't start using e-mail until the latter part of the '90s. That really sped up things. At least we thought it was speedy. It would take about a half hour to download a photo! Times have changed.

I have been privileged to be along for the ride the whole 25 years. Our local support group of adults who have CF began doing the work to start USACFA in February of 1990. At the organizational meeting in September of that year, I was elected treasurer and I've been working on the newsletter ever since. I served as president for a few years and did another turn as treasurer. For the past few years, I have not been a director, but I have continued to volunteer as one of the editors of the newsletter. It has been time well spent.

I am eager to see what new ideas and changes the directors come up with in the coming years. I look forward to being around to enjoy the next many years of the U.S. Adult CF Association, Inc., and *CF Roundtable*. ▲

PHOTO BY MICHELLE COMPTON

Do Not Touch

We touch but only wearing gloves.
Sisters who share a disease, cystic fibrosis.
The special bond has brought us closer together.
But also farther apart.

We cannot touch for fear of making the other sick.
CF and bad bacteria make us vigilant and sometimes afraid.

But as in the past, CF encourages us to be creative.
We see it all as just one more challenge in our lives.
Like all the other challenges we meet it head on.

We can still feel the support,
concern and love we have for each other
even through gloves
even through distance
even through fear.

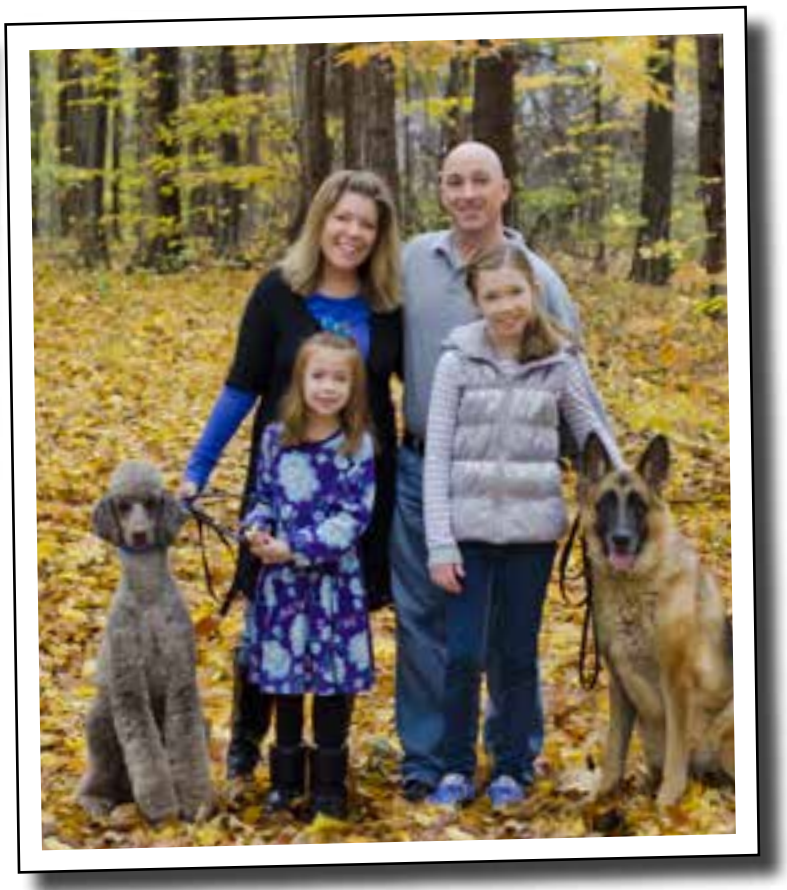
CF cannot take that away.

-Beth Sufian, 1999

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

FROM OUR FAMILY PHOTO ALBUM...

DEVIN WEARING A WETSUIT AS HE LOVES THE BEACH! HE ENJOYS BOOGIE-BOARDING AND WAVES TACKLING HIM.



THE GOVERNOR FAMILY: ANNE, GEOFF, PETEY THE POODLE, HANNAH (7), ALEXA (9) AND ADDY THE GERMAN SHEPHERD.

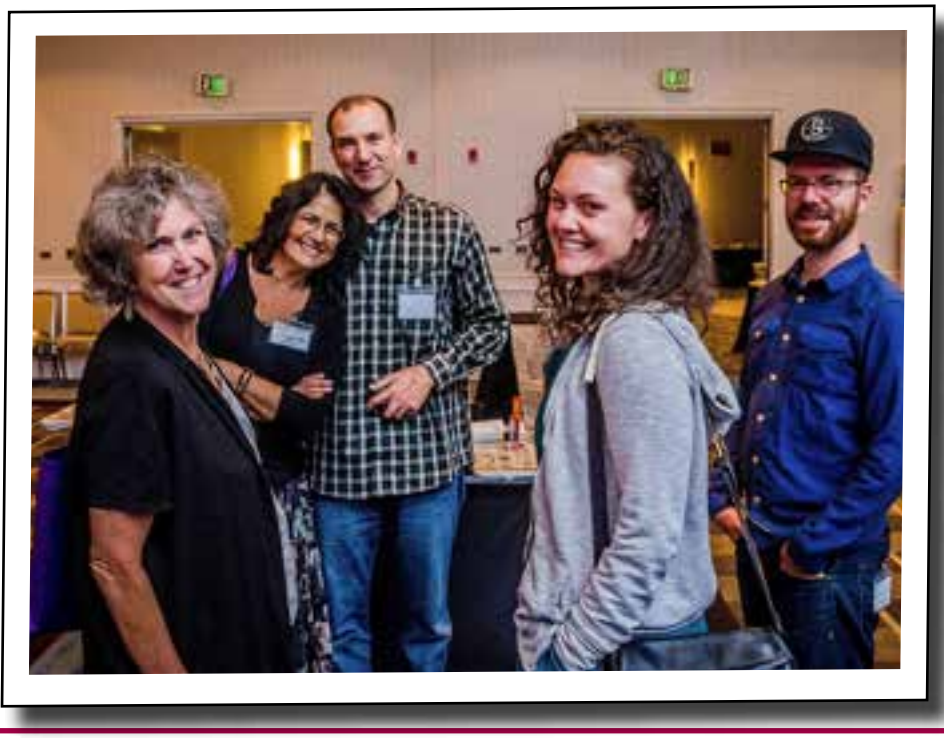
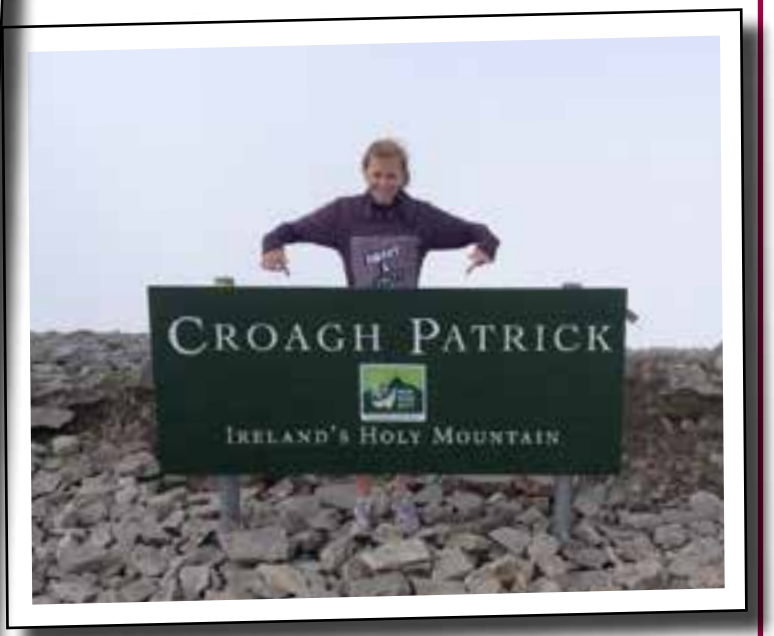


PHOTO BY CRAIG BURLEIGH

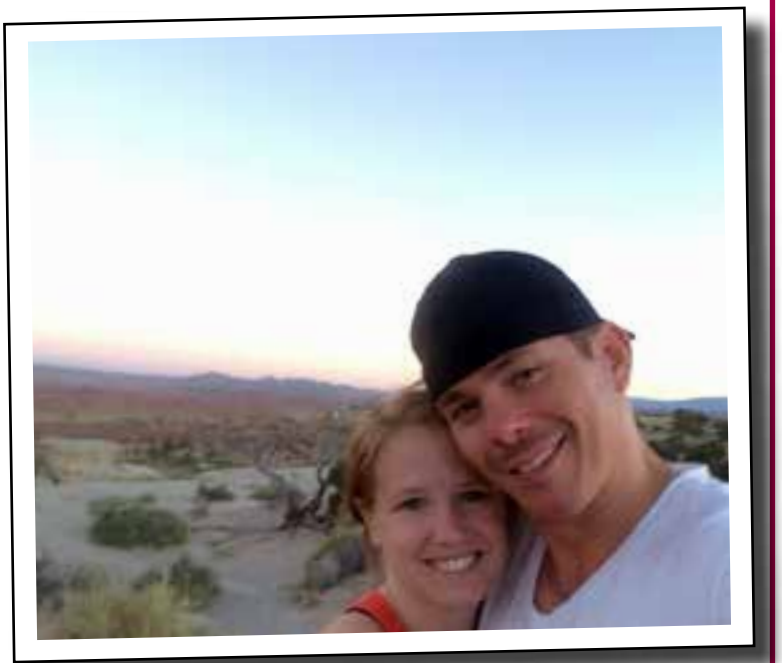
JOHN AND JEANIE HANLEY, LAURA MENTCH, HER DAUGHTER CARA BRODY AND JORDAN ALPORT, CFRI CONFERENCE IN AUGUST 2015

AMY BRAID



JILLIAN MCNULTY AT THE TOP OF CROAGH PATRICK, IRELAND'S HOLY MOUNTAIN. IT'S 2,600 FEET. SHE CLIMBED IT WITH HER BOYFRIEND BILLY AND A GROUP OF FRIENDS.

REID D'AMICO AT THE NASHVILLE ZOO.



KLYN ELSBURY AND JEFF SHELTON POSE FOR A SUNSET SELFIE.

IN THE SPOTLIGHT
With Jillian McNulty

By Andrea Eisenman and Jeanie Hanley

Our newest CF celebrity comes from Ireland, where CF is the most common genetic disease. Jillian comes across as full of energy with a beautiful smile, someone who seems up for anything. This was illustrated when she was chosen to run the Dublin City Marathon to raise money for CF, not having ever done anything close to that. She just started to train and went for it. It surprised her and especially her doctors.

She lives in a place called Longford. When she told me [Andrea], over our Skype conversation, she acted like I would never have heard of such a place. I had to laugh. I was there when I visited Ireland with my husband, who is of Irish descent. When we were there, he wanted to trace his family's roots and they had lived in that region, just west of Dublin. She was thrilled I had heard of it and we got to talking about where I had been on my visit. We talked about our love of good Irish tea and how the people there are so friendly and helpful. We both like the same brand of tea, and when she was visiting the U.S. for her testimonial on Orkambi, she sent me four boxes of it. Too nice!

While Ireland's median life expectancy is slightly lower, around the low-30s, Jillian stays well with a positive attitude and plenty of exercise. She is inspiring to talk to and quite full of life. I found her advocacy work to be inspiring and hope you do, too. Please welcome our newest star. Spotlight, please!

Age: 39

Diagnosed at birth? Why? My first brother, Derek, had CF. He died a few years before I was born, so all of us were automatically tested for CF.

Do you know the median age of survival in Ireland for those with CF? They are saying early 30s but I know so many who have died much, much younger here.

Where do you live? Longford, Ireland.

Do you have any siblings? Two deceased siblings and one still alive without CF. Of the deceased, one had CF and one was special needs.

JILLIAN MCNULTY SHOWS HER "JUST BREATHE" TATTOO.

What are your genetic mutations for CF? I am double Delta f508; it is the most common mutation in Ireland. Ireland also has the highest incidence of CF in the world.

Discuss your "Just Breathe" tattoo and why you got it. I was Facebook friends with the tattooist who did my tattoo. He had done a fundraising event for CF with all funds going to CF Ireland, this is the reason I had it done. It raised positive awareness plus I was forever inked!

I have two other tattoos, "PAL" with my brother Gary's (RIP) date of birth in between and two stars on my foot, one small, one bigger in red and yellow: for the two brothers I lost. One is in red, Gary's favorite color, the larger star, as he was 31 when he died. The other star is in yellow, Derek's favorite color. It is the smaller star, as he was only five and a half when he died.

Discuss your Facebook campaign. Growing up I never discussed my CF. When I started needing to be admitted to hospital at 31, I quickly saw the atrocious conditions for people with CF (PWCF) here in Ireland. We were placed on wards sharing a room with five other sick people, mostly old senile patients. It was then that I decided to get in touch with national media for coverage of this issue. There were many people campaigning throughout the years. Finally after almost 30 years of campaigning, it was agreed we would get a dedicated CF Unit in St. Vincent's Hospital, Ireland's center of excellence for CF. It is also the main CF center for the whole country. The new center was opened in August 2012. We now have the use of 32 single en-suite rooms for CF patients; something which years ago I thought never would be possible. It is such a relief to be able to just walk into a single en-suite room when you're sick instead of fighting for one like so many did for years. [Jillian's Facebook link is: https://www.facebook.com/JillIrl]

Did you set out to be a CF advo-

cate? I never set out to be a campaigner. I kind of "came out" as someone with CF on national radio, something that shocked a lot of people I had worked with for years and they never knew! Since 2009 I haven't looked back really. I am an active campaigner and fundraiser for CF care in Ireland. In the last three years I have raised approximately €15,000 for CF, splitting it between CF Ireland and St. Vincent's Hospital toward CF Care.

Have you experienced positive or negative feedback from being a CF advocate? I've received both. I caught a lot of negative for a couple of years by the same group of CFers that, for some reason, just took a dislike to me. They claim I am only out for myself, for self-praise, for "fame." AS IF!!! If I wanted to be famous I certainly wouldn't be using CF to do so! It doesn't matter what I do, the same group of what I can call only bullies continue to vilify me no matter what I do. But I know the truth; my family, boyfriend and friends know the truth so that's all that matters. I have also received a lot of positive feedback from people all over the world. These people keep me going when I'm ready to give up fundraising or campaigning.

Funny story about CF? Every time I think of this I just laugh! About a year before I started training for the Dublin City Marathon I had been quite sick and the physiotherapists in hospital were trying to build up my fitness level. One morning on rounds, my consultant, Dr. McKone, asked me if I had any questions etc. I said to him in all seriousness (OMG!!!) "I'm kinda worried about exercise." He said "why what's wrong?" I then continued to tell him (in my serious mood) that I was "Afraid the physios were going to give me a heart attack because they were trying to make me jog and I wasn't able to." I can still see his face as he started laughing, telling me they, "hadn't killed anyone yet." Then he began to tell me not to worry, it would be fine! A year later I turned round and told him that I was chosen to represent my county in a national competition and I was going to be Longford's Spartan and was going to do the Dublin Marathon. The poor man, his face was a picture! Here I was doing a complete 360!!!

Do you or did you work? I worked for 11 years in my local radio station, Shannonside Northern Sound. I was a receptionist and I produced a nighttime show for five years.

What do you do for exercise and do you enjoy it? I mostly run, walk and sometimes cycle. I love the thrill of running, mentally and physically it makes me feel so much better!

How has dating been for you? Mostly it was a nightmare. It was very hard to meet someone who would accept CF and its sometimes boring lifestyle!

Tell us about your current boyfriend. Billy is amazing. He's very supportive and understanding in every aspect of CF. He's still learning every day. We had known each other for three years on Facebook before we started going out. He's been there for me in ways I never thought any man would be. He's held a sick bag to my mouth after surgery when I was ill. He's always there when I'm in hospital. I'm very blessed!

What symptoms of CF do you have? I have sinus disease, CFRD, GI issues, hemoptysis, DIOS, and kidney impairment. For me, I find CFRD a nightmare to try to control. I love sweet things and my sugars are forever up and down. Hemoptysis is also the worst thing about CF. I've had major and minor bleeds. Now, most days, I cough up some amount of blood but don't worry unless it's more than an egg-cup full! It's always extremely frightening when I get that gurgling sensation, because I never know when it will stop.

What were your recent surgeries and why? I've had a double hysterectomy and in the last three years I've had two Nissen Fundoplications, extremely tough surgery to recover from!

Why did you need a Nissen Fundoplication? I had severe reflux. My pH readings were some of the worst readings my doctor had ever come across!

Why don't they do sinus surgery in Ireland? They don't do it unless they absolutely have to!

What is it like to not worry about insurance? In Ireland with free healthcare, do you have to wait for care? In most cases we are well looked after especially when we are in hospital. Sometimes we have to wait to see certain specialists and for some non-urgent procedures. If you are in hospital and something is urgent you are seen almost straight-away. I personally can't complain, I've always been looked after very well and in most cases waiting lists have been relatively quick. It's easy to see a CF specialist!

If you can discuss taking new Vertex combo drugs, how have you felt since being in the study? It's been amazing! Orkambi has made such a difference to my life. In fact I'm recently back from the U.S. where I testified for the FDA Panel Advisory Committee. Here is the link: https://www.youtube.com/watch?v=m1TOvZ-GCcw. This drug lessened my DIOS symptoms, gave me more energy and fewer stays in the hospital. Pre-Orkambi I would've been in hospital anywhere from 24-30 weeks a year. In the last year (I've been on the drug almost two years), I've spent just 11 weeks in hospital!

What do you worry about? My future. CF can be very scary. Deaths come so fast, so young. You just never know what can happen.

Are lung transplants being per-

Continued on page 24

MCNULTY continued from page 23

formed for people with CF in Ireland? Yes, we have a relatively new CF Transplant Centre in the Mater Hospital. In the last few years they have been very successful. And yes, as far as I know I will be eligible for one. Please God, that's a very long way off!

What gets you through difficult times? My faith, my family, my boyfriend and my friends.

What do you do for fun? I do like eating out! I don't drink nor do I go to pubs here. I prefer a quieter night. I love SHOPPING! I'm known for my love of the color pink and shopping!!!!

Where have you traveled? I've been to the U.S. three times, Spain, Canary Islands, France and UK. I hope to travel some more in the future. I've been to Lourdes six or seven times, now. I find it the most amazing place on earth. I'm always at peace there. There is just something about it. It's a pilgrimage holiday, a lot of masses to attend. But sitting at the grotto at any time of day or night, even the early hours, is just amazing. I can't even put it into words.

Any words of wisdom for others with CF? Positivity is of utmost importance. If you don't have a positive attitude you're going nowhere. CF thrives off negativity! Positivity and exercise kills CF. ▲

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 53 and is a physician who has CF. She is a Director of USACFA and is the President. Her contact information is on page 2.*

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

TILLMAN continued from page 5

minimizing systemic side effects.
http://tinyurl.com/oky9f66

Novoteris, LLC, Announces $2.8 Million Award from Cystic Fibrosis Foundation Therapeutics to Support Development of Inhaled Nitric Oxide Antimicrobial Therapy

Novoteris, LLC, announced a $2.8 million agreement with Cystic Fibrosis Foundation Therapeutics Inc. (CFFT) to develop an inhaled nitric oxide antimicrobial therapy for people with cystic fibrosis who have airway bacterial colonization. The Novoteris investigators' pilot trial in Europe of this therapy reported encouraging microbiological and lung function changes following two weeks of treatment in patients with CF. Gaseous nitric oxide's potent antimicrobial properties, lack of bacterial resistance, and its small molecule penetration capabilities could provide a promising alternative non-antibiotic approach to treating infections in people living with CF.
http://tinyurl.com/no4vuan
AND
http://tinyurl.com/qzbk6ck

Nivalis Therapeutics' Lead CF Drug Candidate Expected To Restore Hydration To Lungs And Other Organs

Nivalis Therapeutics, Inc.'s, lead product candidate, N91115, represents a new inhibitor of S-nitrosoglutathione reductase (GSNOR) for treatment of cystic fibrosis that could treat the underlying cause of the disease. N91115 is an investigational small molecule that addresses the CFTR defect. Nivalis states that N91115 is the only clinical stage product candidate they know of designed to stabilize CFTR inside the cell and at the cell surface, and that it has shown in preclinical studies that the stabilizing effect of N91115 increases and prolongs CFTR activity when added to other CFTR modulators. The company's clinical program initially targets CF patients who are homozygous of the F508del-CFTR mutation or the F508gdel mutation. Currently, Nivalis is conducting a Phase 1b clinical trial evaluating the safety of N91115 when used as a single CFTR modulator in that cohort of patients. Nivalis Therapeutics expects N91115 to not only increase CFTR function, but also restore the adequate hydration to critical organs. Other preclinical studies had already demonstrated the compound's efficiency in increasing the function of F508del-CFTR. Nivalis has also developed a broad portfolio of proprietary, investigational, small molecule inhibitors of S-nitrosoglutathione reductase (GSNOR), which they say has been shown in preclinical studies to significantly increase and prolong CFTR activity and to decrease inflammation.
http://tinyurl.com/o3drh4p

Spyryx receives award from Cystic Fibrosis Foundation to develop therapeutic peptides for CF

Spyryx Biosciences, Inc., announced that it has received an award from Cystic Fibrosis Foundation Therapeutics that will support development of Spyryx's therapeutic peptides for cystic fibrosis (CF). In the CF lung, the epithelial sodium channels (ENaC) hyperabsorb sodium and water from the surface of the lung airway, leading to dehydration of the mucus layers and reduced airway clearance. Spyryx is developing inhaled peptides, which are designed to potently degrade ENaC, blocking this absorption and returning airway fluid volumes to normal levels, with the goal of restoring proper mucociliary clearance. This therapeutic approach is independent of the genetic

Continued on page 27

Notes From CFRI

By Laura Mentch

Many paths led to Redwood City at the end of July, it seems. We gathered at the Sofitel for the 28th National Cystic Fibrosis Family Education Conference, commonly known as CFRI. As in "Are you going to CFRI this year?" As I entered the conference area the familiar people and place brought a smile. There was a warm welcome at the CFRI table and I had a visit with the vendors to learn what's new and thank them for their support. The U.S. Adult Cystic Fibrosis Association was present with an information table, a copy of *CF Roundtable* in each participant's bag and four of your directors attending.

Settling in Friday evening, I looked through the conference agenda. This year's theme was Partners in Progress. I read that we would hear about supporting our physical and emotional health, developments in medication, therapies and the drug pipeline. We would consider transition not only from pediatric to adult clinical care but also from childhood to the varied experiences of adults living with cystic fibrosis. I was reminded of the infection control precautions we needed to follow to be there together. And we would pay tribute to those no longer with us.

Throughout the weekend I listened, thought, asked questions and wrote. I offer the starred nuggets from my notes:

• Contact the company that makes my enzymes to access the nutrition debit card program.

• Avoid downward dog yoga posture; coming up from the head down position can lead to acid reflux splash into the lungs. (Scott Russell, physical therapist)

• Pancreatic function may improve with Kalydeco. (Margaret Rosenfeld, M.D.)

• The only antibiotic that can interfere with absorption of oral contraceptive pills is Rifampin. (Anna Tsang, N.P.)

• Nine women with CF have had successful pregnancies after lung transplant. (Anna Tsang, N.P.)

• The epidemiology of respiratory organisms is changing. (Lisa Saiman, M.D.)

• Consequences of depression may include poor treatment adherence; anxiety may be a motivator. (Martha Moskovitz and Kimberly Morse, social workers)

• Transition ≠ transfer and requires faith, trust and pixie dust. (Greg Sawicki, M.D.)

LAURA MENTCH

• Adult care centers provide expertise in adult care, experience with long-term effects of childhood diseases and attention to preventive care needs of adults. (Greg Sawicki, M.D.)

• Plan ahead for potential need to apply for SSDI even if you don't think you'll need it. (Beth Sufian, J.D.)

• Watch for heightened scrutiny by insurance companies for all persons with CF related to high cost of medication; especially most recently introduced new and costly medication. (Beth Sufian, J.D.)

On Sunday morning providers on the expert panel were asked what they have learned from patients who are doing quite well. Their responses:

Dr. Margaret Rosenfeld: An appreciation of what it's like to live with CF – how CF fits into a life, priorities and being partners in decision-making.

Dr. Greg Sawicki: Everyone defines their experience of living with chronic disease differently: there is a wide breadth of what individuals perceive as good health for them.

Anna Tsang, N.P.: Patients separate their view of lung function from quality of life. Life is a big circle. CF is a little circle.

These words are echoed in the experience of three presenters and our master of ceremonies all living with cystic fibrosis and actively engaged in the large circle of their lives: Isabel Stenzel Byrnes, Chris Kvam, Beth Sufian and Tess Dunn. Tess, a college student and musician, closed the conference by performing Matt Scale's song, *Breathe*. His 2001 lyrics foretell what we can see in 2015.

I wish I could breathe

Like you, oh, breathe
I know the answer's out there.
Won't you help me...please?

Then one day I'll breathe
I'll breathe, I'll breathe
I'll breathe like you.

I see the future.
It seems so clear.
That one day I'll breathe like you...
like you.

Partners in Progress, indeed. ▲

Laura is 62 and has CF. She is a Director of USACFA. Her contact information is on page 2.

Let's Exercise The Right Way With CF!

By Jeanie Hanley

The CFRI Educational Conference this year was particularly special for several reasons – first, after many years of working on the U.S. Adult CF Assn. (USACFA) board with Beth Sufian, I was finally able to meet this very special person and hang out with our masks and precautions in place. Beth also gave an outstanding talk on legal issues in CF. Second, other board members were in attendance including Laura Mentch and a relatively new board member, Chris Kvam, who gave the opening inspirational talk. Also our own *CF Roundtable* columnist, Isabel Stenzel Byrnes, kept it interesting as the Master of Ceremonies throughout the conference. Third, there were excellent talks by so many speakers. I've always felt there is always something to learn and sure enough I certainly did, as you will soon read below.

Also, I attended the CFRI retreat for the first time this year. Most attendees stayed for the whole week. I was able to stay for only two days, but what an unbelievable experience. Those two days gave me a wonderful glimpse of a fun, warm and engaging environment – one that I'd like to be a part for much longer in the coming years.

Back to what I learned, which is the point of this educational conference. I reiterate that there were many talks worthy of writing about and, fortunately, Laura Mentch's article in this issue will give you a glimpse of highlights on them. The session I want to address was given by Scott Russell, an instructor of Clinical Physical Therapy (PT) at the University of Southern California's (USC) Division of Biokinesiology and PT in Los Angeles. His talk was entitled "Fully Alive: Physical Activity, Exercise and Yoga for CF."

In his talk he covered exercise encountered in CF, causes of exercise limitations in CF, the benefits of exercise and activity, bone health, posture and yoga. It is important to know that an active lifestyle is defined as 10,000 steps per day, which is about five miles. Fewer than 5,000 steps is considered a sedentary lifestyle. Pretty crazy, huh? I believe these numbers need to be adjusted to focus on time and heart rate for those of us with chronic disease and getting on the "less young" side of life.

JEANIE HANLEY

In any case, an active lifestyle improves glucose tolerance and cardiovascular health. It also includes taking the stairs when possible and having a mix of strength training and exercises of moderate and rigorous intensity. An entertaining activity pyramid cartoon was shown that you can view at www.classbrain.com. Basically, TV and sitting—not good; aerobic exercises and recreational sports—good.

Aerobic fitness is determined by the capacity of the lungs and heart, muscle mass and capillary (the tiny gas-exchanging blood vessels) density. An FEV_1 above 60% helps us exercise more like a non-CF adult. Below 60% and especially in severe lung disease, on the other hand, affects our ability to ventilate or breathe effectively thereby reducing oxygen and carbon dioxide exchange. Low FEV_1 also causes air trapping and diaphragmatic flattening. All these cause an unwanted cycle of reduced tolerance to exercise due to slowed oxygen uptake, muscle fatigue, followed by further inactivity.

Time for some good news: Despite these deficits, Scott's presentation detailed how we can overcome them. First, regular exercise will decrease inflammation and the risk of upper respiratory tract infections. Throw in adequate sleep and these benefits will be magnified. In addition, disease progression and decline in FEV_1 will slow down.

Pursed lip breathing is a good method to use. It slows down ventilation, reduces air trapping and helps you control your breathing better during activity, especially activities that make you short of breath. So purse those lips and gently blow out. As Scott suggested, practice by making a candle flame flicker, but don't blow it out. Also I will add to make sure not to hold your breath while exercising. Let that air out!

High-intensity interval training is the most effective kind of physical activity for those with severe lung disease and pre-transplant. This type of training strengthens peripheral muscles and involves anaerobic and pyramid training, where you push yourself hard then ease up, followed by going higher again. Your heart rate should respond accordingly by going high to 180 for one minute then slow to 140 for 30 seconds and back up again, etc. Of course, we are all different and what will make one person's heart reach 180 is quite different from someone else even at the same FEV_1. So you must tailor the exercises to your heart!

As for our bone health, the goal for us adults is to maintain bone mass or slow the bone loss down and ultimately prevent falls and fractures. The best way to do this is — you got it — exercise and lots of Vitamin D. The types of exercise that work best are high impact activities such as jogging, climbing stairs and jumping. Aerobic and weight training several days per week will also keep bones in shape.

Other good news about all this exercise is that it will help our posture. Keeping our backs as straight as possible takes effort. We need to strengthen our upper body and core muscles, but also maintain flexibility. Once we allow our backs to curve (kyphosis), then interference with lung function occurs and we set ourselves up for vertebral fractures. Scott provided many suggestions for exercises – core and scapular strengthening, spinal flexibility and balance training. To see the numerous specific exercises, please watch and/or order his presentation at www.cfri.org.

> "High-intensity interval training is the most effective kind of physical activity for those with severe lung disease and pre-transplant."

Something you can watch immediately that will help you immensely in all these areas is on www.youtube.com entitled LKYT Yoga for CF. At USC Scott conducted a small study with CF patients that resulted in this video. This 20-minute video shows important dos and don'ts during yoga practice. The second step is to dive into a longer session by going to YogisAnonymous.com, click on "Video," then choose under: "teacher," Leslie Kazadi (who is the instructor in the USC video), "length": 90 minutes, "Level": all levels and hit "Search," The only yoga class that pops up can be tried for free to see if it works for you or, alternatively, contact Leslie to help you tailor yoga to your body. According to Scott's yoga study, yoga improved chest wall excursion (the difference between maximal inhalation and maximal exhalation), scapular positioning, lower extremity performance (measured by number of rounds of the sit-to-stand test), quality of life (as measured by questionnaire), balance and ability to do sit-ups (kicking in that core!).

Scott urges us all to set goals, know that exercise will get easier the more you do and, most importantly, have fun! Again, you can find the full-length presentation at CFRI's website. It's worth it! ▲

Jeanie is 53 and has CF. She is a Director of USACFA and is the President. You may find her contact information on page 2.

TILLMAN continued from page 24

mutations that cause the disease and is anticipated to provide a treatment option to the entire CF patient population.
http://tinyurl.com/nlrzly2

ProQR Announces Enrollment Has Started in Global Phase 1b Study of QR-010 in Cystic Fibrosis Patients

ProQR Therapeutics N.V. announced that enrollment has started in study PQ-010-001, a global Phase 1b clinical study of QR-010, a first-in-class RNA-based oligonucleotide designed to address the underlying cause of the disease by repairing the mRNA defect encoded by the DF508 mutation in the CFTR gene of CF patients. The DF508 mutation is a deletion of three of the coding base pairs, or nucleotides, in the CFTR gene, which results in the production of a misfolded CFTR protein that does not function normally. QR-010 is designed to bind to the defective CFTR mRNA and guide the insertion of the three missing nucleotides, thus repairing the mRNA and subsequently producing wild-type, or normal CFTR protein. QR-010 is designed to be self-administered through a small, hand-held aerosol delivery device, or nebulizer, in the form of a mist inhaled into the lungs. QR-010 has been granted orphan drug designation in the United States and the European Union.
http://tinyurl.com/p82evzf

Raptor Pharmaceutical Corp. Acquires Rights to Investigational Bronchiectasis Therapy Quinsair

Raptor Pharmaceutical Corp. recently signed an agreement with Tripex Pharmaceuticals to acquire rights to Quinsair, the first inhaled fluoroquinolone approved as a treatment of respiratory infections caused by Pseudomonas aeruginosa in adult cystic fibrosis patients. Quinsair is administered twice daily and contains levofloxacin.
http://tinyurl.com/q8asd6m

Staphylococcus Aureus Infection in CF Adult Patients Found To Be a Mild Disease Marker

In a new study the authors deter-

Continued on page 29

PARENTING

The Importance Of A Strong Support System

By Anne Governor

As a mother with cystic fibrosis, I am often asked by other CF patients contemplating parenthood what my biggest piece of advice is for successfully balancing parenthood and the inevitable trials of cystic fibrosis.

For my family, an incredibly stable support system is the best tool I have for managing the responsibilities of motherhood and the demands of my cystic fibrosis care.

Like many women with cystic fibrosis, I struggled with impaired fertility. My focus on getting pregnant and staying healthy during pregnancy was so intense, I neglected to plan ways in which our family and friends would be needed to help us once our baby arrived.

Our first born arrived six weeks early. We immediately had to ask our loved ones to step in to help take care of our pets and attend to chores around our home as we spent our days in the NICU.

Once our daughter came home from the hospital, we needed help with meals and everyday chores and people to care for our newborn while my husband worked and I tried to get enough rest to keep myself healthy. We quickly discovered how very important it was to ask for and accept help from others.

When our second daughter arrived, our need for help drastically increased. Not only did we have a newborn, we had a two-year-old daughter who needed care and we still had to take care of ourselves. Finding time to sleep and eat was a challenge. I became an expert at doing CF treatments with at least one child attached to me.

My first major CF exacerbation occurred seven months into my journey of being a mother of two children. I was admitted to the hospital and had no choice but to allow our support team to step in and take care of every detail of my home life. My job was to focus on getting my health in order. It was a humbling but necessary experience having loved ones stepping in to do my job of taking care of my family and home.

Every exacerbation brought a flood

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

Beth Sufian, 50
Houston, TX
On August 13, 2015

Kathy Yoder, 53
Portland, OR
On August 16, 2015

Wedding

Andrea Eisenman and **Steve Downey**
New York, NY
7 years on September 13, 2015

Jeanie and **John Hanley**
Manhattan Beach, CA
29 years on June 28, 2015

DeLayne and **Gerry Santos**
Gulfport, FL
23 years on September 19, 2015

Transplant

Stephanie Rath, 46
Brownsburg, IN
Bilateral lungs
2 years on August 15, 2015

of emotions in dealing with my faltering health and innate need to care for our family. Very slowly, with each exacerbation, my husband and I have become more comfortable with having others step in to help while I focus on my own health needs.

Now that our daughters are school-aged and I have a bit more time to focus on my own care, I still rely on our support system a great deal. I often need childcare help when I have doctor appointments or need some extra rest while doing at-home tune-ups. Our amazing support system is always available to assist in any way possible to help keep life running as smoothly as possible.

Our daughters are old enough to understand cystic fibrosis and how incredibly sick I can get very quickly. We are very honest in giving our daughters age-appropriate answers to their questions about cystic fibrosis. As we answer questions, we reassure our daughters with the fact that so many amazing people love and care for our family so much that they will do everything they can to help us when I am not able.

> *"We quickly discovered how very important it was to ask for and accept help from others."*

As our daughters grow and understand more about life with cystic fibrosis, we have even greater appreciation for each and every person who helps us when my cystic fibrosis care requires even more attention. For our entire family, knowing that we have such amazing family, friends and community members willing to support us in any way possible is of immense comfort.

Thanks to incredible advances in the treatment of cystic fibrosis, parenthood is a dream many of us will be able to achieve. Having a strong and stable support system helps to make the difficult moments easier to work through. ▲

Anne is 35 and has CF. She lives in the Rochester, NY, area with her husband, their two daughters, two dogs and a cat. You can contact Anne via her e-mail: governor@rochester.rr.com

TILLMAN continued from page 27

mined the prevalence of Staphylococcus aureus (SA) infection in adult patients with cystic fibrosis and how this type of infection determines patients' clinical outcomes. The authors performed a retrospective cross-sectional study and investigated the link between the microbial species present in the respiratory tract of CF adults, particularly the presence of SA and Pseudomonas aeruginosa (PA), and lung function. Additionally, the team determined other parameters, including plasma C-reactive protein (CRP, a marker of inflammation) and clinical outcomes. The authors suggest that infection with SA is a marker of less severe disease, and while in children SA infection is associated with a decline in lung function, in adults, SA infections in the absence of PA is a marker of mild disease.
http://tinyurl.com/pyxrlkj

Can Cystic Fibrosis Patient Mobility Be a Risk Factor for Pseudomonas Aeruginosa Infection?

Researchers reported that the mobility of cystic fibrosis (CF) patients between care centers increases the risk of transmission of the associated bacterial Pseudomonas aeruginosa infection. CF patients are thought to acquire P. aeruginosa through person-to-person transmission. Interestingly, cross-infection of shared cystic-fibrosis-specific P. aeruginosa strains has been reported across large geographical distances indicating wide dissemination of the pathogen. In the study, researchers assessed the extent to which the movement of CF patients infected with P. aeruginosa between CF centers contributes to bacterial dissemination. Researchers assessed 983 CF patients (mean age of 25 years) from whom they isolated and identified 531 distinct genotypes of P. aeruginosa. Of these genotypes, 493 were found to be unique strains in 373 patients, whereas 38 were shared strains in 610 patients. Based on their model, researchers concluded that the mobility of CF patients within CF centers is a potential key risk factor for the acquisition of shared P. aeruginosa strain infections.
http://tinyurl.com/pkk4mtp

Cystic fibrosis gene therapy trial offers hope of treatment

The results of a trial provide the first proof of concept that non-viral gene therapy is safe and can benefit lung function in patients with CF. The trial tested a gene therapy where the patient inhales molecules of DNA wrapped in fat globules (liposomes) that deliver a correct copy of the gene into

Continued on page 34

SEARCHING FOR THE CURE

Protecting Your Second Chance:

The Need For Therapies To Address Lung Transplant Complications

By Meranda Honaker and Reid D'Amico

Lung transplantation is the last option for end-stage cystic fibrosis patients. According to the 2013 CFF Annual Registry Data Report, there were 1,291 CF patients being followed and out of those patients 245 were transplanted within 2013. However, this number may be higher due to some patients receiving all of their care at a transplant center. The primary causes of death listed are respiratory, cardiorespiratory and transplant-related complications after a lung transplant. CF patients experience unique post-transplant complications due to worsening or development of CF-related diabetes (CFRD) and persistent CF sinus disease; however, it may be difficult to characterize the specific complications due to unique genetics and the inherent complexity of the transplant procedure. It is also important to note that the chronic infections seen in CF are not beaten with the removal of the diseased lung, since the sinuses may harbor many of the troubling bacteria that led to the damage of the previous lungs. In addition to the typical myriad health conditions (i.e., CFRD, sinus disease, liver disease etc.) CF patients face post-transplant, they also face risk of major health complications intrinsic to the new lungs themselves.

There is a great need to develop therapeutic strategies that can address the obstacles a lung transplant patient might experience. Unfortunately, there is not one simple research focus that can cover all possible complications that can arise with a lung transplant. Ranging from the immune system to molecular interaction, the complexity of organ transplant poses many hurdles to science. The foundation of this research should be built upon the genetics that coincide with the pathologies that manifest from the lung transplant.

For example, bronchiolitis obliterans syndrome (BOS) is characterized by the degeneration of the epithelial lining of the lungs. BOS is most commonly associated with lung transplant rejection, although it is increasingly reported in an occupational setting. Many have described this phenomenon as irreversible, and ultimately leading to a new lung transplant. However, this current prognosis can be sourced to a lack of understanding. Researchers at the National Institute of Environmental Health Sciences and Duke University Medical Center have sought to characterize BOS in hopes of shedding light on strategies that can prevent or even reverse this detrimental outcome from a lung transplant. In their 2015 publication in *Plos One,* they share their findings and shed light on the syndrome. Their experiment was poised to uncover how gene expression differs in the lungs of mice with BOS compared to those without BOS. In other words, are the genes creating proteins differently in the BOS mice when compared to the healthy mice? Their study, in fact, found many genes that were expressed differently. Some genes that code for proteins that are seen in the extracellular matrix (ECM) of the lung were shown to be much higher in mice with BOS than those without. The extracellular matrix is similar to the scaffolding of a building. It provides the collagen, elastin and other proteins that allow the cells to maintain their structural integrity. However, in the lung tissues of mice with BOS, there were some genes that coded for thirteen times the amount of necessary protein that is usually seen in ECM. Some of these genes are known to directly play a role in the irreversible fibrosis seen in lung transplant patients with BOS. Although the magnitude of gene expression does not necessarily correlate with downstream effects, further investigation of these highly up-regulated genes may provide important information needed to unravel the complex mechanisms involved in

MERANDA HONAKER

REID D'AMICO

BOS. The data from this study are being further evaluated to identify key genes and pathways in the pathogenesis of BOS and to facilitate the development of treatment strategies.

This account ultimately sheds light on the strategies that should be adopted in order to progress toward lung transplant treatments. Genetic analysis serves as a starting point to solve the puzzle regarding the mysteries of failed and complicated transplant procedures. By uncovering the molecular and genetic mechanisms, scientists and engineers can then work toward creating new medications to keep the patient alive and healthy. For example, if one of the genes seen above was creating 15 times the necessary amount of a certain protein, it may be possible to create a medication to "knock down" this protein thus resulting in fewer instances of BOS. However, we will be able to tell only if more studies are done to decipher the underlying cause of the pathologies that arise in lung transplant patients.

While a lung transplant may serve as a second chance for many cystic fibrosis patients, there are still many daunting challenges that must be understood and overcome. It is well known that lung transplants may provide life-changing advantages, but science must move to make the procedure successful for every patient. Fortunately, transplant research is a growing field, with more scientists devoting a career to understanding what complications can arise. However, we now find ourselves in a time where treatments and therapies must start entering clinical trials. We must protect this notion that a lung transplant is a "second chance," and this must be done by gaining knowledge and discovering treatments to prevent and reverse complications.

Clinical Trial Spotlight:

1. PET Assessment of Acute of Lung Transplant Rejection
https://clinicaltrials.gov/show/NCT02204202

2. Pro QR Therapeutics: Dose Escalation Study of QR-010 in Homozygous ΔF508 in CF Patients
https://clinicaltrials.gov/show/NCT02532764

3. Prospective Observational Study to Evaluate Biomarkers of Aminoglycoside Nephrotoxicity in Patients with CF
https://clinicaltrials.gov/show/NCT01543620

4. Redox Imbalance and the Development of CF-Related Diabetes (RedoxyMeal)
https://clinicaltrials.gov/show/NCT02202876

5. Predictive Value of PIIINP Urinary for the Development of Chronic Renal Failure in Patients with Cystic Fibrosis After Lung Transplantation (MUCO-IRC)
https://clinicaltrials.gov/show/NCT01572194 ▲

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

Reid is 22 and has CF. He is a Ph.D. student studying Biomedical Engineering. He recently graduated from Duke University and moved to Nashville, TN, to start his doctorate at Vanderbilt University. He is a Director of USACFA. His contact information is on page 2.

Clinical Trials... Helping Yourself By Helping Others

By Brad Aaron

We recently encouraged our readers to share their experiences with CF clinical trials. We selected the following article written by Brad Aaron due to his positive experience in a clinical trial. We hope this article will encourage others to consider participating in CF research!

I am a 30-year-old post-bilateral lung transplant recipient and this is how clinical studies have changed my life. It was a normal day and I got a phone call asking me if I would be interested in participating in a clinical study for a very promising new drug. I did not even have to think about it, the answer was yes. I had always wanted a chance to do my part in helping develop a drug that would help not only me but also countless others.

I went in for screening and was promptly approved to begin the study. There were a lot of tests to determine all the baseline information needed to compare to the results acquired during the dosage. I began the study drug and immediately felt better – better than I had felt in a very long time.

I was amazed at how well this drug worked. In this particular study I was dosed for two weeks, off it for two weeks and then dosed again for two more weeks. There is always a chance that you might receive a placebo. I knew that I had not received a placebo, since I felt so good. The days were long and tedious, but it was all worth it. I'll

Continued on page 36

CONVERSATION CORNER

#MyLifeMatters

By Klyn Elsbury

A few nights ago, I was wrapped in a blanket, lying on top of an RV off of a scenic overlook in Utah, staring up at a sky full of endless, scintillating stars. The air was cool and crisp, delightfully tickling my lungs as they adjusted to the altitude. A handsome man with a beautiful soul was holding my hand and pointing out Venus to the south. Together, we were dreaming about the future. Something that, until Orkambi came, I had all but given up on.

I dropped out of college because I started getting hospitalized several times a year and I believed I would never live long enough to pay off my student loan debt.

I moved to California from Florida for a career in biotech/pharmaceutical recruiting so I could be closer to the companies that were developing the very drugs that would keep me alive. That would give me hope. When I started getting hospitalized every four months, I made the choice to leave my corporate career and preserve my lung function via exercise, diet and adherence to prescriptions that managed the symptoms. I tried to get in on every clinical trial for Orkambi, before it was even called Orkambi, but time and time again I was denied because my lung function was too unstable.

He squeezed my hand excitedly, "did you see that?" referring to a shooting star that emblazoned an almost pitch-black night. My heart skipped a beat. I shut my eyes and made a wish that one day. Someday soon, I would be on this drug. I opened my eyes to see him smiling back at me.

For the first time in a long time, I believed I would have a future again. I was the first person in clinic the day after Orkambi was approved. However, they couldn't write a prescription because I needed to go on IV antibiotics first. My lung function was around 50%. It was my third round of IVs this year alone.

Meanwhile, one of my girlfriends locally who got approved for the drug posted on Facebook that for the first time in years, she woke up without coughing. I can't imagine a morning where an alarm clock wakes me up instead of a violent core-shaking, gut-busting cough.

"Wow!" We both said in unison at yet, another shooting star. Who is lucky enough to see two of them in one night sky — just moments apart? Surely this means there are good things to come. Waking up without a cough became my second wish.

KLYN ELSBURY

I finished my IV round just before taking off on this week-long road trip. I went to clinic and my blood tests revealed that despite three weeks on intense IVs, my kidneys and liver were strong enough to start Orkambi. I just had to fill out two simple forms and they had to fax it in. Simple.

The first fax was never received. I called clinic. They sent it again. This continued seven times. Finally my form went through. I had my initial phone call with Vertex somewhere between Vegas and Utah, where I was assigned a case manager and promised they would send the information to my insurance.

He squeezed my hand, bringing me back to reality. "What are you thinking?" he asked, with his goofy smile and wide, bright eyes. We heard an animal off in the distance and turned our faces to the right, just in time for the third shooting star.

"I once heard that it's bad to wish on a shooting star," I started, "because we are wishing on something that is already dead."

He immediately interrupted with an enthusiastic tone, "Actually, that dead star carries in it all sorts of bacteria and cells and other scientific stuff that is the perfect culmination to create new life. It just needs to land in the right environment."

I thought back to the day my dad had to rent a wheelchair and

> "An insurance company can't really put a price on my life. Can they? Will they?"

push me around 6 Flags on a family vacation. It hurt my lungs too much to walk. I hated everybody having to stop enjoying their time because I had to take "breathing breaks." Maybe soon I will be in the right environment. Maybe my lungs are that perfect combination of bacteria, cells and scientific stuff waiting for the right environment to thrive. My third wish was that he was right.

He kissed my forehead. Marilyn Monroe once said, "The real lover is a man who can thrill you by kissing your forehead or smiling into your eyes or just staring into space." I smiled and turned towards him as shooting star #4 lit up our little world. Wish number four was for my heart, not my lungs.

It was just after 3:00 a.m. and we decided to climb off the roof and go inside. Just as I stood up, I saw the final shooting star streak across the Northwest. Joyously dancing, as if a message from the heavens to tell me my life is worth living and everything I could possibly imagine, would come true. Now I'm not sure on how this whole shooting star, wish thing actually works. But I've had four amazing wishes, so I decided to save my final wish for if I ever needed it.

When I returned from my week-long trip, I was interviewed by KPBS about athleticism, cystic fibrosis and the hope Vertex gave me. The reporter asked, "Have you thought about what happens if you are denied the drug because of cost?"

I have spent a lifetime overcoming obstacles. I have found my greatest strength in the midst of insurmountable weakness. Yes, I have thought about being denied several times. The only thing I haven't thought about was, how I would tell my daddy that the drug I mentioned two years ago while he was pushing me around 6 Flags in a wheelchair, to fill his heart with reassurance and hope, I can't actually have. But then again, a drug company can't really do that. An insurance company can't really put a price on my life. Can they? Will they?

I left the interview and checked my voicemails. "Hi Klyn, this is Chris, from Vertex..." the message began. What followed was a 42-second scripted speech.

I was denied. Some executive, somewhere, decided my life is not worth the cost of Orkambi.

Little do they know, I still have my fifth wish. And I know this great spot in Utah if I need more... ▲

Klyn Elsbury is a former contributor to CF Roundtable *and currently lives in Southern California. Despite being on IVs/hospitalized six-plus months a year, she works part time as a NASM certified personal trainer, internationally licensed Zumba instructor and is a sponsored NPC Bikini Athlete. She believes there is no angry way to say "bubbles" and loves jokes, the people in her life and Starbucks.*

This blog has had over 15,000 trackable shares in the first week since the original publication. Klyn's story has since been featured on KPBS and NPR. Last week, GHPP approved Orkambi for patients with homozygous Delta F-508. She has filled out all necessary forms and had conversations with her clinic, Vertex and insurance. You can follow the story on mylifematters508.wordpress.com

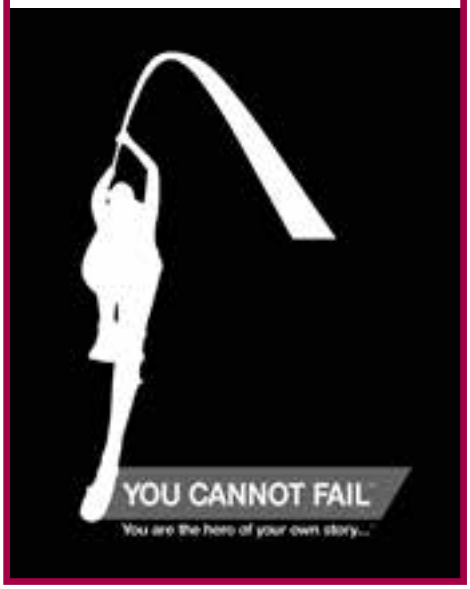
So far, she still has not received the drug.

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.



Voices from the Roundtable

Learning When Enough Is Enough

By Andrea Eisenman

I think of myself as being reasonably in shape. I exercise regularly, like three to four times a week, and bike outside when I can. I recently completed a 35-mile bike ride, to promote organ and tissue donation, on the North Fork of Long Island. It was not that strenuous. I did it at a leisurely pace and had a fun time.

So, when a friend of mine, who is a liver recipient, asked me to join her and her husband for a 35-mile bike ride in New Jersey, also for organ/tissue donation awareness, I thought, "Why not?" She was also at the North Fork ride. Knowing that the LI ride was pretty flat I asked, "Is the NJ ride hilly?" And she thought for a moment and said, "Not really." Well, I should have clarified the meaning of "hilly" or "mountainous" and "not really."

It all started fine. I spent the night in NJ, at my friend's house. I got treated to two cute Portuguese water dogs bouncing all over the place that made me miss my dogs less. In the morning, while I was gathering my stuff, my friend was inflating her tires so she tried to inflate mine and because my bike was still on my bike rack, blew my valve. So, I started stressing. At best, I can change a tire in about 30 minutes, when not panicked (we had a time limit to register for the event and get to the starting line). And luckily, I did carry a spare tire. But more luckily, my friend was a pro at changing tires. One part kept puffing out so that it was not a round tire shape but a bulging odd shape. Eventually it seemed okay and I was ready to get on my bike. The ride was incredibly organized. There were two bike shops to offer support. I had my problematic tire checked and it was fine.

I do not handle stress so well and maybe that set off my adrenals, which put me on edge. And that can set off a domino effect of losing electrolytes and hyponatremia. This happened to me at my first transplant games in 2004 and I was hospitalized for three days!

I tend to lose a lot of sodium due to having CF and all of my immunosuppressants don't help either. They leach away my electrolytes, specifically magnesium, calcium and potassium. So, I came prepared. I ate two bananas that morning, brought and drank Nuun tabs to put in my water bottles and had five 11.5-ounce juice packs of coconut water. Plus, daily, I take two grams of sodium pills and "Salt Stick" buffered electrolyte pills, in addition to my daily regimen of magnesium and calcium plus D.

I was prepared, except for those dang hills! The ride was deep in the hinterlands of New Jersey. It was beautiful but, my goodness, the hills were not what I was used to. My friend enjoyed the downhill aspect after reaching a peak but I was getting exhausted by the uphills. It was not that I could not do them, but I was sweating buckets so I knew I had to keep taking breaks, drinking my coconut water and not drinking plain water. Also, once I would get my momentum to get up a hill, I was faced with other slower bikers and could not pass them as it was a two-lane road with barely room for cyclists on the side. This made it hard-

TILLMAN continued from page 29

the cells in the lung lining. After one year of treatment, compared with placebo, patients who received the gene therapy showed "a significant, albeit modest, benefit" in forced expiratory volume (FEV_1). The researchers conclude the trial is the first to show that repeated doses of gene therapy can have a meaningful effect on cystic fibrosis and change lung function. The trial – run by the UK Cystic Fibrosis Gene Therapy Consortium – now needs to be followed by further studies to assess the optimum dose and treatment schedule.
http://tinyurl.com/qz5pmuj
AND
http://tinyurl.com/q4don6h
AND
http://tinyurl.com/oerh5wa

New Insights Suggest Thiocyanate Could Address Lung Inflammation in Cystic Fibrosis

Thiocyanate is a molecule found throughout the human body. Recent studies in animals have shown that thiocyanate could be used as a treatment for lung inflammation, although more research is needed. These new insights are particularly important for the CF community, as thiocyanate could potentially be useful as a treatment for cystic fibrosis. High oxidative stress is normally found in patients with cystic fibrosis, which contributes to the development of the disease's symptoms. Oxidative stress refers to the production of free-radicals – potentially harmful molecules that damage cells – faster than the body can remove them. Thiocyanate is one molecule that may be able to "mop up" oxidative stress.
http://tinyurl.com/nt6wohu

Lynovex activity in cystic fibrosis sputum and against the emerging CF pathogen, Mycobacterium abscessus

Researchers studied the activity of NovaBiotics' candidate cystic fibrosis therapy, Lynovex. The results obtained demonstrate Lynovex's striking ability

er to keep going. At times I got off my bike and walked up the remaining part of the hill.

After the designated rest stop, at about mile 13, I started feeling my gut get queasy and gurgly. At that point, I told my friend that instead of 35 miles, I could do only 25 miles. She concurred. But in my head I was thinking, I should just call it a day. About five miles later, we stopped at a deli on the route and I used the bathroom and I knew, no way could I go any further on my bike. I was starting to get nauseous and cold. It was about 86 degrees out and I had a long-sleeve sun protection shirt on under my bike jersey. Something was wrong.

I didn't want to quit because I felt like a loser, but also I felt my friend would stop too and she could continue if not for me. We had to find a ride marshal and get a "SAG" wagon to bring us and our bikes back to the finish line. I am not sure what SAG stands for but for me, it was how I was feeling—saggy. I was a bit humiliated and deflated. Basically, they picked us up in a car with a bike rack in the back: A rescue wagon of sorts.

Normally, I would have pushed on and probably been admitted to the ER and given IV fluids. Thankfully I came to my senses as I knew I had to drive home from New Jersey later and why be hospitalized when it could be prevented. I hung up my ego and gave in to common sense.

MICHELE DABAL AND ANDREA EISENMAN JUST BEFORE THEIR RIDE.

While I could not celebrate finishing the bike ride at the planned 35 miles or even 25, I learned we only biked 18. I felt I had to commend myself for not pushing myself into sickness and stopping when my body communicated, "You are done." Learning to listen to one's body is what makes us better patients with CF and transplant recipients...and what keeps us healthier with better communication skills to share our symptoms with our doctors. Also, I didn't want to hear it from my mom and husband that I pushed myself too far et cetera.

While I learned my limits, I will just have to practice on more giant hills. There is another ride coming up in September in New Jersey. But I plan to do only the 18-mile ride—trying to stay realistic. ▲

Andrea is 50 and has CF. She is a Director of USACFA and is the Executive Editor and Webmaster. Her contact information is on page 2.

to reduce sputum bacterial levels within only a few hours of exposure. Lynovex outperformed tobramycin and ciprofloxacin in its ability to kill the complex mix of sputum bacteria within these samples, but the combination of these antibiotics plus Lynovex was even more effective; an important finding as Lynovex is intended as an "add-on" adjunct therapy to be used alongside existing CF antibiotics in order to make them more effective and to counteract/reverse drug resistance. Some of the participating subjects in this study were infected with M. abscessus and these bacteria were isolated and their sensitivity to Lynovex assessed separately, with and without antibiotics currently used to treat M. abscessus. The data generated show that the clinical isolates and one type strain of M. abscessus tested were all sensitive to the antibiotic effects of Lynovex and that Lynovex also potentiated the antibacterial capacity of antibiotics commonly used to treat this pathogen.

http://tinyurl.com/pqs282m

Results from Pulmonary Non-Tuberculous Mycobacteria (PNTM) Study Using Aradigm's Liposomal Ciprofloxacin to Be Presented at ICAAC/ICC 2015

Scientists demonstrated that Aradigm's investigational drugs, Lipoquin® and Pulmaquin®, significantly reduced pulmonary non-tuberculous mycobacteria infection (PNTM) with Mycobacterium abscessus using once-daily respiratory tract dosing in mice that had established colonization with this microorganism. After three weeks of treatment, the number of colony-forming units (CFUs) in the lungs was significantly reduced by 95.2% and 96.1% by Lipoquin and Pulmaquin, respectively; after six weeks of treatment, the CFUs were further reduced by 99.7% and 99.4% for Lipoquin and Pulmaquin, respectively. In contrast, unencapsulated

Continued on page 36

TILLMAN continued from page 35

ciprofloxacin had no effect.
http://tinyurl.com/o5fmtms

Bacteria evolve differences within the lungs of patients with cystic fibrosis

Treatments for the same opportunistic bacteria found in cystic fibrosis patients can work in one area in the lung and be less effective in others. The reason is that bacteria become isolated from one another and evolve region-specific traits. Researchers saw differences in bacterial nutritional requirements, host defenses and antibiotic resistance. The findings suggest that other chronic infections may yield similar bacterial diversity. The team found that while all of the pseudomonas in a lung were descendants of a single strain, each region contained a vast array of sibling bacteria that functioned differently. When the investigators analyzed the genetic codes of the bacteria, the DNA sequences revealed that diversity arose because bacterial cells had become isolated in different lung regions and then evolved locally. The DNA sequences also suggest that traits that evolved over years or even decades may persist in bacteria inhabiting different lung regions and may provide a type of "memory" of past conditions and treatments that strengthen the bacteria.
http://tinyurl.com/nh9as7j

Nutritional Status Shown To Predict Changes in Key Lung Test For Cystic Fibrosis Patients

In both adolescents and adults, nutritional status can play a major role in the rate of disease progression. Individuals with poor nutritional status and low weight retention are at a higher risk for worsening lung function and increased mortality. An additional parameter that the researchers found to impact changes in lung function was diabetic status. Individuals with cystic fibrosis who had impaired fasting glucose tolerance were more likely to experience poorer lung function following a negative change in weight relative to individuals with cystic fibrosis who had normal glucose tolerance. A relative weight gain is positively associated to FEV_1%, while a relative weight loss of at least 2% has a significant negative impact on lung function. Patients with cystic fibrosis may therefore be encouraged to gain healthy amounts of weight to protect their lung function.
http://tinyurl.com/nvl8ezu

Inhaled Mannitol Found to Be Helpful for Those with Cystic Fibrosis

Several studies have recently been released looking at the use of dry mannitol powder in an inhalation device for the management of cystic fibrosis. Mannitol is an osmotic agent that, when dry and inhaled, draws fluid into the bronchial tree. This loosens the mucus in the bronchial tree so it can be more easily coughed up. Mannitol has been found to be safe in cystic fibrosis patients and effective in improving the removal of respiratory secretions. The study participants who used dry mannitol by inhalation had improvements in their spirometry readings, including the FEV_1 and forced vital capacity. Inhaled mannitol also decreased the number of disease exacerbations.
http://tinyurl.com/p3lqkhy

Celtaxsys, Inc., Gains FDA Clearance for Landmark Phase 2 Trial of Anti-Inflammatory Treatment for Cystic Fibrosis (CF)

Celtaxsys, Inc., announced that it has gained clearance from the U.S. Food and Drug Administration (FDA) to begin a Phase 2 clinical trial of its drug candidate, acebilustat (CTX-4430), in adult CF patients in the U.S. Acebilustat is a once-daily oral drug candidate being tested for the treatment of inflammatory diseases. It is a novel small molecule inhibitor of Leukotriene A4 Hydrolase (LTA4H), the key enzyme in the production of the potent inflammatory mediator Leukotriene B4 (LTB4). LTA4H and LTB4 have been strongly implicated in the pathogenesis of many diseases involving inflamma-

AARON continued from page 31

tell you why in a moment.

In the months after my 25th birthday, my health started slowly declining, hospital stays became more frequent and my lung function steadily dropped. Then I had a bout with my right lung collapsing over and over. My doctors and I talked and decided it was time to start the lung transplant process. I continued to get worse and it looked as though I might not have enough time.

Then, the doctor who had headed up the study I had done called and said he had me approved for a compassionate use of that same drug, which had made me feel so much better during the study. I said, "Yes!" I started immediately.

I took the drug for about six months and in that time my health did not get any worse and it somewhat stabilized. I did not get any better, either, but I think if not for this amazing drug that I would not be here today. I owe so much to the scientists, the doctors, the nurses and patients who spent so many hours and sacrificed so much to make this clinical study evolve into an FDA-approved medication.

We are given chances sometimes to help others and we end up helping ourselves along the way. Never think you can not make a difference. If you get a chance to participate in a clinical study, I urge you to go for it. You never know whose life you could save – maybe even your own. ▲

Brad is 30 and has CF.

tion, including cystic fibrosis.
http://tinyurl.com/qavrtac

Columbia Engineering Team Develops Targeted Drug Delivery to Lung

Researchers from Columbia Engineering and Columbia University Medical Center have developed a new method that can target delivery of very small volumes of drugs into the lung. They used tiny liquid plugs of medication and discovered that the level in which the drug absorbed into lung tissue was related to the volume of the liquid plugs. By changing the plug volume and the method of ventilation, medication could be delivered into the deeper airways, forming a micro-film of drug lining the bronchial tree.
http://tinyurl.com/qxomt84

PSYCHOSOCIAL

The psychological impact of adverse drug reactions amongst adults with cystic fibrosis. N. Reid. Journal of Cystic Fibrosis. June 2015. Volume 14, Supplement 1, Page S131

The experience of an adverse drug reaction can be an anxiety-provoking and potentially traumatic experience for CF patients. Appropriate medical and emotional support are essential in order to minimize psychological distress and reduce the impact on future treatment options.
http://tinyurl.com/nvzpjsy

Resilience, intolerance of uncertainty and CF patients' quality of life. H. Mitmansgruber, B. Rabanser, U. Smrekar, T. Beck, K. Niedermayr, J. Eder, H. Ellemunter. Journal of Cystic Fibrosis. June 2015. Volume 14, Supplement 1, Page S131

In several large epidemiological studies, CF patients report equal levels of anxiety and depression compared to healthy controls. They seem to cope particularly well considering their uncertainty and restrictions in living. (1) "Resilience" (the "bouncing back from adversity") and (2) "Intolerance of uncertainty" (IU, a vulnerability factor) have been investigated in their predictive power on CF quality of life. Data support the notion that the majority of CF patients in this sample rather successfully regulate than suppress negative emotions. Perceived personal coping competence has been confirmed as a major focus for intervention.
http://tinyurl.com/pg3svmk

How do men and women with cystic fibrosis think their illness and associated experiences affect their body image, sexuality, relationships and their ideas about parenthood? R. Anderson, A. Pearce, C.A. Graham, R. Ingham. Journal of Cystic Fibrosis. June 2015. Volume 14, Supplement 1, Page S132

This study explored how men and women with CF think their illness and associated experiences affect their body image, relationships, sexuality and feelings about parenthood in an attempt to improve understanding about the psychosocial aspects of living with CF. Body image findings suggested that women with CF are mainly happy with their slim body shape while men often want to gain weight/muscle to feel more attractive. In regards to disclosure, most participants wanted to get to know others before disclosing they have CF as they were concerned people might treat them differently. For most participants the desire for independence was the greatest consideration in the maintenance of a relationship. Factors found to affect intimate relationships included the direct effects of CF, such as coughing, tiredness and other physical symptoms, on sexual relationships. A possible protective effect of parenthood was found; men and women with CF who had children reported they prioritized treatment in order to optimize health as they felt they had a duty to be as healthy as they could for their children.
http://tinyurl.com/q5u39jl ▲

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.

Mailbox

Dear Friends,
Thank you for including my article in the latest issue of *CF Roundtable*. It was great how it was laid out!

Thanks again!
Sincerely,
Johanna Libbert

I want to continue my subscription. Thanks for covering me since last October while we have had no income. My husband finally found a job and we are moving to Maryland for that reason.

We have been married for 34 years as of July 11. I will be 58 in October. I want that to be an encouragement to others with CF. I hope to write for the newsletter after I get moved and settled!

Thanks,
Carol Shepard
Proud to be a CFer and Grandma!

PAY IT FORWARD BY DONATING TO CF ROUNDTABLE

As the holidays are fast approaching, what better way to "Pay It Forward" than by making a tax-deductible donation to the **U.S. Adult CF Assn. (USACFA), the producer of *CF Roundtable?*** You might be wondering why you should make a donation. Please read our mission and why any kind of donation is important to keeping CF Roundtable alive.

Before *CF Roundtable,* there were no means of communication on how adults with cystic fibrosis lived. We never had magazine subscriptions or newsletters geared toward adults and how they lived their lives with CF. We never read about adults beating the odds, inspirational stories on how to live with CF, or successful lung transplants.

This is YOUR newsletter and, because of your donations, YOU have made this newsletter possible for the past 25 years! We can now read others' stories, relate and know we are not alone in our struggle. With your help in making any kind of donation, our mission to provide you with inspirational stories, articles and interviews, as well as new research and events regarding cystic fibrosis can continue.

All work is done by volunteers and 100 percent of every donation goes into the production of the newsletter, *CF Roundtable,* and supporting services encompassing USACFA.

For the holidays, would you like to make a special donation in honor or in memory of someone who has passed? What about making a donation in celebration of a special milestone such as a transplant anniversary, birth of a child, wedding or a birthday? There is no greater way to honor and remember someone than on the holidays, and there is no greater reward than celebrating YOU and YOUR accomplishments. We will publish all donations, memorials, milestones and birthdays in our next newsletter.

To make a donation ONLINE, go to www.cfroundtable.com and click on the DONATE NOW button in the lower left-hand corner. Also e-mail the information below the dotted line to cfroundtable@usacfa.org.

USACFA, Inc., proudly produces *CF Roundtable,* a newsletter for adults who have cystic fibrosis.
www.cfroundtable.com ▲ cfroundtable@usacfa.org

CF Roundtable

Cut at dotted line, fill out form below and mail to: USACFA, Inc., PO Box 68105, Indianapolis, IN 46268-0105

NAME:

ADDRESS:

CITY: STATE: ZIP CODE:

THIS DONATION IS FOR (PLEASE CIRCLE): IN MEMORY/IN HONOR NAME OF THE PERSON:

PERSON TO NOTIFY FOR IN MEMORY/IN HONOR: ADDRESS:

CITY: STATE: ZIP CODE:

DONATION FOR MILESTONE: DONATION FOR ANNIVERSARY:

DONATION FOR BIRTHDAY: AGE:

COMMENTS: TOTAL DONATION AMOUNT:

Please make checks payable to USACFA
Remember USACFA is a 501(c)(3), non-profit organization, so any donation is tax-deductible!

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

AU 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.**

Benefactors

BRONZE

Judy & Ken Greenberg
Johanna Libbert
Janet Meek
Debra Maloney-Evans (In honor of Erin Evans)
Judith and Larry Lafary
Beverly Sufian (In honor of daughters Beth, Sandy & Aviva and of Beth's 50th birthday)

PLATINUM

CF Services
Gilead Sciences
Hill-Rom
Modern Health

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

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/.